The Devil's Disease: A Story of its Impact on Four Generations:

Testimony of Kala Booth
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Good morning Chairman Pallone, Republican Leader Rodgers, Subcommittee Chairwoman Eshoo, Republican Subcommittee Leader Guthrie and Members of the Committee. Thank you for the opportunity to share my family's story, my story and the story of so many HD families.

My name is Kala Booth

I am 34
I am the second generation of my family to be diagnosed with Huntington's Disease, and 4th known suspected generation with the disease
I am a patient
I am a caregiver
and more importantly I am a voice.

Today, I am a voice for the many HD patients who do not have one.

Over the years, I have taught myself to emotionally disconnect. This allows me to be able to separate the Huntington's Disease from the person.

Huntington' Disease is a rare inherited disease that causes degeneration of the brain. It affects each patient differently.
The symptoms are different.
The progression time is different.
But what is not different? It is FATAL and there is NO CURE

While reports on the neuropathology of chorea in adults appeared as early as the 1870s, there was little progress in understanding the cause until the 1960s. From there, advances in genetic sequencing led to the identification of the huntingtin gene in 1993, which led to the ability of individuals to determine with certainty that they had Huntington's Disease.

Huntington's Disease is an autosomal dominant disease, meaning if one parent has the HD gene each of their children has a 50% chance of inheriting the gene. If a person has the HD gene, they will develop Huntington's Disease, if they do not, they will not develop HD or pass on the gene to their children.

Symptoms of Huntington's Disease typically develop in the 40s, the prime of a person's life – highest earning years, middle of raising families, planning for retirement many years off. However, HD disrupts all of that and physically, emotionally and financially drains families. While onset of HD and the progression of symptoms varies in each person, symptoms can include:

- Personality changes, mood swings & depression,
- Forgetfulness & impaired judgment,
- Unsteady gait & involuntary movements (chorea), and
- Slurred speech, difficulty in swallowing & significant weight loss.

Symptoms usually worsen over the course of 10 to 25 years and affect the ability to reason, walk, and talk. Ultimately, HD is fatal.

Most people have not heard of Huntington's Disease, and often even health care professional who have are not familiar with it and misdiagnose its symptoms early in the disease progression. This causes many challenges for families trying to help their loved ones.

Twice I have watched a broken system turn a devastating situation into an almost unbearable one. That is why I am here today. To share my family's story and to seek action from all of you – from all of Congress – to help HD families living across America.

- I urge Congress to pass H.R. 2050, the Huntington's Disease Parity Act THIS year. The HD Parity Act eliminates the six-month Social Security Disability Insurance (SSDI) and two-year Medicare wait periods.

  Even though Huntington's Disease is on the Social Security Administration's compassionate care list, HD families often spend years battling against an uninformed system that does not recognize or understand Huntington's Disease. Individuals often are denied coverage multiple times and forced to hire attorneys to help them navigate the system to secure coverage. Then, when they finally are deemed eligible, many are forced to wait another six months to receive SSDI cash payments and two years for Medicare coverage.

  During this time, patients often lose access to employer-sponsored insurance, cannot afford marketplace policies or their providers do not accept marketplace plans. Ultimately, they go without access to health care services that would blunt some of the symptoms experienced by HD patients improving their lives and helping their families care for them.

  This policy must be changed. Congress made an exception for patients with ALS and the same should be done for patients with HD.

- Expand the focus of the National Neurological Conditions Surveillance System (NNCSS) to include Huntington's Disease.

  The NNCSS is the ……. While the system collects data and tracks disease progression for patients with Parkinson's and Alzheimer's, patients with HD have not been included. This puts the entire HD Community – those with the disease and those family members at-risk – at a disadvantage and hinders research and development into treatments and cures.

  This policy must be changed immediately.

  Require the FDA and NIH to work with companies that are researching HD cures to design trials that recognize the uniqueness of Huntington's Disease. HD is a unique disease that has great variation in onset, occurring as an adolescent disease in some cases, but usually showing noticeable symptoms in patients in the mid-40s with actual symptoms varying widely from behavioral health deterioration, to physical ailments like loss of motor skills and involuntary

movements. Failure to recognize and account for these variations have hindered advancement of research and limited the success of trials.

The FDA must work more closely with the HD community and companies investing in possible new treatments and cures to ensure that these variations are accounted for and do not hinder research that can help patients.

My Family's HD Story:

As a genetic disease, Huntington's Disease likely has unknowingly impacted my family for many, many generations. However, because it was unknown for decades, then very difficult to diagnose and only identified with certainty through genetic testing since 1993, the true impact across my family will never be fully known.

We believe my great grandfather on my mother's side is the first family member suspected to have Huntington's Disease. My papaw was diagnosed in the 1996 and I was diagnosed in 2018. By sharing my story, I hope you will learn more about Huntington's Disease, how it has impacted thousands of families like mine and commit to helping those living with or at-risk of HD.

When I was growing up, I remember being mad and angry. I was angry at Papaw, or so I thought. My papaw was not a normal papaw. He would have violent outburst and uncontrollable behaviors. He was someone I remember locking myself in a room just so I didn't have to be around. I remember walking in seeing Mamaw black and blue, I remember at Christmas having to keep the landline phone beside us in case we needed to call 911. My good memories are outweighed by the bad, but the reality was the man I was mad at wasn't Papaw. It was Huntington's Disease that made him become the man I didn't recognize.

For my brother, that same Papaw was the greatest. They worked on cars and stayed up too late. They rode lawnmowers and go-carts. They could always be found eating sweets and playing Rook. All the normal things you would do with your papaw. That same Papaw was the man that stepped up as a father figure for my own dad. A man that went to Church and drove across country to visit us no matter where we were stationed.

So how could our versions of that same man be so different?

Huntington' Disease or, as I call it, the Devil's Disease. A disease I would not wish on my worst enemy. I have watched HD bring the Devil out of my loved ones. I have watched HD tear families apart. Huntington's Disease is described as having ALS, Parkinson's and Alzheimer's **TOGETHER.** Huntington's disease is a rare, inherited disease that causes the progressive breakdown of nerve cells in the brain.

HD is **FATAL** and there is **NO CURE.**

I didn't understand what was happening to Papaw. I thought it was just him, and at the time doctors didn't understand what was happening to him either. Papaw was born in 1944, and in his 30s he started showing signs of chorea, he started having facial twitches, he started showing mild signs of anger, but no one knew what was to come. By 50, he was in full progression of Huntington's Disease, and by 56 he quit working. In 2005, he entered a nursing home and by 2008 he passed away.

At one point in the late 90s, they thought he had a brain tumor, but when my family took Papaw to a clinic in Chicago they diagnosed him with Huntington's Disease. A new disease, a rare disease, a disease people knew nothing about. And when people don't understand something, they don't know how to help. I remember Papaw being sent to Central State Mental Hospital because of one of his outbursts, not because he was a criminal, but because the care facilities where he lived didn't know how to provide care. He was a danger to himself and the staff they would say.

I watched Mamaw mourn the loss of her husband before he was ever gone. I watched my mom battling court case after court case just trying to keep people from taking advantage of him. I remember seeing the guilt in my mom's eyes when she lost hope and signed him into a nursing home not in our area because that was the only facility in the state that would take him. They say Papaw went downhill pretty quickly when he was in the nursing home. I didn't visit, I was mad. I remember Mamaw canceling his life insurance policy because she couldn't afford to pay it only to have Papaw pass shortly after. I remember my mom trying to keep Mamaw from losing her house as the bills came in. Five years in court is what it took to finally settle his estate.

HD is a rare **INHERITED** gene that has a 50% chance of being passed on to a child. You would think with those odds and all the challenges we faced with Papaw we would have been prepared for what would happen next. But HD doesn't follow a pattern.

Third Generation: Optimist

My mom, Marsha, was born in 1963. By the time my mom was in her 30s we noticed mannerisms that looked just like Papaw. Twitches, tripping, etc. Dad and I would discuss it, but never to mom's face. When asked if she had HD her response was "there is a possibility." By 50, she was in full progression of HD and by 56 she was medically retired.

My mom is the third generation of Huntington's Disease in my family, but her HD has never been confirmed by a genetic test. She took a different approach. She lives each day to the fullest and like it is the last. She never wanted to be tested because there is nothing that can be done, there is no medicine, no cure. She said when it is time God has a plan.

There are no words to describe my mom. She is my rainbow and butterflies. My mom never said a bad word. If you looked in the dictionary you would see her picture by kindness. She had hope and faith. She had hope she didn't have the disease, she had hope there would be a cure or medicine by the time she needed it.

My mom's undiagnosed disease caused its own set of problems. When my mom had her first episode we took her to a local emergency room. They chalked it up to lingering effects of a previous concussion. They sent us home.

A few weeks went by, and the symptoms worsened. She went days without sleeping. We took her to another emergency room where they sedated her so she could rest. We were waiting for a room when the doctor came in and said they had found a room at Norton's and we could follow the ambulance there.

What we didn't know is the room they had was on the psychiatric floor due to her "psychiatric break."

30 days it took to get her home.
30 days we drove to Louisville to see her for 1 hour to be split between all of us.
30 days I watched my dad's heart break in pieces every time he had to leave her

They said she had a psychiatric break
They said she was schizophrenic
They told us it all could have been caused from an untreated UTI

They didn't realize it likely was the progression of undiagnosed Huntington's Disease.

Imagine someone who is already having paranoia to now be introduced to new doctors. If you think someone is after you, do you think doctors will be able to get you to take new pills? I joke the real reason mom never wanted to be tested was she is afraid of MRI machines on her best day. Now we are trying to convince her to agree to one on her worst day to get a diagnosis of "the Devil's Disease."

Even though my dad had power of attorney it did not work when we needed it to get my mom the help she needed. The hospital said my mom was lucid and was refusing treatment. We had an appointment with the judge, a snow storm came and the attorney couldn't make it back in town. The appointment got pushed out. It took three weeks for a judge's signature to sign off to override my mom's decision and deliver the medicine she needed.

We got her home and she slowly was weaned her off the medications that she was given in the hospital. Then, the episodes started again, except this time I wasn't going to let a broken system win. The family suggested taking her back to the emergency room, they suggested taking her to another behavioral facility. My dad sat heartbroken that his elementary school sweetheart would have to go back to a place he never wanted to see again. He felt he was failing her and I felt the system was failing us. I decided in that moment I wasn't waiting three weeks for a judge's signature. I did what I taught myself, I disconnected and forced her to take her medications.

Over the next few years her health slowly declined. She went from being a top real estate agent in the county to needing help calculating figures. She no longer is able to drive, cook, or clean. She has mentally declined to childlike mannerism. My dad and I make sure to always be near. She is losing control of her swallowing and chokes easily.

In November 2019, at the age of 56, we applied for Social Security Disability Income for my mom. It was declined. We appealed and it was declined again. We hired a lawyer and in March of 2021 a broken system finally acknowledged her disability status, and she received her SSDI and Medicare coverage.

Fortunately, our family has always had the financial capabilities to readjust and reallocate. For most HD families, they struggle, they lose at least half, if not all, of their income and health insurance at the same time and additional costs of care are adding more strain and stress.

2nd Diagnosed Generation: Love Your Story

January 2018 my journey started even before I knew My Story began…

I volunteered for a service trip to Jamaica through my employer, the Veterans United. With everything going on with mom, the psychiatric ward and having to force my mom to take medications, I had emotionally disconnected and needed something to spark my soul. Selfishly, I volunteered because I needed a mental reset. I had never been on a service trip and building two houses with strangers for people in need sounded like the perfect remedy.

Even though I was going with my company, I didn't know anybody because I work in a branch, not the corporate office. On the trip, each day you are serving you get assigned to different task, pouring a foundation, working at the store, building a house, and so much more.

On the trip, we were given the opportunity to serve at the Infirmary. The infirmary is a nursing facility for those with family that could not care for them or those who had no family at all. It was opened air, community rooms and no single living space.

There were sign-ups to go back for the second day and I wasn't going to sign up until I heard one of the other guys say "I can lift heavy, I can mix concrete, but that won't challenge me like going back to the infirmary." I thought if he can do it so can I. I signed up

When we got there the second day it hit a nerve, I got quiet and started shutting down. The same guy came over to check on me and I said this could be me one day. I explained to him about my family history, that my papaw died in a nursing home. Even though I have an amazing support system who wouldn't let me be alone, they couldn't be there 24/7. On the bus ride back all of the questions started.

What is Huntington's Disease?

What are HD symptoms?

Does you mom have it?

Do you have it?

Is there a cure?

Why haven't you been tested?

I explained that at a young age how traumatized I was by Papaw's behaviors, and as the years went on I suspected my mom had inherited the gene. This made me realize I had a 50% chance of having it. Something deep down in my heart already knew. I started over-analyzing every time I tripped, every time I jerked, every time I dropped something. The older I get the worse my anxiety, depression and OCD get.

Problem was not only was I living in fear, but I was also battling my mind, I couldn't control the obsession. I wanted to get tested, but my mom didn't want to know and there was no way for me to get tested without her learning she had HD (because I knew I was positive).

I wanted to know for my sanity, I wanted to know to get ahead of it. I wanted to know because I didn't want to have children if I had it. I know that to most people that sounds radical. Some people choose that route, some people have IVF so they know they won't pass on the gene. Some people don't want to know so they can have children without fear. Every path is so different, but so perfect. Everyone has a story and this is mine.

I came back to Kentucky and didn't think any more about it. A month or so went by and I got an instant message on my work computer from the same guy that said "do you have a minute." I said sure, I thought he was calling about the appreciation gifts that I ordered. He said you have been on my mind since the trip and I wanted to reach out to let you know if you want the money to get tested it is yours. The Foundation as already approved it. Our company has The Veterans United Foundation. Employees donate 1% of their pay to the Foundation for those in need. He said think about it and let me know. Well, it wasn't 5 minutes later and I had the tracking number in my email.

I tried to send the money back. There were people in NEED I was not one of them. People need electricity, wheelchairs, food. This was not a need, but they wouldn't take it back. So I called around and found a place that would do genetic testing in a sealed file. I didn't want insurance to know. I also decided that I was going to do this on my own without my mom knowing. I could keep the secret, carry the burden.

To get tested you must see a counselor, they must determine you have the mental capacity and emotional support to handle results. Cleared that. Next step was the genetic testing, which was just a blood draw. Then came the obstacle, they would not give me the results by myself. I tried to convince my dad to go and just not tell my mom. That didn't go over well. So I told mom and I gave her the option to come knowing we would get the answer I already knew in my heart.

Mom wanted to go because she didn't want me to go on my journey alone. But I don't think she fully understood what was about to happen. When we entered the room where they would go over my results, I made sure to explain again that if I found out I had HD then that meant she had it too.

The results: I HAVE HUNTINGTON'S DISEASE. My CAG score was a 44. Everyone has CAG in their DNA, but HD patients have a repeat tail. For me it was a relief. I could live and live without fear. Yet, for my

family and friends, it was devastating. My family was mourning for me and my mom. The shock was for both of us, not one.

Every time I walked in a room you could feel the emotions change. I asked everyone to take the week to feel their emotions but after that we were moving past it. I didn't want to be the elephant in the room. I didn't want to be the disease. I was going to be different.

I wanted to LIVE, really LIVE. I wanted to go on adventures and take every moment in. Time became the most precious gift. I believe whole heartedly that God had his hands on my story because everything was coming together perfectly. I had found a place that would tell my CAG score without seeing my brain. I was able to keep it off the records. I was finding unexplainable comfort.

At 31, I had a plan. I was going to live and go on grand adventures. I did what every normal 30 year old does, I made a death binder. A binder that had my CAG score results, that has my life insurance policies, that has copies of all my bank accounts, the mortgage to my house. It has instructions at which point I want a DNR. It's not normal for a 30 year to be detailing that if I am full blown HD DO NOT bring me back.

My dad had another plan. When we got my results they handed me a folder and inside was information for a clinical trial. He sent the nurse an email. She said she didn't think I would qualify. When I spoke with the nurse she asked questions about symptoms and what stage I was at. I told her everything was mild and unnoticeable to most.

She said unfortunately she thought I wouldn't qualify.

I got really upset. Here was a drug trial 45 minutes from me that could slow the progression down, but I wasn't advanced enough, which is awful because once the damage is done there is no reversing it.

I sent an email pleading my case. I explained I had never had an MRI or PET scan. I didn't know how advanced I was. She reached out to the clinical trial company. They ran a calculation based off my CAG score. They said if I was willing to go through all of the tests, they were willing to determine if I qualified for the trial.

So up to Louisville we drove. I did all the MRI, EKG, PET scan, blood work, etc. Then went home to wait by the phone.

You are waiting for the phone to ring and it felt like it was never going to happen. I wanted the call so badly. I wanted to be part of the trial, BUT what that phone call meant was I was more advanced than we realized. Once Huntington's Disease starts progressing the damage is done and there is no reversing it.

I got the phone call. I got in. Doing this trial meant I was really choosing to change my dreams. It meant putting others first. It meant putting all the people I could be helping, the families who are struggling and my family who could still be affected before my own wants.

What happened next was the HARDEST decision. I made a decision to not have my own children. Instead, I signed a 2-year contract to participate in the clinical trial. There was a 50% chance I would receive the medicine but there was a 50% chance I would receive placebo. In my mind, I was helping thousands of families that were dealing with this disease. So even if I received placebo, somebody had to, I was bringing hope where there wasn't. I was bringing hope to my family, past and future and in clinical trials. I had found peace. I found my purpose. God knew he had a bigger plan for me than I even realized.

I understand everyone has their own opinions on how they would handle my situation. I understand many do not agree with my decision to be tested. Or not to have kids, but this is why we should share our journey to bring light and knowledge. Clinical Trials are not for the faint of heart, but that is where I found my purpose and peace. Yet, it was a double edge sword.

Being in a clinical trial was nothing short of a roller coaster ride. Physically I was exhausted, mentally I was exhausted. It's weird and overwhelming, it's like this clinical trial became a part of me and when it ended I lost a piece of me.

It's constantly praying you are on the meds, but knowing someone needs to on placebo and knowing someone needs the drug more.

It is fear, fear you're on the meds and fear the trial ends without new medication.

It's hopefulness that the drug comes to market for a disease that has no cure, but can slow down time, not just for me but for my mom.

It's isolation

Over the course of my clinical trial, I became known for my shirts. Every appointment I wore a different shirt. The shirts could express feelings I couldn't and thankfully the Blue Rose in town had plenty of options. They were my security blanket. Others called them my superhero cape.

My "Huntington's Disease Warrior" shirt always raises questions. I also have one that says "I love someone with HD (me and my mom)."

I went through the trial only hearing good things about the medicine, the FDA fast tracked it, and people were buying stock in the drug. Alzheimer's gave a grant for the medicine. I never even considered the drug not being approved. I was prepared to hear you were on placebo. What I wasn't prepared for was hearing through the news of the Vaccinex stock crashing because there wasn't data to support the FDA approval.

It wrecked my world. I fell to the floor at work for hours because I couldn't catch my breathe. Hope was yanked out from under me. The emotions that I felt that day. I was angry for mom. I was angry for my future. I was angry for the people I was letting down. I was angry because I was angry.

I was holding on by a single thread.

Mom had officially retired (due to the progression of her HD).

My portion of the trial had come to an end and that was awful. It's hard to explain, but the trial was like a protector, when I was in it, I was helping myself, my family, others. Now I was supposed to sit back and wait. Or should I try to get in another trial. It's like rolling the dice and I don't like gambling.

So what was next? You would think with that ending I would not want to do another clinical trial, but here I am waiting. Waiting for any opportunity I can jump on.

Finally, another clinical trial was announced. However, I DID NOT qualify. It didn't even take them 30 minutes to decide that I was not eligible. On one hand, this means I have not progressed enough to qualify for that trial, so THAT's the positive, but here I was again with the hope taken away. I understand they have to be able to quantify the data for the FDA, which means they need your disease to be advanced. But it is frustrating how hard it is to quantify/qualify the data, because HD symptoms are so different and I feel like it is affecting my behavioral/mental health more than my physical decline.

After that news, my mom and I did our part to always submit our yearly data collection for Enroll HD. Enroll is such an amazing database that allows researchers to be able to access HD patients' data around the world. This year, during the Enroll evaluation, they realized my mom's HD is advanced enough to qualify for the new trial so we are starting another journey, she is starting her first clinical trial.

I also participate on the national HD Advisory board because of my shirts and raising awareness. Helping our Kentucky Chapter of HDSA host a Team Hope walk every year. Hoping to help with Halloween for HD, an awareness event in Louisville. I am looking for clinical trials and making sure to do my yearly Enroll-HD evaluation providing doctors and researchers all over the world access to a database of information.

During the trial, I was posting to a private group page to give friends and family updates. Then, last May with it being HD Awareness month, I decided I was going to make my post public. I wanted to update people and I also wanted to educate people. There shouldn't be a stigma for uncontrollable disease. There shouldn't be a black cloud because someone has HD, or Schizophrenia or any other disease that people feel ashamed to talk about. What we should be doing is having conversations and create places to serve those with compassion and dignity. You can't bring light if you stay in the darkness.

I will fight for my family, I will fight for myself, I will fight for those that can't, but I need and the entire HD community needs Congress' help. Please help us, please pass the HD Parity Act, please add HD to the NNCSS and please improve trials to account for the uniqueness of HD.

Thank you.