

Greetings!

I hope this finds you well. I will keep this brief as I reflect on the 2021 year and my time as serving as the Illinois Chapter President, I am honored to have worked with so many great people. The perseverance and determination to keep working for a cure is amazing. The dedication of the volunteers is amazing, and I cannot think of another group of people that I would want to work with. As my term as President has come to an end, I will still be around volunteering and working towards the HDSA Mission. I hope you all will welcome the new chapter President, Arvind Sreedharan with open arms. Arvind brings great leadership, knowledge, and HDSA experience that will keep the chapter moving forward.



Thank you again for all of your support and dedication! Take care.

Larry Haigh  
Outgoing President, Illinois Chapter HDSA

## President's Message



Dear HD Families,

I hope all of you had a relaxing holiday. Happy 2022! 2021 was a trying year on many fronts, and 2022 continues to be challenging with the pandemic's difficulties and uncertainties.

I am humbled and very excited to support the HDSA Illinois Chapter in my new role. I heard from many people I have interacted with at the national level how the Illinois Chapter is a model for success in supporting the local HD community. I have seen this passion and dedication of Illinois Chapter HD community members with my own eyes over the last two years.

On that note, I want to begin by complementing and commending past President Larry Haigh. Larry has been an excellent leader for the Illinois Chapter. I am very thankful for his past contributions and his tremendous help as I transition into this new role. Thank you again, Larry, for your dedication and commitment to the Illinois Chapter.

I also want to thank Erin Riley for her long service to the Illinois Board. Thank you, Erin, for all your contributions.

Finally, last but not least, I want to recognize the dedication and support of our social worker, Emily Zivin. Emily has provided outstanding support for our local HD community and continues to do so. In late 2021, the chapter received an incredibly generous year-end donation of \$30,000 in honor of Emily Zivin to recognize her service to HD families. This donation is a testament to Emily's outstanding support and care for our HD community. Thank you, Emily, for all you have done and continue to do for the Illinois HD community.

Our mission as a Chapter is to carry out the HDSA mission, which is to improve the lives of everyone affected by HD and their families. I am committed to supporting this effort and thank you in advance for your commitment to carry out this mission at the local level. During these unprecedented times, I hope things will get back to a level of normalcy in time. As always, please feel free to reach out for any support and or help.

I look forward to seeing you in 2022!

Arvind Sreedharan  
President, HDSA Illinois Chapter



Hello!

We would like to invite you to participate in the **PREVENT-HD** research study. The Neurology Department at the University of Wisconsin-Madison is studying the effects of Huntington Disease (HD). This NIH-funded research study is looking at challenges in clinical trials for persons before Huntington's disease (HD) diagnosis. We hope that by addressing them, future clinical trials will be better able to test treatments to delay onset or slow the progression.

If you agree to take part in this study, your involvement will consist of two in-person visits and one "at-home" visit over the next 2 years.

**As a participant in the study, you will:**

- 1) Have an interview about your health.
- 2) Take tests that will assess your language, motor, and memory skills.
- 3) Complete a neurological exam.
- 4) Have an MRI.
- 5) Provide a blood sample.
- 6) Have a Lumbar Puncture (LP).
- 7) Possibly provide a saliva sample.

**Other information:**

- For the in-person visits, your participation will involve a 2 day (~6 hour each day) visit to UW-Hospital in Madison, Wisconsin.
- For the "at home" visit you will be mailed an iPad to use for completing the study assessments.
- You may receive up to **\$350 for completing this study:**
  - **\$50** for completing each in-person visit and blood draw.
  - **\$50** for completing the "at-home" visit.
  - **\$50** for completing the MRI.
  - **\$50** for completing the Lumbar Puncture.
- Meals will be provided for you during your study visit
- If traveling from a distance, we will assist with travel cost or pay for an overnight stay at a local hotel.

**If you would like further information or do not wish to be contacted about this study, please call 833-828-0122 and leave a message. You may also reply by e-mail ([preventhd@neurology.wisc.edu](mailto:preventhd@neurology.wisc.edu)).** If we do not hear from you, one of our research coordinators will contact you in about two weeks by phone to provide further details. We hope you will consider participating in this project as we are very excited about having the opportunity to contribute to the future care of people with Huntington Disease.

Sincerely,

Jane Paulsen, PhD





Deborah Hall, MD PhD  
 Professor, Neurological Sciences  
 Director, Rush University HD Center of Excellence  
 1725 West Harrison, Suite 755  
 Chicago, IL 60612  
 312-563-2030 (main)  
 312-563-2024 (fax)  
 Movement\_HD@rush.edu (email)  
<http://www.rush.edu/services/conditions/huntingtons-disease>

### Update on the COVID-19 pandemic and Vaccine

March 2020 marked the beginning of the pandemic caused a dramatic change our lives. For many with Huntington’s disease (HD), the pandemic has caused increased social isolation and decline in health due to limitations in physical and social activities caused by the pandemic. As we approach the holidays many of us are eager to see friends and family and return to “normal life”. The COVID vaccine is one strategy to help us get there.

### Update on COVID-19 pandemic

Let’s start with an update on what we know about the coronavirus disease (COVID-19). This virus is transmitted by small invisible droplets in the air from an infected individual’s nose and mouth. By wearing a mask, the infected person, who is often not experiencing any symptoms, can reduce the spread of the virus to others and those who are unaffected can reduce the risk of becoming infected. Wearing masks and social distancing are important strategies to reduce the spread of COVID-19 (Figure1).

While most individuals infected with COVID-19 will only experience mild to moderate symptoms, some will experience a severe respiratory illness that requires hospitalization. Severe COVID-19 illness can affect people of any age, but the risk increases with older age. Though the risk of severe COVID-19 illness has not been specifically linked with HD, the presence of chronic illness has been associated with more severe COVID-19 illness. Healthcare professionals recommend that HD patients get vaccinated and receive the booster shot in the appropriate timeframe to reduce the risk of severe illness.

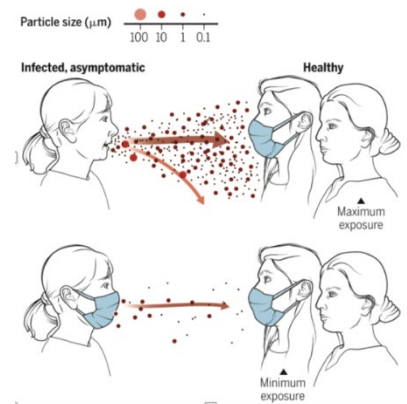


Figure 1: How COVID19 is spread

### How Does the COVID-19 Vaccine Work?

## How a Virus Infects the Body

### Enters

The virus enters the body through the nose, mouth, eyes or breaks in the skin.

### Infects

Once inside the body, a virus infects a host cell by injecting its genetic material.

### Directs

The virus will hijack the cell in order to make many copies of itself and to hide from the immune system.

### Spreads

The host cell dies as the virus multiplies and moves into more cells. This is when the body gets sick.

### Stops

The immune system can protect the body from low-level attacks and adapt to viral infections, so that they won't cause disease again or are less severe. Antiviral medications also can help protect against viral diseases.

To understand how vaccines work, it is important to review what causes infections and how your body fights against them.

Infections occur when a germ, such as a virus or bacteria, invades the body and attacks it from inside while multiplying. From the first time the body encounters a germ, the immune system (the body’s defense) starts building germ-fighting tools to fight the infection. There is delay from when the germ invades the body to when the immune system has enough resources to fighting the infection. Many symptoms associated with the infection such as cough, cold and fever are the result of the immune system “fighting” the invading germs and stopping the infection. During the early part of an infection, people can start spreading these germs before any symptoms appear. Once the body has fought the infection it will keep a “memory” of that germ. This is called immunity.

Vaccines help you develop immunity by imitating the infection. This is done by exposing you to a weakened version of the virus or specific parts of the virus that do not cause the illness. These vaccines help the body build immunity without causing the illness or spreading it to others. Sometimes while the body’s immune system is building its defenses you might feel mild symptoms like fevers, body aches and tiredness for a few days, however it is extremely rare for a vaccine to cause the infection or other illnesses. Over time immunity can fade, therefore, booster shots are recommended to increase the body’s immunity against COVID-19 over time.

**njhealth.org**  
 1.877.CALLNJH (1.877.225.5654)  
© 2020 National Jewish Health  
 7017 12508 MKT 6/20

**National Jewish Health**  
 Breathing Science is Life.

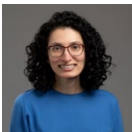
There are 3 types of vaccines available in the United States (Table 1). Depending on where you live, and your medical history one vaccine may be better suited for you than another. It is important to speak with your physician to determine which type of vaccine is right for you. All 3 COVID-19 vaccines are safe for HD patients and we encourage you to sign up for the vaccine and/or your booster shot in the recommended time intervals.

### What about the variants of COVID-19?

All germs mutate or change over time so that they continue to cause infections, mutating allows them to bypass the body's immune system. This is why people who have been vaccinated may get infected with COVID-19 again. Since the beginning of the pandemic there have been several mutations of COVID-19. However, there is strong evidence to suggest that those people who have been vaccinated (or have recovered from COVID-19 infection) are less likely to experience the serious illness associated with COVID-19 from the newer variants. By receiving the vaccine and building immunity to COVID-19, you can lower your risk of experiencing severe symptoms and dying from complications of the COVID-19 virus.

What are the differences?	NUMBER OF SHOTS	HOW IT'S GIVEN	HOW WELL THE VACCINE WORKS	SIDE EFFECTS
<b>Pfizer-BioNTech</b> mRNA Vaccine (Two-dose Vaccine)	<b>2</b> Two shots, 21 days apart	Shot in the muscle of the upper arm	<b>95% effective</b> at preventing laboratory-confirmed COVID-19 illness in people without evidence of previous infection.	Side effects are normal and may start within a day or two of getting the vaccine.  Throughout your body, you may experience: - Tiredness - Headache - Muscle pain - Chills - Fever - Nausea
<b>Moderna</b> mRNA Vaccine (Two-dose Vaccine)	<b>2</b> Two shots, 28 days apart	Shot in the muscle of the upper arm	<b>94.1% effective</b> at preventing laboratory-confirmed COVID-19 illness in people without evidence of previous infection.	In the arm you got the shot, you may feel: - Pain - Redness - Swelling - Rash
<b>Johnson &amp; Johnson / Janssen</b> Viral Vector Vaccine (One-dose Vaccine)	<b>1</b> One shot total	Shot in the muscle of the upper arm	<b>72% effective</b> in U.S. trials at preventing symptomatic COVID-19 after one dose. <small>(The vaccine had high effectiveness preventing hospitalization and death in people who did get sick.)</small>	

Table 1: Types of COVID-19 vaccines available in the US



Article written by Dr. Neepa Patel of The Rush HD Program.

patientslikeme® | Huntington's Disease Society of America

*"Partnering with PatientsLikeMe is an exciting new step in our work to help families affected by Huntington's disease come together to support one another, find vital resources and share their journeys so that care for HD improves as quickly as possible."*

- Louise Vetter  
President & CEO, Huntington's Disease Society of America

[www.HDSA.org/PLM](http://www.HDSA.org/PLM)

Join the **#HuntingtonsDisease** community on PatientsLikeMe to support patients and families grappling with HD and empower them to make their voices heard, learn from others who have faced the same experiences as them and accelerate development of treatments and cures through participation in research.

**LEARN MORE AT** <https://www.patientslikeme.com/join/hdsa>

**#LetsTalkAboutHD #HDSAFamily**

# Memorials and Tributes

## In Memory of Ina Mae Lindgren from:

Kimberly L. Crandall, Reginald R. & Kathleen S. Smidt, Leah Bohlmann, Terry G. & Clara Jo Ogden, Daniel B. & Doris F. King, Kenneth E. & Joyce L. Ropp, James W. & Mabel Gene Davis, James W. & Marsha Sue Ackerman, Marie L. Monk, Becky Shoemaker, Timm & Deann Geiger, Dean & Elizabeth Schafer, Carol & Kevin P Elliot, Kent B. & Brenda K. Geiger, Elsie Mae Schafer & David Schafer & Ann Jensen, David & Marsha Hubert, S W & J G Largent, Russell & Dawn Haigh, Doug & Denise M. Litwiller, Roger Abrahamson, Debbie M. & Timothy E. Cyr, Larry L. & Judith A. Hanson, Thomas G. & Lana K. Frake, Keith L. & Ann T. Jensen, Pamela L. & Lynn D. Wilken, Kimberly N, & Douglas C. Serrurier, Randy & Libby Wilken, JD & Joan M. Cashdollar, Danny L. and Linda M. Grace

## In Memory of Beverly Bentz from:

Erin M. & Sean M. Riley

## In Tribute to Emily Zivin from:

Rebecca Janowitz

## News from



### HDSA Center of Excellence at Northwestern Medicine Virtual Patient and Family Education Series 2022

#### Saturday, February 12th at 10 am via zoom

##### Panel Discussion: Difficult Conversations

Please come join us for a conversation around difficult conversations that include driving, dating, talking to children, social media and much more.

Register in advance for this meeting:

<https://northwestern.zoom.us/join/join?meeting=13droqXYLZeAlhy7I3jW>

\*General HD support group to follow education session

#### Saturday, April 9th at 10 am via zoom

##### Asymptomatic Gene-Positive

Please join Seth Rotberg as he shares his HD journey. This will be an interactive session for individuals who are asymptomatic gene-positive.

Register in advance for the meeting:

<https://northwestern.zoom.us/join/join?meeting=HH455iUoH>

\*General HD support group to follow education session

#### Saturday, August 13th (via zoom or in-person TBD)

##### Couples Retreat

Please join Emily Zivin and she provides an interactive education session for couples to talk about their HD journey together with other couples in the community

Register in advance for this meeting:

<https://northwestern.zoom.us/join/join?meeting=gsqDkuH9fUN4UUJ4jAHjLI7FHw0q8w>

\*General HD support group after education session

#### Saturday November 12th at 10 am via zoom (Date subject to change)

##### Clinical Research Update

Dr. Danny Bega

Register in advance for this meeting:

<https://northwestern.zoom.us/join/join?meeting=iwdeFGriQxTkcU6d47wy>

\*Caregiver support group to follow education session

## CHAPTER OFFICERS

**PRESIDENT – Arvind Sreedharan**

703-599-6000 ~ avs2004@comcast.net

**VICE PRESIDENT – OPEN**

**TREASURER – Andy Hucker**

224-715-0729 ~ ahucker1549@comcast.net

**SECRETARY – Jenny Placek**

630-995-6440 ~ jenniferplacek09@gmail.com

## BOARD MEMBERS

**Karen Bennett – karben22@hotmail.com**

847-212-1240

**Mary Bos - mary\_bos@att.net**

630-830-5329

**Sarah Cozad – cozinn.sc@gmail.com**

309-299-0284

**Wayne Galasek – wgalasek@aol.com**

708-289-1273

**Charlotte Rybarczyk –**

charlotte82963@gmail.com

847-259-3593

**Ann Terry – ann\_terry5@yahoo.com**

312-339-9356

## OTHER CHAPTER MEMBERS

**Emily Zivin – Social Worker**

630-443-9876 ezivin@hdsa.org

**Camille Colletti – Regional Director**

847-849-0680 ccolletti@hdsa.org

**Maryann Moynihan – Newsletter Editor**

708-955-3080 shamrock1959@gmail.com

### *Hopes & Dreams*

is the official publication of the  
Illinois Chapter of Huntington's Disease Society of  
America, Inc.,

P.O. Box 1454, Lake Villa, IL 60046

(630) 443-9876 ~ www.hdsa.org/il

This newsletter attempts to report items of interest relating to the individuals with Huntington's Disease, their families, healthcare professionals, and interested friends and supporters. HDSA and the Illinois

Chapter do not provide medical advice, nor do they promote, endorse or recommend any product, therapy or institution. Please check all drugs, treatments, therapies and products with your physician.

Statements and opinions expressed in articles are not necessarily those of HDSA, Inc. and the Illinois Chapter.



# MEDICAL RESEARCH CORNER

\*\*Study recruitment is impacted by Covid-19  
and will resume as soon as possible.



## **NORTHWESTERN MEDICINE HDSA CENTER OF EXCELLENCE**

### **Hi-DEF Scale Study: NOW RECRUITING**

Individuals with Huntington's disease are invited to participate in the Hi-DEF Scale Study. The purpose of this study is to learn more about impact of Huntington's disease on cognition and everyday functioning. The study involves a one-time commitment that lasts about 2.5-3 hours. Participants will be asked to complete some online questionnaires and two online cognitive tests. Once finished, the participant will be compensated for their time. If you're interested in learning more about the study or how to get involved, please contact Destiny Gomez at 312-503-2778 or [destiny.gomez@northwestern.edu](mailto:destiny.gomez@northwestern.edu).

### **Kinect-HD Study for Chorea**

Northwestern Medicine is recruiting for a study of a treatment for chorea associated with Huntington's disease. The study is of a medication called Valbenazine to treat chorea and is being conducted by the Huntington Study Group and Neurocrine Biosciences. The study involves 9 visits and will last 18 weeks. There is the opportunity to stay on the drug after the first part of the study is over. Participants will be randomly selected to receive the drug or placebo at first. If you or someone you know is interested in taking part in KINECT-HD, please contact our study coordinator ZsaZsa Brown at 312-503-4121 or email [zsazsa.brown@northwestern.edu](mailto:zsazsa.brown@northwestern.edu).

### **Kinect-HD 2 Study**

Northwestern Medicine is recruiting for an open-label extension study of Kinect-HD. The purpose of this study is to continue to gather safety and efficacy data on Valbenazine for the treatment of Huntington's chorea, while also providing study subjects who participated in Kinect-HD continued access to the study drug. In this open label study, all subjects are given Valbenazine, even if they received placebo during Kinect-HD. Kinect-HD 2 is open to research subjects who completed participation in Kinect-HD. For more information on Kinect-HD 2 contact ZsaZsa Brown at 312-503-4121 or [zsazsabrown@northwestern.edu](mailto:zsazsabrown@northwestern.edu)

### **PROOF-HD Study**

Northwestern is recruiting for the PROOF-HD Study. This is a phase 3, randomized, placebo-controlled study evaluating the efficacy and safety of an oral drug called Pridopidine in patients with early-stage Huntington's disease. The objective is to see if Pridopidine can slow down functional decline in Huntington's disease when compared to a placebo pill. If you are interested in learning more about the study and how to get involved, please reach out to study coordinator ZsaZsa Brown at 312-503-4121.

### **Telemedicine for Huntington's Clinical Care**

Individuals with Huntington's disease are invited to participate in the study "TeleHD" to determine the feasibility and value of telemedicine visits for HD patients and their care partners. This research study is conducted by Dr. Danielle Larson and Dr. Danny Bega. Please e-mail research study assistant Robert Modiest at [robert.jr3@northwestern.edu](mailto:robert.jr3@northwestern.edu) or call 312-503-5645 to let him know your interest, or if you have any questions.

## **HDSA CENTER OF EXCELLENCE AT RUSH UNIVERSITY**

### **Uniqure, a gene therapy study for Huntington's disease**

Rush University Medical Center is excited to be participating in a new gene therapy trial for Huntington's disease, sponsored by Uniqure. The therapy is called AMT-130 and will hopefully slow the progression of HD by lowering the level of huntingtin protein in the brain. "Gene therapy" works by targeting genetic abnormalities that contribute to us getting sick. Administration of the therapy involves a small incision in the skull through which AMT-130 is delivered to the brain. Researchers are looking for people aged 25 to 65, with at least 40 CAG repeats in their huntingtin gene, and specific brain structure that will be assessed by MRI. Eligible participants will be randomized to receive the real treatment or a "sham" surgery involving a small mark made on the skin without making an actual incision. Study duration is approximately 5 years, during which time participants will complete physical assessments, treatment dosing, lumbar punctures, blood draws, and MRIs. Assessments and treatment will be completed across multiple sites. If you or someone you know would like to take part in the Uniqure

study, please reach out to Jacob Hawkins at 312-563-5563, or email [Jacob\\_Hawkins@rush.edu](mailto:Jacob_Hawkins@rush.edu). We anticipate being ready to enroll patients in the next few months.

### **KINECT-HD, a phase three drug trial of Valbenazine for Huntington's chorea**

Rush University Medical Center is recruiting participants for a clinical trial evaluating a drug called Valbenazine for the treatment of chorea. Valbenazine is already an FDA approved medication for another type of movement disorder that causes involuntary movements called tardive dyskinesia. The study is sponsored by the Huntington Study Group and Neurocrine Bioscience. Researchers are looking for people aged 18 to 75 with motor manifest Huntington's disease to be randomized to receive Valbenazine or placebo for 18 weeks. Participants will come to Rush for 9 research visits to take surveys, complete physical exams, and have their blood drawn. If you or someone you know would like to take part in KINECT-HD, please contact Jacob Hawkins at 312-563-5563 or email [Jacob\\_Hawkins@rush.edu](mailto:Jacob_Hawkins@rush.edu).

### **KINECT-HD 2, an open label rollover study for continuing Valbenazine administration for the treatment of chorea associated with Huntington disease**

Rush University Medical Center is excited to participate in an open label extension study of Kinect-HD. The purpose of this "rollover" study is to continue to gather safety and efficacy data on Valbenazine for the treatment of Huntington's chorea, while also providing study subjects who participated in Kinect-HD continued access to the study drug. In this open label study, all subjects are given Valbenazine, even if they received placebo during Kinect-HD. Kinect-HD 2 is open to research subjects who completed participation in Kinect-HD up to their week 14 visit and subjects whose study participation was interrupted due to the Covid-19 pandemic. For more information on Kinect-HD 2, please contact Jacob Hawkins at 312-563-5563 or email [Jacob\\_Hawkins@rush.edu](mailto:Jacob_Hawkins@rush.edu).

### **ENROLL-HD, a prospective registry study in a global Huntington's disease cohort**

Researchers at Rush University Medical Center are looking for patients affected by Huntington's disease and their first-degree blood relatives to take part in an ongoing observational study. The data gathered in ENROLL-HD will be used to help doctors and scientists learn more about Huntington's disease and hopefully develop new treatments. Participation involves an annual visit conducted in the Rush Section of Movement Disorders at Rush University, where participants will complete surveys, cognitive tasks, family histories, and a blood draw. in ENROLL-HD, please contact Jacob Hawkins at 312-563-5563 or email [Jacob\\_Hawkins@rush.edu](mailto:Jacob_Hawkins@rush.edu).

### **Cortical Control of Balance and Walking in HD**

A neuroimaging study investigating brain activation during balance and walking under single-task and multitask conditions in people with Huntington's disease. We are looking for individuals with a clinical diagnosis of HD, 30 years of age and older, who can stand and walk unassisted. Participation requires one, 3.5-hour visit to *Rush* University Medical Center. This study is actively recruiting both healthy control and HD participants. Please contact Nicollette Purcell ([Nicollette.L.Purcell@rush.edu](mailto:Nicollette.L.Purcell@rush.edu)) if you are interested in participating and would like additional information.

### **Optimization of Telegenetic Counseling for Huntington's Disease**

Rush University Medical Center will be offering telegenetic counseling services to HD patients and families as part of a new study that aims to assess feasibility and patient satisfaction of a telegenetic counseling program. Lack of access to genetic counseling has been recognized as a critical gap in care for many HD patients and their family members. 35 symptomatic or pre-symptomatic participants aged 18 or older will be recruited for this study and randomly assigned to a group that receives in-person genetic counseling first, followed by telegenetic counseling, or a group that receives telegenetic counseling first, followed by in-person genetic counseling. In-person visits will occur at Rush's HD Center of Excellence, while telegenetic counseling visits will occur via a video platform provided by Rush. Participants will then be administered a post-visit survey with questions regarding content of counseling, format of delivery, and their preferences. During the COVID pandemic, we will be doing telegenetic counseling exclusively. If you or someone you know would like to take part in this telegenetic counseling study, please contact Marc Rosenbaum at 312-563-0665, or email [Marc\\_Rosenbaum@rush.edu](mailto:Marc_Rosenbaum@rush.edu)

## **UPCOMING HDSA RESEARCH WEBINAR**

**THURSDAY, JANUARY 20, 2022 AT 3PM (ET)**  
COMMUNITY PRESENTATION OF NEW ROCHE TOMINERSEN DATA

Please join us for an update from Roche with new key findings from the GENERATION-HD1 study on Thursday, January 20, 2022 at 3:00p EST. This 90-minute community-focused webinar will be an opportunity to hear members of the Roche/Genentech team discuss new data from the study regarding tominersen, ask questions and participate in a conversation with Roche & HDSA leadership. The Zoom meeting will be hosted by HDSA & registration is required at [HDSA.org/researchwebinars](https://hdsa.org/researchwebinars).



## News from Our Illinois Chapter Social Worker Emily Zivin, LCSW

Huntington's Disease Society of America  
Tel: 630-443-9876 or E-mail: ezivin@hdsa.org

Memory loss/dementia are words that can bring up many feelings and questions. Communication is very important when talking to a family member who is experiencing memory loss. Ways to help with communication include the following:

**Speak Slowly:** As Huntington's Disease progresses, your loved one may speak less, but they still understand what you are saying. Remember to speak in clear and concise sentences. Wait for them to respond.

**Prepare Yourself:** Be clear on the goals of the conversation. Do you want to feel connected? Make sure you have the time and energy to communicate with your loved one.

**Environment:** Calm and quiet environments tend to be the best space for conversations. Find a surrounding that helps your loved one function best. Remove extra stimuli.

**Preserve Dignity:** Help your loved one living with Huntington's Disease feel loved and empowered during conversations. Give them the chance to feel like the expert and respond in ways you know will let them feel happy.

**Multi-Sensory Approach.** A gentile touch on the arm or using a favorite scent can help someone who otherwise has difficulty communicating.

Be able to change course. If communication is not successful, try a different approach or try again later.



**Stay tuned for more information.**



## “Seeing” the toxic huntingtin protein in people with HD

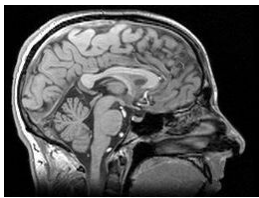
New tools let us “see” clumps of toxic huntingtin protein which build up in the brains of people with Huntington’s disease over time. Tracking these clumps might help us to better understand how HD progresses or how treatments might slow or halt HD.

By Dr. Rachel Harding December 08, 2021 ~ Edited by Dr. Jeff Carroll Originally published on December 07, 2021

Scientists have developed a tool which allows us to “see” the toxic clumps of the huntingtin protein using special scanners. People with Huntington’s disease (HD) make a toxic form of the huntingtin protein which forms clumps in cells of their bodies, which accumulate during HD progression. Tracking how these clumps form over time in people with HD, or how they change when people with HD take different treatments, could help us better understand the progression of HD and which medicines help patients most.

### What are these protein clumps?

We all have 2 copies of the huntingtin gene but for people who have Huntington’s disease (HD), one of their copies has a type of mutation called a repeat expansion. This mutation occurs in a repetitive bit of the huntingtin gene DNA code which has the letters “C”, “A” and “G” repeating over and over. If you don’t have HD, you will have less than ~35 CAG repeats in your huntingtin gene but for people with HD, the mutation means they will have more than 35 CAG repeats in one of their huntingtin genes.



Huntingtin protein molecules with too many glutamines can’t assemble properly so can form toxic clumps which have been shown to build up in patient’s brains over time. However, these clumps are not visible in most types of brain scans like MRIs.

The huntingtin gene is the recipe our cells use to make the huntingtin protein so if the DNA code of this recipe is changed, the protein which our bodies make will also be changed. Proteins are made from long strings of chemicals called amino acids, following the instructions laid down in our DNA. The DNA letters “CAG” are the recipe notes for the amino acid glutamine. This means that if the CAG repeat number gets bigger, the huntingtin protein will have many more repeating glutamines. Huntingtin protein molecules with too many glutamines can’t assemble properly so can form toxic clumps. We have known about these clumps for a long time now and they can be seen in the brains of people with HD when we look under a microscope. However, tracking these clumps in living patients has been challenging and most of our knowledge of them comes from looking at them in post-mortem brain samples from animal models of HD or patients who have graciously donated their brains to research.

### Why do we want to look at these pesky clumps?

Scientists from many labs across the UK, Germany, Italy, Sweden, and the US have developed molecular tools which now allow us to “see” these clumps in living animals, and hopefully soon, HD patients. These tools bind to the huntingtin protein clumps and have chemical decorations, called radiolabels, which mean that they light up when looked at by PET (positron emission tomography) scan.

This type of molecular tools are known as PET tracers and are used in lots of different medical and diagnostic settings to allow doctors and researchers to image specific parts of your body. Different types of tracers can be swallowed, injected, or inhaled depending on what part of your body is being looked at. Once the patient has the PET tracer in their body, they will be scanned, and the part of the body being looked at will light up if the target of the PET tracer is there because the tracer is slightly radioactive. Similar tools have been developed for studying other diseases such as Pittsburg compound B which is used to look at similar clumps in people with Alzheimer’s.

Making PET tracers which allow researchers to see the toxic clumps of huntingtin protein is an attractive idea for several reasons. Firstly, a PET scan can be performed on patients at multiple time points throughout their life so we can track how the clumps accumulate over time throughout the progression of HD. Many of our current methods for looking at huntingtin clumps in patient’s brain can only currently be done at the very end of the disease on post-mortem tissue.

Secondly, PET scans are non-invasive procedures and allow us to look right in the brain whereas more intrusive procedures like measuring huntingtin protein in spinal fluid provide only a proxy for what we think is happening in the brain. Thirdly, the clumps are formed from the toxic form of the huntingtin protein so PET scans will allow researchers to specifically measure changes to this specific form of mutant huntingtin. This differs from most of ways we measure and analyze huntingtin in spinal fluid or blood which measure all the different forms of huntingtin, including the healthy huntingtin protein.

### Development of the first huntingtin PET ligand

Last August, an early version of these tools was published called CHDI-180R – the first time a PET tracer has been made for the huntingtin protein! A team led by Celia Dominguez at CHDI Foundation showed that the tool molecule CHDI-180R was able to bind very tightly to

the toxic huntingtin protein clumps in a test tube. They also used CHDI-180R to show where the clumps of toxic huntingtin were in brain samples from HD mouse models.

In the brains of mice with the HD mutation, clumps of the toxic huntingtin protein could be seen in many different brain regions which are known to be affected by HD, whereas in mice without the HD mutation, these clumps could not be seen, even though they had also been injected with the tool molecule CHDI-180R. Finally, the scientists showed that CHDI-180R spread well through the brain and was also safe with no side effects in both monkeys and rats.



For mice without HD, no brain regions light up, even as they get older, whereas for HD mice, the scientists were able to track the build-up of clumps in the HD mouse brains as they aged using this tool as more and more of the brain lights up over time.

#### Fine-tuning the tools

PET tracer development often takes multiple attempts before an optimal tool is found so the same international group of scientists is also developing other versions of this tracer to have lots of back up options. These (hopefully) new and improved versions of the molecular tool are being tested to work out how they spread in the brains of animals tested.

Other diseases like Alzheimer's also have protein clumps which build up in nerve cells, but these are made up of other potentially toxic proteins, like amyloid beta. The scientists are also checking how specific these tools are for the huntingtin protein clumps which accumulate over time in HD patients compared to other disease protein clumps, like those from Alzheimer's patients. So far, the results have been very encouraging, so the scientists are now keen to start testing the tracers in people.

#### So, what's next?

A clinical trial is being conducted called iMagemHTT study, which will investigate the huntingtin tracer in people. The trial will use PET/MRI imaging to understand how the PET ligand tracks huntingtin in the brain. We previously reported on some encouraging preliminary data from the [Phase I study of this tracer](#) at the CHDI virtual meeting earlier this year. So far, the findings are encouraging, so they are continuing to add participants to the study.

The amount of huntingtin clumps in the brains of people with HD is a good biomarker of disease progression. Biomarkers are objective measurements scientists and clinicians can take to track HD's progression which can be important for working out the best treatment options, as well as if treatments are working properly. It is possible that HD patients in the future might be monitored by PET scan using these types of tools.

If the PET ligands work as scientists hope, it could also be used to track huntingtin-lowering in the brain in future trials. Despite some setbacks, huntingtin lowering is still a promising strategy for treating HD which is being pursued by Novartis, PTC Therapeutics, Wave and Unique, all of whom have clinical trials underway. Regardless of what happens with Huntingtin lowering, these exciting new tools are giving scientists the ability – for the first time ever – to track mutant Huntingtin protein across the entire brain of living HD patients, which is a huge advance.

We look forward to updating you more on this topic soon!

# SUPPORT groups

We invite all those diagnosed with Huntington's Disease, their families, caregivers, and individuals who are at risk to attend our Support Group meetings. Meetings provide a supportive environment where participants can share concerns, challenges, and successes. In addition, participants can lend emotional support to one another and lessen feelings of isolation. Meetings are always free to attend, and all locations are accessible. Your involvement is important for our support groups! At a meeting you might learn about a community resource, discover a new research study, or hear from a guest speaker. Please consider joining us! For further information about any of the support groups, please contact Emily Zivin at 630.443.9876 or email at [ezivin@hdsa.org](mailto:ezivin@hdsa.org).

Cancellations may occur in the case of inclement weather. We will attempt to notify everyone with advanced notice by email. If you are concerned that a meeting may be cancelled, please contact Emily Zivin at 630.443.9876 to confirm.

## **Illinois HDSA Chapter Virtual Support Group** **3rd Tuesday of Every Month (7:00pm)**

Register in advance for this meeting:

<https://hdsa-org.zoom.us/join/joinmeeting/register/tJEufumvpjosHtGoWGckn3GCqwnTf8Ftn6>

Questions? Contact Charlotte Rybarczyk at  
[charlotte82963@gmail.com](mailto:charlotte82963@gmail.com)

## **MUNSTER, IN**

**2nd Tuesday of Even Months (7:00 – 8:30pm)**

**2021 Meetings: Contact Cindy Rogers for specific dates/format**

Southside Christian Church, 1000 Broadmoor Avenue  
Contact: Cindy Rogers (219-836-2369); [clrogers111@comcast.net](mailto:clrogers111@comcast.net)

## **Northwestern Medicine Virtual Support Group**

**2<sup>nd</sup> Saturday of Every Month (10:00 – 11:30am)**

Odd months - Caregiver Support Group

Even months - General HD Support Group

Register in advance for this meeting:

<https://northwestern.zoom.us/join/joinmeeting/register/tJlqf-2vqT8qE9KddIKdiOkVyTijeHFvyhYL>

Questions? Contact Emily Zivin at [emily.zivin@northwestern.edu](mailto:emily.zivin@northwestern.edu)

## **Rush University Medical Center Group**

**4<sup>th</sup> Saturday of Every Month (10:30am – Noon)**

**For more information and Zoom details please reach out to the following support group leader:**

Sarah Strait, RN (312-563-2900); [sarah\\_strait@rush.edu](mailto:sarah_strait@rush.edu)

## **Illinois HDSA Chapter Caregiver Support Group**

**Feb/April/June/August/October/December**

**Upcoming Meeting: February 23<sup>rd</sup> (7:00pm)**

Winnetka Library, Community Room, lower level  
768 Oak Street, Winnetka

Questions? Contact Emily Zivin at [emily.zivin@northwestern.edu](mailto:emily.zivin@northwestern.edu)

**Meeting Guidelines** - We read the guidelines before each meeting to remind us that we are all responsible for following and committing to the group standards, which are in place to keep this group a safe place to share.

**Share the airtime** - Everyone who wishes to share has an opportunity to do so. No one person should monopolize the group time.

**One person speaks at a time** - Each person should be allowed to speak free from interruptions and side conversations.

**What is said here stays here** - This is the essential principle of confidentiality and MUST be respected by all participants.

**Differences of opinion are OK** - We are ALL entitled to our own point of view.

**We are all equal** - We accept cultural, linguistic, social, and racial differences and promote their acceptance.

**Use "I" language** - It's important to use "I" language because you are talking about yourself and not a vague person or group of people.

**The use of "I" helps avoid someone feeling like they are being attacked** - Examples include: "I feel like you handled that difficult situation the best that you could have" "I had good experiences with antidepressant meds in my family"

**It's OK not to share** - People do not have to share if they do not wish to.

**It's everyone's responsibility to make the group a safe place to share** We respect confidentiality, treat each other with respect and kindness, and show compassion.

 SAVE THIS DATE!

May 15 <sup>th</sup>	HDSA IL Chapter Team Hope Walk – Naperville, IL
August 6 <sup>th</sup>	HDSA IL Chapter Baggo Tournament
September 10 <sup>th</sup>	HDSA IL Chapter Team Hope Walk – Central Illinois

**<https://hdsa.org/il>**



**JANUARY 2022**