# VHL Family Forum

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# A Family's Milestone: Kidney Transplant

*Adapted from an article by Frederick Melo, Assistant Editor, the Brookline Tab, August 1, 2001**

If wishes were horses, Joyce and Damon Graff would own a herd to ride down the leafy streets of Brookline, beating a swift retreat from words like tumor, metastasize and malignant.

Instead, on Wednesday, July 25, this mother and son capped a milestone in their 15-year struggle to educate themselves and the medical community about von Hippel-Lindau (VHL). In a three-hour operation at Beth Israel Deaconess Medical Center in Boston, doctors removed Joyce's left kidney and successfully transplanted it into her son, ending his two and a half years on dialysis.

It was a mother's sacrifice, accomplished after a year of dieting, soul searching, meditation and prayer.

VHL has plagued their family the past three decades. A bout of nausea and vomiting once raised the question: "Is it the flu, or is it a brain tumor?" Fortunately an MRI assured them it was the flu, but questions like this are not unusual in families grappling with VHL.

In 1977, Graff's husband, Frank, 35, died after a 20-year battle with Von Hippel-Lindau syndrome, an often misdiagnosed and overlooked multi-system disorder that had claimed his own father's life as well. VHL is a genetic tendency toward formation of vascular tumors of the retina, brain, spinal cord, kidney, pancreas, or adrenal gland. Some of these, especially in the kidney or pancreas, can progress to cancer if not treated appropriately.

Then, little more than a year after her husband's death, doctors discovered a cancerous lump in Graff's right breast.

With her 6-year-old son, Damon, having just lost his father, Graff opted for a mastectomy to spare him the sight of her exhaustion and uncertainty under chemotherapy.

"I needed a clean answer," remembers Graff, fighting back tears for the first time in a series of interviews on her family's health. "I needed to hear 'OK, you're fine. We got it all.'

The operation saved Graff's life, but her troubles weren't over. In 1986, Damon, then 15, was diagnosed with Von Hippel-Lindau syndrome. In Damon's case, the disease attacked his kidneys.

Doctors urged Graff to allow them to remove both of Damon's kidneys and put him on dialysis while awaiting a kidney donor. She resisted, even when doctors emphasized she might be playing roulette with her son's life. He was a healthy, teenager with no symptoms, and she did not want to put him on such an arduous course at such a young age. She needed more information, from doctors who had seen more than one or two cases, and who knew more about the growth and development of kidney tumors.

Determined to know her enemy, Graff began urgent research into Von Hippel-Lindau. It was a decision resulting in the establishment in 1993 of the Von Hippel-Lindau Family Alliance, a network of more than 10,000 people afflicted with the illness in 67 countries. The VHLFA has established a toll-free help line, a website, and an international support group.

*cont'd on page 2*

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But in those early days, little was known about the growth and natural history of kidney tumors. An article was written about her son's case, noting that he was the youngest person in which they had ever seen kidney tumors. But Graff found that at that time he was one of the youngest they had ever screened for VHL in the kidney. She found doctors in Kansas, England, and Germany who were able to assure her that a more conservative course of action was not stupid, it was in fact the right thing to do. Nonetheless, Damon underwent multiple surgeries to forestall the cancer's advance. He lost one kidney at age 19. Two and a half years ago, another operation failed to save his second kidney, forcing him into a regimen of dialysis that didn't end until last week.

"He's had four kidney surgeries. The organ can only take so much before it fails," Graff explained. With better surgical techniques, it is possible now for a kidney to survive as many as 3-4 surgeries, but the trick to staying on your own kidney power is to minimize the surgical assault on the kidney – delay surgery until the largest tumor is approaching a dangerous size, and/or treat tumors with less invasive techniques like Radio Frequency Ablation (which uses a heat probe inserted through the skin, much like a needle biopsy). In Damon's case, early efforts to minimize the requirement for blood transfusion caused extensive scarring to the blood vessels feeding the kidney, and this scarring complicated the future surgeries

After his second kidney failed, Damon went on dialysis three nights a week, five hours at a stretch, while his name lingered on a waiting list for a kidney donor that in New England was nearly five years long.

Dialysis was clearly impacting his health. Tumors on his retinas had already cost Damon his left eye, and laser surgery to remove a tumor from his right eye left a permanent blind spot Damon likens to having just stared at the sun. His best chance for long-term transplant success was a living related kidney donor.

Graff offered to donate, but at first was rejected. Graff, who had long struggled with obesity and high blood pressure, was in her late 50s and, at a shade over 5 feet-tall, weighed well over 200 pounds. Even a trip up a flight of stairs left her winded. Damon's Aunt Susan Leonard from California volunteered. A DNA test confirmed she did not have VHL, and she was accepted as a kidney donor, but at the last minute, literally on the day of the scheduled transplant, with suitcases in hand, they were told that the doctors wanted to postpone for six months to watch a cyst on his spleen, to be certain that it was not metastatic cancer. The disappointment was devastating to all. Six months later when the cyst had disappeared and Damon had medical clearance, Susan's husband was facing heart surgery and three joint replacement surgeries, and she was unable to proceed.

*Damon Graff*

Joyce went to two other transplant teams, trying to understand what it would take for her to qualify as a donor. Dr. Martha Pavlakis and the transplant team at Beth Israel Deaconess Hospital in Boston helped her set realistic goals to ensure that donating a kidney would not be harmful to her own health. They told her she would have to lose 70 pounds and get her blood pressure under 150/90 on one blood pressure medication, not two.

In the meantime, dialysis was claiming a large section of Damon's time and energy. Waste unable to be cleaned out through the procedure would seep into his skin, causing him constant itchiness that resulted in welts as he scratched in the night. The treatments forced him to limit his liquid intake, causing constant thirst.

The dialysis also became a scheduling nightmare for Damon, who traveled frequently in his position as an advertising salesman for high-tech magazines. He would contact hospitals around the country weeks in advance of his arrival, only to hear that they would not be able to confirm a dialysis treatment until a few days before the appointment. Nonetheless he continued to work and travel in the U.S. and Europe.

Graff, meanwhile, set to work on herself. "I had to face the fact that I was using food to manage stress. I had to find other healthier ways to deal with it."

Under the advice of nutritionists and personal trainers, Graff avoided starvation diets, water-loss diets, diets that promised instant miracles and lifelong dreams achieved overnight. She knew that sudden, drastic weight loss could damage her kidney, weaken her and make her useless to her son. Doctors warned that losing more than two pounds per week could be dangerous, or result in her regaining the weight.

Graff went through the Cardiac Wellness program at Beth Israel, a combination of exercise, nutrition, and

meditation, under the direction of Dr. Herbert Benson, designed to control blood pressure and other cardiac risk factors without medication. This program was funded in part by her health insurance. She also began a progressive program of weights, low-impact aerobics, and a host of exercises prescribed by trainers at the Fitness Connection in Coolidge Corner, a fitness site that became one of her regular haunts. She met with a personal trainer an hour every 6-8 weeks to adjust her program as she progressed.

"I'm exercising muscles I never knew I had," said Graff, who now benches 60 pounds regularly.

Last week, on the day before the kidney transfer, she was ready.

"When the doctors set those goals," Damon chuckles, "I don't think they thought she would do it, but I knew from the first that she would. They didn't know who they were dealing with." Nonetheless, Damon was almost afraid to be excited at the prospect of the transplant. He knew all too well that there are many things that can go wrong. A cousin who volunteered and went through six months of testing was disqualified due to factors that might put him at risk in the future with only one kidney. "In October 1999 before I met Aunt Susan at the hospital I had taken flowers to the dialysis nurses to thank them, and made a big deal of ending dialysis, only to wind up back there the following week. This time I don't want to do anything to jinx myself. I'll believe it when it happens."

Joyce looked almost cheerily at the surgery ahead.

"People ask me if I'm afraid of the surgery. I'm actually very excited. It's almost like having a baby," she said, sitting in the living room of her Brookline home hours before going under the knife. "You don't look forward to labor and delivery, but you do look forward to a great outcome ... There's a purpose to this, there's a mission."

On the day of the operation, Damon walked the mile from his apartment to the hospital. Joyce wanted to walk the two miles from her home as well, to enjoy the fresh air. Her sister Margaret Smith insisted otherwise and drove her to the hospital.

Three hours after the first incision, it was over.

"They had us in beds that were next to each other in the recovery room," Joyce said afterward. "My sister was standing between our beds, holding one of each of our hands, and I said, 'Tell him I'm sending a hug his way.'"

For the first time in two-and-a-half years, Damon could eat the foods he and his doctors have long since relegated to his list of "forbidden fruits" — gourmet coffee with cream, pizza, bananas, oranges and ice cream, hot chocolate with breakfast, macaroni and cheese and more ...

It had been more than two years since he had peed on his own.

"But the best thing of all," Damon says, "is that I no longer itch. I have been itching like crazy for years now, and I am now much more comfortable in my own skin."

Today, Graff and her son are back home in Brookline, stitched up and healing — both literally and figuratively.

"The new kidney wasted no time in getting down to business," says Damon. "It's rather like when my mother comes over to my apartment and immediately starts cleaning up. One of the big measures they use to determine your kidney function is to test your creatinine level. A normal person's creatinine scores between 0.5 and 1.2. Just after surgery, mine was about an 11. The next day it was down to 6, then 4, then 1.7, then 1.4. The creatinine reflects that the kidney is working great."

Now Damon has between 20 and 30 years ahead of him living on his mother's organ, and he's successfully chipped away at his core symptom of VHL. Hopefully by then there will be an ever better answer – maybe using stem cell technology to grow him a new kidney of his own cell structure!

"The VHL is not out of my system. I still have to deal with it in other areas — it's still in my eyes, and there are other areas I have to keep checking — but the biggest problem is out of the way. It feels like I can finally get on with my life," Damon said.

After the death of his father, if there was anything VHL robbed of him in addition to his eyesight and physical comfort, it was time. With his lengthy dialysis treatments hopefully in the past for good, Damon has enrolled in a part-time graduate business program to further his career. Shortly before the operation, Damon took his graduate school entrance exams and scored 770 out a possible score of 800, or among the top 1% of those taking the exam.

"It's still going to be a lot of hard work for him," says Joyce. "He still has to take a number of medications, and learn when to take what in order to avoid kidney rejection. But he'll do it. He has always taken responsibility for doing what it takes to maintain his health."

"It's my life," says Damon. "Overall I am feeling great, ready to begin my new life."

And so far? They both smile. "His new, slightly used, kidney is serving him well."

### *Research Campaign*

This year the Annual Report replaces the Research Report. We hope that you will share copies with your family and friends, and encourage them to help us meet our goal of funding one more research grant in full next year! Let's cure VHL in this decade!

Write us in on your United Way campaign!

Federal workers, choose # 1098 in the CFC campaign!

# Q&A About Kidney Transplantation

*Question: Are people with VHL candidates for transplantation?* -- Yes.

*Question: There is a general recommendation for a waiting period of two years. Can this be shortened?* -- Yes, as long as the kidney tumors removed were small enough (under 3 cm) and well confined, to present a low risk of metastatic disease. Where there are questions about the progress of the cancer, a waiting period of two years is a wise thing to do. If there is a cancer anywhere in the body, the immune suppression required to keep the transplanted kidney will cause the cancer to spread. When in doubt, wait.

*Question: If a new kidney is transplanted into someone with VHL, won't that kidney also be at risk for VHL tumors?* -- No. VHL tumors are not carried in the blood to the new kidney. VHL tumors are "neoplasias", new growths, arising from a single cell whose VHL protein is missing. The cell structure of the new kidney is that of the donor, not the recipient, and therefore is not at risk for VHL tumors.

*Question: How well do people with VHL do on transplantation?* -- A study published in 1997 in the Journal of Transplantation showed that people with VHL do at least as well as people with other conditions. The average age of a kidney recipient with VHL is 20 years younger than people with other conditions, and people with VHL generally lose kidney function abruptly as a result of surgery rather than going through long-term degradation of kidney function. There are continuing advances in the anti-rejection drugs used, and there are experiments with using stem cells from the blood of the donor to accustom the recipient's body to the donor's tissue.

*Question: The wait for a cadaver donor[1] can take three to five years. How can that wait be shortened?* -- The need for organs rises every year, but the number of donors has not risen significantly in the past five years. The best chance of early transplantation and the best long-term outcome is a living related donor. Next is another living donor who may not be related but has the same blood type as the recipient and a satisfactory match. A spouse or a friend can also be a donor. Unrelated living donors give essentially the same results as a related donor, and significantly better than a cadaver organ.

*Question: If a healthy relative is an excellent match and is eager to donate a kidney, why all the delays and tests?* -- It is essential to assure the donor that everything after the donation is going to be the same as before. The donor undergoes a very thorough medical evaluation, beginning with DNA testing to confirm that this person does not have VHL. If we find anything that poses a potential risk to that individual, even if they might be the best possible match, we can't do it. The most important thing is to preserve not only the life, but the quality of the life of the potential living donor. The kidney that remains in the donor must be absolutely normal. We have to be able to carry out the procedure successfully with the recipient and also not compromise the donor.

*Question: What does it entail for the donor?* -- In some cases, the kidney can be removed laparoscopically, through a small incision, minimizing scarring for the donor. In other cases it may entail a 3-6 inch incision and open abdominal surgery. Depending on the surgical approach required, the recovery may take 2-6 weeks.

*Question: If a transplanted kidney would not grow VHL tumors, shouldn't we simply take out both kidneys and move directly to transplantation? Isn't that a "cure" for VHL tumors in the kidney?* -- No, transplantation is not a cure. It is a serious medical path under which you are a patient your entire life, taking an expensive set of medications every day to keep the organ from rejecting. If you can stay on your own kidney power, you will be healthier and have fewer side effects. Transplantation is better than dialysis, but it is almost always better to be on your own kidney power, even if you have only part of one kidney, as long as the risks of metastatic cancer can be kept sufficiently small.

*Question: What is the best way to manage VHL tumors of the kidney?* -- The keys to managing VHL kidney tumors are to find the tumors early, use laparoscopic cryotherapy or radio frequency ablation where possible, and delay any invasive surgery until the largest hard tumor is approaching the 3 centimeter level. At that point, the preferred treatment is usually a partial nephrectomy using techniques to "scoop out" the tumors, minimize vascular damage and leave as much functioning kidney tissue as possible. It is important to continue keeping a close watch on the kidney, and there will likely be additional tumors within 5-10 years which will also need to be followed and treated appropriately.

*Question: Is there anything the patient can do to minimize or slow down tumor growth?* -- We know that smoking causes kidney tumors to grow more rapidly. The most important thing is to keep the body strong and healthy so that your body's normal self-repair mechanisms are working at peak performance. Other than that, we have not yet identified any risk factors that would be under your control.

1. A cadaver organ comes from a person who is brain dead due to an accident or stroke, and who has signed an organ donor card. Once the heart stops beating entirely, the kidney cannot be used.

2. Our thanks to Dr. Oscar Salvatierra, Dr. Andrew C. Novick, Loriann Marquardt, and Damon Graff for their assistance in the preparation of this article.

# Stem Cell Research and VHL

*By Joseph Verdi, Ph.D., London, Ontario, Canada*

A great deal of interest in the scientific literature and in the printed media has focused on "stem cells" and their potential therapeutic application for the replacement of diseased or damaged tissue. It is not too far in the future where effective treatments and cures for such diseases as Parkinson's, ALS and Diabetes may be able to utilize stem cell replacement therapies to correct the disorders. VHL is another primary candidate for benefit — perhaps not so much a total systemic cure, but certainly help in battling and reducing the need for repeated surgeries.

Stem cells are defined in scientific circles as multipotential, self-renewing cells capable of giving rise to multiple specialized tissue types. In plain language, this means a set of undifferentiated cells that have not yet "specialized" to become any particular tissue or organ. Such cells can continue to divide until division gives rise to one daughter cell that can form a specialized cell (kidney, liver, spleen, blood or brain) and a second cell that is a copy of itself. In this way this process can go on indefinitely. It is obvious that one potential use of stem cell technology is to create an unlimited supply of healthy replacement organs for transplantation. In the case of VHL, it means eliminating finding donors for kidney transplants, long waits and countless disappointments before such transplants

> "There are legitimate concerns about the potential misuse of stem cell research. . . . Just as there are careful controls on the ethical treatment of laboratory animals, and bans on the sale of organs, so too there should be careful ethical controls on the sources and use of stem cells."

can be performed. Moreover, it is not a pipe dream or wishful thinking to say that within the next five years,[1] if funding continues, that science will understand the mechanisms by which stem cells choose one lineage or organ type over another and be able to manipulate stem cells to generate whatever tissue type is required. Furthermore, with advances in gene therapy it is not far in the future (perhaps only 5-10 years) when we will be able to harvest stem cells from an afflicted individual, use gene therapy to correct the defective gene and differentiate those stem cells into any organ required for transplant. This would eliminate the need for immunosuppressson therapy to block rejection since the donor would be the recipient him or herself. Better than that, the VHL mutation could be repaired in the new kidney, so it would not be at risk for VHL tumors. A new kidney, without the VHL mutation, with no risk of rejection!

Unfortunately, stem cells have also become synonymous with embryonic stem cells, which are derived from the inner cell mast of miscarried or aborted human embryos. But this is not the only source of stem cells. Embryonic stem cells retain a greater ability to form almost all specialized cells in the body, but stem

> "Stem cells may also be found in the umbilical cords of healthy babies, or even in adult tissues like hair or skin."

cells may also be found in the umbilical cords of healthy babies, or even in adult tissues like hair or skin. There are legitimate ethical issues to be dealt with, but imagine being able to grow a replacement kidney from a single strand of your hair! Imagine repairing spinal cord damage to free someone from chronic pain or help them walk again!

There are legitimate concerns about the potential misuse of stem cell research. One is the fear that scientists may clone people, or that people might fertilize human eggs only to "farm" fetuses and harvest stem cells. There are already laws that forbid such reprehensible acts. Nearly every powerful new technology can not only be used for powerful good, it also has the potential for misuse. Should we give up all the potential benefits because of fears of the dark side? Or should we instead learn to control the power and harness the benefits? Just as there are careful controls on the ethical treatment of laboratory animals, and bans on the sale of organs for transplantation, so too there must be careful ethical controls on the sources and use of stem cells.

While enthusiasm for the potential medical benefits of stem cell therapies is high, and justifiably so, the development of such technology still requires a greater understanding of the basic biological properties of stem cells. There is a great deal of expensive research to be done to achieve the goals, which is why governmental funding is so very important. In my opinion, the pros far outweigh the cons, and the potential benefits for people with VHL are enormous.

1. While scientists will likely understand these mechanisms in another five years and be able to perform these procedures in the laboratory, FDA approval for mainstream treatments would likely take at least five years beyond that.

Stem cell therapy (now still experimental) can help in diabetes, stroke, cancer, spinal cord injury, and burn repair. It can help regenerate or repair kidney, pancreas, nerve, adrenal, and retinal tissue. The development of stem cell research is a major scientific breakthrough. -- *US National Institute of Diabetes, Digestive Disorders, and Kidney*

# Palo Alto Meeting a Great Success

On a beautiful sunny California day, 146 people gathered in the Fairchild Auditorium, at Stanford University to talk about von Hippel-Lindau disease.

There were 20 physicians, 6 nurses, 2 genetic counselors, and a physician's assistant. The remainder were people with VHL, their families and friends. Nearly half (67) were from California. Ten people were from Canada, two from England, and one physician came from Colombia in South America.

"I was astonished," said Raeanne M. of California, "to be in a room filled with people who have the same surgery scars that I do! Where else could I find so many people like me? This was my first conference and it exceeded my expectations. Meeting people was great. And, I also got to meet and directly question the top doctors and scientists in the field of VHL intervention and study. Too bad you missed the lecture on killer ubiquitin chains! They are working on developing drugs that will intercept and hopefully correct the errant cell biochemistry on at least four levels."

Dr. Michiel Innes, a geneticist from Alberta, Canada, commented that he "enjoyed it tremendously. As a geneticist I often find myself playing the role of case coordinator and patient advocate so hearing and learning so much from a patient perspective was very rewarding. Things have already paid off as two days after my return I saw a woman in my general genetics clinic who almost certainly has VHL and I certainly felt prepared!"

Hetty DeVroom, clinical research nurse for Dr. Oldfield at the U.S. National Institutes of Health,

Dawn Cerf (left), California Chapter Chair and Chapter Chair of the Year, led the group in simple Tai Chi breathing exercises and movement on Sunday morning, a wonderful stress management technique.

reported on the VHL Clinical Program at NIH which has now seen nearly 800 people with VHL. She presented the screening protocol they recommend to local physicians. Joyce Graff explained about the VHL Clinical Care Centers program, which helps to carry information from the expert centers, like NIH, out into the field.

Dr. Ronald Bachman, Chief of Genetics at Kaiser-Permanente Oakland Hospital, reviewed the genetics of VHL. Dr. Wayne Fung, Clinical Professor of Ophthalmology at University of California San Francisco (UCSF), reviewed the ophthalmological issues in VHL. He stressed the importance of careful screening, beginning by age 3 and continuing as often as every six months through puberty. "The smaller the lesion is when we discover it, the more favorably it will respond to treatment."

Dr. John Adler, Professor of Neurosurgery at Stanford and Medical Chairman of the meeting, explained Stereotactic Radiosurgery (SR) and where it is most effective. He stressed that the "gold standard" for treatment of VHL hemangioblastomas of the brain and spinal cord is still traditional microsurgery. However small, well-qualified lesions can often be treated with SR. Dr. Mitchell Berger, Chief of Neurosurgery at UCSF, explained the approaches used in microsurgery. The cyst must be drained when necessary to relieve pressure, but the cyst walls may be left in place as they are not the problem. The cyst is created by the tumor, so until the tumor is removed or treated with SR, it will continue to refill the cyst. He explained that the emerging field of brain mapping allows the surgeon to locate the areas of the brain that control speech and motor functions prior to surgery, and plan a surgical approach that will avoid these areas and preserve function. He went so far as to say that if surgery will involve the supra-tentorial area of the brain, that patients should "demand" brain mapping, which is now available at most large medical centers.

Dr. Randall Hawkins, Radiologist at UCSF, explained the state of diagnostic imaging, especially Positron Emission Tomography, or PET scanning. PET scanning allows us to see the metabolism of glucose or other substances by the tumors, giving the physicians better information about the activity level of different tumors. It is less useful at this time in the kidney, but can be a helpful adjunct to CT and MRI outside the kidney. PET scanning has become a great deal more available in the last year since it has now been approved for Medicare reimbursement.

Dr. Yasser El-Sayed gave an excellent talk on Pregnancy and VHL, which will be published as an article in a future newsletter. There are two primary areas of concern for women with VHL during preg-

nancy: pheochromocytoma and lesions of the central nervous system (CNS), or brain and spinal lesions. It is always best if the mother has been checked prior to the pregnancy, and any lesions requiring treatment have been appropriately treated. The reality is, however, that many pregnancies are not planned, and that many women do not have a diagnosis of VHL before the pregnancy begins. The more is known about the mother's condition, the easier it will be for the medical team to monitor the situation and avoid critical problems. Communication is critical. "The patient must be a real participant -- counseling, consent, understanding of the ambiguities." There is some evidence that there may be progesterone receptors in VHL tumors of the CNS that may cause the tumors to grow during pregnancy, but the situation is unclear. Most women do not have good "before" pictures, so it is hard to tell how long the tumor had been there. It is also hard to tell whether the tumors grew because of the pregnancy, or because they were simply going to grow that season.

*Left to right: Earl and Patricia Rasmussen, Jill Shields, Joe Verdi, and Kathy Braden. Patricia is our new Director of Clinical Care, Jill and Joe are Directors of the Canadian VHLFA, and Kathy is Vice-Chair of the Alliance.*

In his review of the literature, he finds that pregnancy should not be discouraged in women whose lesions are asymptomatic or have been adequately treated prior to the pregnancy. However it is extremely important to know what you are dealing with, and to make sure that the entire team is in communication.

Myriam Gorospe, Director of Research, announced this year's VHLFA grant awards (see page 14).

Dr. Graeme Eisenhofer, Director of the Clinical Neurochemistry Laboratory at NIH, explained the test for plasma metanephrines which is a much more specific test for pheochromocytoma than the 24-hour urine collection. It can be very useful in identifying pheos which are less active, or which are intermittently active and therefore difficult to diagnose. His group has also been working with PET scanning using fluorodopamine, which is proving to be even better than MIBG in locating pheos.

Dr. Fabiàn Fonseca Guzmàn, urologist from Colombia, presented the work he did for his doctoral dissertation in Costa Rica on the diagnosis and treatment of VHL in the kidney. Dr. Oscar Salvatierra, Jr., Director of Pediatric Renal Transplantation at Stanford, explained the state of the art of kidney transplantation, and issues specific to people with VHL (see page 4).

Tom Rodenberg, attorney from Blue Springs, Missouri, gave an excellent talk on dealing with insurance companies, which will be published as an article in a future newsletter. His most important messages were "read your contract" (a policy is a legal contract) and "don't give up!"

Dr. Robert Williams, Medical Director of the Gould Medical Foundation, talked about "utilization management" in the Health Maintenance Organizations (HMOs). In order to gain approval for scans or tests, he stressed that you need your doctor to write the right codes and justification for the tests. Without that information, the funder will probably reject the expense. But if the physician explains what is suspected, what they are looking for, or that this scan is essential in the responsible surveillance of this condition, then it should be approved. It may take some persistence.

Dr. Peter Jackson, Assistant Professor of Microbiology at Stanford, gave a very clear explanation of the state of our knowledge of the function of VHL in the cell. The VHL protein joins with elongins B and C to form the VBC Complex. This complex functions as a kind of garbage collector in the cell, marking a number of substances for deletion, similar to marking trees in a forest that should be taken down. This process serves as a kind of "off" switch for a number of functions in the cell, controlling those processes. Without functioning VHL protein, these cell functions cannot be turned off and go out of control. There are a number of very promising lines of research that will likely lead to drug therapies for VHL in the near future.

Debbie M. from Florida, found the meeting "wonderful and very encouraging. The conference is a must for anyone who has never had the chance to meet someone in person who is handling all the medical issues and the stress related to them with VHL and to see how everyone really strives to go in positive directions."

"What an experience it was meeting everyone at the symposium, and how emotional!" said Sue L. from Canada. "When you think that you are the only family faced with this disease, you feel so isolated. Hearing the experiences of all the families that were there, you realize that we were all in the same boat, and thank goodness for the VHL Family Alliance."

# New Canadian Leadership

*by Jill Shields, Director, Canadian VHL Family Alliance*

The VHL Family Alliance is just that - We're a family. People helping people, people reaching out and giving a hand where strangers will not. The new board members reflect this family affair. By way of introduction let me describe to you how we all came to be involved. The four of us come from distinct walks of life, different socioeconomic backgrounds and different careers but are galvanized by the spirit and family of our incoming Co-director, my sister, ***Susan Lamb***.

Sue has three wonderful children, two of whom are afflicted with VHL. Brent, 25, Devon, 23 and Kellaway, 21 are her life, along with a very supportive husband and two stepchildren. She retired last year due to health problems of her own, after working for 34 years in the financial industry. She is a tenacious, no-holds-barred, find-the-truth type of individual when it concerns finding facts about VHL. Her friends call her Susie WaWa, in reference to her ability to ask the right questions and get answers like Barbara Walters, who was satirized on TV's *Saturday Night Live* as "Baba WaWa". Susan is the most down-to-earth person one could imagine and her greatest asset is that she truly cares.

Our first introduction to VHL was in 1973 when my late brother-in-law, Bill, was diagnosed with von Hippel Syndrome after tumours burst in his eye. Unfortunately, laser surgery had not been perfected at that time and he lost sight in that eye. It wasn't until Sue was pregnant with her first child, Brent, that doctors informed us about the Lindau part of the disease and were told any of Bill's children had a 50% chance of inheriting VHL. The ophthalmologist that Bill was seeing explained that Bill could develop benign brain tumours and so he had annual brain scans. Each scan petrified my sister, not knowing what the outcome would be, but she was strong and reassuring to her three young children. In 1991, after many appointments with various specialists, Bill was diagnosed with metastasis in the spine caused by renal cell carcinoma and passed away at the age of 43 in December of 1991.

At this time, knowledge pertaining to the disease was limited, but Sue researched what she could to try to understand this life altering disease. She has a good friend that went to her local University library to find out what she could on VHL. I too canvassed the halls of The John P. Robarts Research Institute, where I work as an administrative assistant, to find answers. Needless to say, we were shocked at what we read

*Susan Lamb and Joe Verdi*

from various medical publications concerning all of the parts of the body that VHL could affect. Sue then set out to find doctors in the Southern Ontario region who would ensure that her children, my nieces and nephew, were being given the correct screenings. We were appalled at the number of physicians who hadn't heard of VHL.

Five years ago, one of Sue's daughters was found to have an eye angioma, and through a literature and Internet search we thankfully found the VHL Family Alliance website, and made our first phone call. Sue and I spoke separately with Eva Logan, (a hot line volunteer). She was truly wonderful and had all the current information and latest literature sent to us. I personally saw how Sue's frustrations and fears were lessened from just one person lending an informed ear. What a great resource the VHL Family Alliance is! It was then that I decided that the best way to help my sister and her kids was to become more involved.

I have resided in Southern Ontario for the past 30+ years. I have worked for the past 12 years at The John P. Robarts Research Institute in London, Ontario, the only privately funded, non-profit, medical research institute in Canada. Working so closely with such dedicated scientists has given me a unique perspective and appreciation of the unbelievable costs, time involvement and benefits of medical research. I thought the best way to become involved was to search out top-flight scientists interested in researching the cause of this disease and putting research dollars to work toward finding a cure. I was surprised when almost no one knew of the disease. However, one person with whom I work, ***Joe Verdi,*** had heard of VHL and said he would try to learn more.

Joe is 100% Californian. He was trained at UCLA and Caltech and is a leader in the field of stem cell biology and tumour biology. He sits on the scientific advisory board of several biotechnology companies and

*Sylvia Honselaar*
*Treasurer, Canadian VHLFA*

runs a very productive lab dedicated to understanding mechanisms of human disease. He is a passionate, don't-take-no-for-an-answer individual and family man, who gives his heart and soul to everything he undertakes. In June 2001, Sue, Joe, and I attended the VHL Patient Care Provider Conference at Stanford and were thrilled to meet all of the dedicated, wonderful, family and friends of the VHL Family Alliance. The three of us stood in awe of the great work being done by the Alliance, and felt we had the nucleus and dedication to help here in Canada. After the meeting, I approached Joe about becoming more involved and before I had finished asking, and in typical Joe fashion, he said, "I'm in, let's rock!" We made several calls to the Co-chair Tania Durand, whose term was coming to an end, and she asked if we were interested in taking over the reins. The three of us decided, what better way to help than to lead?

During our initial discussion, we realized that we had a formidable task in front of us and that none of this could be achieved without the help of a professional finance accountant. To my delight, my best friend, ***Sylvia Honselaar***, who knows Sue and her story, immediately said, "Yes" to my request for assistance. Sylvia obtained her designation of Certified General Accountant in 1997, after graduating with a Bachelor of Business Administration degree from Lakehead University in Thunder Bay, Ontario. She holds the position of senior manager with the London public accounting firm Partridge Skrypnyk LLP. Sylvia is active in both professional development and volunteer work. She has been an elected Director of the London Chapter Board of CGA for the past 5 years. Sylvia is also a Co-chairperson of the CGA committee formed to raise funds for Hospice of London, a local charitable organization dedicated to caring for and helping terminal cancer patients. Now, with all the pieces coming into place, we made the official call to Tania to transfer this responsibility to our hands.

Speaking on behalf of the new board, I would like to acknowledge and thank the outgoing board led by Tania Durand, Paul Bonneau and Michelle Elliott for a job well done, and for setting the bar that much higher. We feel we are ready to fill your huge shoes and are hoping that by the end of our tenure there will be a cure for this disease. Until such a glorious day our main goals will be to continue to aid in a greater recognition of VHL in the medical and public communities, improve the diagnostic capabilities of doctors, lend greater support to the families afflicted and improve the quality of life for people afflicted with VHL. We will continue to keep you updated through our Canadian Newsletter. Stay strong, we are all in this together - We're a family.

The Canadian VHL Family Alliance is an official Canadian Charity. Donations go to support local programming and pay for newsletters delivered to Canada. Volunteers are needed to extend services within Canada. Canadians may call 1-800-767-4845, or write to canada@vhl.org.

# Nicotine makes new blood vessels grow

*Reuters Health newswire, 29 June 2001*

In findings that suggest a new theory on how smoking causes disease--and raise concerns about long-term use of nicotine replacement therapy--California scientists have discovered that nicotine can trigger the growth of new blood vessels. This process, called angiogenesis, has been implicated in the spread of cancer, the growth of angiomas of VHL, and the build-up of plaque in heart arteries.

In experiments with human cells and mice, researchers at Stanford University found that nicotine prompted new blood vessel formation--a process believed to help tumors and artery-clogging plaques thrive and grow. They report the findings in the July issue of the journal *Nature Medicine*.

'This is the first evidence that nicotine promotes angiogenesis, and more work is needed to understand how it is related to tobacco-related diseases,' lead researcher Dr. John P. Cooke said in an interview with Reuters Health. What it does suggest, he said, is that nicotine-replacement therapy should remain only a short-term therapy to get smokers off of cigarettes--and not a long-term fix for nicotine craving--since nicotine on its own may have health consequences. 'This shouldn't be taken to say that nicotine replacement therapy should be stopped,' Cooke stressed. 'But it should be used only in the short-term.'

*Nature Medicine* **2001**;7:833-839,775-777.

# Meetings in 2002!

***In the U.S. . . .***

There will be a Patient/Provider meeting in Cleveland, Ohio, in June-July 2002. This meeting will be designed for primary care physicians and families, empowering them to provide good coordinated care for VHL, knowing where to go as needed for additional information and coaching. Family members who have not attended a meeting before should enroll in "VHL 101" the morning of the first day, a great way to get to know people and become familiar with the terminology that they will hear.

***In Europe . . .***

There is a meeting in Germany in October (in German), in France in November (in French), and the Medical Symposium in Italy 6-9 June 2002 (English).

*If you are a family member* -- please come to a meeting when it is near you. Best to choose a Patient/Provider meeting in local language so that you will gain the most from the meeting.

*If you are a physician* -- please come to any meeting you can manage. The Cleveland meeting will offer CME credits at a low rate, to encourage you to join us.

*If you are a VHL expert or research scientist* -- you are welcome at any meeting, and in 2002 we would particularly like to invite you to Padua for the Symposium. This is where you will meet with your peers, an opportunity to network and collaborate. We need the benefit if your expertise at that meeting, where we will work to improve the screening protocol and recommendations for treatment.

# Travel Assistance

*by Ed Boyer, President, Mercy Airlift*

Mercy Airlift organizes free travel to medical treatment for patients. People often ask if this free air travel can also be used to attend medical conferences about their condition. None of the several fully free airline programs for patients allow for the tickets to be used for conferences. These are rules from the airlines.

However, Mercy Medical Airlift recently arranged a very deep discount airline ticket program that can be used for conferences. This can save a lot of money.

If the travel to the conference is less than 1,000 miles, the travel can be for free through the Angel Flight America network -- service provided by private aircraft. Access this type of help through the National Patient Travel Helpline at 1-800-296-1217.

For free patient travel or deep discounted airline tickets for conferences, call the National Patient Travel Center in Virginia Beach, Virginia, at 1-888-675-1405.

*Peggy and Don Marshall were honored on their retirement from the Board of Directors at the Palo Alto meeting. They have been key volunteers in VHLFA since 1993. One or both of them has served on the board for the past seven years. They continue to serve, Don as Publications Chairman, and Peggy as Chair of the 800 Line Committee.*

# Expanding the possibilities

***Medical Symposium 2002, Padua, 6-9 June 2001***

Dr. Giuseppe Opocher is proceeding with plans for the Symposium. The Call for Papers is going out this fall, and we look forward to having excellent representation from the worldwide community of physicians and researchers involved with VHL. The meetings will be held in the Aula Magna, the Great Hall. This has been a famous institution of learning since the 15th century. Galileo taught here. During the first half of the 19th century the Aula Magna was used as a room for drawing, or design. The current decorations were carried out in 1854-56, with a ceiling by Giulio Carlini.

There are plans to have a special focus group in diagnostic and treatment issues in neuroendocrine tumors of the pancreas. The primary focus of this meeting is for the physicians and researchers to share ideas and collaborate. The language will be highly technical. Family members are welcome to attend on the understanding that for the first two days they will be listening in to a medical meeting.

Beginning Saturday there will be sessions designed for family members, to which the doctors are also invited. Sunday morning there will be a workshop for VHLFA leaders from all countries.

Padua is very near Venice, and an optional side trip to Venice will be offered to attendees. There is a virtual tour of the university, including the Aula Magna, at http://www.unipd.it/esterni/visiteweb/english/pagine/credits.htm

# Defeating Depression

*Adapted from Milestones*

Feeling happy one day and gloomy or frustrated the next happens to many patients following surgery. Such mood swings are often a normal and temporary part of the post-operative recovery process, but depression is a different condition — and much more serious.

### *Depression is an Illness*

Depression is a specific illness that can be caused by a combination of genetic (inherited), mental and medical influences. There are different kinds of depression, but they have certain symptoms in common:

- Uncontrollable feelings of sadness and grief, and the inability to cheer up even for a brief time
- Feeling hopeless, worthless or guilty for no particular reason
- Loss of pleasure and interest in activities you formerly enjoyed
- Sleep problems — trouble falling asleep, and either waking up too early or constantly oversleeping
- Less energy than usual, always feeling tired and sluggish
- Difficulty concentrating, thinking or making simple decisions
- Either a loss of appetite and weight loss, or urges to overeat
- Frequent thoughts of death or suicide

You may not have all of the symptoms at once but, depending on how serious the depression is, those that are present can last for days, weeks or even months at a time.

### *An Ignored Problem*

Depression can be a complication of long-lasting diseases and the suffering they cause, so it may start even before surgery. After any surgery there are many new stresses and certain medications that can contribute to depression, especially steroid drugs.

Sometimes patients and their loved ones try to dismiss symptoms of depression as just a natural reaction to dealing with recovery or a temporary setback. Some people still view depression as a sign of mental weakness, rather than a specific medical problem.

### *Caring for the Caregivers*

It is not just the patient who is at risk for depression. The rest of the family may also be at risk. Caregivers and other members of the family may feel their own needs have to be put on hold in order to serve the patient. The unaffected siblings may feel they are ignored, or may even feel left out, or guilty that they have been spared.

### *Help is Available*

The good news is that, once it is recognized, depression is treatable.

Emotional support is a first step, so professional counseling is usually recommended. There are also a number of medications — called antidepressants — that may be helpful for relieving symptoms. It usually takes over a month for these antidepressants to work effectively, and, meanwhile, it may be necessary to closely monitor levels of other post-surgical medications to make certain they are not affected.

Your medical team should be told about any symptoms of depression so that they can recommend the best treatment.

Milestones: Post-Transplant Issues and Information is a publication of the Transplant Patient Partnering Program, an educational service of Roche Laboratories. Patient Partnering Program, 1-800-893-1995, P.O. Box 1464, Bluebell, PA 19422. http://www.tppp.net

"For a long time it had seemed to me that life was about to begin - real life. But there was always some obstacle in the way, something to be got through first, some unfinished business, time still to be served, a debt to be paid. Then life would begin. At last it dawned on me that these obstacles were my life."
*-- Father Alfred D'Souza*

# Audio and Computer Versions

The *VHL Family Forum* is available on audio tape for people who need assistance in reading it. It is read for us by the Lighthouse for the Blind and Physically Handicapped in New York City. It is also available on the internet as a set of web files which can be searched.

One of our readers who uses a text-to-speech reading device has expressed a desire to have a computer text version of the newsletter, a set of text files on a diskette which can be searched and read using a text-to-speech program. It is not beautiful visually, and has no pictures. If this would be helpful to you or someone you know, please contact the VHL Family Alliance at info@vhl.org or 1-800-767-4VHL and ask for the computer text version.

The National Cancer Institute has just updated its fact sheet on national organizations that offer help to cancer patients and their caregivers.

You can find it at http://cancernet.nci.nih.gov

Additional NCI info on resources (including how to find community-based support services) can be found at http://cancernet.nci.nih.gov/support_resources.html

# VHL in Germany

## *Dealing with Psychological and Social Aspects of the VHL Disease*

*By Gerhard Alsmeier, Chair, German VHL Alliance*

After several years of local activities we managed to establish our German-speaking VHL-Support Group in October 1999. Currently, we have more than 140 members from Austria, Germany and Switzerland.

Thanks to the efforts of Prof. Neumann at the Freiburg University Hospital, treatment of VHL in Germany seems to have reached a comparatively high level, but since Freiburg is in the Southwest corner of Germany, one of our aims is to spread information out among all the professional members of the health care system, throughout every part of Germany, Austria and Switzerland. Major vehicles are our newsletter (published quarterly), our annual meeting in autumn and our homepage (http://www.hippel-lindau.de). To keep in touch on a personal level, we chat online every first and third Sunday.

However, our experience is that information on the professional side is not enough – especially in the case of our disease. That's why a large part of our effort deals with educating the members themselves on the regional level. For that reason we have put a lot of effort into organizing regional spring meetings this year.

Last year we only had one regional meeting for our members in the North and West of Germany. At the end of last year we decided -- together with the members of our medical advisory board Prof. Neumann and Prof. Schmidt -- to hold four regional meetings: (1) North and West, (2) East, (3) Southwest and Switzerland, and (4) Bavaria and Austria.

| Psycho-Social Topics of Greatest Concern to the German VHLFA Membership | |
|---|---|
| Fear of screening results | 37% |
| Making healthy choices | 32% |
| Coming to grips: it never stops! | 31% |
| Life and Health Insurance | 29% |
| Coping with "health decline" | 24% |
| Finding good physicians | 24% |

*Table 1: Compiled by the VHLFA in the German-speaking countries (Germany, Switzerland, Austria)*

In the last one and a half years we found that for many of our members, the psychological and social aspects of VHL in daily life are of great importance. So we decided to focus on this topic in our newsletter and in our annual meeting, in addition to the medical aspects of VHL. In order to learn what the main problems were, we organized a broad discussion process at each of our regional meetings about the social and psychological aspects of VHL.

As a first step, we collected the problems our members have in dealing with VHL. Afterwards we asked them to write down the three or four topics they personally considered most disturbing. In that way we found out what the main problems were in each region. At the end of every regional meeting we talked about possible solutions.

After all our four meetings were held we compared the results. Out of all topics raised, the ones most often mentioned are shown in Table 1.

Our annual meeting, will be held this time in Berlin from 26-28 October, 2001. Among the speakers will be a psychologist, giving a speech on the topic "Coming to grips: It never stops!" At the Padua Symposium 2002 we hope to learn more about making healthy choices (diet, vitamins, exercise, not smoking) and we will try to cover the other topics in our next meetings or in our newsletter.

We would like to hear from the other VHL-groups, if they have experienced similar problems and how they are dealing with them. Don't hesitate to contact us.

Gerhard Alsmeier, Chair of the German VHL-Alliance,
g.alsmeier@hippel-lindau.de

## *Sigma Phi Gamma supports VHLFA*

Jay Platt attended the convention of Sigma Phi Gamma in Baltimore in June, where he accepted their check for $11,000 to support VHLFA.

VHLFA was chosen as their international charity 1999-2001. Over this two-year period their local chapters in major cities, states, and provinces in the U.S. and Canada raised money both for VHLFA and for their local charities through a series of local events: dinners, auctions, bingo, raffles and more. This check brings their two-year contribution to more than $21,000.

We are very grateful for the support of all the good women of Sigma Phi Gamma International. Special thanks to Toni Brunk of Ohio, International Secretary, who sponsored our candidacy.

# New Alabama Chapter

*Linda Darden is our new chapter chairperson for Alabama. She shares her story, and her goals for the chapter.*

I am 48 years of age, one of seven children, with me being number seven and twin to number six. There are 4 boys and 3 girls in our family. I graduated High School and went into the paramedic program at the University of Alabama, Birmingham, Alabama.

I worked for Carraway Hospital in the emergency room at the time and wanted to train for the Life Saver Helicopter that was just being introduced. In my second level of paramedic training I was sent to NIH in Bethesda, Maryland, for evaluation of muscle problems. It was then that I was diagnosed with Muscular Dystrophy -- a very rare form called McArdles Disease (phosphorylase deficiency). This ended my dream of becoming a flight medic or even a paramedic. Since I could not be a paramedic, I worked as an office manager with several companies, and then started my own cleaning business approximately 8 years ago.

I have two children. My son, Patrick, is a Staff Sgt. in the Air Force in Louisiana. My daughter-in-law, Brandy, graduated from the Police Academy in June. My daughter, Candi, lives near me and helps me with my cleaning business. I have two grandsons, Justin (5) and Hunter (1), the joy of my life.

During 1971 my oldest sister, Peggy, became very ill -- brain tumor, spinal tumors, but no diagnosis. For years she has gone without a diagnosis. In 1988 my brother became ill with brain lesions and vision problems. Peg went 29 years without a diagnosis, and our brother 12 years. It was not until 2 1/2 years ago that we were told for the first time that von Hippel-Lindau was in our family.

I have always helped my sister Peg. Before her first brain surgery she lost her ability to walk and do things like she could before. The von Hippel-Lindau diagnosis was like a bomb hitting us. I researched on the internet almost night and day. I do have medical background from working in the hospitals, and doing this research made me realize that my family was hit with a condition that would be with us through eternity. That no matter what happens now, the genetic ramifications could always be there, in us, our children, our children's children, on down the line.[1]

I got on the internet and researched VHL and found the home page for the VHL Family Alliance. It was a great relief to hear other people's stories, hear them tell about their surgeries and vent their frustrations and problems, and get comfort from the other VHL members. It was like finding Heaven on-line, just to know that others knew what we were going through. Words of encouragement -- a little "how ya doing?" -- go a long way in the life of people fighting each and every day to stay well. Some may ask me, "Why don't you worry about your condition with McArdles?" I do, it is under control; I am at high risk for a lot of things, but my life with McArdles is so much simpler than what people with VHL experience.

Why I want to do the Alabama VHL Chapter? For my family. It is as pure and simple as that. I want to help my family and others to know that VHL does not have to be as severe as our family's experience. With the proper diagnosis in the beginning you can manage and live a very useful, productive life. So, who will get the word out about VHL here in Alabama? Me. I could turn and walk away and say let someone else do it. But then I am passing the buck, so I thought about it, I prayed about it, and then I looked at my brother and sister and realized the rest of our family is at risk, and if I passed on this, then who would do it? Who would make sure that the information on VHL has gotten to the doctors in the state of Alabama? It is simple -- me. I don't want other families to go through all that our family has. The 29 years Peg spent without a diagnosis took an enormous toll on her both mentally and physically.

I will strive to do the best possible job I can, I will continue to educate myself and work hard to reach as many physicians as I can to make sure they are well informed on VHL. I don't want them to just know a little, I want them to know A LOT!

If you know anyone in Alabama who has VHL, please call the hotline for my telephone number. 1-800-767-4VHL.

1. VHL is a dominant gene, so a person with VHL has a 50/50 chance of passing the VHL alteration to a child. Each child has an equal 50/50 chance of inheriting the gene, like flipping coins. Someone in the family who does not have the VHL alteration (diagnosed through DNA testing) cannot pass VHL to a child. It does not skip generations and appear later on. If we look only at symptoms, VHL can appear to skip a generation if the person in the middle had few or no symptoms. That's why DNA testing is the only way to clearly identify who does and who does not carry the genetic alteration in their VHL gene.

The most important thing we do, the greatest gift we can give, is to let each other know that we are not alone.

***Can you help in your own local community?***

Please call or write to volunteer.
*In the U.S.:* Kathy Braden, Chapters Committee, (317) 894-3909, info@vhl.org
*In other countries:* Your country chairperson (if any), or Joyce Graff, +1 (617) 277-5667 or info@vhl.org

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# Triathlon

*by Alice C., Washington*

Today, August 20, 2001, was the Seattle "Danskin Triathlon"!!! 1/2 mile of swimming, 12.4 miles of biking & 3.1 miles of running. I finished it!!!!

I would never in my wildest dreams classify myself as an athlete. I have had two operations for bilateral ELST's,[1] and one surgery on my right kidney. A group of people at my work began by walking several miles at lunchtime, and four of us competed in the Triathalon.

I have been training since February, and believe me I had lots of moments when I was ready to throw in the towel. My first practice swim I lost my balance. My first time out on the bike I ran into a wall and cried. The first time I tried to run a mile I had to stop and walk. In fact even yesterday when I lined up for the swim start and saw 4,500 women surrounding me I almost quit.

At first Dr. Jeff Kim at NIH advised against the swimming for fear I might get kicked in the head. For someone who has had a few surgeries on the head, that would not be a good thing! He offered to write a note for me, but I told him this was a competition, not a gym class! So instead I found a "swim angel" through a wonderful organization called TeamSurvivor, for physically challenged women who want to be physically active.[2] I explained the problem and asked if there were a volunteer who could assist me. My "swim angel" swam next to me and made sure I stayed on course, and away from the other participants so I wouldn't get kicked in the head. There is no doubt with 4,500 participants I would have had a few whacks in the head without her. I was in the second wave of 125 participants to kick off and swim the half mile. I stayed toward the back and side, with my swim angel watching out for me.

A year ago I couldn't imagine myself biking 12.4 miles -- with my balance?? But through this six months of training my balance has definitely improved.

I can't even begin to describe how exciting it was to cross the finish line! It was the most thrilling moment I have ever experienced in my life.

I was so proud of myself that I wrote all my current and past doctors. Dr. Dan Choo replied, "I always think of you when people ask me the simple question 'am I sick?' Being diagnosed with VHL certainly does not mean that you are ill. Your vigorous lifestyle is a testament to that." Dr. Choo was one of the surgeons who removed my bilateral ELST's at the NIH. He is now at Children's Hospital Medical Center of Cincinnati.

As always, thank you to my VHL family for all your support and your "can do attitude". For me it took having VHL to realize I can do anything!!! It may require some modifications, but I can do it!!! I can't wait to do it again next year.

1. Endolymphatic sac tumors (ELST's) are tumors on the outer edge of the brain that can interfere with hearing and vestibular function (balance).
2. TeamSurvivor was originally founded to assist women who had survived breast cancer. It is now open to all women cancer survivors, including women with VHL. See their website at http://www.teamsurvivor.org

Photo by Brightoom, http://www.brightroom.com

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