

***MY STORY written by Mandy Newton***

***This is the story of my father, Graham Brown's, death by fCJD as experienced by me. Dad passed away on August 23<sup>rd</sup> 2004. He was just 63 years old.***

***I send love where there is only fear and well wishes to those of you who, like my family and I, find themselves in this difficult circumstance.***

I remember as a teenager growing up in the 1980's seeing a disturbing report on the evening news one night. It was about a devastating disease that had struck a significant number of cattle herds in the UK. I remember watching the footage, taken aback, as the ill-fated animals persistently staggered and fell down. I don't think I gave it any more thought until some 20 years later on August 6<sup>th</sup> 2004, as I sat clasping my father and mother's hands and a neurologist told us my father was dying of a variation of the same disease.

CJD sets a rapid course and in the 8 weeks or so prior to receiving this devastating news, my family and I had spent many, many hours conducting what seemed to be futile searches, pouring over the Internet, looking for answers as we wondered what was happening to Dad. To finally know the name of the monster we were facing was in a strange sense a relief and yet the knowledge that it was invariably fatal was obviously harrowing. I will not forget the look on Dad's face as he cried and whispered to me, "I never thought I'd get that". It was probably one of the last times, I believe, that he was lucid enough to fully comprehend what we had just been told.

As Dad recovered from a hip replacement surgery in late May 2004, my mother began to notice changes in Dad's behaviour and memory. On one occasion during a discussion about South America, Dad was unable to recall the name of Argentina, a country he had been to himself. While it is certainly quite normal for us all to suffer from temporary lapses of memory at times, it was unusual for Dad who was a well travelled, well-read, highly intelligent individual to forget something as simple as recalling the name of a place he had visited. Mum had also become concerned that Dad was becoming forgetful regarding daily activities, unreliable and vague.

On the 23<sup>rd</sup> of May 2004 my son had a birthday party and Mum & Dad made the 2-hour drive from Toronto to Sydney to attend the party. Besides the need for a walking stick due to the hip replacement Dad was in high spirits and thought he was recovering well. I remember thinking that he didn't look at all well. He had a ghastly complexion but I put it down to the surgery and assumed that it was quite normal following surgery of that gravity. People have asked me since, "Did you ever have a feeling that it was something very serious early on"? The truth is that I had a strong sense or an "inner knowing" that day that something about

Dad was amiss. That evening, sitting with my husband after we had tucked the boys into bed, heart racing and stomach churning, for the first time I identified and articulated the fear I had felt since seeing Dad that day. "Dad will die one day," I said. I had not experienced any sort of prophetic vision that enabled me to see what would happen in the next 2 months but I had for the first time in my life a very strong sense of my fathers' mortality. Isn't it peculiar? The average person spends very little time contemplating this. We are strangely conditioned to believing that our parents are unrivalled beings that will live on and on, ever present, ever able to guide and direct us. And yet, we know that it is the very nature of human existence that we will all eventually depart, no matter who we are. We must all eventually separate from one another, whether you believe it is only temporary or permanent.

And so it went. Over the next few weeks Mum made regular telephone reports to my sisters and I that Dads memory-loss was worsening and he was behaving increasingly more uncharacteristically. He had also become over-tired and had taken to sleeping for long periods during the day. Mum was also concerned that Dad, who usually exhibited the characteristic of extreme generosity but with parsimonious caution, had begun to buy things that didn't seem particularly well thought out. On the weekends when my sister's families and my own family visited we noticed that Dad, who was a very "hands-on" Grandpa and adored by all 7 of his grandchildren had become the antithesis of the interested, all-fun-loving Grandpa we knew him to be. He was noticeably less interested in his grandchildren and frequently became quite irritable and argumentative, which was extremely out of character for him.

I suppose we were fortunate that Dad certainly was aware of what was happening to him and that he agreed that he was becoming very forgetful, particularly his short-term memory. Mum suggested that he visit their doctor with her and he agreed that it was a good idea. Apart from his memory loss, as the days passed, he also became unable to do a number of daily tasks unassisted and Mum had begun laying his clothes out on the bed each day in the specific order they were to be put on. He was extremely restless in bed at night and had developed an odd kind of twitch in his legs. He was also losing weight rapidly and expressed a concern to Mum that he might have had "some sort of cancer". On June 30<sup>th</sup>, during their first consultation to discuss Dad's condition with their doctor, Mum mentioned that when she and dad were first married, Dad's maternal uncle had a similar experience with memory loss and that he had died after a rapid illness at only 58 years of age. The doctor thought that this was fairly irrelevant and saw no point in investigating the matter further. After performing several basic neurological tests he concluded that Dad may possibly be in the early stages of Alzheimer's disease and advised Mum and Dad that he would arrange for Dad to undertake further, more extensive, neurological examinations. When Mum broke this news to my sisters and I we were devastated. Dementia was the one illness that Dad dreaded. At the time his mother was still living in a

nearby nursing home with dementia and he had frequently remarked that he didn't care to suffer the same fate.

On July 2<sup>nd</sup> Dad had a CAT scan and we were advised that the results were inconclusive and it would be necessary for Dad to have a second CAT scan the following week. Desperate for answers, this was incredibly frustrating for us because each day his condition was worsening. He had begun to shadow Mum around the house and she compared his timid behaviour around others to that of a frightened child.

Despite retiring from Pharmacy several years earlier when Mum & Dad moved from Gilgandra (NSW) to Toronto (NSW), Dad liked to keep active and he occasionally worked as a relief pharmacist in the Newcastle area and for his business partner in Gilgandra during the school holidays. As it was, Dad had been asked to work on July 9<sup>th</sup> at a pharmacy in Newcastle and in Gilgandra the following week. Mum was sensitive to that fact that he simply was not up to this at this stage and delicately advised Dad that she didn't think he was capable of working. After all, dispensing drugs required considerable responsibility. He reacted badly, terribly offended that Mum doubted him but ultimately he agreed not to work.

The second CAT scan was taken and there was no indication of any problem at all when Mum and Dad received the results on July 9th. I remember asking if I could speak to Dad on the phone that evening although I certainly doubted if he would be able to do so. With a little guidance and direction during the conversation though; he coped extremely well. "So I hear you got the CAT scan results"? I asked him. A few seconds ticked by and finally, "Err...yes," he answered. Then after further probing, in a truly characteristically "Dad" way, he remarked, "yep, the doctor said oh what a beautiful brain you have". Amazingly, Dad certainly hadn't lost his wonderful sense of humour at all at this stage!

Mum and Dad were very active members of the Toronto Anglican Church and on a Sunday morning in mid-July Mum advised their parish of Dad's condition. Mum was aware that it was becoming quite obvious and she felt strongly that this was the most responsible thing to do given that Dad would no longer be able to fulfil some of his responsibilities on the Parish Council but more importantly because she knew that others were beginning to worry about him and cared about him. After Mum made the announcement, Dad reacted very badly initially. Dad identified himself as fit and capable, he rarely complained about illness and on the few times I knew him to reluctantly see a doctor, he was terribly ill or injured and by then usually in serious need of medical attention. I suspect now that his being publicly labelled with such a degree of "finality" challenged his strong, competent self-concept. He was embarrassed and displeased about the announcement but later explained that he was not angry with Mum for making the announcement in church. The fact is, he probably couldn't remember being

angry with Mum. His failing short-term memory was preventing him from doing so. What he did remember though was the extent of his love for Mum.

On July 15<sup>th</sup> Mum made a private appointment with their GP. Dad was spending some time with their Priest and so she decided to seize the opportunity to discuss her concerns regarding Dad's rapid deterioration without him. This particular GP had been both Mum and Dad's doctor since their arrival in Toronto 5 years earlier and so he was professionally familiar with them and had prior knowledge of the sudden onset of Dad's symptoms from the earlier consultation on June 30<sup>th</sup>. Unfortunately, what Mum hadn't banked on during the consultation was the doctor's reluctance to discuss another patient – in this case Dad – with a third party. Their doctor was following his ethical nose, committed to patient confidentiality. This was extremely frustrating for Mum who understood the ethical issues the doctor faced but still had hoped given his insight into the matter, that he might view it more as a moral dilemma and not necessarily an ethical barrier that threatened his integrity. Sadly, no amount of cajoling, even begging would change his mind. Instead, he insisted that Mum and Dad make a double appointment and the earliest Mum was able to do so was in 3 days time. At this stage this was like being told to wait 6 months because every day Dad was getting worse.

Later at home, Mum enjoyed the pause, appreciative of the narrow opportunity to take some time to herself before being thrust back into the reality of what had become life with Dad – fear, confusion, agonizing worry. Mum & Dad's Priest dropped Dad home later that night and made a regrettable revelation that Dad had been unable to remember how to get out of the car. Usually a lover of good food, good wine and good company – we were also to learn that Dad had spent a majority of the evening at a men's parish dinner in a state of disinterest. The things that previously brought Dad a great deal of pleasure were becoming increasingly insignificant.

On Monday 19<sup>th</sup> July, Mum and Dad had their appointment with their GP and just when it seemed like things couldn't get worse, they arrived to discover that their usual GP was unavailable that day and they would have to see a locum instead. Mum explained the nature of their visit to the locum and of their earlier consultation with their usual GP. Instead of acknowledging her uncertainties regarding Dad's now very obvious problem, much to her dismay the locum went on to make several derogatory remarks about his own wife, insisting that she regularly accused him of forgetfulness. The consultation concluded and Mum still had no answers.

Fortunately Mum and Dad's neighbour was a neurosurgeon and he was able to expedite the testing procedures their GP had arranged for Dad. Had he not been able to do so, Dad would have had to wait another week before the first of a series of tests began. Given that by now he was unable to remember the names

of objects or the meaning of words; unable to interpret things he saw; unable to make judgements; and unable to "find the words" when he communicated, waiting another week was simply not an option. The following day Dad had a Magnetic Resonance Imaging (MRI) test and an Electroencephalogram (EEG) and by July 26<sup>th</sup>, the results of one of the tests confirmed our fears - there was something quite seriously wrong. The cerebrum frontal lobe was deprived of oxygen; known as anoxia. This particular part of the brain controls our movements, aspects of thinking, feeling and decision-making. On July 30<sup>th</sup> an ECG was taken and July 31<sup>st</sup> yet another MRI. Dad visited his neurologist on August 2<sup>nd</sup> and was placed in Lingard Private Hospital on August 3<sup>rd</sup>.

If the doctors treating Dad had any suspicions that Dad had CJD at that stage, they were certainly not saying so. Even Mum & Dad's neighbour was still not prepared to "suggest" a diagnosis. Dad's neurologist arranged for a lumbar puncture to be performed immediately, the result of which would come back to us later in the week. By now Dad was in a seriously confused state and he was moving very slowly, unsteady, shuffling along in baby steps, entirely dependent on the aid of others to compensate for his loss of balance. On August 5<sup>th</sup> Mum took Dad by car to Hunter Imaging as his neurologist has also asked for an MRA.

On August 6<sup>th</sup> I made a sudden decision to drive to Newcastle to see Dad in hospital. I remember the nurse directing me to his room and as I approached I noticed Mum coming from the other direction. We exchanged a hug and Mum nudged the door open. A consulting neurosurgeon was just finishing up some tests on Dad. He asked Mum to come with him to his office while I stayed with Dad and fed him his lunch. This was the first of many occasions that I would feed Dad and certainly the most confronting and emotional. I certainly had not expected to do so for many, many years to come. Mum returned shortly afterwards and I could tell that she had been crying and was quite clearly distressed but as always, she remained the cool, calm, cookie she is. She didn't speak of her discussions with the neurologist but I knew the news must have been bad. Instead, she approached Dad, gently kissed his forehead and questioned, "how are you darling"? Shortly afterwards, his original neurologist knocked on the door and asked me if I would like to stay while he spoke to Dad about his condition. I agreed and although I can recall certain aspects of what he said it was somewhat like I had stepped outside of myself momentarily, as though I was watching someone else receiving this most devastating of news. After he left I threw my arms around Dad and sobbed. I told him how much I loved him and that I was eternally grateful for everything he had ever given me and done for me. Distinctly he replied "oh my beautiful baby bird". Baby bird - a term of endearment for all 3 of his babies. After I had composed myself I called my sister Lisa and broke the news.

After the news was broken to the extended family, Dad's cousins confirmed that this was the same disease that their father had died of in 1965. We knew then

that our family had been struck by the even more rare strain of CJD - genetic or familial CJD. We were informed about genetic counselling and the possibility of undertaking predictive testing to see if we had inherited the mutant gene. The neurologist has asked Mum how she would like to have him cared for now in this palliative state and she chose to have him taken to Toronto Private Hospital with the hope that he could be taken home for short periods through the day with help from friends and family. This would not happen as his condition was deteriorating rapidly before our eyes.

On August 12<sup>th</sup> Dad was transported to Toronto Private Hospital, which was conveniently close to Mum & Dad's house. My sisters, mother and I spent each day and night with him, sleeping on fold up beds in his hospital room, aware that it was just a matter of time. Days, weeks - we didn't know but knew that it couldn't be much more than that.

Dad began to have terrifying hallucinations. On the evening of August 13<sup>th</sup> I walked into his room sing-singing "hello Daddy" as Mum & my sister Susan were helping him back into bed. He stared me down and became very distressed, questioning "Daddy"? He thought I was something else, a demon, a monster, I don't know but to him I wasn't me and I honestly thought he might die of fright that night. I have never seen another human being more petrified and this particular hallucination went on for hours. We did our best to calm him and convince him that what he was seeing wasn't real and that his mind was playing tricks on him. Earlier that evening Mum and Susan witnessed another distressing incident. As Dad sat on his bed he began to grab at his pyjama pants as though something on his skin was irritating him. They pulled his pyjama pants down to investigate and were shocked to find every hair on his legs sticking directly out at 90 degrees and the skin on his legs had taken on the texture of sandpaper. This lasted a while and was extremely distressing for Dad.

The GP visited the following morning, August 14<sup>th</sup>, and Susan and I requested a private consultation with him. We described the events of the previous evening and I begged him to do whatever he could to ensure that Dad didn't have to suffer that way again. I told him that we had always promised Dad that we would do whatever we could to ensure this never happened. The GP agreed to put him on an IV combination of Morphine and Midazolam, a drug that is used to produce sleepiness and relieve anxiety. My sisters and I contacted our husbands and instructed them to come to Toronto immediately with our children to see their Grandpa for the last time. Honestly, we had not expected Dad to live into the next day and assumed that the IV combination may have been enough to set him free. Consequently getting our families to Toronto had become a matter of urgency.

How do you explain to a child that after today they will not see their grandfather again? How do you tell them his body is broken and can't be mended? I used whatever spare time I could in the next few hours rehearsing in my mind what I

would say to my sons and later when they finally arrived in Toronto everything I had planned on saying to them seemed unnecessary. They were too little to comprehend their loss and it seemed more important to simply allow them to be with Dad. And so, 7 children ranging from 11 months to 15 years filed into their Grandpa Graham's hospital room that day to give him one last cuddle. At that moment, as I watched each child crawl on top of Dad and kiss him goodbye I have never felt more cheated, more exposed. Outwardly as we - the adults - watched on we must have all appeared to others to be very composed. Inwardly we were dying.

Much to our surprise Dad clung to life for over a week after the IV was introduced. He was much more settled and slept for most of the day. At sunset though the hallucinations often returned but we discovered that playing soothing new age music and singing his favourite hymns and tunes was a great comfort to him.

Around August 19<sup>th</sup> Dad had his last meal. Swallowing was now almost impossible and he existed on thickened juices and thickened lemonade we fed him for a few days. We relieved his thirst to the best of our ability by regularly spraying his mouth and using swabs to clean his tongue. As a family we decided that we would not give him fluid intravenously as this was simply going to prolong his suffering and we knew that he wouldn't have wanted this. We knew that we could expect renal failure within days.

Dad developed pneumonia and was having difficulty breathing. On the evening of August 21<sup>st</sup> overwhelmed with fear and uncertainty as Dad struggled for breath, Mum approached the night-nurses for assistance. Sadly the attention and care we'd come to know from the excellent regular nursing staff was not offered with casual Agency staff over the weekend. Mum was anxious because a plan of care had previously been established by the doctor and general staff to ensure Dads comfort. It seemed obvious that the casual staff were unaware of what they were facing. Siphoning fluid from his lungs offered some temporary relief and they eventually agreed to administer the "cocktail" which had been ordered 3-hourly if necessary. Dad had a restful night and became much more settled.

In the early hours of August 23<sup>rd</sup> Dad's breathing changed, becoming very shallow. I would describe it as similar to a young child's short gasps after a serious crying bout. We knew that it would be soon and I remember that I felt a strong sense of the presence of Spirit or God with us as we prepared for Dad's passing. We held hands around his bed telling him we loved him and that it was okay if he felt had had to go now. At 10 past 4 that morning, encircled by love and only the love of "his 4 girls", Dad found peace.

It is almost exactly one year to the date at the time of writing. Strangely in many ways it seems like a lifetime ago and at times I feel like only days have passed. My sister Susan has remarked since that often she catches herself anticipating

his return as though it's all just one big joke. In he walks straight through her front door. He smacks her a cheeky grin and sniggers "ha - gotcha"! Me though, I spend my time dreaming - vividly - about him, "flying around with him", I tell my kids because this is what I believe. I feel his presence frequently and I know he is guiding me through the ever-present obstacles of life that cross my path. I am a firm believer that everything happens for a reason and I believe that Dad has gone ahead early, at only 63 years of age, to further the awareness of CJD in Australia. Dad donated his brain tissue to science and I know that his death has not been in vain and from this donation he has been able to contribute considerably to the cure jigsaw.