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genetics [2] brain [3] disability [4]

When illness strikes, the impact can be widespread and profound. There’s one disease that has more far-reaching consequences than most, penetrating deep into families and parading itself in front of strangers. But is it being shunned by science?

What is Huntington’s Disease?

It’s a progressive incurable inherited condition in which brain cells are gradually destroyed. Symptoms may be mild for a time but award-winning neuroscientist/novelist Lisa Genova [5] describes how, for one woman, Huntington’s “destroyed her ability to walk and feed herself. It mutilated her good mood, her patience and reasoning. It strangled her voice and her smile. It stole her family and her dignity, and then it killed her”.

I have nursed people with Huntington’s in the hospital, the hospice and the care home. The most striking feature is the loss of control over movement: the body can seem to dance to a music that no-one else hears, always on the move. It can look like you’re fidgeting – or fighting a fearsome (invisible) enemy.

Impact on the Spouse

Promising to care for each other “in sickness and in health” is tested in Huntington’s as the spouse becomes caregiver; the jerky body movements meaning help is often needed with activities of daily living [6].

Harder still are the emotional changes [7] that form part of Huntington’s: apathy and depression are common – and difficult to talk about because speech can become slurred and trains of thought tangled.

Irritability [7] can accompany Huntington’s, with Genova’s fictional wife saying, “He’s got this short fuse. You never know what’s going to set him off, and he can go from like zero to sixty in nothing….He’s got this weird temper, and it’s not like him to be like that”.

Impact on Siblings

I once cared for two siblings with Juvenile Huntington’s [8] (early onset). Both in their twenties, the sister could only watch as her brother succumbed to the disease and died. She had undergone genetic testing [9] and knew that a similar fate awaited her: hard to believe while she was still a beautiful young woman working in a bank.

For those siblings not carrying the mutated gene, their ‘freedom’ often doesn’t taste so good. Genova sums up how, for one sibling, the freedom “feels unfair, tainted, rotten. She feels utterly unworthy of that freedom”.

Impact on Successors

If your parent has Huntington’s, you have a 50% chance of inheriting it. Choosing to find out whether you have the mutated gene is incredibly difficult. Genetic counsellors [10] are trained to provide information to support people’s decision-making regarding the genetic test.

90% of those at risk [11] of Huntington’s choose not to find out whether they carry the gene: fear of the future, loss of hope and absence of an effective cure are common deciding factors.

Choosing whether to have your own children is another difficult decision to make.

Impact on Strangers

Since Huntington’s can cause you to slur your speech, fall over, lose your temper and drop things, it has often been mistaken for drunkenness. 24 year old Ben Jones [12] was refused entry to a bar for this reason, despite displaying his card [13] which explained his medical condition. In Genova’s novel, the main character takes to wearing t-shirts with the words “You are staring at a man with Huntington’s” or even “I have Huntington’s disease. What’s your excuse?” to overcome such prejudice.

“I have Huntington’s disease. What’s your excuse?”

I’m guilty myself though. Arriving on a shift one afternoon, I thought that a father was assaulting his son and called for help. I didn’t know the Huntington’s diagnosis.

Strangers can, however, become involved in a very positive way – by wearing their jeans to work [14] and raising money to help affected patients and their families.

Impact on Scientists

In 1993, scientists discovered the genetic mutation that causes Huntington’s. 24 years later, despite many attempts, families still live just in hope since no treatment or cure has been found.

Genova suggests that drug companies are more interested in researching diseases such as Alzheimer’s and breast cancer, which affect millions of people each year (whereas 37,000 people have Huntington’s in the United States, and approximately 5,700 in the UK).

Scientists are, however, working hard to find a cure. Oxford Professor Matthew Wood [15] and Cambridge Professor Roger Barker [16] are among those researching gene therapies; potential treatments are being tested in trials [17] today.

A Positive Impact?

Marjorie Guthrie, widow of the famous American folk singer, Woody Guthrie [18] (who died from Huntington’s), didn’t like the emphasis on the fatal nature of Huntington’s. She said, “Fatal implies that your life is over tomorrow – it’s not. Life itself is a fatal disease and being born is the first symptom”.

With Huntington’s, there can be many productive years between diagnosis and death. Diagnosis can help re-focus on living, not dying: for some, priorities change, dreams are dreamt, relationships restored. With the right help and support, it may just be possible to light one candle rather than curse the darkness.

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