PRACTICE

A PATIENT'S JOURNEY

Motor neurone disease

Diane Sackett experienced the first symptoms of motor neurone disease in 2004, and died in September 2009. Diane's husband, Brian, was her main carer.

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This is one of a series of occasional articles by patients about their experiences that offer lessons to doctors. The *BMJ* welcomes contributions to the series. Please contact Peter Lapsley (plapsley@bmj.com) for guidance.

To share more people's experience of depression go to: http://www.healthtalkonline.org/Nerves_and_brain/motorneuronedisease

In 2004 my wife Diane noticed that her arms and shoulders ached and that she tired easily. We put this down to repetitive strain and consulted our general practitioner. The physiotherapist at the practice referred her to a specialist, who after extensive tests broke the news. In May 2006, Diane was given the diagnosis of motor neurone disease.

We had never heard of the disease before. We were both devastated, even more so when the specialist told us that Diane might die within three months. We came out of the specialist's office in a state of disbelief and shock. From that terrible day I became Diane's full time carer; not quite what I had in mind for my retirement.

The hardest part was telling our children, family, and friends. We both felt hurt that friends stopped visiting or making contact. Why? And we both agreed not to tell anyone that Diane might have only three months to live.

Within days of the diagnosis the professional team started to visit and introduced themselves to us. Our local general practitioner took on looking after Diane and learning about motor neurone disease for the duration. The entire team became more than specialists in their own fields; they became great friends, making regular visits that we could rely upon. When members of the team linked up and made joint visits, these were of great benefit to Diane, as she could save her breath and energy by not having to repeat herself to each individual. She looked forward to these meetings eagerly and although she found them exhausting, her spirit and wellbeing was greatly improved by each one.

Diane and I had a mantra we would say together, "If this is the best I am going to feel, then I'm going to make the best of it."

In becoming her carer, I had to be careful not to treat her as someone different or as an invalid. Instead, I needed to let her do whatever she could, so she could keep her independence for as long as possible.

Diane got very frustrated when she realised she could no longer do even simple things for herself. Appearance was always part of her life, and she always looked beautiful, so we talked to both her hairdresser and beautician, who both agreed that if she was not up to visiting either of them, that they would visit our home and do whatever was needed. People often remarked on how soft and smooth Diane's skin looked. We both put it down to the fact that every day after her shower, I would give her a full body massage using emu oil. She enjoyed the massage sessions because they provided contact between us. A little retail therapy also gave her a boost. Her appearance was maintained right up to the end. It was also important that I made an effort to always look my best for her. I could not allow myself to become ill.

Washing and bathing was the next experience we had to overcome. My lifting Diane in and out of the bath was becoming a worry for her, and I was worried that I could drop her. We agreed to take the bath out and install a shower, and we had many fun showers together.

Saliva built up in Diane's throat making it difficult for her to breathe and eat properly, and causing her to choke often, which exhausted and frightened her. The only thing I could do to help was to rush her to the bathroom while supporting her, turn on the cold tap and get her to gargle. Then I would place my fingers in her mouth and pull the thick saliva away, which was very unpleasant for her but necessary. This procedure used a lot of her energy and scared her, but it worked. One solution was for her to drink plenty of sparkling water and take carbocysteine (Mucodyne syrup), 10 mL morning and night, prescribed to reduce the thickness of the saliva.

Medicines in tablet form became a problem as she was having difficulty swallowing; liquid medicines provided the answer. We devised a cocktail, mixing all the different medicines

together so that she only had to take one dose instead of the four prescribed.

Breathing was becoming more and more difficult, and a bilateral positive airway pressure machine was provided by the William Harvey Hospital, Kent. Diane's breathing improved beyond recognition, as did her sleeping. She regained her appetite, food tasted better, her speech improved, and her choking had reduced; this was an important factor in prolonging her life.

Eating became more of a problem as her arms became weaker. The solution was to share our meals together. I would feed her a spoonful, and then I would eat a spoonful. This encouraged her to eat and increased our sense of togetherness. We kept it up until the very end of her life.

Always thinking about what was going to happen next, we discussed, among other things, the possibility of Diane losing her voice. She was given a light writer, a text to speech device, to programme well known phrases and sayings, but she never used it in earnest—she was determined to use her voice for as long as she could.

Sleeping was another big obstacle. She could not sleep on her back because that put too much pressure on her lungs. So we experimented using various pillows, finally solving the problem by using a shaped foot wedge under a v-shaped pillow, which supported her at a 45 degree angle.

Shortly after Diane was diagnosed with motor neurone disease in 2006, we went to Cairns in Australia. She was not really strong enough but was very determined and insisted on going. It was a big learning curve for both of us, in terms of understanding the illness and each other, particularly as we alone knew that Diane might die in three months. I looked after her every need, which we both recognised later as a mistake on my part because I had taken away her independence by not allowing her to do even the simplest things for herself.

Early in 2007 Diane's condition was deteriorating. She was very aware that she was losing weight and what little strength she had. She needed to wear her neck collar more often as her neck muscles were weakening. Walking took its toll. I located a lightweight wheelchair, "Oh this is comfortable Brian, what do you think?," she said. "You sure love?" I asked. "Yes, let's buy it." The chair was a wonderful purchase because it opened up a whole new world for both of us. Folded up it fitted into the boot of our car, and we made many happy excursions together.

In 2008 Diane was showing signs of weakening, was very frightened, and was losing weight very fast. We carried on with life as best as we could and purchased an electric scooter to provide a bit more independence for her. At the end of July, the Pilgrims Hospice in Canterbury (<http://www.pilgrimshospices.org>) opened a therapeutic labyrinth garden and we were the first couple to walk it, which we were both very proud of. We were both wearing our "MND" t-shirts and were hoping to raise awareness of this terrible disease.

In September 2008 Diane gave up and wanted me to help her end her life. Using all my strength, I refused her request and

quickly got her into a hospice, where she stayed for two and a half weeks and they allowed me to help look after her.

At Christmas, she arranged a surprise birthday party for me at our favourite restaurant. I asked her "Why this year and not next, when I will be 70?" She replied, "I don't think I will be here next year."

One of the worst things about motor neurone disease is watching your loved one slowly fading away day by day, while becoming more and more dependent on their carer. By mid 2009 Diane was putting on a brave face, still smiling, but despite her efforts she looked very weary; drawn, her face sunken, tired most of the time, and sleeping more.

On 5 July, Diane decided to have a percutaneous endoscopic gastronomy feeding tube fitted. The temporary tube was fitted in adequately. We returned after a few days for the permanent tube to be fitted. It was unsuccessful, caused her great pain, and was removed. In August, Diane seemed to be picking up, looking good, eating well, and smiling a lot. On 14 August, the doctor visited our home and informed me, "I don't think she will make it to the end of August, she is near the end." When Diane and I had a discussion about the end of her life, she said, "Change the radiator in the bathroom please. That is all I want, I have always hated it." Diane lost control of all her body functions, was not sleeping well, and not eating. She was very unwell, tired, upset, and irritable, lost her temper easily, was very frightened, and was losing her voice.

On 1 September she did not want any food and slept all day. I sat just waiting for some response from her. On 2 September she was confused and frightened, unaware of where she was or what she was doing. She did not recognise me. I called the hospice, where we arrived around midday. The staff made her as comfortable as they could. The doctor took me aside and gave me the bad news that Diane had only a short while to live. She died early the next day.

Only now can I admit that every day of Diane's illness scared the life out of me.

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A doctor's perspective

As the consultant, I was responsible for leading the multidisciplinary motor neurone disease team in formulating the care plan throughout the course of the patient journey and I visited Diane and Brian at their home for ongoing assessments.

Specific training is needed to break devastating bad news such as a diagnosis of motor neurone disease. Clinicians need to exercise caution before giving individual patients an average time frame of life expectancy.

Carers value the continuity of care, which helps build up trust and a relationship with clinicians.

Inability to cure a patient should not mean that clinicians abandon hope or fail to address distressing symptoms, such as the thick saliva problem that arises in motor neurone disease.

Decision making in relation to a relatively simple procedure, such as percutaneous gastroscopy, becomes emotionally difficult and may impact on a patient's capacity to consent and agree to such interventions.

I believe that carers need to be given more formal consideration by society. Has the time come for amending the legal duty of care to include the carer's view? At the moment, doctors owe legal "duty of care" only to patients, if they are an adult and have got a clear mind to consent.

The system compels carers to go through an identity change from "lover" to "carer" to "professional"—for example, carers have to learn how to use and look after various assistive technologies. Being a carer brings increased responsibilities without any real authority being reflected in British law, which does not give the carer any legal authority in the decision making process for those interventions.

Medical professionals must consider the danger of psychiatric injury when providing information to relatives and carers and carry out an appropriate risk assessment with their peers.

A professional minimum standard should be set, against which the response to family carers should be judged. It should include a duty to impart guidance to carers that deals with what to expect and how to deal with a condition, and should include a duty to inform carers where follow-up information can be found.

Judgment of the standard of care towards carers should not be left to the common law and judges to codify. Case law accepts the views of a reasonable body of professionals. The medical profession is therefore well placed to guide duties of care towards carers.

Setting a standard towards carers could avoid arguments along the lines of those that arose after the Hillsborough disaster, by defining whether a carer is a primary or secondary victim and addressing the additional problems connected with proving duty of care to a secondary victim.

Consideration of our duty of care will help to prevent carers from becoming the cared for.

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