Huntington's Disease Society of America
Tennessee AFFILIATE

Summer 2016

CHATTANOOGA TEAM HOPE WALK

HELP FOR TODAY. HOPE FOR TOMORROW

Watch Workshops and Presentations from the Thirty-first Annual Huntington's Disease Society of America from Home

The 31st Annual HDSA Convention just wrapped up. If you were not able to attend or there was a presentation you missed or would like to hear again, you can **still** be a part of this great event. Many of the presentations were recorded and can be watched online from the convenience of your home at http://hdsa.org/about-hdsa/annual-convention/2016-convention/.

The convention is always informative and uplifting. This year's convention was no exception. Although there were a few sessions that were not videoed, you can view the majority of the sessions plus the PowerPoint presentations for some that were not recorded. Videoed sessions available include:

- The opening ceremony with brief comments by Chief Executive Officer Louise Vetter and Board of Trustees Chair Jang-Ho Cha, MD, PhD, followed by an entertaining and encouraging discussion of caregiving by Tennessee's own Peter Rosenberger.
- Information you can use every day such as "I've Got the Music in Me" (an enlightening presentation about the positive effects of music), "Strategies for Managing Depression," "Living Positively with Huntington's Disease," "Making the Foods You Love Work for You," "Role of the Speech-Language Pathologist and Huntington's Disease" (discussing both speech/communication and swallowing), and "Strategies for Dealing with Aggression in Huntington's Disease."
- "Effective Strategies for Educating Law Enforcement and Medical First Responders on Huntington's Disease," including suggestions for being proactive and for communicating one-on-one with law enforcement, first responders, and 911.
- The Research Forum, "The Best Time to Plant a Tree", and the Clinical Trials Showcase – a total of four videos that explain and provide valuable and promising information about the current state of HD research.

- The question that stumps all of us – "Why is HD Still Diagnosed as a Motor Disease?"
- And MORE!

Many of the PowerPoint presentations from the sessions that were not recorded are available and provide valuable information. Among the slide presentations offered are "Between a Rock & a Hard Place: Helping Young People Cope with Grief, Loss, and the Reality of HD," "Long Term Care-What to Expect," "Exercise and HD," "Family Planning," "Talking About HD," "Legal Issues," and several others.

While HDSA works hard to make many of the convention sessions available by live streaming during the convention and recorded sessions online after the convention, there are other helpful sessions that HDSA is not able to record. More importantly, you cannot duplicate the experience of being with 900 plus people who know and understand HD. Also, the exhibit hall offers many opportunities to learn about services and products available to HD families. Plan *now* to attend next year's convention, the **32nd Annual HDSA Convention** beginning June 22, 2017, in Schaumburg, Illinois. Scholarships are usually available for First Time Attendees to cover the cost of registration, lodging, and transportation to convention. Watch the HDSA website (www.hdsa.org) or our affiliate website (www.hdsa.org/tn), or follow HDSA nationally or the Tennessee Affiliate on Facebook to learn details about the convention and scholarships when more information is available in January or February 2017.

Huntington Study Group to hold annual meeting in Nashville

The Huntington Study Group (HSG) will hold its 2016 annual meeting November 3 through November 5 at the Gaylord Opryland Resort and Convention Center. HSG is an organization of compassionate professionals dedicated to finding Huntington's disease treatments that make a difference and improving the quality of life and outcomes for HD families. To accomplish those goals, HSG brings together families, medical professionals, clinical researchers, HD advocacy groups, and sponsors to raise awareness of HD, share knowledge and best practices, and develop innovative treatments. The HSG annual meeting is an internationally-recognized forum for training and education of HD researchers and for presentation of new research findings and treatments. The meeting draws HD researchers and clinicians from throughout the United States, Canada, South America, Europe, Australia, and New Zealand. Many of the sessions are open to the public, particularly those on Saturday, November 5.

The annual meeting offers continuing education courses designed for HSG members, specialized programs for experienced HD clinicians and investigators, continuing medical education for local practitioners, and education for families and HD community members.

- Thursday, November 3 – These sessions and a number of sessions on Friday, November 4 will be focused on HSG members and researchers.
- Friday, November 4 – In addition to the sessions for HSG members, there will be a full day of continuing medical education program specially designed for regional health care providers, including physicians, genetic counselors, psychologists, nurses, social workers, and other health care practitioners. Encourage your health care providers to plan to attend the practitioners' continuing education program.
- Saturday, November 5 – HDSA Family Education Day. The day will begin with the highly-acclaimed HD Clinical Research Symposium, which draws worldwide recognition with an annual following of over 300 leaders in HD. The symposium includes information about the latest findings in research and treatments for HD. HSG wants families, patients, and HD community

members to attend the symposium as it actively engages the entire HD community (researchers, clinicians, and people affected by HD) in discussion about the future of HD care and research. The rest of the day will consist of practical workshops and sessions designed for the HD community and HD family members.

There is no charge to attend the HSG annual meeting so make plans now to attend. Through the generosity of Blue Cross Blue Shield Tennessee Health Foundation, the Tennessee Affiliate will offer scholarships to assist families with the travel and respite care costs for the HDSA Family Education Day. Like us on Facebook and check our website, www.hdsa.org/tn for more information about the scholarships. For more information about the HSG annual meeting, go to http://huntingtonstudygroup.org/about/our-annual-meeting.

Letter from Nancy Hale, HDSA Tennessee Affiliate Chair

The past year was another great one for Tennessee Huntington's disease families and the Tennessee Affiliate of the Huntington's Disease Society of America. We were able to provide education days for families, patients, and caregivers in Memphis and Nashville and a conference with Social Worker Carol Rabideau in Knoxville. We raised HD awareness and funds with Team Hope Walks in Nashville, Lewisburg, Mt. Juliet, Knoxville, and Chattanooga. In addition, we raised awareness through a songwriters' benefit in Dickson, a golf tournament in Memphis, and a fashion show/half-marathon weekend in Knoxville. Through these events, many people who were not familiar with HD learned about the disease and many HD families were introduced to HD resources in Tennessee. Our new website debuted in April and we continue to add information about HD resources, HD support groups, and HD-related events to the website. A part-time HDSA social worker will assist families across Tennessee. Through our fundraising efforts and donations, we raised a little more than $25,000 (net of all expenses) to be used to support HDSA's research, advocacy, and patient/family support efforts. This great year was capped by the announcement that the Huntington Study Group, an international group of HD researchers and clinicians, will hold its 2016 annual meeting in Nashville. Read more about that fantastic opportunity elsewhere in this newsletter.

Special thanks go out to Joy Sexton, who produced the elegant Knoxville fashion show and half-marathon and raised general community awareness to new heights; Tiffany Adams, who coordinated the first Lewisburg Team Hope Walk, introducing our cause to an entirely new community; Kim Sellers, who organized the outstanding Singing and Supper – A Celebration of Hope, giving us a fun opportunity to celebrate the advances that promise to brighten the futures of HD families; Gina Becker, who led her sixth Team Hope Walk, continuing the great tradition she began in 2010; Mark Bascom, who led the Knoxville Team Hope Walk, reaching and giving hope to newly-diagnosed families in East Tennessee; Don Saemenes, who chaired his SEVENTEENTH annual Cure HD Golf Tournament, which was both our highest grossing and highest netting fundraising event this year, and our regional development manager, Kim Brammer, who stepped in and coordinated the Chattanooga Walk on our behalf at the beautiful Riverpark.

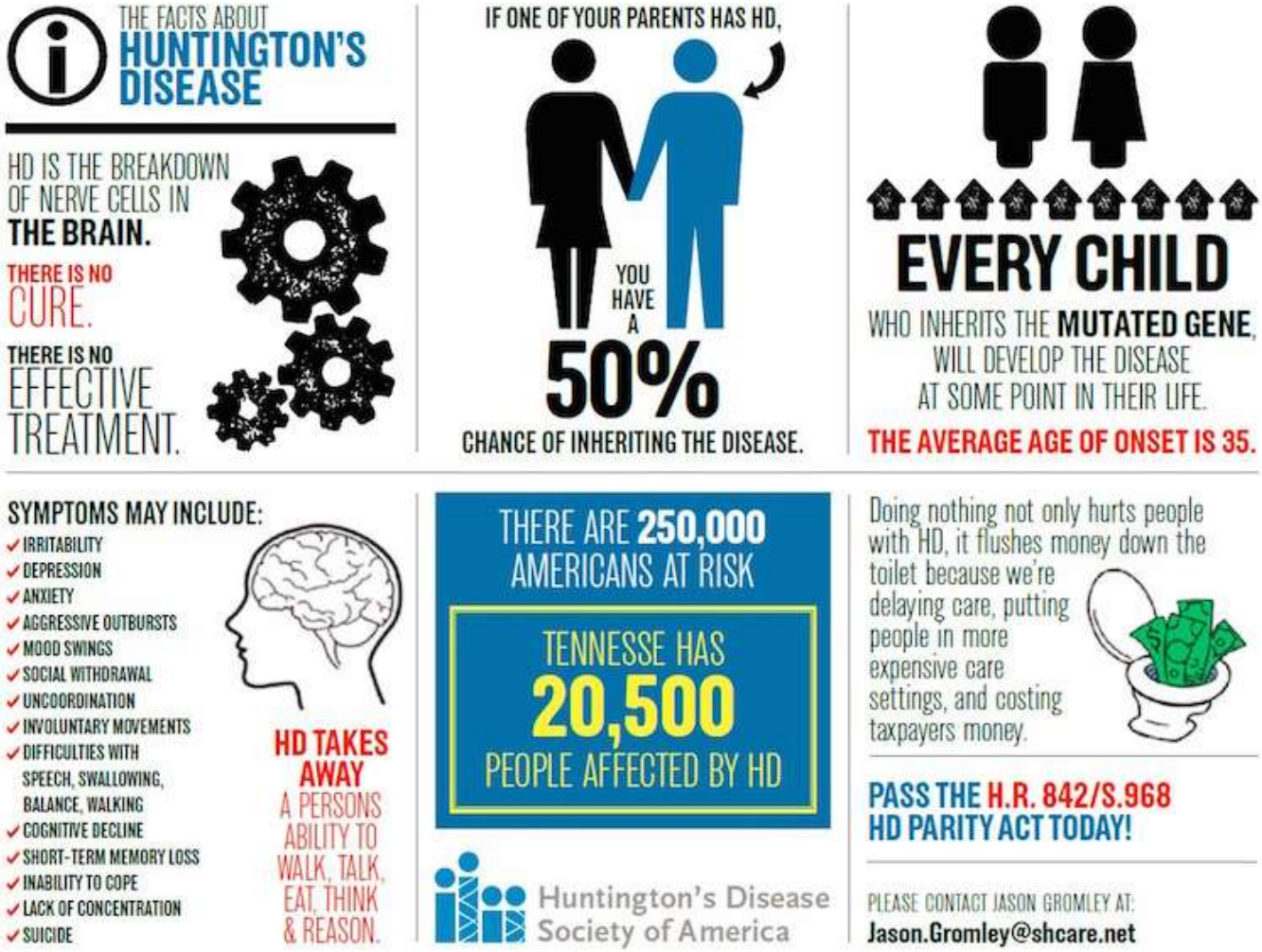
We all know there are tremendous needs in the HD world – for research, awareness, advocacy, support for HD families and patients, and education for medical professionals and the community. It takes all of us

working together to meet those needs. We are fortunate that there are a number of organizations that address some of those various needs, some on a local or regional level and a few on a national level. HDSA, however, is the only national organization that works across the board

- providing education for patients, families, medical professionals, law enforcement, long term care facilities, and the community at large,
- advocating for HD families,
- providing support to people and families affected by HD, and
- supporting HD research.

The adage, *there is strength in numbers*, is more than mere words. We need the unity and broad nationwide impact we get from HDSA to support education of the community and our families, remain abreast of the latest research and clinical developments, and advocate for the needs of our patients and families. Our mission, as the Tennessee Affiliate, is to use the vast resources of HDSA to identify and address the needs of Tennessee HD families. To accomplish that mission, we will continue to sponsor education sessions and support groups, seek to provide information to the community about HD, and advocate for the needs of HD families. We welcome suggestions regarding the needs that should be targeted and ways to meet the needs of Tennessee HD families.

Facts about HD



Looking to the Future

By Carol Rabideau, LCSW, Vanderbilt Medical Center, HDSA Center of Excellence

Throughout the year, there are times you or people in your life want to give gifts – whether in the form of an item or an act of kindness. Supportive gifts are always a good idea. This could be a consumable gift of food, a nurturing gift like a massage, or a rejuvenative gift such as providing some respite, or a trip to the grocery store. These can be win-win gifts for all – the givers and the receivers.

Another meaningful gift is assistance taking steps to plan for the future. Here are some ideas:

1. Does your family member with HD have a "co-pilot" for
    a. Making medical decisions?
    Durable Power of Attorney for Health Care (DPoAHC) – can be effective immediately, along with the patient, or can become effective when the person becomes incapacitated.
    Health Care Surrogate (HCS) - designates someone to make health care decisions if you are not able.
    b. Taking care of financial responsibilities?
    Durable Power of Attorney – Gives "co-pilot" the legal power to manage financial affairs. The person giving the power to a PoA does not lose power themselves. PoA is different than just putting someone else's name on a bank account which only allows a person to write checks on that account. And a Durable Power of Attorney is distinguished from a plain Power of Attorney in which case the agent can only act as long as the person/patient has legal capacity. A Durable Power of Attorney says "the authority granted survives my subsequent incapacity."

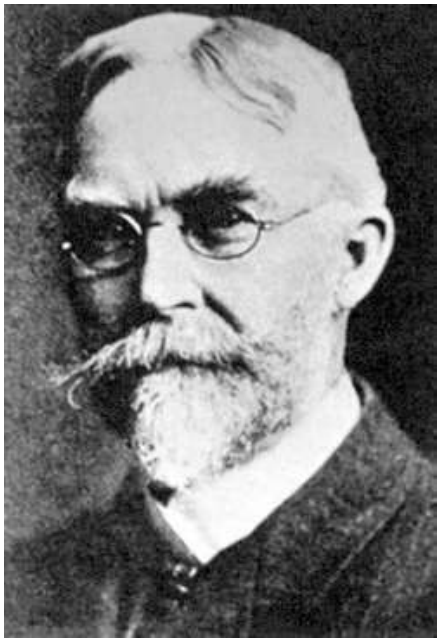
2. Have your family members given others a roadmap about their care?
    a. **Living Will** – "If any of these conditions occur, this is what I want or don't want." Might be tube feeding, fluids, pain management, or other measures. This gives the HCS/DPoAHC a roadmap. Without a living will, a court battle can go on for years.

These descriptions and discussions of legal documents are provided by **Sean W. Scott, JD, 727 539-0181, Elder Law Attorney, Largo, FLA swscott@virtuallawoffice.com**.

Making these decisions and executing these documents with the help of an attorney can provide peace of mind and make going through difficult times a little easier. Whether we have HD or not, none of us know what tomorrow brings. Taking these steps can be another meaningful gift for family members to give one another, an important expression of love in this journey through life.

Huntington Disease - Back to the Basics

By Mark LeDoux, MD, PhD



(Photo of Dr. George Huntington)

The eponymic terms Huntington Disease or Huntington Chorea arose from an exacting and prescient description of the disorder by George Huntington, MD. In 1872, Dr. Huntington wrote the following about the disease that now bears his name, "…It is attended generally by all the symptoms of common chorea, only in an aggravated degree, hardly ever manifesting itself until adult or middle life, then coming on gradually but surely, increasing by degrees, and often occupying years in its development, until the hapless sufferer is but a quivering wreck of his former self…" It should be noted the common chorea is a passé term for Sydenham's chorea which is associated with strep throat and rheumatic fever.

Huntington disease (HD) is caused by an abnormal trinucleotide repeat (CAG) in the gene encoding huntingtin (HTT). CAG encodes glutamine and repeated stretches of CAGCAGCAGCAGs. . . encodes a protein containing a polyglutamine tract. HD is an autosomal-dominant neurodegenerative disorder that typically results in midlife onset of progressive motor abnormalities (positive and negative), behavioral disturbances, and cognitive impairment. Although the average age of onset is around 39 years of age, some individuals develop HD before 10 years of age or after 80 years of age. The oldest age of onset reported in the medical literature is 89 years. United States prevalence estimates for HD range from 4-10 per 100,000 persons. At present there are over 30,000 affected individuals in the United States and 140,000 people at risk of developing HD.

Motor features of HD include chorea, dystonia, myoclonus, tics, dysarthria, bradykinesia (slowed movements), rigidity, postural instability, dysphagia, parakinesias, motor impersistence, tongue protrusion (darting tongue), and milkmaid's grip. Chorea is the most common and characteristic feature of HD. Motor abnormalities such as myoclonus are uncommon and may never appear during the lifetime of an affected individual. Cognitive features of HD may include poor judgment, impaired recent memory, decreased attention and concentration, abnormal visuospatial processing may be impaired earlier in the disease course, and impaired recognition of emotional facial expression. The most common HD neuropsychiatric symptoms in descending order of frequency include dysphoria, agitation, irritability, apathy, anxiety, disinhibition, euphoria,

delusions, and hallucinations. In fact, hallucinations are quite uncommon in HD. Patients with early HD are at increased risk for suicide. Risk of suicide is higher in males than females. Individuals with robust family support are less likely to commit suicide.

The HD gene (*HTT*) is located on Chromosome 4. With rare exception, people with 35 or fewer CAG repeats will not develop HD during their lifetime. Individuals with 36-39 repeats may develop HD whereas persons with 40 are more repeats will likely develop HD if they live long enough. In rare cases, repeat number may exceed 80 and these unfortunate individuals will typically develop childhood onset HD. HD is the most common hereditary cause of chorea. However, there are over 10 other hereditary disorders that can also include chorea as one disease manifestation. Isolated cases of chorea may be due to non-hereditary causes such as medications and immune disorders such as lupus and antiphospholipid antibody syndrome.

At present, tetrabenazine is the only FDA approved treatment for patients with HD. Neuropsychiatric manifestations such as depression and anxiety often respond well to medications approved for these specific indications. Patients with HD may benefit from moderate exercise, physical therapy, speech therapy, occupational therapy and a healthy diet rich in fruits and vegetables. Support from family and friends is essential. Patients with HD should remain integrated in their public circles including church, hobbies, and work as long as reasonably possible and avoid social isolation. Current research holds genuine promise for viable treatments that will delay disease onset and/or slow disease progression.

Cure HD Golf Tournament 2015

Team Hope Walks set for October

Make plans to attend and bring your family and friends to the Chattanooga, Knoxville, or Mt. Juliet Team Hope Walks (or make it a grand tour of all three!) in October. Team Hope Walks provide a time to raise the profile of Huntington's disease for the community at-large -- breaking down stereotypes, building understanding, and engaging more people in our work to fight Huntington's disease. The month of October will begin with the Saturday, October 1, Chattanooga Team Hope Walk at the Fry Center at Riverpark. The walk itself will be a beautiful riverside stroll beside the Tennessee River. One week later, join us on Saturday, October 8, in Knoxville, again beside the Tennessee River at Volunteer Landing. The next week, on Saturday, October 15, we will run and walk in Mt. Juliet. Although the Tennessee River does not go through Mt. Juliet, the 5K and Team Hope Walk at Charlie Daniels Park will be scenic with the flat 5K route through a residential area and the short walk totally contained in the park.

For more information and to register:

Chattanooga Team Hope Walk – Saturday, October 1 – www.hdsa.org/thwchattanooga

Knoxville Team Hope Walk – Saturday, October 8 – www.hdsa.org/thwknoxville

Mt. Juliet 5K and Team Hope Walk – Saturday, October 15 – www.hdsa.org/thwmtjuliet

Bring all your family and friends! In addition to the short scenic walks (and the 5K race in Mt. Juliet), there will be loads of fun activities for the kids, entertainment, and more.

Meet the Schmidt/Pace Family

My name is **Gary Schmidt**. I lost my lovely wife, **Eileen,** to Huntington's Disease in February 2012 at age 58. She struggled with HD for approximately 15 years. Living with HD, Eileen feared that she would miss out on two big life events: (1) seeing our daughter, **Brionne,** graduate from college and (2) seeing her walk down the aisle on her wedding day. As you see by the picture above, God blessed her with witnessing both milestones. In addition to myself and our daughter, she left an awesome son-in-law **William** (who was always her favorite), her father **Rue**, sister **Laura**, brother **Rue** (whose battle with HD ended April 14, 2015), niece **Nikki** (with HD), nephew **Josh,** and a great-niece and great-nephew (at-risk). Since her death, she has become a grandmother and would be so proud of the cutest little man **William III.**

Eileen loved life, studying the Bible, serving God, and don't get in the way of her chocolate. She was always the nicest/sweetest person in the room. You could always count on her for some encouragement.

We miss her daily, but thank God for the time we had with her. We now rejoice that she is with our Lord in Heaven and look forward to the day we are all united again. We support, hope, and pray to find a cure for HD.

In the first picture below, Eileen's mother, **Coravallene Pace,** is sitting with Eileen's siblings and Nikki. Pictured, from left, are **Eileen**, **Corinne** (whose battle with HD has also ended), **Laura** (who has not shown any

symptoms), **Rue (**whose battle with HD ended in April 2015), and **Nikki** (Corinne's daughter). The second picture is of Nikki and her two children.

Cure HD.

(Coravallene Pace Family)

(Nikki Schwartz Family)

Time to visit with HD support group?

By Melissa Darnall

When it comes to attending a support group, what is your first reaction to the idea? Yes, mine too. Do I have to talk and participate or can I just sit and listen? Will someone ask me questions I really don't want to answer?

These are all normal reactions to the idea of attending a support group. Coming from a family with HD, caring for a father with HD and being at-risk for HD are the reasons the Nashville Support Group was founded. After many years of caregiving and having an HD support group I continue to learn from others in our HD community. Our support group welcomes anyone affected by HD as a place where you can feel comfort in knowing you are not alone.

Share what you want, if you want, and know without a doubt that many others in our community are there to support you.

The Nashville Support Group meets the second Thursday evening of the month 6:30PM-7:30PM at Skyline Medical Center in Auditorium A on the ground floor. The facilitator is Melissa Darnall (HDadvocate.Melissa@gmail.com, (615) 714-6519).

The Memphis Support Group meets the fourth Saturday of each month, usually from 10:00 AM – 11:30 AM at Cordova Public Library, Meeting Room A. In September and December 2016, the group will meet from 3:30 PM to 5:00 PM. The facilitators are Dr. Misty Thompson, PhD, (misty.thompson@gmail.com) and Amanda Nolte (anolte@uthsc.edu (901) 448-6180).

We are currently looking to expand our support throughout Tennessee. If you or someone you know is willing to start a support group, please email: HDadvocate.Melissa@gmail.com. For any additional information, please contact Melissa Darnall (615) 714-6519 or Anne Leserman, LCSW, at aleserman@hdsa.org or (212) 242-1968, ext. 240 .


(Misty Thompson)


(Amanda Nolte)


(Melissa Hall Darnall)

Predictive Testing for Huntington Disease: What am I getting myself into?

By Vickie Hanning

Many of you may be considering having predictive testing for HD, but are not sure of the cost and time commitment involved. Therefore, I have summarized information about the testing process below.

1. **Testing typically consists of three appointments on 2 different days**. We encourage you to come to all appointments with a support person such as your spouse, significant other or a friend.

a. You meet with a genetic counselor (me) for about an hour and on the same day,

b. You meet with one of our neurologists who specialize in Huntington disease, usually Dr. Katherine McDonell, for about 30 minutes.

The purpose of the first 2 appointments is to obtain information about you and why you want to be tested, provide genetic counseling about HD, discuss benefits, drawbacks and limitations of predictive testing and talk about how results might impact your life. In addition, Dr. McDonell will examine you for signs of HD.

Records confirming a family member's diagnosis of HD are helpful, but not required. The main reason for confirmatory records is to ensure that we are testing you for the correct genetic condition.

In most cases, individuals wanting testing will have their blood drawn following these appointments. The results will go in your electronic medical record, but are not sent to anyone, including your primary care doctor, unless you request this.

For individuals who are especially concerned about the privacy of their results, there is an option of being tested under a false name. If you prefer this option, it is necessary to provide a check to pay for the laboratory cost at the time your blood is drawn. Your appointments with me and Dr. McDonell will be in your own name, but details of our discussion, including your risk to inherit HD, can be left out of the medical record.

If you plan to purchase insurance products such as whole life or long term care insurance, we recommend waiting to receive your test results until your purchase is completed.

c. Approximately 2 weeks later, if testing was done, you meet again with me and Dr. McDonell to discuss your test results and receive a copy of the result to take with you. **Results will not be given by telephone.** The length of this session varies a lot, depending on the test result and what questions each individual has.

If you cancel your disclosure visit or no show, we will not contact you, since we will assume you do not want the results yet. The results will be in your medical record or in the medical record under a false name and will not be sent to you or anyone else, until disclosed to you in person.

2. **Cost:** Accurate cost estimates cannot be made if you are paying by insurance, as each policy is different. If

you decide to private pay the charges, best estimates are below:

Initial Genetic counseling visit	$ 130
Neurologic Evaluation	$ 150- $300
DNA testing:	$ 220
Follow up visit	$ 150

If you choose to private pay and are seen at Vanderbilt for other services for which you use insurance coverage, a separate private pay account can be set up for you.

3. **Who can be tested?**

At present, we test asymptomatic individuals who are 18 years of age or older. Children are sometimes tested if they have neurologic symptoms, to rule out HD as the cause of their symptoms. The reason for not routinely testing asymptomatic children is that we know many individuals at risk for HD do not want to know their test result.

Since an asymptomatic individual's test result does not affect medical care, the standard of care is to test only individuals who can legally consent. This is an imperfect rule, but it is the best way we have to be sure that individuals who don't want their result aren't tested. It is understandable for a parent to want to know their child's result, but there is potential harm if a child is tested early and found to be positive. This rule will change when medication slowing the progress of HD is available and found to be safe for use in children.

Testing could potentially be postponed for an individual due to mental health concerns. However, this is extremely rare.

Adults at-risk for HD can be tested even if their at-risk relative chooses not to be tested, since they have the legal right to request predictive testing. We usually talk during genetic counseling about how they might handle a positive test result with their family, since this would automatically diagnose a parent or an identical twin.

4. **Prenatal testing:**

Prenatal testing for HD is accurate and available at some medical centers. The concern that medical professionals have is that a gene positive child diagnosed at birth might be negatively impacted by the result, even if the child were not informed. Also, if the result were known and not shared, the child would very likely resent the secrecy when he or she found out. However, if a couple arranged to have prenatal diagnosis for HD, they would be at liberty to make their own decisions about continuing or ending the pregnancy if the result were positive.

5. **Questions?**

If you have more questions, you can contact me or another genetic counselor by telephone or email, or if you prefer, you can schedule an appointment. My contact information is:

Email: Vickie.hannig@vanderbilt.edu

Telephone: 615-322-7601

Schedule an appointment or request a copy of the Vanderbilt HD Predictive testing protocol: contact Lisa Robinson, 615-875-1850

Knoxville Team Hope Walk 2015

Coming HD Events in Tennessee

(Details and Additional Events can be found on Facebook – (HDSA-TennesseeHDCommunity) and the Affiliate Website (www.hdsa.org/tn)

- Saturday, October 1, 2016 – Chattanooga – Riverpark – Hubert Fry Center – Team Hope Walk Chattanooga www.hdsa.org/thwchattanooga
- Saturday, October 8, 2016 – Knoxville – Volunteer Landing – Team Hope Walk Knoxville www.hdsa.org/thwknoxville
- Monday, October 10, 2016 – Memphis/Millington – Mirimichi Golf Course – 18th Annual Cure HD Golf Tournament www.hdsa.org/18tngolf.
- Saturday, October 15, 2016 – Mt. Juliet – Charlie Daniels Park – Team Hope 5K and Walk www.hdsa.org/thwmtjuliet
- Thursday, November 3, through Saturday, November 5, 2016 – Nashville - Huntington Study Group meeting – HDSA Family Education Day on Saturday, November 5. More Information - http://huntingtonstudygroup.org/about/our-annual-meeting
- Saturday, November 12, 2016 – Dickson - Singing and Supper – A Celebration of Hope – A benefit concert and dinner. www.hdsa.org/singingnsupper.

In Memoriam – Remembering those we lost in 2015

Our loved ones leave the world, but never leave our hearts.

Richard D. Allen III

Richard D. Allen IV

Carolyn Hayes Baker

Harold Ted Ledford

Rue Pace Jr.

Karen Romberg