EDITORIAL

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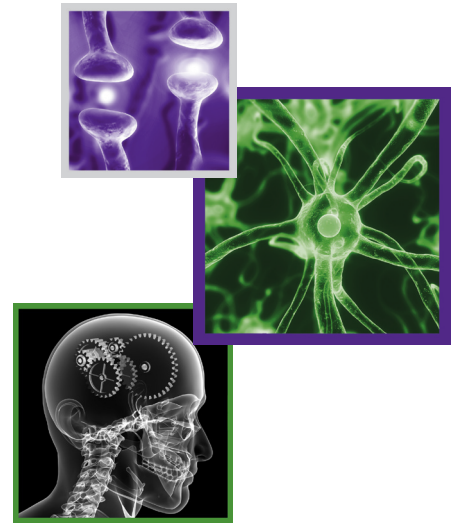
A standard of care for Huntington's disease: a patient and family perspective



Charles Sabine*

"The sight of family members, carers and institutions with, by their own admission, little real idea of how to cope with the debilitating symptoms of sufferers, only serves to compound the horror of the next generation who are themselves at risk of carrying the mutant gene..."

There are several significant benefits to emerge from the growing consciousness and understanding of the nature of Huntington's disease (HD) in the 21st century.

The most immediate of these to exert a material effect is the understanding of the importance of implementing consistent standards of care to a condition that, until shockingly recently, suffered shameful treatment more often than not.

Michael Wooldridge, who now runs Rapkyns, a care home for people with HD in Sussex, UK, related to me the moment when he knew the path his life would take. It was in 1969.

"I had just started nursing in one of the large mental hospitals and was being shown around a geriatric ward. In the distance, I could hear screaming coming from down the corridor – it got louder and louder until we came to a side room and the person opened the door.

"Sat on the floor was a young lady, screaming. The person with me said she had Huntington's chorea, as it was known in those days, and made a lot of noise, but had no idea what was going on; so I was not to worry about her.

"The room had plain walls, with a very high ceiling and no windows. The door had a peephole, so you could look in on the person without opening it.

"I had just started nursing, so had no idea about how the brain worked, but I couldn't believe someone didn't understand what was going on around her.

"As a child, I had a hearing problem, so I was used to having to listen very carefully whenever anyone was speaking.

"I was able to spend time listening to what she said, and discovered that, far from not knowing what was happening, she told me all about herself and her family, and explained the reason for the screaming was that she got very painful headaches, which felt like someone was crushing her head.

"I told the nurse in charge and they told the doctor and so, instead of being given an injection, she was given paracetamol, which helped her."

Now, remember; those words relate to 1969, not 1869.

The green shoots of change in the profile of HD over the last three decades, which have brought about care homes like the excellent one that Michael Wooldridge runs, have also brought challenges; not least the

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growth in the number of families known to the Huntington's Disease Association and its outstanding, but already overstretched, regional care advisors. The apparently growing prevalence of the disease, as more people dare to emerge from the shadows, is also, in part, a product of many of them being cared for with greater dignity. What person at risk of the disease would have been able to openly confront or admit a destiny of the type that so shocked Michael Wooldridge in 1969?

This subtle but highly important factor demonstrates how the European Huntington's Disease Network Standard of Care working group guidance papers [1–7] could affect the HD community in more ways than the obvious provision of accumulated knowledge to the health services.

From the time it was named by George Huntington in 1872, and indeed, before that, when it was commonly known as 'St Vitus Dance', this devastating neurodegenerative disease has left a lost community in its wake; a section of society hidden by its own shame and made transparent by a vacuum of self-esteem.

"HD has had an almost unique power to challenge the human spirit because of its terrible paradoxical mix of finality and perpetuity; finality for its individual victims and perpetuity for the misery it imparts on a family."

The reasons for this are manyfold; from the stigma derived from associations with bad character, criminality and even witchcraft and evil, to a self-imposed social exclusion caused by family members wishing to keep hidden the presence of the disease among them.

However, whatever the causes, the factor that has combined with the unique genetic nature of the condition in an evil partnership perpetuating the misery caused by HD, has been the absence of guidance in how to care for those suffering from the disease.

The sight of family members, carers and institutions with, by their own admission, little real idea of how to cope with the debilitating symptoms of sufferers, only serves to compound the horror of the next generation who are themselves at risk of carrying the mutant gene, and their reticence to face the reality of the disease.

This is a particularly troubling phenomenon at a time when the possibility of clinical trials in the coming years necessitates ever more 'at risk' family members joining a proactive collaboration with researchers.

As my father reached the last stages of his illness in the 1990s, my mother, who personally cared for him at home until the month of his death, knew little of the benefits of guidance in nutritional, occupational, speech and language therapy, dental care and physiotherapy. For the next generation of my family to witness this wisdom being applied to my own future care will give support to their efforts to confront the future knowing that they are not beyond the radar of health services.

However, it is not just for the next generation of patients that this guidance is crucial. It is, of course, imperative for the current body of patients themselves. In my career as a journalist, I discovered that human beings lose their social equilibrium when you take two things away from them: hope and dignity. There is the possibility that a glimmer of hope – so valuable when set against a terrible darkness – might be emerging from the laboratories searching for treatments for a thus far untreatable condition. But what about dignity? What might provide that to sufferers of such an undignified illness?

The positive effect on a patient of knowing carers have a plan is difficult to describe. The only way, perhaps, is to explain what it is like to perceive the absence of one. For families to ask health professionals what the best advice is with regard to an aspect of practical care and be told "Well, no one is really sure about that" is not only demoralizing to those who love the person with HD, but undermines the already wafer-thin dignity of that sufferer. "How can there not be a standard of care for people with this disease", one wants to shout; are they really that far down the food chain that no one can be bothered to lay down a code of best practice of how to look after them? It is hard to imagine a less dignified place in society to inhabit.

And then, of course, the last group to benefit from the Standard of Care guidance is the body of carers; the secret army that suffers the greatest collateral damage of any disease known to man. These are the people who demonstrate that in our world it is in the darkest moments that the greatest qualities of the human spirit shine brightest and, in doing so, they give us all hope.

HD has had an almost unique power to challenge the human spirit because of its terrible paradoxical mix of finality and perpetuity; finality for its individual victims and perpetuity for the misery it imparts on a family. However, its greatest evil has been the way in which it sucks hope into a vortex. It has not defeated the human spirit, because the very best of humanity surrounds it; the superhuman patience and tirelessness of the families, and other carers. They deserve not just our untold admiration, but also all the tools that can be afforded them in their Herculean task.

The professionals that have taken yet one more tireless step beyond the call of duty to put these guidance papers together can never

be shown enough gratitude by every member of the community I represent.

On their behalf, I say; thank you.

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