CBGD
Caregivers Report
Corticobasal Ganglionic Degeneration

Compiled by Alan G. McIlvaine
March, 2000

CBGD

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The "CBGD Caregivers Report"
may be downloaded from the Tornado Design
web site at www.tornadodesign.com/cbgd
For those without Internet capability who are
concerned with CBGD, a printed copy like this
may be obtained at no charge on request.

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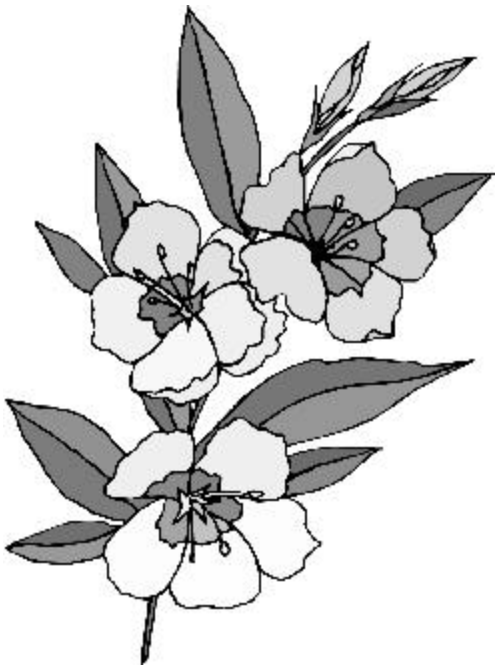
Donations sent to
any medical research facility that has an active
program for these rare brain diseases
will be appreciated.

IT WILL HELP THE CAUSE

Dedicated to my wife Barbara
and all those other stalwart souls who have
braved the ravages of CBGD.



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Special thanks to:

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for encouraging me to complete this report and to all of you who cared enough to contribute to it so that others may benefit from your experiences.

Alan G. McIlvaine
Caregiver

Foreword by Richard J. Caselli, M.D.

Corticobasal Ganglionic Degeneration, or CBGD, is not a common illness. It is difficult to pronounce, let alone to understand. In a world full of neurologic disease oriented foundations, such as the United Parkinson's Foundation, Alzheimer's Association, Multiple Sclerosis Society, and Amyotrophic Lateral Sclerosis Society of America (to name only a few), there is no, nor is there likely to soon be, a "Corticobasal Ganglionic Degeneration Organization." Yet this disease exists. It impacts real people and real families. It presents overwhelming challenges to all it touches. It is a profound condition that effects our ability to communicate through spoken and written word, and gesture. Yet it typically leaves our comprehension and insight intact. A common theme which emerges for the patient and their family is isolation. The patient can feel progressively cut off from their ability to communicate with the outside world. Their family feels isolated from the medical and caregiver community because few people understand this disease, and there are few support groups or organizations dedicated to it.

This remarkable monograph is a caregiver's attempt to fight back against the isolation of CBGD. Alan Mcllvaine shares generously, the experience of his wife Barbara (who was my patient) and himself in an effort to break that isolation. He is a humanitarian magnet who has attracted other caregivers to share their personal stories of CBGD and what we are given, then, is a collection of true case histories, as personal as they are medical. The goal is to bridge that isolation. Those of you who suffer with, or are close to someone who suffers with CBGD are not alone. We know what you are experiencing. Alan Mcllvaine, Darcy Croissant, Sandra Till, Robert Hall, Louise Davis, Sandra Roberts, Theresa Roberts and a caregiver who wishes to remain anonymous share in these pages their experiences with CBGD. Additional contributions come from, Dr. Geri Hall, Dr. Bradley Boeve, Attorney Kevin McFadden and Financial Planner Tom Mills. General Douglas McArthur taught us that greatness is defined by ordinary people confronting great challenges. These are very great people.

To Alan and his late wife Barbara, to my other patients and their families with diseases I lack the power to cure, I would like to say, that their illness and how they handle it matters enormously. The world is watching. The world is vulnerable to the very illness you are now confronting and the world is afraid of it. Teach the world to be brave. You have that opportunity.

Richard J. Caselli, M.D.
Scottsdale, Arizona

Caregivers Case Histories

My Wife, Barbara, Had CBGD

From Alan G. McIlvaine, Scottsdale, AZ

This is a story of the small victories and distress that we experienced in caring for Barbara through the relentless assault of CBGD. It is not a guide on what to do but rather, since this disease treats everyone differently, different symptoms, how we coped with each new devastating symptom. Hopefully it will give others some ideas on what to expect and how to handle it.

Some years before our introduction to CBGD, Barbara and I had decided that neither of us would be comfortable in a nursing home if we became ill enough to warrant it, and the other would do whatever was in his/her power to maintain the ill one at home. I know that Barbara would have felt deserted and would not have received the tender loving care in a nursing home that she did in our home. If you have a close family and there is any way possible to keep the patient at home with adequate care, I strongly urge you to do it. You will all feel better for it. I realize that sometimes it is not possible in which case the caregiver should devote as much time as possible to overseeing that the care being provided is appropriate, adequate and loving.

I have also learned from letters received from others concerned with CBGD and from our own experience that this disease is very difficult to diagnose.

In October of 1992 Barbara and I went back to Eastern Pennsylvania, where we had grown up, to attend the 50th anniversary of my high school graduation. Both of us had attended Cheltenham High School in Elkins Park, Pa. She was a year behind me so her reunion came up in 1993. We visited friends and relatives in the area and then drove up to Danbury, Connecticut where we had lived for about 10 years before our return to Scottsdale, Arizona in 1987. In Danbury we visited with old friends and on one occasion played golf with Ellie and Coleman London.

After playing golf that day, Barbara, with a concerned look on her face, said that she had had trouble gripping her golf club correctly. She had to concentrate abnormally to place her hands correctly on the club. This was the first symptom that we recognized of the early stages of CBGD although we did not know what it was for another six months.

To give you a time frame, most of what follows started in mid 1994. However her first symptoms were noticed in Oct 1992 when she realized that she had to concentrate to

make her hands, particularly the left hand do what it was supposed to do. But, it was Oct 1991 that we first noticed her carrying a filled glass on a tilt. Her hands became progressively worse with tremors when ever she attempted to use them, particularly her left hand. It wasn't until early '94 and numerous doctors that the doctors at Mayo diagnosed the disease correctly although as early as December '92 they said it appeared to be CBGD. She died Jan. 20, 1997 at 71 years old.

In spite of the onslaught of CBGD, we tried to enjoy life to its fullest as long as we could. In May of 1994 we took a two week trip to Hawaii including a cruise of the islands. In August of 1994 we exchanged houses for three weeks with good friends in England. We took our grandchildren, Chris and Brook, our daughter Barbie's children, with us. Barb had a bit of a problem getting into busses and trains but Chris just got behind her and picked her up and placed her in the bus or train as though it were a normal occurrence. Brook handled the cooking when we weren't out for a meal and the house chores, which were not much of a burden. At Christmas time in 1994 we flew to St. Louis to be with Barbie and her family, who lived there at the time. All during 1994 Barb could barely use her left hand for anything. We went to restaurants fairly often and I would instruct the waiter to cut her food into bite size pieces before he brought her plate to the table.

Barb was an excellent bridge player. In fact she was certified by the American Contract Bridge League as a Director to run duplicate games and she had a ton of master points. She continued to play competitive bridge until about the end of 1994 and played in neighborhood and senior center games throughout 1995. I had made a card holder for her and the later part of 1995 she needed assistance arranging her cards. This convinced her she could no longer play and it was a bit emotional when she had to tell some of her bridge partners. I point these things out only to demonstrate how quickly her condition deteriorated.

Barbara started falling before most of the other symptoms showed up. She would just suddenly lose her balance and fall, usually backwards. While she was still quite mobile and seemingly good shape she took several vicious falls. Once while she was walking around the neighborhood for exercise by herself she fell and hit her face and left arm on the curb. Fortunately a neighbor saw her fall and rushed to her aid while her daughter called 911 and me. She had a large gash right at her left eyebrow and her arm, face and knee were bruised and skinned. There also was a slight skull fracture at the eyebrow line. The doctor at the emergency room did a great job stitching up the eyebrow wound. After about a month it was barely discernible. Barbara said that as she came around the corner the sun caught her right in the eye and she could not see, resulting in her tripping over the curb. In retrospect, we know this is not what happened, she lost her balance. Another disastrous fall was when she fell backwards off about the third step in a tri-level friends home. She did a back swan dive onto a ceramic tile kitchen floor, flat on her back and her head whacked the floor in a sickening

thud. It turned out that she had fractured 3 vertebrae between her shoulder blades. This hurt her for the rest of her life but she rarely complained about anything. She showed very little evidence of osteoporosis so her bones were not in jeopardy from minor bumps. We soon realized that we could not let her stand or walk alone. Someone had to be close enough, usually holding her, to prevent further falls. She still had a few backward pitches. Barbara was small to start with, 5' 4" tall and about 130 lbs., so it was not difficult to hold her or pick her up. She lost weight rapidly over the last 10 months-down to 80 lbs. She also would miss the chair, usually to the left, when trying to sit down. I finally had to tell her that she could not get up and walk without someone with her. It was too dangerous. At first it was OK for someone to just hold her elbow and keep her steady but also be prepared to grab her if she started to fall. The control of her walking increased to where we walked behind her and with hands holding her on the side of her ribs, practically carried her. This progressed to where she was totally wheelchair bound to move around the house.

We leased a wheel chair, the type they call companion chairs because someone must push it, there are no large side wheels for self-propulsion, because Barbara couldn't use her hands or arms. Also, the chair was narrower with only 4 small wheels. Didn't wreck the house as much as a larger one would have. Medicare paid for the lease of the wheel chair through Osco Drug where we got it.

We then bought a reclining chair that had a motorized lift that would lift her to a standing position and vice versa, she would stand against it and be lowered to a sitting position. In spite of her light weight it was tough on my back and our daughters back to have to pick her up many times a day. The chair came in handy. When we asked if that could also be charged to Medicare they said no, because we already had a wheel chair. It would have been better to charge the recliner chair to Medicare because it was more expensive than the wheel chair.

All during this time I would occasionally take her, using the wheel chair, food shopping or a few times to dress shops to just look at the new styles. Quite frequently I would take her over to our daughters house for a meal or several hours visiting the family. After my operation when I could no longer manage and Barb was becoming even more dependent, Barbie's husband, George and her son, Chris, both big strong guys, would pack Barb in their van with all necessary chairs and equipment and take us over to their house for an afternoon and dinner. Granddaughter Brook and grandson Chris would help their Mama with eating and what ever else was necessary. I believe the last time was January 16, 1997 with oxygen tank, wheel chair and all.

There was a period when she had to go to the bathroom constantly, sometimes as often as every 3 minutes for a period of hours. By this time she could do nothing for herself.

Barb's left arm became practically unusable by the end of 1993. Her left hand had tremors when ever she raised it and sometimes her entire body appeared to shake. She said that she felt shaky at times. Barb could not do things that she could do a month earlier, for instance, pouring juice from a pitcher or putting instant coffee into a cup. Putting on jackets or sweaters was impossible. She needed assistance getting dressed. She could, with great difficulty, take hold of something with her left hand but then she could not let it go. It had to be pulled from her grasp. She also seemed more susceptible to temperature variations and was frequently fatigued. Barb's left hand later, 1995, became rigid and sore. It was frozen against her breast with the hand clenched and the forefinger sticking straight out. We did not think about Barb's rings until it was too late. We had to have them cut off her left hand fingers. The local fire company rescue squad did it competently and cheerfully.

Finally in late 1996 Barb was given Botulinum toxin shots in the muscles of her left arm and hand. This eased the tension in the arm and hand and lessened the pain but of course, could not improve function. When her hand became frozen in a clenched position it was necessary to keep the skin on her palm healthy and from being pierced by her fingernails. It was difficult to keep the nails trimmed due to her clenched hand. We had to force it open enough to allow us to trim the nails. It pained her when we did this but it was necessary. We constantly kept skin ointment or powder on her palm and after several attempts at making a brace to keep her hand partially open failed we kept a thick, between 1/8" and ¼", piece of soft but tough plastic between her palm and fingernails.

Sometime along the way, when Barb could no longer wash herself, because her hair covered her ears, I neglected to keep her ears clean. I was shocked when it was discovered, you could have planted potatoes in her ears they were so dirty. You can bet they were cleaned every day after that.

From March through May of 1995 Barb suffered from Obsessive Compulsive Disorder. Apparently this can occur when the ganglia is attacked. This manifested itself with Barb in a manner that caused her to be constantly picking specs of dust or imaginary specs of dust from her clothes and dropping them on the floor. Another thing she did constantly during this period was to struggle to take a Kleenex from her pocket, brush the tip of her nose with it and then struggle to get the Kleenex back in her pocket. Many times she did not even touch her nose, just passed the Kleenex past her nose. It drove the rest of nuts until we realized what was occurring, we asked her frequently not to do that and she, after about three months, eventually did stop. We were all relieved, including Barbara.

At about this time, May '95, I reported to Dr. Casselli that Barb was usually quite tired, not much energy and didn't sleep too well. She had problems eating; said she felt as though she had to regurgitate after eating only about a quarter of her meal. Her

weight was down about 25 lbs. in the last year. She had also been afflicted with diarrhea frequently and her speech and responses were deteriorating.

We, our daughter Barbie and I, decided in early 1996 that we needed help with caregiving. Barbie was home schooling her two kids and spending a lot of time relieving me from caregiving. The schooling suffered a lapse but I needed relief, mostly to get some sleep and food marketing. The social services offices at both Mayo, Scottsdale and Scottsdale Memorial Hospital provided recommendations about caregiving organizations. We were fortunate in contacting a local, small organization called McCormick Ranch Caregivers. It was founded by two retired R.N.s who did it because it was needed. They ran it with some minor assistance from a local church. They screened the patient's requirements and provided a caregiver who fit the requirement. The two R.N.s took no pay, they recruited the caregivers, trained and supervised them. The caregivers were also women who were more concerned with caregiving than pay. They charged $7 per hour and were excellent. (For reference, I believe the federal minimum wage was about $4.50/hr.) We hired them for several afternoons a week and gradually increased it to 5 afternoons per week. About that time I hit the wall and was dragged, kicking and screaming, into the hospital for an emergency heart bypass operation. Due to complications I took much longer to recuperate than anticipated. Consequently I could not take an active part in caregiving again. In fact I became an added burden to the already overextended daughter and caregiver crew The McCormick Ranch Caregivers could no longer support the amount of help we needed so at this point, end of Oct. 1996, we hired full time, around the clock, caregivers. At our daughters suggestion, our doctor at Mayo recommended that Hospice get involved. Again we were fortunate. Hospice of the Valley here in the Phoenix area helped us tremendously. We also found a very capable woman at our church, Mrs. Joan Page, who had a good deal of experience in caregiving. She took care of her young grandchildren during the day and took care of Barbara from about 5P.M in the afternoon, when she relieved the daytime caregiver, until 9 A.M. in the morning when the daytime caregiver arrived again. The night time caregiving consisted of: preparing dinner for and feeding Barb. Dinner at this point consisted of Ensure and pureed food (found a small, about 1/3 regular size, blender that was perfect size for small qty). She took Barb from the recliner chair, put her in the wheel chair and wheeled her into the kitchen and up to the kitchen table. Barb could not help herself in any way at this point in time. She took Barb to the bathroom as frequently as Barb had the inclination, which was often. Transferred her to the toilet seat, cleaned her and brought her back to the recliner chair. We usually had Barb's favorite TV shows on but don't know if she saw them. Actually she was not a big TV fan but did have a few shows she liked, Trivia, Wheel of Fortune , Wall Street Week and news. She did listen to Rush Limbaugh on the radio. She was basically unable to speak during this period so we ended up almost playing charades trying to figure what she wanted. At night she would wash her and handle all the necessary toilet requirements and put her into bed. There was another bed right next to Barbs in which the caregiver slept,

so she could attend Barbara's needs during the night. In spite of taking her to the bathroom at her slightest indication there were many accidents. We had to resort to Depends (adult diapers) and disposable bed pads which were used not only on the bed but also on the recliner chair. Both caregivers were qualified to dispense medicines so they handled that also. The first duty the day caregiver had was to get Barb up from bed, shower (on a special chair supplied by Hospice-it was on wheels, had arms for safety so she wouldn't fall out and the seat doubled as a high toilet seat), dressed and wheeled her into kitchen for breakfast. Same thing for breakfast, Ensure and pureed food. Then to the recliner chair and bathroom as required. She then, when time permitted, washed the sheets and bed clothes and any other laundry. She took care of Barb's lunch and sometimes read to her, we think Barb listened. The daytime caregiver was from an agency so we paid $15.00 an hour to the agency. The night caregiver was independent and we paid her $10.00 an hour (which was more take home pay than the agency CG made at the $15 rate). We had to fire several of the daytime caregivers from the agency until we found one that was capable and dependable. The one we settled on was a young woman who was uneducated in the classical sense but had been trained very well in caregiving and dispensing of medicine. She was very considerate, compassionate and happily tackled any job that was required and some that were not.

Hospice of the Valley provided all sorts of assistance. Barb's Medicare was signed over to them and they assumed all charges from then on (not including the cost of caregivers-we had insurance for long time care but it only paid $50 per day for care at home whereas the total came to $285 per day). First and foremost they provided very competent R.N. service as often and as frequent as needed. A regular nurse was assigned to us. She was excellent and came once a week on a regular schedule. She kept in contact with Barb's doctor, Dr. Caselli, and kept him up to speed concerning Barb's condition. He in turn would advise her, Mrs. Debra Traub, R.N. of specific care or medications. We would call the Hospice nurses for help and advise or instructions at any time and if necessary, they would come to the house. Hospice provided, at Mrs. Traub's request, a fantastic air bed with a motor control that rolled Barb over to one side, then returned to her back and then to the other side at variable time intervals that you could preset. Barb was unable to move in bed and this was a tremendous relief for her. Barb began to have trouble breathing and Hospice provided an oxygen generator and necessary tubing and nose pieces for administering the oxygen. They also provided emergency tanks of oxygen and appropriate tubing and face/nose assembly in case of a power outage or if we wanted to take Barb out of the house. Barb started to accumulate mucous in her throat and lungs to the extent that on at least one occasion we had to take her to the hospital emergency room for suctioning. They were able to relieve the problem satisfactorily. Hospice then provided a suction machine for us to use when necessary. All the caregivers including daughter Barbie and I were instructed in use of the various equipment that Hospice provided. Hospice provided all medicines required whether prescription drugs or not and had them

delivered to our door at any time of day or night.

Medicines: During the last month or so the pain from her back and rigid, tensed muscles became quite severe and we resorted to Morphine to ease it. Prior to the morphine we used ULTRAM for pain relief. We used a patch with slow release medication, Scopolamine, that helped control the mucous and we doubled up on the sleep medication, CHORAL HYDRATE liquid, so Barb could sleep through most of the night. Thereafter she slept about12 hours at night and a good deal sitting in the recliner chair during the day

Barb had difficulty swallowing and problems with gagging when eating for at least the last year. Pills were crushed and put in the Ensure so she could get them down. It was necessary to make sure she was getting enough liquids and calories.

During the last few months she had great difficulty both urinating and with bowel movements. On one occasion she became so constipated that we had to take her to the hospital emergency room for medical assistance to get her relieved. It was necessary to use laxatives and to keep track of her progress. To do this we devised a chart that the caregivers filled out. Barbs tummy also became extended and hard during this period.

For some unexplainable reason, during the last 2 weeks Barbs speech improved to a point where she could communicate again which was a surprise to us all and she seemed to feel much better. We even perceived some movement in her left arm.

The afternoon of Jan 20th Barb was sitting in the recliner chair. Lasandra Fields, the daytime caregiver, was sitting on one side of her gently rubbing Barb's tummy and I was sitting on the other side rambling along talking about what ever entered my brain when Barb announced, with more than a chuckle, more like a laugh, that she thought that she was going to have a baby. Lasandra almost fell off her chair laughing and I announced that I was too old for that kind of nonsense and if she was going to do that she would have to throw me out and get a younger man. All three of us had a good laugh. I only bring this out to show you that even at this late stage of CBGD she had alert perception of what was going on and still had a good sense of humor.

All through this ordeal I never heard Barbara complain. She was more stoic and courageous than mere mortals have a right to be. I'm sure her strong faith in God helped her through this traumatic time. We were married 48 ½ years. She was a Perfect Lady in every sense of the word and I loved her dearly.

After her death, her brain was donated for medical education and research. In excess of $5000 in lieu of flowers was donated to same for research on dementia.

TIME LINE DEPICTING APPROXIMATE PERIOD WHEN DIFFERENT SYMPTOMS FIRST BECAME APPARANT WITH BARBARA

YEARS	SYMPTOMS
0 **1991**	• Carried glass of water on a tilt- not realizing it
1 **1992**	• Extreme concentration to make left hand do what it was supposed to do • Gradual deterioration of handwriting • PRELIMINARY DIAGNOSIS CBGD BY MAYO PHYSICIANS
2 **1993** **By end '93**	• Hand tremor when attempting to use, particularly left hand • Left hand and arm unusable • Unable to dress herself • Unable to navigate stairs • Loosing appetite and strength in general
3 **1994**	• CONFIRMED DIAGNOSIS AS CBGD • Started losing balance on occasion. Falling backwards • When attempting to sit down, would miss chair to her left.
4 **1995**	• Occasional body tremors • Unable to get up or sit down or walk without assistance • Left hand and arm became rigid across breast • She could not release things that she had grasped • Obsessive Compulsive Disorder for several months • Excessive need to urinate and bowel movement • Problem eating and swallowing- drastic weight loss • Problem sleeping • Gradually losing ability to speak • Increasingly less involved with outside world
5 **1996**	• Could no longer walk • Complete loss of speech • Pain from rigid arm and hand and fractured vertebrae • Trouble breathing- required oxygen • Difficulty urinating and bowel movements • Excess mucous in throat and lungs required suctioning • Unable to sleep at night without medication • Slept much more during day
6 **1997**	• Body very rigid • Tummy became extended and hard • Suddenly could talk in sentences • Succumbed Jan 20, 1997 from CBGD AUTOPSY RESULTS — CBGD

Following are letters or excerpts of letters received from caregivers of patients with CBGD. Permission has been granted to copy them.

From Theresa Roberts

Long Beach, California 2/15/2000

This guide has been written for those sufferers, families, and anyone else involved with CBGD, CBD. I hope that it can be a possible starting point for those who, after having been diagnosed with this disease, have nowhere to turn. "Nothing can be done and we don't know what to expect" were the common responses from the medical community. Well, it is true that what one family goes through may not be the exact path another takes. However, through over 5+ years of correspondence with others diagnosed with this disease; common situations, events, symptoms, patterns begin to take place. This guide is not meant to be set in stone. It is simply that - a guide to help those about to embark on a journey. I write this in memory of my father, Joseph F. Harvey, and dedicate it to all those who have or will travel a similar path as he.

This is a story of the Harvey family. It is how 2 brothers, 2 sisters, and 2 parents lived through a horrible disease. A little background is necessary before we begin our journey of CBGD. While I was the one caring for my dad daily, it was each of my family members, including aunts, uncles, neighbors, and friends that also helped.

I realize this guide is to help others cope with this disease, and as I walk you through my experience, please keep in the back of your mind, my Dad through this entire process, was 100% fully capable of thinking, making decisions, and knowing what was going on. In other words, he was competent. The only things that were not were his hands, feet, and at times his words—he knew what to say, but could not get the words out.

While my involvement came later in the disease, I can touch on a bit of the prior years events. It was in 1989 the signs of CBGD first came about. Dad had a hard time tying his tie before work. It would eventually become something my mother would do for him without any questions as to why he could not. He then had trouble writing, his print once perfect, now was shaky. He had a hard time remembering how to write his name. He couldn't remember how to swing his golf club, something he had enjoyed for 40+ years. He could not throw a ball anymore because he could not quite figure out how to let go. When mowing the lawn, he did not remember how to get the grass clippings out of the basket and into the trash. Things were not the same.

I have outlined some of the symptoms associated during this time. They are as follows:

Phase 1 — (1989 - 1994)
Handwriting
Forget how to do things
Perception problems (missing steps)
Judgement problems
Couldn't throw the ball

Mom saw it. Mom watched. However, we kids, were busy with our lives, stopping in and out of the house, never really taking notice. We would think she was being overprotective, but later I learned she was just worried. I remember going shopping at Costco and she was scared he would get lost and I would jokingly say, "Ah, Mom, the store has 4 walls, how can Dad get lost?" It wasn't until I began to care for him did her words ring true. Dad tried putting the car into reverse and did so with the blinkers, thus breaking it. He gave up his work and the car keys. He went for long walks. Later, he began losing his balance. One weekend, he fell off a 4-foot ladder trying to trim the peach tree. When he fell, he fell straight back on his head. This is when my mother began to worry more. He had been seeing a neurologist for a stroke he suffered in the 80's but this time, Mom wanted to know more. The results were that he had some type of progressive brain disease that would worsen. Without more testing, they could not say exactly. Dad opted not to know.

My mother died on February 15, 1994 from emphysema. After my mother's death, my Dad's symptoms worsened. We believe that this emotional trauma in his life helped to expedite the disease processes. I was at a point in my life where I could move back home and care for Dad. So, my husband, my 3-year-old daughter and I moved into the back part of the house. From this time forward, I was the primary caregiver for my father with my entire family as backup. Without them, I could not have done what I did. And without my Dad's sense of humor, good nature, and just plain being easy going, none of us could have gotten through this. And, it is from here that my journey begins. 3 years, 10 days of caring for my father who had Corticobasal Ganglionic Degeneration a.k.a. CBGD/CBD.

Dad could still get around, only falling occasionally, and could still do most things for himself. New symptoms begin to appear. He would move his right hand sometimes when he spoke. Other times, he would pick up the TV changer or a Kleenex on the table beside him hundreds of times. He would pick it up, put it down, pick it up, put it down, etc. When asked why, he said he did not know "It's like it has a mind of its own". Dad during this time began to get depressed and cry all the time. His neurologist suggested Dad take Prozac. This helped with his emotional problems. We still had

no answers regarding his neurological problems.

However, seven months later, Dad got worse. I began having trouble-picking Dad up from the falls. I was 5'2" and my Dad was 6'. He had a harder time helping me. Rather than pushing up, he would push back and we both would end up on the floor. It was at this time, we decided as a family to go ahead and do the additional tests. My sister was a nurse, and we thought it would be easier to face this if we knew what lay ahead.

So it was in November of 1994 we received the diagnosis. CBD, Corticobasal Degeneration. It was suggested we put Dad in a home because it would become too hard to care for him. But together, as a family, we decided this was not an option. We were fortunate to have resources available to keep Dad at home. And it is a decision every family must make and each decision is correct for each family situation. For us, we would take care of him.

Dad was in phase 2 of the disease; approximately 6 1/2 years into the disease. I have outlined it below.

Phase 2 - (1994 - 1995)
Increase falling
Alien hand (picking up TV changer constantly without knowing)
Problems eating (food finding the mouth)
Neglect to one side (would not realize doorway and walk halfway into)
Help putting on shirt/shaving/brushing teeth/tying shoes

My sister suggested we get home health to come in and help Dad with possible occupational and physical therapy. We had a supportive neurologist who wrote the prescription for home health. However, the nature of the disease was that Dad was going to get worse, not better. After 8-12 weeks, they would release him from the program because they did not see any improvement. They did give us helpful hints, however, because of the rare disease, they did not know what they were dealing with. No one had ever heard of CBGD. I had downloaded abstracts regarding CBGD and would give this to them to read. However, this strange neuro disease baffled them.

Dad would still go for walks to the golf course and would go on small trips with my brother. He spent most of his days listening to books on tape. My sister had told us that if we get a doctors note indicating Dad could no longer read, that the Braille institute would "loan" a special tape recorder (one with big numbers) and books on tape at no charge. We could either rent the books at the local library or have them mailed to us free of charge. Dad must have listened to hundreds of tapes during this time.

It was in December of 1994 when things would drastically change. A hard fall on the

sidewalk, straight back on his head. Dad said he was okay. We took him to doctors, who said he was okay. He ended up in the hospital with prostate problems. We urged them to do a catscan. They did and found he had a subdural hematoma caused by the fall. They had to operate and put tubes in his head to drain the blood and relieve the pressure. He was in rehab for 6 weeks. When he got out, he was not able to walk well anymore. This is when Dad's world slowly started to shrink.

He was now in phase 3 as outlined below.

Phase 3 - (1995 - middle of 1996)
Walking w/support or wheelchair bound
Can no longer do the things for ones self - (dressing, eating, showering)
Communication getting harder and slower (Couldn't remember words)
Alien hand – Arm would go up in air for hours like asking a question
Thinking he had to go to the bathroom all the time
Muscles stiff (every inch of my father was hard as a rock)
No reflexes

Because of Dad's size and strength, I needed help. We called some agencies and found a woman, Margaret, who was 6'1" tall. She came and lived with us 5 days a week. Margaret was with us until Dad died. She was an invaluable person in this story. She was always there, always wanted to learn, and above all, respected "Mr. Harvey." The other 2 days I was on my own. Thanks for my brother and husband, because there were days I could not move Dad. My father's muscles for almost 2 years straight were rock hard.

During this phase, we tried various medications. Symettrel and Sinemet the Parkinson's drugs, Flexerill for stiffness, Neurontin and Permax for movement, Klonipin for his restless feet at night, Tofranill for relaxing, Ambien for sleep. If we thought it could help, Dad was willing to try. Life was like this for approximately 1-½ years. He would have periods we called "lean to" when he would lean to his right side (bad side) for approximately 1 week. After this he would have lost something, meaning, his speech would be worse, his alien hand would act up, he would get stiffer, etc. These would come every once in a while, but later would happen increasingly often.

We used a gait belt to transfer Dad. A gait belt is a cloth belt that goes around the upper body that allows you to have control when transferring the person. Dad could not move his hands or feet the way he wanted to. During this time, we would take Dad for long walks on his wheelchair, go get ice cream, etc. My father's attitude through this disease was unbelievable. When I would comment on how positive he was, he would say, "What else can I do?"

Near September of 1996, Dad began withdrawing increasingly. We thought it was

because he had such a hard time talking. He would get as frustrated as would I. He slept more often. He began having problems with his bowels; either constipation or diarrhea. He had a hard time eating, at times taking almost 45 minutes to feed him. But Dad loved to eat, and he wasn't going to give this up very easily. He pushed on.

We are now entering the final phase. I have outlined it below.

Phase 4 - (middle 1996 - 2/25/97)
Speech problems / unintelligible speech / mumbling
Drooling
Bowel problems
Unproductive cough
Swallowing problems
No gag reflexes
Withdrawing/Sleeping
Dementia-at times (Would think we had a 2-story house, thought he had just gotten back from golfing)
Pain but not able to identify area

Phase 4 obviously was the hardest. It lasted for 6 months, but the last 2 months were the hardest. However, what did make it easier was the fact that both my parents made it known their wishes for dying. I can not stress enough the importance of a Living Will, Durable Power of Attorney and Do Not Resucitate orders. This made the process easier as ***I*** was not the one making the decision.

In January of 1997 he seemed to be getting less interested in having us wheel him around the neighborhood in his wheelchair. He was more quiet than usual - hindsight being 20/20. In addition, one night in particular, he choked badly on some food. Oh, we had other choking problems, but this night his face was purple and it was not until the last moment he remembered HOW to cough... see his stomach muscles were so tight that doing a Heimlich was almost impossible. He seemed to be okay but was having more and more problems taking meds and he was either constipated or had diarrhea. We had that problem in the past but nothing like this. He also began complaining about pain in his left shoulder. My father had never complained of pain before this. When we would transfer him, he would shout "Ow,ow,ow,ow..." from the pain.

We had recently gotten hospice in and they felt that Dad was doing okay...(although no one had ever dealt with CBGD). I would urge everyone to call hospice and have him or her evaluate your situation. They were only with us 6 weeks, however, they were a great source of emotional help, medical help, and spiritual help. Because of Dad's shoulder pain, we left him in his hospital bed and he began sleeping most of the day. Eight days prior to his death, he had to have an enema due to a possible

compaction of the stool. What I saw in his eyes was when I knew he was giving up...He realized this was what lies ahead and I feel it was then he decided to call it quits. Dad was complaining so much about pain they began morphine; which of course makes one constipated! He began to sleep continuously. He started filling up with mucus and it went from clear to dark green in a day. No antibiotics were given, as that was my father's wishes. Even when I tried to suction him, he would bite the tube and look up at me as if to say, "NO MORE" I AM READY....

He died in his sleep after 5 days of not getting out of bed on 2/25/1997. The cause of death was aspiration pneumonia. He had all of his kids around him during the day/ night. He passed at 5:00 am (about the time he used to get up to go golfing) and UCI -University Irvine came and took his body in order to remove his brain for donation. He then went to the mortuary where he was cremated and his ashes were spread on the ocean like my mom's.

It was confirmed approximately 8 months later from the brain autopsy that dad did have Corticobasal Ganglionic Degeneration. My dad felt strongly that he donate his brain to help others. By donating his brain, he has helped researchers in all areas of neuro studies. I also feel that Dad did not die in vain.

My mom used to say, "Things happen for a reason". And I try to find reasons my Dad went through this horrible disease that robbed him of his movement but not his mind. And I have come to the conclusion, that while this disease is awful, there are others out there just as terrible if not worse. But we turned it around. CBGD brought our family together in ways I never thought possible. My siblings and I learned so much about my Dad that we never would have talked about. My dad and daughter were able to know each other because we all lived together. And finally, because of the CBGD Support Network and cbgd-support listserv, I have met hundreds of people in similar situations and hopefully have touched their lives with my experiences with CBGD.

Life goes on. Nevertheless, a day doesn't go by when I don't think about my Dad and remember a fond moment in our lives or a funny antidote and laugh. He was such a good sport through this disease. I hope this has helped and urge everyone to talk with others. You are not alone.

The following is an outline of "Tricks of the Trade:"
Bicycle Helmet (for falls when they walk)
Plate Guard (Goes around plate so food does not go everywhere)
Gait Belt (Belt that goes around waist for transfers)
Daily / Weekly Log Sheet – (Help track status)
Hospital Bed – (Easy to get in and out, won't fall out of bed)

Lift up Chair – (Lifts up to help keep independent)
Books on tape (from the Braille Institute)

THE FOUR PHASES

Phase 1 — (1989 - 1994)
Handwriting - Sloppy and not remembering how to form the letters. Forget how to do things
Perception problems (missing steps)
Judgement problems
Couldn't throw the ball

Phase 2 - (1994 - 1995)
Increase falling
Alien hand (picking up TV changer constantly without knowing)
Problems eating (food finding the mouth)
Neglect to one side (would not realize doorway and walk halfway into)
Help putting on shirt/shaving/brushing teeth/tying shoes

Phase 3 - (1995 - middle of 1996)
Walking w/support or wheelchair bound
Can no longer do the things for ones self - (dressing, eating, showering)
Communication getting harder and slower (Couldn't remember words)
Alien hand – Arm would go up in air for hours like asking a question
Thinking he had to go to the bathroom all the time
Lack of reflex muscles
Stiff all the time - Rigidity

Phase 4 - (middle 1996 - 2/25/97)
Speech problems / unintelligible speech / mumbling
Drooling
Bowel problems
Unproductive cough
Swallowing problems
Lack of gag reflex
Withdrawing/Sleeping
Dementia-at times (Would think we had a 2-story house, thought he had just gotten back from golfing)
Pain but not able to identify area

From Theresa Roberts

From Darcy Croissant

Glenwood Springs, Colorado. 12/5/1997

I have had a difficult time sitting down and trying to put things into words. It is pretty jumbled, but I hope it will help. I think as caregivers - we have to be grateful for each and every moment we have. As overwhelming as the days can seem I don't want to wish them away.

In December of 1991 Mom began having difficulty writing, she couldn't explain it but she just didn't seem to be able to write well. She then started to develop a tremor in her right hand. and her speech started to slur.

She was first diagnosed at the Mayo Clinic, in Rochester, MN. In July 1994.

Increased tremor which has also now shown up in the left hand, inability to control speech, body movements, alien limb, occasional double vision (seemed to go along with some of the medication). Mom can think of something she wants to say but then it doesn't come out of her mouth. She can't recall numbers, letters, dates, etc. She knows them in her head but can't speak them. She often says yes for no and vise versa. She is unable to write. – She has great difficulty feeding herself due to tremors and lack of motor control in the limbs. She shuffles when she walks and tends to bump into things on her right. She has had numerous falls. She has had some difficulty with swallowing but that seems a bit better. She also has frequent loss of bladder and bowel control. She can no longer dress or bathe herself. I stitched the buttons on her blouses with elastic thread so they would be buttoned but she could slip them on and off. We sewed Velcro to some of her buttons, and bought all pants with elastic waste as she can't work zippers, buttons, etc. We also bought elastic shoe laces. She wears supportive tennis shoes and with the elastic, they don't have to be tied.

Mom has a wonderful physical therapist. Medicare refused to cover the therapy any more because there was no improvement. We strongly feel the physical therapy helped— Her therapist continued to treat Mom weekly on her own time. In addition she does speech therapy and occupational therapy.

Equipment:
We have a LaBoudet for the bathroom, Mom has great difficulty wiping and this helps. She has a swivel spoon to feed herself, a (motorized) recliner chair with a button to raise the chair to get in or out, a medical alert button (which works GREAT). The occupational therapist made a special splint for her right hand to help the rigidity of the hand and the thumb.

Medications:
Neurontin- at first Mom felt it really helped the speech however, her speech is increasingly more difficult to understand.
Proponal- this seemed to help the tremor.
Zoloft- she says it helps her focus.

Mom is remarkable in her attitude, she laughs a lot. She looks at this disease as a challenge. I think her positive attitude has made a big difference in how fast –the disease — has progressed to this point. She is truly an inspiration. Our family is very close. Our father passed away from Melanoma and we know how cherished life is. We all pull together to make things work and we have a strong faith in God to help us through every obstacle. – As a caregiver, it is hard for me to follow my own advise, but if you can get help, take it. It is so easy to get so overwhelmed.

Caregiving, Medicare and Hospice:
Medicare wouldn't cover physical therapy any longer and we thought it was very beneficial. They also didn't cover her medications which would cost around $400-$500 per month. As a caregiver, wife and mother of two young children it became very difficult for me to do all the day to day care for mom, work full time and care for my own family. Medicare refused coverage for home health because they said she didn't need a nurse, it was primarily personal she needed. I tried to contact a home health aide on my own and they wanted $50 per hour for 1-2 hours a morning. I finally hired an outside provider, but had to get liability insurance and pay unemployment taxes, as well as, withhold taxes for federal and state. It became a bookkeeping burden and very overwhelming.

Mom has now qualified for Hospice through our local hospital. They help a couple of hours each morning and then I take care of her at night and weekends. Hospice has been an answered prayer. I would like to get more help but I don't want to give up my time with Mom. It is something I struggle with at this point.
Medicare is now covering her physical occupational and speech therapy, medications and her home health aide through Hospice.

Mom has a Durable Power of Attorney, Medical Durable Power of Attorney and Living Will. She also has donated her brain to Harvard for research when she dies.

Darcy Croissant

Another caregiver, who for personal reasons, prefers to preserve some privacy, but is willing to share their experience if it might help someone.

The first slight indication of any problem for this patient, a healthy, athletic male in his mid-seventies, was in late 1991. He had been a good golfer but noticed increasing difficulty with his golf swing. A year later, during a routine physical examine, his doctor prescribed medication for a mild tremor in his left hand. Later that year he developed spatial problems while driving.

In 1993 he gave up trying to play golf, as his left hand was becoming increasingly hard to use. His internist referred him to a neurologist, but MRI'S, x-rays and various other tests revealed no specific problem. He was also checked by a neurosurgeon. Parkinson's Disease was suspected. He was referred to a neurologist at the UCSD clinic in San Diego who confirmed the possibility of Parkinson's and started him on Sinemet. That was discontinued because of adverse reactions. By December his speech was a bit deliberate but not enough to be too noticeable to others.

A second MRI and hand therapy in 1994 produced no positive results. He gave up driving the car. The doctor decided to start him on another course of Sinemet which also had to be discontinued. Following a third set of MRI'S, still inconclusive, he was started on Parlodel which also had to be stopped. His left wrist became stiff.
In early 1995, the neurologist at the UCSD clinic gave the possible diagnosis of progressive muscular palsy and started him on Permax. This was followed by six weeks of hand therapy, as his handwriting was getting much worse. Speaking required increasing effort although his speech was distinct enough.

From September 1993 through March 1995, he had been given Sinemet (twice), Parlodal, Artane, and Permax. None helped. All caused adverse reactions such as dizziness or a drop in blood pressure resulting in many falls, sometimes three or four a week. Several times he started walking rapidly and could not check himself until he fell.

He was finally referred to a Motion Disorders specialist at the UCSD clinic, Dr. Clifford Shults, who, after a very thorough examination, said he thought he knew the problem and diagnosed corticobasal ganglionic degenerative disease. We still knew very little about it.

In late May of 1995, he became very ill with a sepsis and was hospitalized. After the infection subsided, he was unable to walk and speaking was more difficult. He spent a month in the hospital undergoing intense therapy. I was also instructed in how to help him, as from then on he could walk only with assistance and the use of a gait belt. He could not get out of a chair by himself.

When he was released from the hospital, occupational, physical and speech

therapists came to our home. He was also provided with the services of a Home Health Care Nurse and a person to assist him in showering and bathing. We installed grab bars in the bathrooms, acquired a seat to place over the bathroom stool and a wheelchair which at that time we did not need to use in the house. We also bought a small bike for exercising his legs, a brace for his left hand and a plate guard.

He began to experience some swelling in his left hand and leg and was put on a diuretic. His left ankle began to turn outward, his left leg was weakening, his back became more bent, and he listed to the left when walking. A comprehensive eye exam revealed that while his eyes worked independently, they would not coordinate for reading. He could not write or use the computer and could only watch TV in a limited way.

In October, 1995, he was invited to spend two weeks at the National Institutes of Health in Bethseda, MD to participate in a study of this disease. He had been a very healthy man—took no medication—so any problems were because of the disease. So little is known about it and there seems to be no clue as to what causes it. The doctors at NIH supplied as much information as they could about it and provided as much help as they could in how to cope with it, but the sad reality is that this disease is untreatable and terminal.

Upon returning home, he was assigned to Hospice. Every morning someone to help him shower and dress, a nurse made regular visits and we could summon help at any time. All this was under Medicare and Blue Cross and the supervision of our doctor, an internist. We were not in an HMO. Most of the therapy was discontinued as he became too rigid. He wore both day and night braces on his left hand and a brace by day and a Podus boot (supplied by NIH) at night on his left foot. They also supplied me with a back brace to wear when working with him. By December he was using the wheelchair in the house most of the time, and later we got a hospital bed and bedside table.

By early 1996 a caregiver was sometimes supplied by Hospice to come for a couple of hours so I could get out to do errands. Our children stayed as much as possible and friends helped a lot. Our oldest granddaughter, in her final year, of nursing, came to live with us for the summer and help.

His mind stayed alert and active although speaking became more difficult. He enjoyed visits with friends and retained a remarkably good attitude. As he could no longer read, he spent hours listening to “talking books’ which were supplied, along with the machine, at no cost by the Braille Institute. (Arranged by the doctor.) This provided many hours of pleasure for someone who was a great reader.

His condition seemed relatively unchanged for a few months but by early June, 1996, it was apparent that the disease was finally exhausting him—just wearing him out. He was definitely weaker and no longer wanted visitors. It was too wearisome to try to follow conversations and make responses. He no longer wanted to leave the bedroom. Chewing became an increasing problem, so foods were cut in small pieces. No longer could he manage feeding himself.

Because in various parts of his body he began to experience nebulous but still very

real and distressing pains for which there was no visible cause, he was started on small doses of morphine and ativan. He also developed extreme sensitivity to noise—the clatter of dishes, the shutting of a door, loud laughter or noises—all of which normally would have gone unnoticed but now caused him distress.

For some time he ran a low-grade temperature for which we were unable to find any cause. Another unexplained condition was that quite frequently one limb, usually his right leg, would be very cold to the touch clear up to his knee while the rest of his body was warm, yet the pulse in that limb was normal. About the last month he had episodes of "pumping" his right leg which could only be quieted with doses of morphine. Toward the last this leg became drawn up and almost impossible to straighten.

Because swallowing was so difficult and choking a real concern, he was started on soft foods. Until the last two weeks, he continued to sit up in a chair for a while each day. His ability to speak became so limited that he mainly communicated by squeezing our hands, but he understood everything said to him. Doses of morphine and ativan were increased, as he obviously had pain and discomfort.

The last five days he was unconscious. This later deepened into a coma. He was unable to swallow even medications which had to be administered by syringe into his mouth, so he rapidly lost weight. We turned him every two hours. He died in July, 1996.

Added thoughts:

I can't stress enough how helpful Hospice was and the importance of a good doctor who cooperates well with them.

Almost all of the equipment we had to acquire was supplied either through Medicare or our insurance.

Early on he dressed in his regular clothes every day. As he became stiffer, we went to sweat suits and later pajamas. Toward the last we used colored short-sleeved tee-shirts for tops and split them down the back for ease in putting on.

It would be wise for anyone faced with this disease to consult their attorney to be sure all legal matters are in order.

Contributor remains anonymous.

From the UK, Sandra Till reports on her Dad, Jim Pemberton.

I am not the main carer in this case but hope I can give you some insight into the progression of my Dad' illness up until the time of his death Oct., 18, 1997.

Dad was first diagnosed 6 years ago, at the age of 64. He had been experiencing strange sensations in his shoulder for a short period of time but was not too concerned about these. His job was a Postman, so his shoulders had received a lot of punishment over the past years. He was retiring in nine weeks so this he thought would help. His GP thought that the cause was probably a frozen shoulder. Prior to this Dad had one or two falls that nobody could explain and to this day we do not know if these were connected to CBGD or not.

One day whilst at work he picked up a letter he was sorting out into his mail bag and was unable to let go of it. His thumb and forefinger would not part.

When he went back to his GP concerning this he was sent to see the Consultant Neurologist at the local hospital. Certain tests were carried out to ascertain dexterity and also a mini mental score test was carried out. An appointment was made at the university hospital in the neighboring town for further tests. As Nurses, my husband and I were starting to suspect Parkinson's Disease but never expressed these fears to Mum or Dad. The Consultant at the Queens Medical Centre University Hospital in Nottingham diagnosed CBGD saying that it was a new illness that was part of the Parkinson's family that very little was known about. It was known that the illness would progress and that there was at this time no cure for it.

The numbers in the UK were very low - Dad was the only one in our town with it and there were only 15 cases in the whole of the Midlands. Dad was asked to donate his brain on his death and this he agreed to do.

After that point we were basically left to get on with things and discover the illness as it gradually took more and more of Dad's dignity, it never could attack his brilliant sense of humor though, this stayed with him right up to his death and I'm sure this is what helped him quickly become one of the favourites in the nursing home where he spent his last four weeks.

After the incident of Dad's thumb and forefinger he eventually lost use in that arm. Before this though he went through a stage where his arm and particularly his hand would not obey him. He would shake your hand or pick up his glass of beer and not be able to let go. His grip was like iron and I'm surprised that the glass never shattered under the pressure. Never one to give in, it took him a long time to give in

and use his left hand and when he did it was very clear that this hand had lost some of its movements.

Whilst the GP was very good and supportive to Mum and Dad he admitted that there was nothing that he knew about the illness so was unable to offer any advice.

Dad was seen periodically by the consultant (or rather his staff) at the local hospital but I always thought that this was more for their benefit rather than ours. Each new problem put to them was answered by the restatement that Dad was never going to get better and nothing could be done. You only need to be told this once, maybe twice to help it sink in, but every hospital visit, it started to get Mum down. They prescribed Selegiline three time daily to see if these would help. They never seemed to but Dad carried on taking them; I think it was more because at least it felt as if something was being done. Later on in the illness they also prescribed Baclofen but this was stopped when no improvement was noticed.

Dad's legs were the next to be affected. He began to drag his right leg and his walking became unsteady. He began to have numerous falls. When walking he would be slow to start but once started, impossible to keep up with. The hospitals way of helping with his unsteadiness was to give him a walking stick. Bearing in mind the fact that he had already lost control of his arms. The stick was carried more like a magic wand going every where but where intended so was soon put in the cupboard.

As the illness progressed both legs became affected. Dad became increasingly rigid and seemed unable to command his body to do as it was told. When Mum tried to lift him, instead of coming forward, his body would lean backwards, increasing the strain and frustration on both of them. But still they kept laughing.

Dad's speech had started to go at this point. Occasionally he would use completely the wrong word. Sometimes he was unable to say the word at all. His speech deteriorated to the level where only occasional words could be understood and our lives became a constant game of charades, we asking questions and he answering either yes or no until we got there. Often, though the moment had passed before we understood what Dad was showing or telling us. It was this part of the illness that I found the cruelest. He tried so hard to tell us things and struggled until it was understood. Along with the loss of speech came the loss of his ability to swallow. He suffered a bout of aspiration pneumonia and was advised to only drink liquids that had been thickened. At one point he could only manage teaspoons full of this thick gunk that resembled none of his favorite drinks. Slowly he improved and was able to drink his thick liquids from a beaker.

Dad's package of care consisted of one day a week at a day hospital and two weeks every eight weeks in a hospital ward to give Mum a break. Whilst at home a care aid

would visit every morning to help wash and dress him. They weren't allowed to lift him in case they injured their backs, so Mum continued to do most of the lifting. Late on in the illness a nurse would visit to bathe Dad or to help him have his bowels open as this had become a problem.

On June 12th, 1997 Dad went into hospital for his regular respite care. It was here that he suffered the aspirational pneumonia. Whiles his ability to tolerate drinks from a beaker improved his general strength did not.. His stiffness became more pronounced making it impossible for Mum to be able to care for him at home.

Once well enough to leave hospital we found him a nursing home near to where Mum and I live. This was a hard decision to make. Mum said it felt as if she was signing him over. Once in the home Dad's charm soon won the carer's affection and during the month he stayed there he received excellent loving care. Mum visited daily and I as often as I could.

Dad took ill during one night. The GP was called who diagnosed bronchial pneumonia and prescribed more anti-biotics. Dad had never really been free of them since his illness in June.

This was his last illness. His breathing just deteriorated and the cause of death was put as pneumonia. Whilst it may not seem important what caused the death, I regret that the GP did not put CBGD on the death certificate as I think this would help towards research in the future as regards the amount of deaths caused by the disease. But, at the time, you don't think of these things.

I hope that this is of use to you. It has helped me just sitting down writing it all down. As an ex-carer (although I must say Mum did most of the care). The CBGD family has been helpful to me and I hope to remain in touch in the future.

Sandra Till, Allenton, Derby, England

Robert Hall from South Bend, IN. reports
November 1997

CBGD patient: Judy Hall (51)
First symptoms: September 1989 (42)
Birth date: 10/15/1946
Has misfortune of being youngest-to-date to be diagnosed with CBGD (according to Cleveland Clinic).

Caregiver: **Robert Hall, husband**
Address: **59580 Saturn Ct., South Bend, IN 46614**
Telephone: **(219) 291-9171**

Patient was a literature teacher. Began experiencing difficulty holding items in her left hand, dropping erasers, chalk, books during class. Pain was minimal at the time. Could not wave at grandchildren with left arm or hand.

October 1989
Family physician refers patient to a neurologist.

January 1990
Neurologist tests for Carpal Tunnel and other potential muscular problems. None found.

Spring 1990
Neurologist diagnosis: MS. This diagnosis achieved through MRI disclosure of small spot on right side of brain.

Treated three and half years as MS patient. Given usual symptomatic drugs for MS.

Symptoms now expanding to involve the entire left side of patient. Left hand and arm becoming very rigid, fingers beginning to curl, left leg and foot involved to where the patient would stumble and fall regularly. On one occasion, patient fell down a flight of stairs breaking her right wrist (requiring a cast) making her life more difficult with no use of the left hand and only limited use of a few fingers on the right hand during the healing process. After three months the cast was removed. Unfortunately within a week the patient fell again breaking both bones of the right leg and the ankle bone cracked. This required surgery to insert a permanent rod from the knee to the ankle, with two permanent pins and two screws for the cracked ankle bone. The patient was required to wear a knee high boot for nine months, undergoing extensive therapy for both the wrist and leg.

Patient never regains full use of the right arm or leg. Before this was possible, symptoms of CBGD began to affect the right side of her body.

After more than three years without the usual remission periods consistent with most MS patients and the realization of the symptoms now beginning to effect the right side of the patient, neurologist does a second MRI. The original spot on the brain was discounted as the cause of symptoms.

July 1994
After four day stay in local hospital and all new testing proving negative, neurologist admitted he was mystified, patient referred to Cleveland Clinic Foundation in Cleveland, Ohio.

July 1994
First appointment at Cleveland Clinic.
First diagnosis: ALS
Medication: Baclofen to ease spasms.
Patient cried often during return trip home.

August 1994
second appointment to Cleveland Clinic, different neurologist, clinic not satisfied with first diagnosis. New diagnosis: Primary lateral Sclerosis. Continue Baclofen. Patient cries often during return home.

November 1994
Third appointment to Cleveland Clinic, yet another neurologist, clinic still not satisfied with first two diagnosis. This neurologist is at the top of his profession, works with an assistant. An entirely new assortment of tests conducted, lasting the better part of two days. New diagnosis: Corticobasal Ganglionic Degeneration

December 1994
Fourth appointment to Cleveland Clinic.
Diagnosis confirmed: CBGD
Extensive range-of-motion therapy prescribed. Clinic contacts South Bend therapist to discuss program.
Medication: Continue Baclofen, add: Sinemet and Klonopin
Continued trips to Cleveland clinic on an every other month basis to monitor disease progression and evaluate medication.

September 1995
Patient is admitted to Cleveland Clinic for two and a half weeks of occupational and physical therapy. She is measured for new wheelchair she can propel herself with the little use she still has in right leg. she is taught means to be as independent as

possible.
After January 1996 trips were cut back to every three months as the Clinic's involvement had been reduced to strictly monitoring the disease and medications.

July 1996 was to become our last trip to the Clinic up to current date. Our South Bend Neurologist had no knowledge of CBGD. Upon our return, after diagnosis, we informed him. He has since received all reports from Cleveland Clinic. Patient has infrequent appointments with the local neurologist as he is now prescribing new medications as the disease progresses. She is currently taking Baclofen, Klonopin, discontinued the Sinemet, added Valium and Vicodin for pain. Vicodin turns her into a zombie. She takes Pepcid AC one hour before bedtime to control her stomach acid. melatonin is also taken to help her sleep through the night. Occasionally we use a nebulizer with Albuterol Sulfate Inhalation Solution to help clear her lungs of congestion. This also requires physically hitting her with a cupped hand to dislodge the mucus in the lung. This is done to both front chest areas, back, and both sides.

At the present time (after eight full years), she is unable to speak, has difficulty chewing, often chokes on food (I have had to reach into her throat numerous times to retrieve small chunks of food. A few times I had to do CPR to save her). Has also choked on liquids. Eyes are basically fixed (can move slightly), eyelids hard to open and close, cannot turn her head (neck pain is almost constant), jaw locks open often (when she yawns to retrieve more air in her lungs), and at times cannot hold her head up (before the Valium, her head would often be drawn backward), has no use of fingers, hands, arms, feet and legs. Recently, this has all been compounded by another diagnosis that she is menopausal. Add Prempro to the list of medication. Some pills are crushed and given to her with applesauce or yogurt. Not all medicine is effective if crushed.

The Patients days are generally spent: Stays in bed watching TV until around noon (this enables caregiver time to do household chores). Caregiver bathes and dresses her while she remains in bed. Transfer from bed to wheelchair followed by trip to bathroom for personal hygiene care, including hair and some make-up. A squirt of perfume and she's ready for lunch at the table. Rigid jaw muscles need some coaxing to open for even soft foods. Drinking straws are most often flattened by the clinched jaws during drinking process. Breakfast is often soft, moist oatmeal or Carnation liquid breakfast mixed with fruit via the blender. Lunch is yogurt, any food without a lot of consistency. There are some days where she can manage more chewy foods. Her afternoons and evenings are spent in front of a TV set she seldom watched before the disease altered her life. We do manage to get her into the car for short trips around town (just for the ride), and/or visits to local relatives homes for special occasions.

Our local Hospice entered our lives in April 1997 when the caregiver needed serious

surgery. The long three month recuperation period made it necessary for the patient be cared for by Hospice nurses with assistance from a local home care group to bathe, cloth, feed her. A second surgery extended this care into early August of 1997.

The purchase of an adjustable bed (not a hospital bed) has made it easier to keep the patient comfortable for her long hours in bed and for watching her TV in bed. The use of the usual raised toilet seat makes giving her enemas easier (her bowels do not function on their own). We have an oversize shower stall in the master bath.

Foley catheters are a way of life for her also.

We attempt to keep her surroundings as normal as possible. There are no ramps, lifts, handicap van or other visible items to make her uncomfortable when company arrives. Her weight is now down to less than 120 pounds. The caregiver weighs 190 and physically lifts her in most transfers.

Unfortunately, with only one exception, the patients former friends have ceased to visit or call. Most have told our relatives they are uncomfortable with the situation (understandable, I guess). The lone, remaining friend will endure till the end.

Despair, Depression, Disappointment are the words for an average day. She makes every effort to be pleasant and cheerful. Her wish is for the end to come, while at the same time her main concern is not for herself, but the future for me, her children and grandchildren, without her. Divine intervention is our only hope.

Robert Hall

Louise Davis, New South Wales, Australia, reports on her husband -

Roger Davis, born 1933 Australia (age 64 in 1997)

First symptoms and diagnosis:
1992, Roger seemed depressed and less confident. He was made to take early retirement from his senior executive management career. It seemed to me that he was in "shut-down" mode. His usual exceptional sense of humour was very flat and he had lost of lot of confidence.

1993 on world trip he lost his memory in Orlando, totally without warning. He was in hospital for 10 days. This was perceived to be "a schematic event". The scan showed small infarcts and a possible blockage from the heart to the brain. He was diagnosed by the US doctor as an alcoholic! This was amazing considering the doctor (psychiatrist) was a well known expert on Alzheimer's Disease in Florida!)

1993/4 visits to psychiatrist, counselors (in Australia), needless to say he was not an alcoholic, and no depression was found. The heart specialist found no cause for concern re his heart. Eventually, in early 1994 a psychologist sent him to another psychiatrist who had an MRI done. This showed brain cell atrophy, and some small infarcts. The diagnosis was Alzheimer's Disease.

I**n 1995/96** a neurologist and neurological consultant expert diagnosed cortico-basal degeneration or possible Diffuse Lewy Body Disease. CBD was eventually confirmed due to Roger's alien limb syndrome and general apraxia. His left arm seems to not belong to him.

Symptoms: Since general slow down and some memory loss, symptoms have included severe aphasia and apraxia, inability to plan or organize, wandering (not "away" but from one room to another).... There has been no treatment prescribed, only TLC, calm atmosphere and routine.

He has had considerable "care" since 1995, including home visits by government subsidized carers, outings with carers; day respite twice a week; eventually daily carer for dressing/shave etc. Roger did not like people coming to the house to dress him or shave him. He thought he could do it by himself.

1996 week-long respite on occasion and hostel placement August 1996.
Nursing home placement is likely this month (November 1997).
No medicines at this stage

Helpful Hints:

Contact the Alzheimer's Assoc. from day 1.

Seek advice from anyone with knowledge of the disease or as a caregiver for someone with CBD (for early onset spouse it is very important to seek knowledge, form a bond with someone in a similar situation). I found that comparing notes with carers of parents, or older spouses was not very useful as their situation is so very different.

Always make a list of questions and ensure the doctor answers every one, each time you visit a doctor or specialist.

Always take notes during doctor visits.

Establish a good relationship with all medical professionals and care providers.

Seek legal and financial advice immediately.

Keep a journal of the disease progression and your own health and feelings.

Keep a circle of friends... you will need lots of TLC.

Make a list of the things the patient likes, before the disease progresses.

Do a family history if possible before the patient stops communicating.

Check out all care support options including day care, respite, hostels and nursing homes.

Do not resign from your job if you like it, if there is any other alternative.

Think of others who need you to be OK (apart from the patient) e.g. children.

I was able to get my husband a disability pension, which pays for the weekly hostel/ nursing home care. I had to pay a bond, which was a significant amount.

Regards,
Louise

Corporate Community Relations Manager, IBM Asia Pacific, c/- IBM Australia (FC32)
55 Coonara Avenue, West Pennant Hills 2125 NSW Australia; Tel: 61 2 9354 4596

Sandra Roberts from Norfolk, VA Reports

I am Sandra. My husband, Bill, died November 1, 1999 from CBGD (or rather, the complication thereof). In regard to needing help and "where is the church when you need it" - The Bible says "Ask and you shall receive"... A friend of mine set up a support network for me with my church called "Funny family." She called about 30 people active in the church who had worked with Bill in various ministries and asked them to come to our house for a meeting- 25 showed up. They were told what the situation was - that I needed someone to come in at various times to sit with Bill (who was total care by then and having great difficultly communicating) so I could go to church services- run to the store for milk and bread- or just go for a walk. Everyone filled out a form indicating what days and times they might be available and what they were capable of or willing to do. No one was scheduled for anything- but if I needed someone, I could just check the list, see who would do what and when and give them a call. Since they had already committed to something there was no hesitation on my part to call them. I knew that if they were not available, they would say so. I got the information on how to set this up from a book called "Share the Care: How to organize a group to care for the seriously ill" by Cathy Caposella and Shield Warnock.

I also had a caregiver that I hired to work while I worked part time (approx 20 hours a week). I paid her $7 and she was God-sent. I paid her straight cash which worked out best for both of us, although I could not deduct the expenses from my taxes. She was faithful and excellent with Bill and I could not have existed without her.
In October 1998 I called in Hospice - the doctor said Bill couldn't last much longer. In April 1999 the insurance company started wanting to know when he was going to die. Well, so did I, and so did he. The nurses, God bless them, wrote the notes in a way that the Hospice program continued until he died in November, 1999. In truth a real answer to prayers - many of them.

The thing about this horrible disease, in my opinion, is that while there are many similar symptoms from person to person, there is no "course" or direction. Bill had no other health problems, his heart and blood pressure were great - never had a serious illness, so that probably prolonged his existence (I can't bring myself to say "life" because for the last 18 months, he did not have one.) His death came from starvation and dehydration (not as bad as you might think). He had made important decisions long before he lost everything - no feeding tube, no antibiotics, no anything. Just let him go. And letting him go was the easiest part of the whole ordeal. I had the privilege of keeping him at home - not so much an altruistic move, but mostly lack of finances to do anything else. And I was with him when he died. I held him and told him I loved him. I told him how lucky he was to be walking into the arms of Jesus, and I prayed with him. He watched me to his last breath and I saw his eyes go blank and knew his soul was gone. I always felt I had lost Bill in May of 1998 when he became

total care and the first few weeks were based on that- but I find now, after two months, that I even miss taking care of him. I am OK, though. My family, my friends, and my church are still my support. I think the hardest part is realizing that I can stay home if I want. At first, I was going everywhere because I had been a prisoner for 18 months. I am beginning to slow down and take care of ME.

There is only one good thing I can think of about this horrible disease. It has connected me to many wonderful people throughout the world who have helped me and whom I have been able to help. It showed me the depth of the love of my family, my friends, and my fellow parishioners at my church. I experienced their concern, their care, their pain at our suffering. I have been truly blessed.

If I can be of any further help, please ask. My email address is sandrabill@erols.com Keep the faith and ask for help from any source you can find. You cannot do this alone.

Sandra

Update on Corticobasal Degeneration From Bradley F. Boeve, MD, Rochester, MN Rare Dementia Registry

Corticobasal degeneration (CBD), also known as corticobasal ganglionic degeneration (CBGD), was first described in the late 1960's by Drs. Rebeiz, Kolodny, and Richardson. Following a lengthy period with no additional reports, several more patients were identified and their symptoms and autopsy findings were described in the 1980's and 1990's. Patients typically have symptoms reflecting dysfunction in the cerebral cortex (thus the term "cortical" or "cortico-") and basal ganglia (thus the terms "basal" or "basal ganglionic"), and symptoms are usually worse on one side of the body. Specifically, cortical dysfunction is manifested as poor coordination of the arms or legs (apraxia), tendency for the arm "to act as if it has a mind of its own" (alien limb phenomenon), numbness or odd sensations (cortical sensory loss), poor comprehension and/or expression of language (aphasia), and quick jerks (myoclonus). Slowness of movement (bradykinesia), stiffness in a limb (rigidity), fixed muscle contractions such as when the fingers curl into a fist (dystonia), and tremor are presumed to reflect basal ganglia dysfunction. Some patients develop memory impairment and/or personality/behavioral changes. Problems with walking eventually occur in almost all. In our studies the duration of illness from onset of symptoms to death has ranged from 3-13 years. The vast majority of patients do not appear to have any family history of dementia or parkinsonism, although there are rare cases in whom a hereditary process may be at play. The cause of CBD is not yet known.

This illness is frustrating to patients, their families, and the physicians who care for them. Since insight and memory tends to be preserved throughout most of their illness, depression is common and should be treated when it evolves. Physical, occupational, and speech therapy can be helpful although as the illness progresses third party payers tend to not reimburse for these services, unfortunately. Medications provide little benefit, but agents such as Sinemet are worth trying. All sleep disorders such as sleep apnea and restless legs syndrome should be evaluated and treated as improvement in quality of life for patients and their loved ones can occur.

I realize I have not painted a pleasant picture to those suffering from this illness and their loved ones, but I must be honest in what research thus far has taught us. The frustration over misdiagnosis is problematic for patients and families as many are diagnosed with Parkinson's disease or a stroke. Misdiagnosis for clinicians and researchers adds to the confusion regarding CBD, as our research has recently shown that only half of those diagnosed in life with CBD are actually found to have CBD when brain tissue is examined after death (by autopsy). Other disorders that can appear identical to CBD during life (also known as CBD mimickers) include Alzheimer's

disease (AD), Pick's disease, progressive supranuclear palsy (PSP), nonspecific degenerative changes, and rarely Creutzfeldt-Jakob disease. Thus a definitive diagnosis of CBD requires examination of tissue after death. The high misdiagnosis rate makes research on patients suffering from presumed CBD during life difficult to interpret.

However, recent research is shedding light on CBD. When brain tissue is prepared appropriately and examined by an experienced neuropathologist, the prominent abnormalities in a protein called "tau" as well as other findings helps establish the diagnosis of CBD. The functions of tau in nerve cells are complex and not fully understood, but it is clear that tau is required to bind to structures called microtubules for normal functioning to occur in brain cells (neurons). When something goes wrong in tau functioning, neurons eventually die. As more neurons die, symptoms progressively worsen, and usually focal atrophy in the brain becomes apparent on a CT scan or MRI scan. SPECT and PET scans can show abnormalities when CT and MRI scans are rather normal.

Therefore, abnormalities in tau protein are now thought to be the critical factor in the pathogenesis of CBD. Interestingly, tau dysfunction also is critical in the pathogenesis of Alzheimer's disease, Pick's disease, and progressive supranuclear palsy. It is highly possible that a treatment for one of these disorders involving tau processing will be beneficial for some or all of the others (although treatments for Alzheimer's disease that influence amyloid, which is an abnormal protein in Alzheimer's disease but not CBD, Pick's disease, or PSP, may not be effective for the non-Alzheimer's disorders).

How will a disease-altering or preventative treatment for CBD be developed? Let's consider what has already occurred in Alzheimer disease, where the identification of genes has led to major breakthroughs in our understanding of the pathogenesis of AD. Approximately 5-10% of patients with AD have a hereditary form in which roughly half of the members of each generation of a family develop AD. Three such genes have been identified through 1999 (and there are several more not yet identified). Mutations in which a single error in the DNA has occurred in these different genes all act to increase levels of a form of amyloid in the brain, which form structures known as amyloid or neuritic plaques. It is believed that these plaques somehow cause neurons to die and neurofibrillary tangles to develop (neurofibrillary tangles consist of abnormal tau, but this abnormal form of tau is different from that in CBD). Scientists have placed these genes into mice so that they develop amyloid plaques and thus they appear to develop Alzheimer-type changes. Developing strains of mice with abnormal human genes offers great opportunities to test various medicines to see if any prevent or delay the development of disease. This line of research has already led to one major discovery in AD (the vaccine against amyloid) and many more discoveries are likely to follow.

A similar approach is being applied to tau-related disorders. There are members of several families around the world who have developed what is called "frontotemporal

dementia and parkinsonism" or FTDP, in which abnormal tau is found in the brain tissue of those who have granted permission for autopsy. The findings are quite similar to CBD. Scientists from various institutions around the world pooled their efforts and identified several mutations in the tau gene that cause this illness (as of early 2000, there is no genetic test available for clinical use, but this may become available in the future). Clearly, there are other genetic and probably environmental factors involved in the pathogenesis of CBD, but strains of mice carrying the abnormal tau gene are being developed, and research with a variety of medicines will begin soon. Most researchers are quite optimistic that preventative and/or disease-altering medicines will be developed, but when this will occur, what side-effects will be present with treatment, and how costly treatment will be, are not yet known.

This is where patients and their families can contribute to CBD research. Talk to your local physicians to help identify a nearby institution where research on CBD, Alzheimer's disease, Parkinson's disease, etc., is/are being conducted, and consider participating in research. If you can't find anyone or any institution nearby, keep searching. It is clear that research involving "biologic tissue" such as blood samples, cerebrospinal fluid samples, autopsied brain tissue, all have enormous potential for advancing knowledge in this area. Considering whether to grant permission for eventual autopsy is not a pleasant issue to ponder, but this is one of the most important aspects of research in CBD and other disorders. Tell your legislators your thoughts on continuing adequate funding for neurodegenerative disorders research. Get involved in your local Alzheimer's Association chapter-the staffs at the national and local offices of the Alzheimer's Association are dedicated individuals who want to help. Let others know what has and has not worked in your journey with this illness. And maintain emotional and spiritual support for all those affected by CBD.

I strongly commend all those individuals have supported the Rare Dementia Registry and provided their words of wisdom in this monograph. Personally, I'd like to thank Alan McIlvaine, Theresa Roberts, and Darcy Croissant for their assistance to me and my colleagues who are actively involved in research on CBD (I'm sure my friend and colleague Dr. Caselli shares these thanks), as well as for their perseverance in the fight against this illness. Do not underestimate what impact highly motivated individuals can make in the fight against any illness—this superb monograph exemplifies what can be developed by dedicated and caring individuals.

Please realize that many gifted scientists from around the world are devoted to finding the cause of and cure for corticobasal degeneration. Through the Rare Dementia Registry we will keep you abreast of significant advances in CBD research.

Thanks to all who strive to optimize quality of life for those confronting this illness.

Brad Boeve, MD

Thanks to Dr. Boeve for the foregoing information.

Caregiving Attitude and Philosophy

There is a terrific book that covers the caregivers attitude and philosophy. *Your Name is Hughes Hannibal Shanks* by Lela Knob Shanks is a caregivers guide to Alzheimer's. Although the requirements of caregiving for CBGD patients is far different than Alzheimer's and their symptoms are far different, the book is well worth reading for the humaness intellectual insight and the real reason for personalized caregiving. I highly commend this book for your intimate knowledge of the real meaning of caregiving. It is available at your public library or local bookstore. If they don't have it, either can surely get it for you.

IT IS NECESSARY FOR US, THE PATIENTS AND CAREGIVERS, TO UNDERSTAND WHAT OUR DOCTORS, THE MEDICAL PROFESSION, ARE UP AGAINST AND WHAT THEY ARE TRYING TO ACCOMPLISH SO THAT WE CAN JOIN THEM IN THIS BATTLE TO LICK THESE INCURABLE BRAIN DISEASES. WE MUST FACE UP TO TASKS AND PROCEDURES THAT IN THE PAST HAD SEEMED INTOLERABLE.

This leads into a subject that is difficult, not only to think about, but very difficult to write about. "AUTOPSY" . Implied in all of our communications is the advancement of knowledge that will eventually lead to the prevention and cure of CBGD. For sufferers of CBGD who succumb, arrangements should be made to have the brain donated to some medical research institution for study and research in furtherance of our goal. Arrangements prior to death should be made through your physician.

Alan G. McIlvaine

Hospice

The first Hospice program in the U.S. was established in Connecticut in 1974. Today there are more than 3000 programs across the country. Hospice is historically a not-for-profit organization but with increasing health care costs, there are increasing numbers of for-profit hospices. Currently approximately 80% of hospices are not-for-profit or- as with V.A. hospices, are affiliated with a government agency. All of their services are palliative (to reduce the violence of or to moderate the intensity of a disease), based on the philosophy that every patient has the right to spend his or her remaining days in comfort, with dignity and pain free.

Their services vary to some extent from area to area but as a general rule, they may include some of the following: nursing care, social services, home care aides, medicines (prescription drugs and OTC), special equipment, bereavement support, inpatient care and respite for the caregiver. They also pay any doctor bills. Typically, as in our case, we just turned Barbara's Medicare over to Hospice and that took care of the financial end. If the patient does not qualify for Medicare some medical insurance plans cover it. Hospice usually will help if there is no way to pay.

Hospice was very helpful to us, especially in giving us advice and equipment that we would otherwise not know about. My advise is to get them involved as early as possible. This disease is not predictable so you cannot state a specific time frame so have your doctor request hospice services early on.

To learn more about hospice check out their web site on the Internet at the National Hospice Organization Internet Home Page **http://www.nho.org**

General information from their web site:
Considered to be the model for quality, compassionate care at the end of life, hospice care involves a team-oriented approach of expert medical care, pain management, and emotional and spiritual support expressly tailored to the patient's wishes. Emotional and spiritual support also is extended to the family and loved ones. Generally, this care is provided in the patient's home or in a home-like setting operated by a hospice program.

In recent years, many hospice care programs added "palliative care" to their names to reflect the range of care and services they provide – as hospice care and palliative care share the same core values and philosophies. Defined by the World Health Organization in 1990, palliative care seeks to address not only physical pain, but also emotional, social, and spiritual pain to achieve the "best possible quality of life for patients and their families." Palliative care extends the principles of hospice care to a

broader population that could benefit from receiving this type of care earlier in their illness or disease process. To better serve individuals who have advanced illness or are terminally ill and their families, many hospice programs encourage access to care earlier in the illness or disease process. Health care professionals who specialize in hospice and palliative care work closely with staff and volunteers to address all of the symptoms of illness, with the aim of promoting comfort and dignity.

- Medicare, private health insurance, and Medicaid in most states cover hospice care for patients who meet certain criteria. In addition, many hospices depend on charitable contributions to cover the costs of care for terminally ill patients who cannot afford to pay for their care.

In addition to this web site, the National Hospice and Palliative Care Organization offers information on local hospice and palliative care programs across the country and operates a toll-free Helpline at (800) 658-8898.

The National Hospice and Palliative Care Organization has a General Information Packet (GI Packet) with information on hospice and palliative care. If you would like a packet mailed to you simply call the HelpLine.

Alan G. McIlvaine

Legal Issues

"Estate Planning Close To Home Proves Its Worth" - by Tom Mills, Estate Planning, Napa, California

I have done estate planning work for over 30 years. For the first 17 years, it was all theoretical. It was what my grandfather called "book learning. None of my clients ever died. This was understandable since many of my clients were younger, and death was far off.

In the last 15 years, it has changed. The estate planning I do today is not based on theory or classroom training, it is built on real life experience. Until someone actually uses the strategies and appropriate documents that make up a major part of their estate plan, it is just a stack of papers. However, when the time comes to put them in force, they become so important.

In the past few days, I have personally sat with my own family as we sorted through these critical documents for my very ill mother. Forgive me for the personal references, but today they are the best teacher of the essentials of estate planning that I can pass on to my readers.

My mother died just after midnight on Jan. 4. The events, both before and now after her death, have again taught me the value of proper planning. About eight years ago she and I had a heart-to-heart talk about her plans. She had a very modest estate and saw little need to do much planning. Luckily, I convinced her otherwise.

As a single woman, she needed planning more than when she was married.. There was no spouse to pass things to by right of title. Her family, my brother and sisters and I faced a certain probate and many other problems if she did not set things up correctly. How grateful we are that she did the planning.

Mom's health had been declining for several months but after Christmas it accelerated. In a hastily scheduled medical appointment, her physician immediately hospitalized her. She had congestive heart and kidney failure compounded by pneumonia. He couldn't imagine how she had made it through the big family Christmas gathering at her home just a few days before. This annual family event was so important to her. I imagine she was toughing it out on grit and determination only to plummet afterward. Her condition worsened. Everyone knew where she kept her important papers, but now they were critically needed. initially three documents came immediately into

play. The first was her "health care power of attorney." Although she was fairly alert when initially entering the hospital, she quickly lost most of her cognitive powers. My sister, who lives close by, was given the power to make all medical decisions. She produced the document and the medical staff never questioned her authority.

Mom also had completed a "living will." This document basically stated her wishes, to be allowed, if it came to that, to die without artificial, life-prolonging assistance. I remember our extensive discussion about this matter. She was vehement about it. She did not want to be kept alive by a machine. She felt it was demeaning and too expensive to only prolong the natural events of life.

The 'living will" implementation was not as easy however as we wanted. Controversy arose when the physicians posed different treatments. There was some confusion as to what was considered life prolonging versus life saving. It is terribly important to be certain of the difference in the two. After the facts were out, several treatments were used that met mom's wishes.

The third document that was immediately used was a "durable power of attorney." There were bills to be paid and other expenses. Mom never wanted anyone to know about her assets. She was funny that way but also normal, by most standards. Again, my sister had been given the power of attorney. She went to the banks, and they almost immediately honored the document. Business life goes on even when health problems change everyday living.

Each of these documents served a valuable purpose as each was implemented. Now our family has moved to the next stage. The legality and use for the first three documents is finished. Now her 'revocable living trust" goes to work. Actually, since she has died, it is no longer revocable. It is a valid trust and expressed mom's wishes from the date of her death and beyond.

The next phase of her estate plan deals with the orderly distribution of her assets. Since her assets were quite modest, you may be thinking that a living trust was overkill. This is not true. Like all families, there are members who have different needs and financial abilities. A trust not only circumvents the expensive probate and legal oversight, it also allows for the special needs of any family member.

It will take some time to finish all the work ahead. Decisions regarding personal effects and furniture are necessary. Real estate values need to be established and decisions regarding a possible sale. IRA, saving and checking accounts, life insurance policies and pension and Social Security benefits all must now be coordinated with the trust. I expect it to go rather smoothly thanks to the estate planning and clarity of the trust document.

Thousands of dollars and countless hours of work will be saved because she Implemented the estate planning documents outlined for her years ago.

My mom has been In this column before. You recall she was the lady who had all her Christmas shopping done by Labor Day, She was a special person to me. Thanks to her, my family can focus on the legacy she left to the world of her children, grandchildren and great-grand children and not on the bickering about her assets. She put it in writing.
Where appropriate, it was correctly witnessed or notarized. We will miss her deeply, but her love for all of us was demonstrated again by relieving her family of problems. This was her lifelong pattern, and she was true to it to the end.

Thanks Tom for putting it all in perspective.

Legal Considerations

From Kevin McFadden, JD, Scottsdale, AZ

GETTING YOUR AFFAIRS TOGETER

1. Complete a will or revocable trust. A revocable trust will enable your estate to avoid probate yet will require you to transfer ownership of your assets to the trust. This may be time consuming but it can also ease the administration of your estate upon your death. Money spent now may greatly reduce costs and frustrations for your family upon death.

2. If you already have a revocable trust, work with an advisor to verify that all assets intended to be placed in your trust are in fact in your trust. He or she will also advise you on beneficiary designations for retirement accounts and life insurance policies.

3. If your estate is of a certain size, i.e. greater than $675,000 in the year 2000, you may want estate planning to avoid the pitfalls of the estate tax. See an estate-planning attorney to advise you of your options.

4. Prepare powers of attorney, both financial and medical. These will enable your family to assist you when you no longer can assist yourself. An attorney can assist you with these documents.

5. Prepare living wills. Living Wills are written directives to your doctor instructing him or her to remove all life supporting procedures if you are comatose or in a vegetative state. An attorney can assist you with these documents.

6. Call your insurance agent and make sure your life insurance policies are in order.

7. For those families that are in need of liquidity, there are a couple of options. You may consider the sale of an existing life insurance policy, the reverse finance of your home, which pays you the equity on the house, or greater withdrawals from you retirement accounts. Since these are very complex, visit with a professional on the advantages and disadvantages of these and other methods to create liquidity.

8. Organize your assets and bills. To avoid the time and difficulty for your family of determining what assets you own and what bills you owe, consider placing statements and bills in a folder. You will likely want to create a summary to

explain the statements and bills to your family. This will assist them in organizing your affairs upon death.

9. Make a list of all advisors, attorneys, accountants, insurance agents and other professionals to be contacted by your family to settle your affairs upon your death.
   Make sure your family knows where all of your records are and where the key to your safe deposite box is kept.

10. Go to a funeral home and make your request for burial or cremation. States usually have laws regulating these choices.

11. A great resource for forms and assistance is the local Hospice organizations, legal service centers for low-income individuals, libraries, and free seminars by attorneys and other estate planning professionals.

LEGAL AND TAX DUTIES TO BE PERFORMED UPON THE DEATH OF A LOVED ONE

Below you will find a typical list of steps necessary to resolve the legal matters that arise after a person passes away. While each individual case will vary, and every state has its own requirements, this will provide a broad guideline to assist the individual dealing with the loss of a loved one.

1. Nothing has to be done immediately. Take your time to grieve. You will find that most events are not emergencies and if they are, they are sometimes reversible. Typically, you have 6 to 9 months before you have to seriously look at the legal issues of an estate.

2. Find a friend, family member or reliable professional to assist you with those financial and legal decisions. Remember that nothing has to be done immediately.

3. Unfortunately, you will be contacted by salespeople, attempting to sell you all types of items they claim you will “need.” This is not the time to make those decisions.

4. While Social Security needs to be notified, funeral homes often now notify the agency. Eventually you will need to contact the Administration to set up your own plan. There is no rush but they should be contacted as soon as you are

Thanks Kevin for the guidelines.

Glossary of Terms

by Geri Hall, Ph.D., ARNP, CNS, Associate Professor and Associate Director for Outreach, University of Iowa Center on Aging.

Abulia - Functional errors of omission: failing to perform activities to meet basic human needs: inability to make decisions; lack of will or willpower.
Acalculia - inability to do simple arithmetical calculations
Anomia - inability to recall or recognize names of objects
Aphasia - loss of power of expression by speech, writing, or signs and/or loss of comprehension of spoken language or written language due to brain injury or pathology
Apraxia - loss of ability to carry out familiar, purposeful movements in the absence of paralysis or other motor or sensory impairments, especially the inability to make proper use of an object
Apraxic agraphia - inability to express oneself in writing due to apraxia
Asimultanaanosia - Inability to visually integrate the components of ordinarily complex scene into a coherent whole
Atopoaraphaonosia - inability to recognize familiar faces
Aural comprehension - understanding of stimuli perceived by the ear
Constructional praxis - inability to copy simple drawings or reproduce patterns of blocks or matchstick constructions
Dysarthria - imperfect articulation of speech due to muscular weakness resulting from damage to the central or peripheral nervous system
Echolalia - stereotyped repetition of another person's words or phrases
Executive function - ability to set a goal, make decisions, and implement appropriate activities towards meeting that goal.
Ocular apraxia - inability to voluntarily direct their gaze to a target of visual interest
Optic ataxia - the inability to benefit from visual guidance in reaching for an object
Paraphasia— speech defect characterized by disorderly arrangement of spoken words
Phonemic - speech sounds that are the basic units of speech (i.e. "leviator' instead of 'elevator;' or .grontologs" instead of "gerontology")
Praxis - the performance of an action; "doing"
Prosody - the variations in stress, pitch, and rhythms of speech that convey meanings
Prosopagnos'ia - inability to recognize faces
Semantic paraphasia - substituting a similar word for an object, i.e., "staple" for 'paper clip' (Caselli, 1995, p. 3)
Semantic precision - use of words appropriate or significant to the meaning of the intended communication. i.e. substituting "machine" for "automobile".
Verbal memory - ability to remember speech
Visual memory- Ability to remember what is seen

Thanks Geri for unscrambling this verbiage for us.

We recognize that the word **dementia**, as defined by Webster, has many people associated with CBGD deeply concerned. Dementia, in the form of memory or intellect loss is not as prevalent in CBGD patients as it is with Alzheimer's or some other similar brain diseases.

Geri Hall has a much more acceptable definition of **dementia**. The word **dementia** "describes the set of symptoms that occur when the cerebral cortex, the thinking, acting, doing part of the brain is damaged permanently. Symptoms may or may not include memory loss, judgment, consciousness and orientation, visual loss, difficulty in motor planning, use of language, behavior and intellect.

With this understanding, the word **dementia** is much more acceptable to me considering the degree of **dementia** discernible in my wife Barbara's behavior in the latter days of her illness.

Alan G. McIlvaine

Network Informational Resources

I urge you all to join the **'Rare Dementia Registry'** of the Phoenix Chapter of the Alzheimers' Association, **NORD's Rare Disease Registry** and if you have e-mail capability, join **Theresa Roberts' CBGD support group.** Details follow. The more CBGD people on these lists the more attention we will attract. We need attention in order to encourage medical research in CBGD. It will also be another avenue for you to use in soliciting advice, assistance and support and in providing the same to others. It is extremely important that ex-caregivers join because we have the experience and knowledge that others are searching for.

CBGD Online Support E-mail List (Updated 7/12/2001)

A mailing list created for anyone affected by CBGD (Corticobasal Ganglionic Degeneration) a rare progressive degenerative brain disease. I hope this mailing list will make it easier to communicate with others who are also dealing with this disease. I remember the first time I spoke with someone else dealing with this disease was that I/my dad wasn't alone! Someone finally could relate to what was happening. Please join in!

You can join this list by going to the following web page:
http://groups.yahoo.com/group/cbgd_support
There are approximately 255 subscribers to the list as of July 12, 2001

Kristine Manion
List Manager
kahm99@yahoo.com

Note: Please join, there is no charge for anything.

National Organization for Rare Disorders, Inc

NORD* PO Box 8923# New Fairfield, CT. 06812-8923
(203) 746-6518 FAX (203) 746-6481
e-mail: orphan@raredis.org

Nord maintains a Rare Disease Registry which includes CBGD. Contact them for specifics. The database is confidential and will be shared only with your permission.

Rare Dementia Registry
Greater Phoenix Chapter, Alzheimer's Association

This registry is made up of individuals who have been diagnosed with a rare dementing illness and their primary caregivers. The registry functions as a telephone support group. This database is confidential; names will be shared with other registered families only.

Rare Dementias that are listed on the Registry.

* **Cortical Basal Ganglionic Degeneration**
* Pick's Disease/Frontotemporal Dementia
* Progressive Aphasia
* Progressive Cortical Visual Syndrome
* ALS - Dementia Complex (Rare)*
* Other

In order to support the families dealing with these rare diseases that are related to, yet different from Alzheimer's, the Alzheimer's Association felt the need to create a new type of support system. The Alzheimer's Association already manages programs such as Support Groups, Care Management, and Helpline for families affected by Alzheimer's disease and related disorders, and is particularly sensitive to the support needs of those dealing with aging-related issues.

If you, or a family member, are diagnosed with one of the rare dementias listed above, simply apply to the Alzheimer's Association. Your name will be added to the registry and given to other registered families when appropriate. There is no fee for this service. When your application is received, you will be contacted by the Alzheimer's Association. Information is available on the related dementias and will be offered to you. Names of registered caregivers who are coping with a similar disorder will be shared with you when available.

Alzheimer's Association, Greater Phoenix Chapter

1028 East McDowell Road
Phoenix, Arizona 85006-2622

Call their Helpline
(602)528-0550
1-800-392-0022
toll free in Arizona