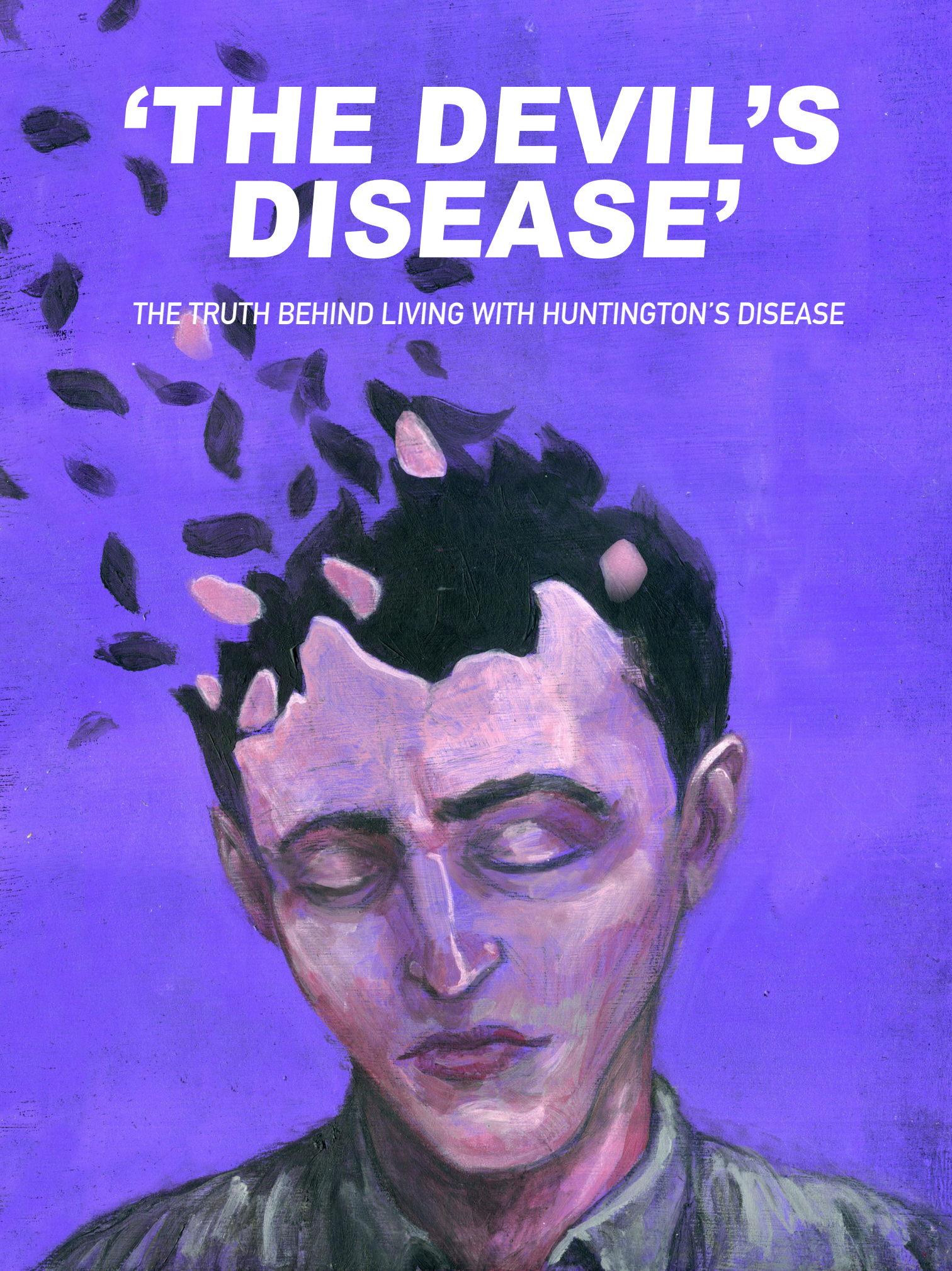
‘THE DEVIL’S DISEASE’

THE TRUTH BEHIND LIVING WITH HUNTINGTON’S DISEASE



“We call it the devil’s disease because it takes everything from you.”

Despite 32,000 people in the UK being at risk of carrying Huntington’s disease, why have professionals, the press and the public not heard of it? *WH takes a closer look at the sinister disease affecting thousands of lives across the UK.*

By Bethia Wyborn

It starts with a few mistakes at work, a few too many smashed mugs, or forgetting a child’s birthday. For carriers of the gene mutation and those who aren’t aware that it runs in the family, these mistakes can be dismissed by medical staff with the symptoms blamed on growing older or menopause. But the reality of carrying the Huntington’s gene mutation is a lot more sinister.

Compared to similar and just as serious diseases, Huntington’s gets a fraction of the attention on all forms of media. Many of those who have Huntington’s believe this is due to the disease not being ‘glamorous’ or because there hasn’t been a big figurehead who has publicly shared their experience of carrying the gene mutation such as Stephen Hawking for ALS (Amyotrophic Lateral Sclerosis) or Muhammad Ali for Parkinson’s disease.

But despite Huntington’s disease (HD) not being as visible, around 8,000 people in the UK have the disease and a further 32,000 are at risk of inheriting it and these numbers only represent those who have been diagnosed. It is unclear how many people actually have Huntington’s disease.

As well as being a life-debilitating disease, those who carry the gene mutation are currently having to fight for changes in areas from the NHS to the armed forces.

Vicki Rutland, head of communication and marketing for the Huntington’s Disease Association (HDA) said: “ It’s frustrating that not many patients or professionals have heard of Huntington’s disease and that we have to fight for changes that should have already been in place.

“We’re campaigning for five things and we have already achieved two of them. We managed to get a benefits review that allowed people with Huntington’s and conditions similar to have an increase of benefits to reflect inflation. We also have improved mental health access for patients with psychiatric symptoms. Previously we saw people with Huntington’s be excluded from community services because they were told because it was too specialist.”

Vicki went on to say that the organisation wants an Armed Forces review as currently people who are known to be at risk of Huntington’s need to have a negative test. Which Vicki claims is denying young people a chance in this field.

She added: “NICE Guidelines (recommendations for health and care in England and Wales) exist for most diseases, so why can’t Huntington’s have it too? We need these specific recommendations to ensure there is consistent care for this complex illness.

“Finally, we want an increase of Specialist Care Coordinators in each area. Huntington’s is really complex and patients tend to need multiple services such as physiotherapy and occupational therapy. We want more specialist coordinators so people with Huntington’s can navigate the correct care they need with professionals who know about the disease.”

Awareness

This May, for Huntington’s disease awareness month, members of the HD community have asked people to take five minutes out of their day to educate themselves.

The campaign started because Huntington’s disease is frequently misunderstood by patients, professionals, and the public.

Despite 1 in 10,000 people being diagnosed with Huntington’s disease in the UK, a frequent misconception is that people who carry the faulty gene are drunk or on drugs. Because the disease affects a person’s movements it can appear that they’re unsteady or unsafe. Unfortunately, this has caused people with Huntington’s to suffer abuse or discrimination in spaces from hospitals to aeroplanes due to their uncontrollable movement.

But most of all the Huntington’s community ask for compassion and understanding for families going through an all-consuming and impossibly tough time.

As Huntington’s disease is genetic you have a 50% chance of inheriting it if your parent has the faulty gene. This is a high probability that affects generations of families. Even for those who test negative, it is likely that they will care for a parent, sibling or another relative with the disease.

James O’Connor is a specialist youth worker for Huntington’s disease and knows first-hand what it’s like to work with young people who have Huntington’s in their

family.

He said:"I think physically it can be obvious that someone has Huntington's, but it's the cognitive symptoms that are the hardest to manage for the person, the family and anybody around them because you can't see what they're struggling with.

"Young people tend to find it difficult to talk about Huntington's with their family members because they don't want to upset them. So having an independent person from the HDA is an opportunity to express themselves without fear of upsetting anyone."

Even when Huntington's disease awareness month comes and goes, those living with the faulty gene experience the reality of the disease everyday.

James added: "It is tough, and it's hard not to take on their anxiety, their sadness, and their grief. But it's incredible to work with such resilient young people. I'm inspired by them every day."

For more...

Advice and support:
Helpline: 0151 331 5444
Email: info@hda.org.uk

Information:
www.hda.org.uk

To make a donation:
HDA: 0151 331 5445
Email: fundraising@hda.org.uk

To join a HDA group:
HDA: 01909 518942

Support for carers:
Advice line: 0808 808 7777

Celebrity spotlight

Despite Huntington's disease not receiving the airtime it deserves, one show brought to light what living with the disease is actually like. BAFTA-winning actor *George Rainsford*, spoke to Women's Health about playing a character on Casualty who had Huntington's and why more people should have a better understanding of the disease.

▲ George Rainsford

What did you know about Huntington's disease before you took on the role of Dr Hardy?

"Shamefully, I didn't know anything about HD before the show. The character didn't know he had inherited the faulty gene, it was the outcome of a storyline involving Ethan's brother researching his birth mother and discovering she was HD-positive."

What was it like playing a character with Huntington's disease?

"I've always wanted to make sure it's an authentic presentation. Within a long-running drama, there are certain constraints which aren't necessarily realistic such as how quickly something advances or how long testing takes, but in terms of character intentions or emotional truth, that was the stuff as an actor that was very important.

I felt responsible for carrying an important storyline and one that needed to land in an impactful way. I also didn't want it to be seen as wholly devastating, I was trying to find the light and shade, to show hope in terms of living with it. To reflect the lives of those I'd met in similar positions."

Why is awareness of Huntington's disease so important?

"I think because so many people still don't know much about the disease and shows like Casualty have the opportunity to educate the masses. Having met lots of families living with HD they are keen for people to have a better understanding of it. Ordinary people aren't aware of what others might be coping with and make ill-informed judgements.

An example of this is when people assume HD sufferers are drunk due to their movements"

Hannah Dickson, 35, from Weston-Super-Mare thought life was over when her husband was diagnosed with Huntington's disease. But it was just the beginning...

I thought it would have been like every other night out in Weston-Super-Mare. Pub, club, chippy on the way home and then bed. But I couldn't have been more wrong. Standing outside the club in the smoking area on a chilly December evening with my girlfriends, I saw a tall man with dark hair looking solemn. Being typical of me, I went up to him and said "Mate, what's wrong with you? Smile".

I could never have guessed what he'd say in response, "I've just lost my dad" the man replied.

I felt awful and all I could do was keep apologising. We carried on chatting throughout the night. I discovered his name was Stu and he was alone because none of his friends wanted to come out with him, so I grabbed him by the arm and said "I'll be your friend".

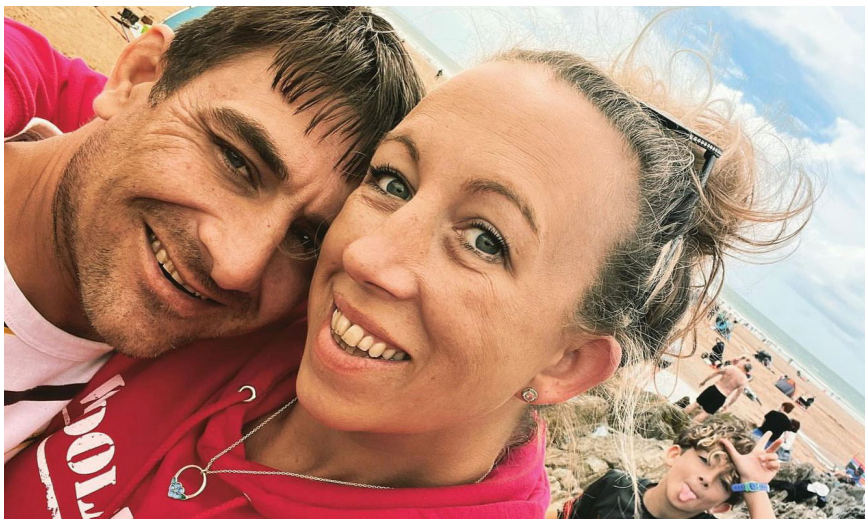
From that moment on, we kept seeing each other and he opened up about how his father had Huntington's and he might carry it too. At that time I worked in a psychiatric hospital so I immediately pictured some of the patients I had worked with who carried the disease. At that point, I didn't realise it progressed in stages, so I was shocked to hear he might carry the Huntington's gene. All I pictured was patients who were incontinent and were unable to feed themselves. But that didn't put me off and 3 months later I fell pregnant with our baby boy, Harry.

At fifteen years old I was told I was infertile, so the last thing we expected to happen was to fall pregnant. Worries about passing the Huntington's gene crossed our minds but we were so thankful to have our miracle on the way that we just decided to ride the wave. Then we fell pregnant with Bella three months after I gave birth.

This was a very intense time for us. Two children within 16 months is hard for any family but after a while, Stu started to notice something was wrong with him. Coming back from a shift at the building site, Stu sat me down and said something that I hoped he never would.

"I had an accident at work, there's something wrong with me," Stu murmured. I felt sick to my stomach. Instantly I tried to come up with

▼ Hannah and Stu



What is HD?

- A disease which is caused by a faulty gene in your DNA called the Huntington gene.
- It affects the body's nervous system; the network of nerve tissues in the brain and spinal cord that coordinate your body's activities.
- Some symptoms of Huntington's may include: Involuntary jerking or fidgety movements, difficulty speaking, slow or rigid movements, personality changes, and breathing problems.
- Symptoms will gradually get worse over 10 to 25 years until the person dies.

How do you get Huntington's?

- It is inherited.
- Every child conceived naturally to a parent who carries the Huntington's gene mutation has a 50% chance of inheriting it.
- The children of someone with Huntington's disease can take a genetic test after the age of 18 to see whether they've inherited the faulty gene. They have to undertake a year of genetic counselling before they can know the result.

Is there a cure?

- There is no cure for Huntington's disease or any way to stop it from getting worse.

reasons why it wouldn't be Huntingtons disease. Surely it's just because of being new parents, or stress at work. But the severity of the accident wasn't like normal Stu. While doing some weekend work he got into the wagon and instead of going into first, he went into reverse and smashed down a whole wall. He also nearly knocked out a colleague by dropping a spanner on his head.

My mind cast back to other instances where his Huntington's might have shown.

Only a month previously I woke up with bruises all over my body because he twitched in the night and accidentally clipped me, another time I lost a tooth because he smacked my face in his sleep. He also would do silly things like smash my expensive mugs, which looking back I regret getting annoyed at him for. As soon as that conversation came to a close, we both knew that he needed to get a test.

Driving up to Bristol on the 10th of January for a consultation, I was in denial. Despite all the evidence in front of me, Stu having Huntington's was the last thing we needed as a family. Most people who are at risk of Huntington's and want to find out if they carry the gene have to go through a year's worth of genetic counselling with a medical professional before they can find out the result. But as we walked into the hospital, they took one look at Stu's twitching and movement and decided it was in Stu's best interest to find out the truth.

"I tried to come up with reasons why it wouldn't be Huntington's disease"

On the day that we were due to receive results four weeks later, we decided to go for lunch in Weston-Super-Mare but we both felt so sick we couldn't eat anything. We both jumped when the phone rang.

That whole moment was a blur but all I can remember was the genetic counsellor saying "We're sorry to inform you that the test came back positive." I don't even know how I kept it together.

"What does this mean for us?" I stammered in response. I felt pathetic because the main thing that worried me was if Stu could go back to work. Since his accident, he had taken time off because of anxiety which meant we had already got ourselves into a bit of debt.

"Don't worry, there's help out there." That's when we were told about the Huntington's Disease Association and support groups that were available to us as a family.

After the diagnosis, I tried to find other people in my world because I felt very isolated. At that time my nearest support group was over an hour away and by the time I had come back from work as a SEN teacher, sorted the kids and driven over my whole evening was gone so it wasn't sustainable for me. So I decided to start my own group in Weston-Super-Mare.

I contacted a few families in near me and we started meeting regularly. It was like a massive weight was lifted off my chest.

People in my group call Huntington's 'The devil's disease' because it strips everything away from you but despite the hardships, it was liberating to meet people in the same position as me.

We started fundraising for other people in the community who needed support. So far we've raised over £4,000 and helped multiple people in the area purchase stair lifts and safety helmets when they've had violent twitching.

There are so many misconceptions about Huntington's disease. When we go to the hospital staff and patients think Stu is drunk, when we go on planes no one understands why he twitches the way he does.

He may not be the same man that I married, but I feel lucky that I get to spend whatever time we have left together.

▼ Hannah, Stu, Harry and Bella

Charlotte Conn, 26, from Teeside has seen first hand the affects of Huntington's disease on her family members. But now she's using her experience to help others...

Leaving my corporate job in strategic HR I strapped into my gown, face mask and goggles for the weekend to work at my Nan's care home. It was during the pandemic, so working at her residential centre was the only way I could see her. It was my 23rd birthday and one of the worst days of my life.

"Nan, what's happened? Are you okay?" I stammered.

I had just walked into her room in the care home and I could instantly tell something was wrong.

Her face was drooping and her speech was slurred but the nurses on call didn't even notice.

"Help, Help!" I belted. It was horrendous and I immediately rang an ambulance.

Unfortunately, this wasn't the first time my Nan Lin had been let down by those trying to care for her.

My nan was misdiagnosed with Parkinson's and severe Alzheimers.

I remember being seventeen and visiting her in North Yorkshire. She was wheelchair bound and all she could eat was mashed banana which she would chuck at staff. Her symptoms progressed so severely that she sadly starved herself to death. After she passed away, my family had an inkling that her diagnosis was wrong due to how abnormal her symptoms of Parkinson's were.

It was in her post-mortem that it was discovered that she carried Huntington's. It's awful to think back and know that she wasn't receiving the correct care. Despite thousands of of people across the UK living with Huntington's, in my experience, it's very rare for patients to receive specialised and appropriate care.

▼ Charlotte and Lin

Since my mum started developing symptoms in 2010, our lives havent been easy. It started with little things like making mistakes at work or breakdowns but then it turned into physical outbursts on my family. She would often hit, kick or punch me. During this time I would just have to rember that it's not her, it's the illness.

I knew I had to test when the time was right.

Sitting in my family home, I had driven over as soon as I got the result.

"Mum, I got the test back and it's positive," I said slowly. There was silence while my mum and dad processed the information.

Then it started.

"It's all my fault" she howled. I've never seen her scream and cry so much. I felt awful. But there was nothing I could do to get rid of that feeling for her, in no way was she accountable for my gene-positive status.

In some ways getting a positive result brought me and my mum closer. It was either a sink or swim situation. I could cry about my diagnosis, stay sad and wait for the inevitable or I could get up and make the most of my life. We both were going through the same thing at different points in our Huntington's journey and all we could do was support each other.

For the past few years, I've been passionate about spreading the word and educating more people about Huntington's. I like to post my reality on social media as well as provide training in residential homes.

In my nan's care home after she passed away, they asked me to come in and teach the staff about the intricacies of Huntington's because they had a new resident come in with the disease.

Making small differences like this brings me joy, even if I can make one person's life with Huntington's easier, it makes everything worth it.

Ashley Delgado, 34, from Jersey was diagnosed with Huntington's disease in 2016. But she hasn't let her gene positive status hold her back...

For the past month, this day was all I could think about. Walking up the steps to the hospital on a sunny day in May 2016, I was lucky I had my partner, stepdad and mum to join me when I got the result. Sitting down in the stuffy hospital room my mind was racing. I squeezed my mum's hand in anticipation.

When the genetic counsellor walked in we all held our breath.

"I'm sorry to inform you that the test came back positive, you have Huntington's disease," He announced. They gave us a moment to process the information.

"I know." I said in response. I don't know what came over me at that moment, despite my future being uncertain, I always had an inkling that I carried the disease that my dad and my grandad had before him.

My mum and stepdad were really supportive. They checked I was okay and gave me time to understand what the next few years of my life might look like. But my boyfriend was a different story.

Receiving my positive result made me remember being fourteen years old and visiting my grandad in the hospital. He struggled a lot through the last few years of his life with mental health and mobility problems but it wasn't until he passed away that they realised it was Huntington's. Looking back I wish he got the support he needed, as a family it was awful seeing him go through that without an explanation.

After my grandad passed away, my dad and I decided to look into testing for Huntington's because he started to show symptoms. Like my grandad, he was clumsy and would often lose his balance. I struggled to a lot for me so I stopped seeing him for a while.

I was born and lived in Jersey so for any specialist test or treatment we would have to catch the plane over to London. But before I was able to take the test, I had to undertake a year's worth of counselling with a genetic counsellor like the majority of people who are at risk of Huntington's and want to be tested.

After my dad's condition progressed, I knew I wanted to find out if I carried the disease. I also wanted to know if I had the gene mutation because me and my boyfriend of eight years were thinking about starting a family and if I carried Huntington's we would have to reassess our options.

I was in my late twenties and now just felt like the right time to find out where I stood.

My partner and I had already started to face problems but getting diagnosed with Huntington's exacerbated our issues.

Walking out of the hospital after getting my positive result, I instantly knew he was worried.

"I know you must be very upset but would you be able to support me?"

I said gently. Even though I wasn't crying, I wanted to be the one receiving comfort.

"I won't be able to have kids with you." He replied. My heart broke at that moment.

From then on we kept going around in circles. He'd say he would change and support me through my Huntington's journey but then he'd revert to his old ways. I wanted to live new experiences while I was well and he wouldn't have any motivation to.

We broke up four years ago and since then I've struggled to find someone who's going to be able to support me through my illness.

There's been a few times when as soon as I tell people I'm dating that I'm gene-positive for Huntington's, they act as if they don't want to know me. It makes me feel awful. I think they're just scared.

I'm lucky that I have a great group of friends and family who support me through tough times. I'm 34 now and my condition has progressed over the past five years. I have high anxiety, twitches, struggle with focus, tiredness and irritability but running has recently been a positive outlook for me.

I've always wanted to run the London marathon for the Huntington's Disease Association because they've given me so much support over the years and after 6 years of trying, I ran the 26 miles this May.

Having Huntington's has made me want to raise as much money as possible to help other people like me or to find a cure. I'm lucky that I've got a great support system around me and the opportunity to go on trips and travel but many other people with Huntington's aren't as fortunate.

I struggled at the last five miles but all I could think about was my grandad, my dad and everyone else who has Huntington's in their life.

▼ Ashley