

PREFACE

Please be advised that the following is not a unique account of one person's death due to Creutzfeldt-Jakob Disease (CJD). It is experienced by victims and their families and friends 300 to 400 times a year in the United States. It is extraordinarily rare, about a one in a million diagnosis. A finer cut since, while not exclusively, the disease generally strikes persons over 50 years of age is 1 in every 7000 deaths annually in the United States. It and associated neurodegenerative diseases are classified as a group as Prion Disease.

"Tom, this is not MS. This is a once in a career diagnosis for a Neurologist. It is an extraordinarily rare disease for which there is no treatment or cure," said Dr. Leonel Limonte as he was advising me of his suspicion about what was causing Sherry's sudden onset of rapidly progressing dementia. He had already made arrangements for her transport from Carlsbad, NM Medical Center to Texas Tech Medical Center, where he serves as adjunct faculty, for a thorough confirmation of his suspected diagnosis, Creutzfeldt-Jakob Disease (CJD). This conversation was on January 4, 2018.

Sher started exhibiting some subtle changes in behavior a few days before Christmas 2017. Having been her care giver for 25 years since she had been diagnosed with remitting relapsing Multiple Sclerosis in 1993, I was becoming concerned that she may be relapsing. The symptoms of MS are wide ranging and one, loss of mental acuity, had robbed Sher of her career in 2007. She loved her job but when she recognized that her memory and ability to sharply recall, retain and analyze facts had eroded, she grudgingly enforced on herself a disability retirement. A perfectionist and driven professional, she had no other choice and, frankly, it was an act of courage and sacrifice.

On December 26, Sher awoke confused and disoriented failing to recognize familiar things, where the bathroom was, how to dress and some others I can't recall. This was a person in severe crisis. I called Dr. Limonte's office that morning and got an appointment on the 28th upon his return from a holiday trip. By the 28th, Sher had lost the ability to communicate verbally (Akinnetic Mutism). Sher and Dr. Limonte had a five year physician-patient relationship treating her MS. That relationship was special and I am convinced that Sher was in love with this handsome, classy gentleman. I'm equally sure that the good doctor cared for her deeply. He had asked her to fill a vacancy on the Board of Directors of Carlsbad Medical Center (CMC), where he was Chief of Staff, but she declined for the same reason that forced her early retirement.

On December 28, 2017, Dr. Limonte did a standard cognitive examination which pointed out evidence of what appeared to be a rapidly progressing dementia. He sent us immediately to CMC with orders for an MRI, EEG, full Blood Panel and Spinal Tap. My understanding was that we were taking the approach to find out what it is not to give us a starting point from which to figure out what it is. He also told us to stop her daily MS medication. The next three days were reasonably peaceful with Sher sleeping a lot but also visibly continuing to worsen in terms of caring for herself. As she declined, my ability to provide a safe level of care was stretched to its limit, and I was not some guy who knew nothing about the care giver role. I was damn good at it with 25 years of practice.

On January 1, 2018, Sher awoke and was unable to get out of bed and was unable to walk, something that she had battled in the 90's and early 2000's. Symptomatic medication specifically targeting ambulatory issues associated with MS had been developed and she worked her way out of a wheelchair confined life style to a daily regimen of a 1 to 2 mile unaided walk around our neighborhood. We kept her wheelchair for moving through airports and "just in case". This day "just in case" had happened. After I helped her with some hygiene issues, I gently lifted her from the bed to her candy apple red ultra-light chair and wheeled her into our sun room with a view of her pool. She smiled.

Dr. Limonte had instructed me to call him if I sensed that there was any significant change in her condition. He was in Miami visiting his family for the New Year and when I called that morning I caught him at breakfast with his family. After a brief description of Sher's situation, he instructed me to get her to the medical center emergency room and that he was contacting them to be ready for her immediate admission. He indicated that he would be returning to Carlsbad the afternoon of the 2nd and that he would see us immediately upon his arrival. I packed a bag for Sher and then gently transferred her from the

chair to our SUV and we left for CMC in less than 30 minutes. This was the last time Sher saw our home. When we arrived at CMC after the 5 minute ride (small town) a team was waiting for her and I transferred her care to a group of excellent medical professionals. What followed after Sher was settled in her room was another EEG, an enhanced MRI and IV fluids.

The afternoon of New Year's Day I started communicating with the family. For 30 years Sher and I were the family most distant from the rest. I called our daughter Beth, son Bob and my sister Sarah together in Bowling Green, KY so that I could let them know that Sher was experiencing a bit of a crisis and that she was in the hospital for diagnosis. They were shaken but I told them just to go to standby mode and that I would provide daily updates. I called Sher's sister Karen, a career ICU Nurse, in Sidney, OH and described what was going on. She was shaken as well and I asked her to tell Sher's youngest sister Marcia what was going on. And then was how to approach Sher's 87 year old mother Patsy? Karen and I agreed that she would handle that communication as we had those times in the past when Sher relapsed. Karen is one of those wonderful nurses who did not choose her profession, she was chosen. I then called my brother Bob in Bedford, VA. He had spent his career as a Nursing Home Administrator and Corporate Executive in the Health Care industry. Shaken as well, Bob was also on standby and ready to help me plow through any process required to assure Sher's care.

Next, I called some of Sherry's closest local friends in Carlsbad. First on scene was Sher's closest friend, Lisa Watson and her husband Kerry. Lisa credited Sher as her mentor and role model and attributed her successful career in part to Sher's influence. Next was Keith Nelson our longest duration friend in Carlsbad having raised our children together since 1988. Then followed a gentle flood over the next few days of dear friends and former colleagues just to briefly sit with her and share stories or just hold hands. I never felt overwhelmed.

On January 2, 2018, Dr. Limonte arrived. When he entered her room he greeted Sher in his wonderful baritone voice, and charming Cuban accent, "Sherry, what is going on here?!" She greeted him with a coy, flirtatious smile further evidence of her love for her doctor. He examined her and told me that he had already reviewed the initial MRI, EEG and blood work. He indicated to me that his first notion was to eliminate MS as a cause for her condition. As I look back, I'm pretty darn sure that this highly skilled, recognized medical expert had more than just an inkling of what was at play.

So back to the first paragraph, on January 4, 2018 Dr. Limonte and I had the aforementioned conversation about his suspected diagnosis. At 0400 on January 5, Sher was transported via ambulance to the University Medical Center at Texas Tech University. I followed them to Lubbock, 188 miles a trip that was very familiar, in our family SUV.

When Sher and I arrived at Texas Tech on January 5 she was greeted by the Chief of Neurology, several specialty Department Chairs, the Director of the MS Clinic, several Attending Physicians, Residents, Medical Students and Nurses. Sher's best friend Lisa Watson arrived that evening and stayed by our side throughout our time at Tech. This was an extraordinary team. Batteries of tests, scans and EEGs followed as there were visible changes in her condition daily.

The afternoon of January 8 the same team that greeted us entered Sher's room. The Chief of Neurology then proceeded with utmost care to explain in detail their findings and that Dr. Limonte's diagnosis was confirmed. I had steeled myself the best I could leading up to that moment determined not to fall apart. Surprisingly, I succeeded. The only relevant question I could think of was, "I know that you folks are not gods, but can you give me an idea of how long Sher may suffer with this hideous thing?" The answer was, "days to weeks." That one pierced my armor over the heart but I did not flinch, I had a job to do. I thanked the team and each of them embraced me as they left the room, several with tears in their eyes.

When we were alone, I sat on the edge of her bed and held her hand. The essence of the one-sided conversation that we had then was calm and frank. I told her of my enduring love for her and that we were faced with something that was going to take her life; that I would be with her every step of her final fight; that her courage was incredible; and that I would make sure that the end would be peaceful and pain free and that she would be surrounded by those she loved. At one point she just closed her eyes. I kissed her cheek and called the nurse. I then took a long walk around the huge medical complex to collect my

thoughts and reach some level of composure. It was time to calmly make a plan. And I remembered what one of the wisest men I have ever known, brother Bob, told me about plans. "If you want to hear God laugh, make a plan."

During those few days I had established a text message group of family and friends that I titled "F-TROOP" (the old sitcom from the 60s) as a means of updating them on important information. On the evening of the 8th I called the family and told them about Sher's diagnosis and prognosis. I also informed F TROOP via text.

On January 9, 2018, at 1000 Sher was discharged and transported back to the Transitional Care Unit at CMC via ambulance during an intense West Texas sandstorm with 100 foot visibility and tumbled the size of calves pummeling both vehicles. Not in the plan and I heard Him laugh. Upon arrival Sher was readmitted and settled in. Dr. Limonte was there within an hour. We discussed Sher's transfer to a local Skilled Nursing Facility, Landsun Homes a half mile from CMC. I made those arrangements the following morning.

The next day our daughter Beth arrived to be with us for a couple of days and stayed with Sher for a "girl's sleepover". We held each other as we always had and privately cried together. Lisa was frequently there to hold us up and Kerry and Keith made darn sure I had their strong shoulders on which to stand when I needed it. On the 10th I visited my physician and told her what was going on and she prescribed a mild antidepressant to help keep me level. That proved to be a very good thing. Beth left on the 11th and our son Bob arrived that evening. My guess is that that was a sibling hand-off of Dad. Bob stayed through the 13th and both children got to spend quality time with their Mom while she was able to interact a bit. They both have their mother's strength and courage and both of our caring hearts.

During one of Lisa's visits while Bob was still there she was teasing Sher about how good a dancer she was. We were sitting in a lounge area on the ward. One of our family wedding traditions during receptions is that all the women in the family get a glittery wooden spoon to use as a microphone and dance with the bride to "It's Raining Men" by the Weathersgirls, followed by "We are Family" by Sister Sledge. Lisa found "It's Raining Men" on her phone and played it. Sher immediately started to dance in her wheelchair with Lisa sitting beside her dancing with her. They had a ball and Bob captured a video of the performance on his phone. That evening when Dr. Limonte visited we played it again. It and "Family" became Sher's anthems for the rest of her battle. Something inside her was telling this damned disease that, "while you might you might kill me, you Ph@****G disease, it's not going to be easy and it's going to be one helluva fight!" Sher had the face of an angel, the genius of a Nobel Laureate and the mouth of a sailor, but only when necessary.

On January 15, 2018 a bed opened on the terminal care wing of Landsun Homes. Sher was discharged from CMC that morning and was transported by ambulance to her room there with a window overlooking the banks of the Pecos River. The staff there was excellent top to bottom. The Chaplain visited immediately after Sher was settled in. She helped me get in touch with Lakeside Hospice and I engaged them that afternoon. The routine set in immediately and Sherry's charge nurse took one look at me and said, "We have this! YOU go home! Get yourself something substantial to eat. Call a friend over and have a couple of beers if you want to. Get in bed by 2200 and I don't want to see you here until breakfast at 0700. Got it?" Me, "Yes Ma'am!"

On the 16th I greeted a new day resolved to be smart in taking care of myself and conserving energy when I could and expending it when necessary, having no idea about how long Sher would need me. I left for Landsun at 0700 as ordered. When I arrived the staff updated me that Sher had had a peaceful night and had already eaten a light breakfast. Her hair was in a beautiful French braid and we quietly spent the morning together. Around lunchtime Lisa and a few of Sher's close circle of friends visited briefly. We did a lot of hugging and I assured them as best I could that her care was extra special. We played "Raining Men" several times and the dance party was on. For the next two weeks each day followed pretty much the same pattern. Dr. Limonte visited daily, as did the Hospice nurses. Sher's decline was gentle and persistent. I informed F-TROOP via text and phone about how things were morning and evening.

Sher's close circle of friends spontaneously started delivering meals to our refrigerator at home so that I did not need to cook. They were angels. This group for years had done Ladies Night Out together monthly and two to three times a year Sherry hosted cookie decorating parties at our home during holidays. I was the bartender for the cookie parties that were all day affairs starting with Bloody Marys, then Margaritas, ending with Midnight Expressos. I used to tease them that they were "The Coven". Nothing could be farther from the truth, angels all.

On January 30th, when Dr. Limonte visited Sher appeared to be in rapid decline. I asked him if I should let the dispersed family know to make arrangements to travel to Carlsbad. "Sooner is better than later," was his response. I made those calls to our children and Sher's sisters and they mobilized quickly. Bob arrived late the next day. The day after the sisters Karen and Marcia arrived with Sher's mother Patsy and one of our nieces, Amy who had spent a summer with us interning at WIPP during her undergrad program at Ohio State. Beth arrived the following day and I had a house full.

All of them were concerned that Sher may not recognize them. I am convinced that she did and for the next couple of days the dance party was on again. We set up a round the clock rotation that assured we were by her side 24/7. We held each other up.

On February 5th it was evident that Sher was laboring and not responding well. Hospice had begun morphine and anti-seizure medication a few days before. I was sent home at 2200 to get some sleep if I could and Bob had the watch. At 0300 Bob called me and said, "Dad, it's time to get everybody over here." I woke the rest of the family and we were there in fifteen minutes. When we arrived the end was approaching and we surrounded her bedside. We whispered to her softly and each of us in turn would take a minute outside the room for composure. The Hospice nurse had told me that the last sense we lose before death is that of hearing. As her breathing became more labored, I continually whispered, "It's OK to let go dear. Let your spirit be free. White sandy beaches, white sandy beaches." I was gently holding her pulse. As she breathed her last, I felt her last heartbeat, kissed her gently and nodded to the nurse. Sher was pronounced at 0436 on February 6, 2018.

We held onto each other and cried. Patsy suffered the most having been there for her first child to precede her in death. That is not the natural order of things.

What followed then was pretty a standard operation after a person's passing. The aides put Sher in clean clothes and did her hair, the funeral home was called, we each spent some private time by her side, I asked Beth to cut a couple of braids of her hair and each of us started making calls to the extended family and friends that the end had come. The hearse arrived just as dawn broke. I thanked all of the aides and nurse for their gentle care of Sher. The hearse left the nursing home and after we had composed ourselves the family went to the house.

Within an hour, who should show up at the house was Lisa and baskets of pastries from the Blue House Bakery. As time passed, a gentle stream of neighbors, friends and former colleagues stopped by paying their respects and by late in the day Sher's wine collection came in handy. My sister Sarah arrived from Kentucky around noon. The 6th had pretty much turned into an impromptu Irish wake. That's how small towns in southeast New Mexico work. That evening's dinner turned into a pot-luck with enough food to feed a regiment. Hugs, tears, food and laughter were the order of the day. We finally retired for the night and I, while grieving, welcomed a strange but welcome sense of peace.

The next couple of days were devoted to family time. Sher never wanted a funeral and wanted to be cremated. All she wanted was for us to have a great party sometime after her death whenever that may have been. Sher's family prepared to return to Ohio on the 9th and my brother Bob arrived from Virginia that afternoon. Bob and Beth went home to Kentucky on the 10th and Sarah returned to Kentucky on the 11th, and then it was me and my brother Bob.

Bob and I had some great quality time together. While we were talking in our garage sitting in Sher's wheelchairs I was assessing some next steps to bring order to life moving forward. This wise man said to me, "Tommy, you are a committee of one. You make all the decisions and there is no such thing as a bad one." After a while I looked at us in the chairs and thought that her good one should be put to good use.

As luck would have it Bob told me that an old friend of ours, one of fraternity brothers and fellow Viet Nam Veteran, had recently broken his and need one. My first decision opportunity! The next morning we took the candy apple red ultra-light to UPS and shipped it to his brother Al. Sher was going to have an impact. Her spirit smiled.

In a few days Bob returned to Virginia and I went about the process of normalizing a routine and dealing with the stages of grief.

As time passed with the wonderful support of distant family and local friends the healing process started to take hold. We had planned in October of 2017 to relocate to Bowling Green, Kentucky in January 2018 to join the family there expressly to close family ranks around Beth and our grandson Ethan. Our son-in-law, Brent, died in an automobile accident in March 2017. We had purchased a home there that I sold during Sher's CJD battle not knowing how long I would have to provide for her care. Early in April I made a house hunting trip to BG and found a beautiful home on the first day there. I could then start the relocation process fulfilling a promise to Sher that I would follow through with our plan.

Late in April, Lisa served as the hostess for Sher's party at our home. Sher's pool was open and spotless. Lisa's family set up our backyard just as it had been two years before for Bob and Mindy's wedding and reception. The caterer did a great job on the food and we had plenty of refreshments available for children and adults. It was a family affair. Most of the children swam along with some adults. The air was filled with the sound of children splashing and laughing with 60's and 70's music in the background. More than 100 people attended during the afternoon into the evening. Only a core group remained around dusk, mostly the Coven and their families and some other close friends. The last three songs that we played were "The Parting Glass" by Celtic Woman, "It's Raining Men", and "We Are Family". Her spirit smiled and she danced.

In June 2018 I moved into my new home in Kentucky surrounded by my family. I settled in and became a full-time Poppie, Dad, Brother and Uncle. Her spirit smiled.

Early in December 2018 I accidentally became aware of the CJD Foundation. I had seen on the internet that a young man in the NFL, Byron Jones of the Dallas Cowboys, had dedicated a pair of game worn cleats to the memory of a young woman, Diana Hunter, who had died as a result of CJD. They were being offered through the NFL Auctions Program called My Cause My Cleats. The proceeds from the auction would go to the CJD Foundation in Diana's name. I decided then and there that I would win those cleats whatever the cost. I figured if Sher could not beat that damned disease I could at least throw a bucket of money at it. As the on-line auction progressed I was in the hunt. On Christmas Day the pace had slowed a bit and it seemed that there were only a handful of bidders still in the game. I then registered a high bid that folks were going to have to chase to beat me. Every bid thereafter was immediately countered by my high bid. At midnight December 30th mine was the prevailing bid! I had won! Her spirit smiled but I distinctly heard a whisper in my ear that said, "You crazy old man. What are ya, 12?" I smiled. I had met the enemy on ground upon which I could fight and help however I could in the effort to find a cure for CJD.

Sometime in early January I contacted Debbie Yobs, President of the CJD Foundation, and introduced myself as the guy who won the cleats. She thanked me and asked what attracted me to the CJD Foundation. When I told her that Sher had recently died as a result of CJD she welcomed me to the club "that nobody wants to be in." We talked a while. Her husband died due to CJD some years ago. I asked Debbie if I could get in touch with the Hunter family to wish them my best. She said she would get in touch with Vickie Hunter and she felt sure that Vickie would reach out to me.

A few days later Vickie Hunter and I connected by phone. We had a wonderful conversation. I've since gotten to know Vickie and Clarence Hunter through frequent phone calls and finally got to meet them in July at the Annual Family Conference in D.C. I have adopted them and they me. They are amazing people. So are Debbie, Lori Jo Nusbaum and the staff at the foundation.

Speaking of amazing people, meeting 200 other families who had lost loved ones to CJD at the Family Conference was as close to a life changing experience as I can get at my age. Just when you thought you

were alone in your grief; just when you thought it was a lonely fight with one measly bank account and a 401K: just when you thought yours was the only story, hundreds of amazing people get poured into your life. I was overwhelmed.

Along with those families were the dedicated scientists who are in the research trenches and fighting holes putting in the work pushing research forward. All research is an evolutionary process, generally not a revolutionary one. One day these are the great minds who will give us our own BIG BANG. There will be a revolutionary event and we must provide those researchers every necessary resource to reach that outcome.

In closing, as I have reflected on the entire sequence of Sherry and my life together for 47 years and 8 months, I am struck by a few things. In 1993 Sher's MS diagnosis drove us closer together. We were in fact and deed a couple with great courage and determination to battle MS every day. Our love was immense. Our immediate and extended family including friends was how we were most blessed. Sher's legacy is our children and grandchildren. Her fighting spirit, noble bearing, and determination in all things marked a remarkable life. In the end when the CJD ambush struck, our metal was tested. Sher died as she lived giving no quarter and asking none. I have described her final fight as her "Alamo", a hopeless unwinnable fight. My role in the final fight was to stand at a distance with my hands tied behind my back hopeless and powerless to save her. The only thing I could influence was her comfort in the end. I pray I succeeded.

Creutzfeldt-Jakob Disease Foundation (CJD Foundation)

What is CJD?

Creutzfeldt-Jakob disease (CJD) is a rare, rapidly progressive neurodegenerative disease, one of several Prion Diseases caused by prion proteins that misfold in the brain. There is no treatment or cure and the disease is invariably fatal.

Mission

The mission of the Creutzfeldt-Jakob Disease Foundation is to support families affected by Prion Disease, raise awareness, and support medical education and research. We carry out this mission through:

- Family support including a 7-day HelpLine, referrals, support groups, and teleconference speaker series
- Education and information for families, caregivers, medical professionals, and funeral professionals
- Advocacy with political representatives and public policy makers
- Annual Family Conference that brings together affected families and prion disease experts
- Family Workshops held around the country
- Collaboration with scientists, clinicians, medical centers, health authorities, professional organizations, and international patient associations
- Communication of current research and Foundation activities via newsletter, social media, emails, and family teleconferences
- Research grant program, with international awardees selected by our Scientific Advisory Committee

Philosophy

The Foundation's philosophy is to be proactive by establishing collaborations and linkages, communicating with family members, researchers, physicians and political representatives and informing the broader community about CJD.

This Cook Book

In January 2012, Sher got the idea to compile a Family Cookbook. She wanted to do it for the family and to keep her mind occupied and exercised in her struggle with mental acuity. It was the beginning of a five year process. She compiled family recipes, recipes from great chefs (Julia, Martha, Ina, Emeril and others), recipes from various cooking magazines that fit her taste and recipes from friends (mostly the

Coven). She diligently tried them all and would modify them to her standards. It was a great process to watch.

The book went through a series of revisions over the five year span and she finished the last revision in early September 2017.

SPOILER ALERT: While I was editing this version, I found headers, footers and a cover that she developed in January 2012. If you care to read the book cover to cover please help yourself and ENJOY! If you would like to see the essence of the fighter that she was, go to the back leaf of the book first and there you will find my SHEroe! Either way her spirit will smile.

I bid you peace.

Tom

CURE CJD!!!!!!



