Learning to live with Huntington's Disease book

Chapter 1. The eye of the storm: Sandy's story

My name is Sandy. I'm 47. I have Huntington's Disease. That sounds like one of those introductions at an Alcoholics Anonymous meeting (I would imagine). I have two boys, Bromley (25) and Danny (15). Each has a 50% chance of inheriting the gene. We'll know one way or the other when they are well into their thirties (probably), as that is usually when the symptoms appear. I myself inherited the Huntington's gene from my dad Brian, who died from the illness in 1995. My brother Geoff also inherited the gene. He's a couple of years younger than me. His three children are therefore also each 50% at risk. Our younger sister Wendy, the baby of the family, did not inherit the gene. Her two girls are, therefore, not at risk of inheriting it as it cannot skip a generation. Phew, some good news to end that introduction with, at least.

Before this chapter starts properly, I need to write you a quick note about the photo at the top. It's me before I developed the symptoms of Huntington's. I wanted you to see me as I am myself, not with any changes that may have been imposed on me by the illness. The rest of the family say this is a ruse to get away with a picture of myself as I was twenty years ago and that in fact it is the change imposed on me by age that I want to wish away. What a cynical lot.

Oh, and a quick explanation of the title of this chapter is needed too, in case it's not obvious. Again according to the rest of the family, I like to sit around relaxing with a cigarette in my hand like some still, small, centre of calm, while they all run around dealing with the fallout of Huntington's Disease. Well, it works for me. So, on we go with my story. I'd like to start with being stuck inside an enormous tube…

Inside the machine

"You'll have to lie still. If you move, it will affect the image and we'll have to do it again," says a voice. I'm lying on a table. My head is at the opening of a vast, noisy machine. The table, with me on it unfortunately, is about to be sucked into the mouth of the machine. Gulp. I feel like Sean Connery in that James Bond movie where the villain clamps him to a table and the ray from a giant industrial laser is working its way up the table towards him.

"You need to lie still," repeats the voice gently as my head twitches. "If I could lie still, I wouldn't need to be here," I say from my horizontal position. "Ah, I'd forgotten about your bizarre sense of humour," smiles the consultant. "Yes, I do still

have one," I think, while trying not to move. "Despite the holes in the brain your scanner might show up, wherever my sense of humour is located in there, it's doing a good job of avoiding the damage caused to the rest of my brain by Huntington's."

That's what Huntington's Disease does to you, gradually destroys brain cells and the connections between them. When you have HD, the medical people like to get a snapshot of your brain as early as possible. They can then compare it with later snapshots (providing you can keep still, I suppose) to see how the disease is progressing.

On this occasion, I was on the table in the MRI Unit (Magnetic Resonance Imager, but you probably knew that already) of Hammersmith Hospital in West London. Dr. Puri, the consultant and owner of the disembodied voice, needs a picture of my brain because he is leading a trial for a drug that may help the symptoms of the illness I have. I'm one of 100 people taking the trial drug. It's a bit like taking cod liver oil – two light amber coloured gel capsules taken twice a day. The deal is that if I spend a year on the trial without taking any other medication apart from the anti-depressants, HRT therapy tablets and sleeping pills I was on prior to the trial, then I am rewarded with an endless supply of the drug before it becomes available to the general public.

The downside is regularly having to go through tests and scans every few months to see how my illness is progressing compared to others on the trial. The trial is over now. The first year was 'blind': half the participants were on placebos and half not. We didn't know which was which. The second year everyone on the trial was on the drug. The trial finished a couple of years ago. We had high hopes at the time for it. But, after two years, the results were not conclusive. There seemed to be a beneficial effect, but it wasn't statistically significant enough to push the drug through the fast track that the Federal Drug Administration in the US and its equivalent in Europe had prepared for it.

So, they are giving it more trials. In the meantime, I keep on taking my free supply. You never know.

Poked and prodded

If you are not careful, life with Huntington's Disease can become a constant round of visits to, or being visited by, an endless stream of health and social care professionals. The list has included at various times: ongoing trips to the family doctor (obviously); a dietician (half a dozen visits); a genetics counsellor (lots of visits); a physiotherapist (one visit was enough); a psychiatrist; an occupational therapist; several consultant doctors specialising in genetics; a movement disorder clinic; various voluntary agencies; social workers; two very nice social care assistants who came to the house wanting to give me a bath – I sent them packing - ; a neuro-psychologist (I think. May have got that one wrong); a speech therapist whom I didn't actually get to meet but had several appointments made before she gave up and sent me a note saying I was no longer on her list…and lots of other people who tried to intervene in one way or another with varying degrees of success. Plus the drug trial visits, of course. Oh, and a homeopathist my sister put me onto, and a faith healer who lives near my mum, is a very nice lady and helps me feel better by focussing on colours. There are lots more but that gives you the general idea.

You have to resist the invasion sometimes. So, I quite often, as with the speech therapist, just refuse to show up if I'm too worn out or disinclined to go when the appointment time comes around. This is known in Huntington's jargon, I believe, as 'challenging behaviour'. I call it trying to keep some level of control over my life.

Seeing the funny side of things is absolutely essential with Huntington's Disease, as it is with any serious condition. It can surprise the medical people and is quite useful for getting them to realize you are a human being, not a lab rat. As anyone who has to make regular hospital trips to be assessed or for ongoing treatment knows, medical professionals tend to split into two groups – the ones who will laugh with you and those who look astonished when you offer a joke or an opinion, as if they'd assumed you were just a subject with an interesting condition, not a fully-rounded human with the power of speech.

Sometimes, the lack of communication can just be silly. When I was still able to drive, I had an appointment with a hospital physiotherapist. I had to park quite a way from her building and was a little late, so ran to get there. She greeted me at the door to her office. I was panting, having run all the way from reception. "I don't know much about Huntington's," she said immediately. "Can you walk unaided?" "Well, yes, since I'm standing here on my own, and you're quite a long way from the reception, let alone the car park, and I gave up flying ages ago, I guess you could say that I can walk unaided, yes," I replied. She didn't see the funny side of it. I didn't go back to see her after that. There's that challenging behaviour again. But, you see my life is shorter than the average. So, I exercise my right to choose who I spend time with. Would you want to spend one hour a week in a room on your own with a physiotherapist who has no sense of humour, when the total number of hours you have at your disposal for the rest of your life is pretty precious to you?

Anyway, I've always been challenging, with or without Huntington's Disease. Let's look back a bit to prove it.

Finding out I have the disease

A couple of years after my dad died I was on a course at Oxford Brookes University, training to become a lecturer in Journalism. That's when I first had to face the fact that I had indeed inherited Huntington's Disease. By then we knew that that was the illness my dad had. In fact, he had died from it a few years previously. We had spent a lot of time, described elsewhere in this book, helping him get into a care home before he died. I knew I was 50% at risk of inheriting the illness. I had chosen not to take the genetic test which would tell me if I had the gene or not.

I was giving a presentation of a lesson plan, when my tutor pulled me aside and pointed out to me that I shouldn't wear bracelets, because my movements made them jangle and create too much noise. She said I appeared to have 'a motor problem' and asked me what it as. I was devastated, as I had not noticed any symptoms myself. I burst into tears and explained, between sobs, that my father had died of Huntington's Disease and the movements she described were a symptom, so I must have inherited it, too.

It was then I decided teaching would be too stressful a job for me and I gave up the course. When I came home from the college session where the lecturer had pointed out my movements to me, I was obviously devastated. I was in tears – I don't know how I drove the 20 miles from Oxford to Bloxham, the village where we live, in that state. My husband Phil sat me down, made me a tea, and asked me to tell him what was wrong. When I told him what had happened, his face fell.

I went on to discover later that Phil knew that I had the illness but hadn't felt able to tell me. I had made it clear at the time that I didn't want the blood test, as there is no cure. The majority of people don't want to know. If there was a cure obviously that would be different.

The blood test – confirming it for sure

The worst fear is the fear of the diagnosis – not knowing. So, the five years or so after that – knowing I had a 50% chance of inheriting it, had been full of worry. A needle pricking my finger was the first step in finding out for sure that I had Huntington's Disease. The neurologist then undertook several co-ordination tests on me, similar to when the police test motorists to see if they are drunk. You'll be familiar with this from the movies. Or, perhaps from your own experience. You have to touch your nose, walk in a straight line and so on.

For some reason, having been quite stoical up to this point, I suddenly found this process horrible. I felt humiliated. Phil was there with me and confirmed afterwards what I thought as I went through what I came to think of as an ordeal. The doctor was blunt, cold and clinical. I felt I was being treated more as a lab assessment, a curiosity or a clinical exercise than a person.

I had to wait an agonizing month for the blood test results and when they arrived I had to go back to the hospital to see the same cold and clinical neurologist as before. She said, 'Bad news…', and went on to tell me I had not only the Huntington's gene, but that my reactions to the cognitive and movement tests showed I had the symptoms as well.

When I found out, it was actually a relief to know, in a bizarre kind of way. My view had been, over the seven previous years during which we had learnt about HD and seen its effects on my dad, that if I knew I had it, I wouldn't be able to live with that knowledge. That's what I told my husband whenever we discussed having the test.

But as often happens when you have shocking news, you don't react the way you think you would. You find strength from somewhere. After a cursory "We would advise people in your situation not to have any more children," which I interpreted as "We don't want any more like you around, do we," we went home.

Telling the Children

Lots of couples who know they have Huntington's in their families choose not to have children in case they inherit it. I already had two sons when I found out. But I don't regret having them. I'm glad for many reasons that I did not have to make that choice. I like to think my choice would have been the same. I have no regrets at all. Both my

sons are the best things that happened to me. I love being a Mum. I couldn't imagine a world without them.

I was 41 when diagnosed with the illness. Both my sons Bromley (now 25) and Danny (now 15) have had to learn to live with the fact that they not only will lose their mother to this illness – unless the high living I engage in gets me first. I'm doing my best. That's a joke, by the way. But they also have had to adapt to the fact that there is a 50% chance that either or both of them might have inherited the defective gene from me.

I had already told them we had Huntington's in our family. You'd be surprised how many people in our situation don't. Some families don't talk about it because it's too painful, or they feel defeated by it. A parent's natural instinct is to protect their children, perhaps by withholding information. But, we should never underestimate our children's capacity to cope, particularly with information like this.

Research shows that it is, in fact, better to be honest about it as early as possible, because children find it harder to deal with the shock of discovering they or their family are 'different' later in childhood than if they have known about it for as long as they can remember.

Developing the symptoms

When I first started to develop the symptoms of Huntington's, I didn't try to hide them, though it is easy to feel self conscious, it affects one's movement, concentration and memory. When I am tired I forget words in the middle of sentences, which is particularly frustrating for me as a writer and as I have always had a very good memory. I realized that I was forgetting little details that normally I remembered, such as……er, I forget where that sentence was going. Yes, that was for dramatic effect. Sorry about that. I also get angry and frustrated about things that normally wouldn't have affected me.

The symptoms take hold progressively over a ten to fifteen year period, so it's a gradual decline. People with Huntington's are often accused of being drunk, because of the combination of slurred speech and a tendency to fall over! The illness affects my balance and co-ordination. Though I find it difficulty I can still walk at the moment. People in the latter stages of the illness can't, so often they have to use a wheelchair.

I can't protect them from it

I know that if either of the boys do get it, there will be nothing I can do to protect them or help them, as I probably won't be around. Any mother's natural instinct is to protect their children and it is hard to imagine that situation, and also that the illness would have come down through me. So, I try and live a full and active life, and to be as positive and cheerful as possible. If I can make it clear I value and enjoy every minute of time I am with them, it shows them that if they do get it, life goes on. That's the best thing I can do for them.

Involuntary movements are just one of the symptoms of my illness. Losing weight, due to the energy taken up by the continuous involuntary movements, is a problem for people with Huntington's. So, I am on a high calorie diet (the envy of most women I know). I can eat what ever I like, and do! From fry-ups to Chinese food to ice-cream, my eating habits are the envy of my friends. As Huntington's is a disease of the brain I make a point of eating food such as fish, cod liver oil and blueberries, all reputedly good 'brain food'.

It is dreadful to always be dropping and breaking things. I can't cook anymore, which is why Phil does it. We always shared mundane tasks like housework, ironing and cooking, but now he has to do all of that, with the help of an occasional cleaner plus my mum and his own mum coming in to help to stop things getting on top of him, as I find these things too difficult.

I can only concentrate on one thing at the time, this is a symptom of Huntington's and multi-tasking has become impossible, Also my tiredness is a problem, I've always had lots of energy but by the early afternoon these days, I can feel exhausted. I am much slower in my ability to think and make decisions, I used to write fast and think fast, but I can't do this now. I was very decisive.

I will be on anti-depressants and sleeping pills for the rest of my life. When I don't take the anti depressants I lose interest in life, everything seems pointless, I feel lethargic and obviously if I'm depressed it affects my concentration. I've always had a very good sense of direction, but as the Huntington's developed I found that I was getting lost more when I was driving.

Sleeping through the earthquake

I also used to be a really heavy sleeper. When we were working in California I had slept through an earthquake in San Francisco that had woken Phil up and made all the newspaper front pages the next day. I even slept through the famous 1987 hurricane in England. But sleeping now can be difficult which is why I am on sleeping pills. I can't remember my dreams anymore, oddly, and I used to have nice ones!

It could be easy to be self conscious as I do drop things in shops. People tend not to intrude too much which is good. I can't read my own handwriting anymore. I originally said no thanks to a disabled badge for my car because I felt like a fraud. But, more recently, after losing the right to drive and having to be driven around everywhere, I have had to accept a disabled parking disc for whoever is driving me around at the time.

Losing my Independence

When you are forced to face something inevitable like this, there's an initial period of shock and disbelief, anger, fear, a sense that your life is over. But, you either collapse and take to your bed, or you keep getting back up every time the illness progresses a bit more and knocks you back. Like when I was told I could no longer drive.

I lost my licence after suffering from Huntington's for a couple of years. You don't legally have to tell your employers if you have a disease but you do legally have to

tell the driving licence authority in the UK and the car insurance company. I had to take an annual driving assessment, which seemed to me harder than the original driving test itself. It's a serious point, actually. If 'ordinary' people had to retake their driving test every year, how many of them would pass every time?

I twice took the assessment. I passed the test the first year. But, the second time, a year later, even though everyone thought I would pass, I failed. When the man conducting the test told me I'd failed, it was a major change to my life and I felt everything crashing down around my ears. I couldn't nip to the shops anytime I wanted, visit friends, go clothes shopping, pick the boys up from school if they needed it…all the things you take for granted. In a stroke of his pen in the 'failed' box, the world in which I could operate independently had suddenly shrunk to the house and how far I could walk from there. And walking wasn't getting any easier.

So, I stormed out, leaving him standing there in the reception area of the test centre. He was trying to say some consoling words, but anything he said after the words "I'm sorry, you've failed" were a blur. I just had to get out of there. I was so shocked and furious at having my independence taken away. I just hadn't expected it at all. I remember staggering outside, fumbling for a cigarette, with tears of anger in my eyes. I instantly lost my freedom to drive and Phil had to bring me home. You can't appeal. That's it. No more car.

This loss of freedom and independence was really hard for me to deal with. From that moment on I had to rely on everybody else for lifts. After being able to drive myself to Oxford to buy clothes, to London to see friends, even do mundane tasks like visiting a supermarket, I found myself stuck in the middle of the countryside in Oxfordshire, with no means of transport, but the local bus service. I had spotted one bus in the ten years or so we had lived there.

My work suffered too. After I was diagnosed with Huntington's, I continued to write a few articles but I found the ability to carry on writing, to the degree I had done before diagnosis, increasingly hard to maintain. I think the shock of finding out I had Huntington's dampened much of my creative capability and I also suffered from extreme tiredness which affected my capacity to write; Having said that, I did write a few articles for different papers about my battle with this disease. Realistically though, Huntington's pretty much ended my writing career.

There have been so many misunderstandings about Huntington's. Most people in the UK had never heard of it until recently, when it was featured on the TV soap opera Eastenders, where it was presented in such a negative way, with the introduction of a character in the final stages of the illness, bed-bound, helpless and unable to speak. That's not me.

Live Life to the Full: The Positives

Nowadays, after years of adjusting, I try not to think about my illness but rather to concentrate on what I can do. I spend much of my time at a local riding school, having taken up horse riding again after many years. It was a passion of mine – I had a pony as a teenager as I mentioned above. And I found it's a hobby I still can do. My body seems to just remember how to control the horse, Caroline, the owner of the

stables where I ride, told me. She had been concerned my movements – my arms and body muscles tend to move constantly – would confuse the horse. But, it turned out not to and so I have rediscovered a skill and passion I had not found the time to keep involved with. And I am grateful to her for giving me the chance to try and prove I could do it.

A couple of years ago I got it into my head that there was one thing I needed to replace my little sporty car that had been taken away. It was taken away shortly after I opened the driver's door into the path of a car coming up behind me and as the car passed it virtually ripped the door off. The car couldn't be economically repaired and shortly after that I failed my test. What I needed to win back some independence, I realized, was my own horse.

Phil almost had a heart attack (that would have helped pay for a horse, actually – he's nicely insured, whereas my life can't be because of the Huntington's). He came up with all kinds of arguments as to why we couldn't afford to buy and then maintain a horse. He used words like 'maintain' as if it was a car. My sister Wendy came to the rescue with a creative solution just as I was about to find something really heavy with which to convince Phil that, compared to a number of blows around the head, the cost of a horse was painless. That's a joke too, by the way. Wendy's solution was a rent-a-horse scheme where you can ride a horse a certain number of times a week for a monthly fee. We found our local stables, Turpin's Lodge, offered this 'horse for hire' type of programme and tried it for six months.

It was brilliant, but I wanted more. I still wanted my own horse. Phil and I clashed a few times more over this. Then Caroline at the stable came up with yet another creative solution. One of her horses, Charlie, who happened to be my regular ride and whom I loved dearly, was due to retire. They would keep him on for a year or two and treat him as mine for that time if we paid the livery (that's his upkeep, vet's bills, food and so on). This is hugely therapeutic for me and I try to ride at least once a week. I have fallen off a lot, but I always get back on eventually!

Another plus thing to come out of not being able to drive, as well as rediscovering riding, is I have a great new friend called Jo from an organization called The Clive Project, who pops around twice a week. Jo says her job is partly to make my dreams come true; to find out what I want to do in life and act as my facilitator to make it happen. She is amazingly good at it! I enjoy the freedom and independence this gives me and the sense of being in control. The fresh air and exercise of riding helps me counter depression and is good for balance and co-ordination. On a broader level, Jo simply makes my life richer and I look forward to her coming over.

The family counts

We make a point of taking more family holidays together – conscious of the fact that time together as a family is more precious as it is likely to be shorter than for most people. I love the adventure of visiting new places and different cultures. One unexpected positive thing about my diagnosis is that we now, as a family, take adventurous holidays that we might otherwise have postponed until later life, including a trip to Southern Australia a couple of years ago, where we met the in-laws

– my son Bromley married a lovely Australian girl, Chantel – and travelled the Great Ocean Road in a rickety old motor-home.

Phil now works from home so he can be around to keep me company and we can do things together, like nipping out to the local pubs for lunch. I get far more tired than I used to, so he has gradually taken over some things that we used to split 50-50, like making sure our younger son has clean clothes for school and someone to see him off in the morning, shopping and stuff – my mum pops around to help with things, too. He helps me deal with all the social services, health professionals and other bureaucracy that crops up when you have a disabling illness. And he picks me up when I fall over.

Not fade away

I realize that through my diagnosis of Huntington's I have lost my future. I will not live to be old. I suppose that I have always known, certainly since my Dad was diagnosed with Huntington's, that the outlook for me was quite bleak. I live with this by distracting myself I suppose. Phil helped me cash in my small employee's pension recently because I felt that I should be able to enjoy the benefits of it now rather than when it is too late, because I'm not going to make retirement. Phil now tells me it has all gone on paying for Charlie for two years and on covering the cost of my cigarettes. I'm not sure I believe him. He's swanning around in a rather expensive-looking shirt at the moment. I'll have to hunt out the receipt and see if I'm paying for it. That's another joke, by the way.

My Dad didn't make it to his 60th. He died when he was 59. When you know you won't live to be old, memories are more important than material things. I love doing things together as a family, spending time with the people I love, celebrating Christmas and birthdays and heading off on family holidays. We drag all the family away with us at least once a year, whether they like it or not.

Though I have this disease I will not let it overwhelm me. I intend to live my life to the full, to enjoy every day, spend happy times with my children and the people I love, plus Charlie the horse. I am not going to go quietly into that still night, or whatever the phrase is. I am going to continue being as noisy and challenging as possible. I still bleach my hair. Yas, my brilliant hairdresser, still spikes it up and makes it look pretty formidable for me. Challenging behaviour has always been my specialty. I see no reason to stop now.