



VHL Family Forum



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Ophthalmic Issues in VHL

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We will be talking today about how a VHL lesion can cause damage to the eye. How do we diagnose the presence of a tumor? What can the patient do to help us diagnose the lesion early so that we can treat it most successfully?

We have a lot of treatments for the retinal tumors which accompany VHL, but our treatment results are much better if we can diagnose the presence of a tumor when the tumors are small. I'll give you some examples of some diagnostic techniques, like fluorescein angiography. We'll touch upon laser therapy and cryotherapy which are the two most widely used means of ablating these tumors at the present time. And I will briefly touch on photodynamic therapy (PDT).

The retinal lesions caused by VHL are capillary hemangiomas (CH). They can start out as little teeny nubbins in the retina that are very hard to see, but eventually they grow to a size where they are fairly obvious. There are a few characteristics that accompany these lesions. Generally speaking there is a dilated arteriole (small artery) which is draped bringing blood to the tumor, and a dilated vein that is taking blood away from the tumor. If the tumor has been present for some time, the tumor will leak serous fluid, protein and fat into the subretinal space, the space beneath the retina. These collections of protein and fat can sometimes signify the presence of a very tiny tumor.

In the normal retina you see the macula in the center, the most sensitive part of the retina. Half of the light-receptors in the entire retina are collected in this one little area. Even though VHL tumors may be way out in the periphery of the eye, they can cause reduction of vision. The products from the tumor gravitate toward the macula, and fat and other proteins can accumulate in this region.

The normal retinal vessels branch gracefully like trees; around the tumor there will be abnormal retinal

vessels, swollen and tangled. And sometimes these dilated vessels will signal to the ophthalmologist that there is a tumor lurking out in the periphery. Near the center of the retina is the optic nerve (see Figure 1).

The discovery of new lesions is very important. The smaller the lesion is when we discover it, the more favorably it will respond to treatment. What can the patient do to help us discover these lesions early?

Anyone with a family history of VHL should have an ophthalmic examination at least one a year. This examination should consist of dilating the pupil, and examining the retina with an indirect ophthalmoscope. This is the instrument that the ophthalmologist will put on his or her head and shine a light through a prism or lens held in the hand.

For those without a family history of VHL — in other words, for everyone in the general population — anybody that has a reduction in central vision should have an examination to determine why there is a central blur in the visual field?

In VHL, the most common cause of reduced central vision is the accumulation of proteins and fats in the region of the macula. These proteins and fats actually travel from the periphery to the center of the retina, accumulating within the macula. So when a lesion is seen, especially if it is associated with dilated retinal vessels, one must suspect VHL. To confirm the

cont'd on page 2

Inside this issue!

Making a Difference	How Could this Happen?
Audrey on the Line	Healthcare Proxy
Valentine Pageant	Ask the Family
Women and Stress	Volunteering for Health
Eva Logan's Voice	Carrying the Torch

Padua Meeting - review your hotel choice
Cleveland Meeting now October 11-13, p. 7

presence of a tumor, and/or to diagnose new lesions, we will frequently use a test that is called a fluorescein angiogram. This is a test where sodium fluorescein dye, which is a very inert dye, is injected into the patient's vein in the arm. The dye reaches the retina within 19-20 seconds. This test is usually done in the company of a fundus camera, positioned to take video of the back, or fundus, of the eye. The camera then begins to record the dye running in the vessels and going toward the tumor, approximately 20-25 seconds after completion of the injection.

After 3-5 minutes we see dye begin to leak out of the tumor and enter the vitreous cavity (the large space in the center of the eye, normally filled with a clear gel). In the later stages of the disease, you may see a whole quadrant of the vitreous cavity lighting up. These lesions, then, can be seen either with the fundus camera or with the indirect ophthalmoscope which covers a much larger zone, and with the proper filter in place on the ophthalmoscope the ophthalmologist is able to see even the tiniest lesions out on the periphery of the retina. They look like stars in a midnight sky.

Once diagnosed, then, we want to eradicate the tumor. The most common modality is the laser or photo-coagulation with the so-called "hot laser". If the tumor is so large that we cannot use laser, then we will rely on cryotherapy. The difference between the two is that laser therapy is thoroughly innocuous. It does not involve any invasive cutting. The laser energy is directed through the patient's dilated pupil, and you can do multiple sessions of treatment without having the patient admitted to the hospital or actually having to go through a cutting procedure.

Cryosurgery, on the other hand, is the application of cold energy through the wall of the eye. Cryosurgery is used when we can no longer use laser therapy. Sometimes cryotherapy needs to involve an incision in the lining over the white of the eye in order for us to get the cryo probe back to the lesion, and for us to deliver the cold energy through the wall of the eye to the lesion. That obviously involves creating a sterile field, and making an incision in the membrane. It usually involves admission into the hospital or clinic.

You have heard talk about the involvement of Vascular Endothelial Growth Factor (VEGF) in the process of development of a VHL tumor. We think it is precisely VEGF which also initiates the wet form of age-related macular degeneration where we see the invasion of capillaries beneath the macula, growing toward the central macula or the fovea. Many of the new drug therapies being developed for macular degeneration seem to have applicability for VHL lesions as well. There are several now in clinical trials, so better treatments are on the way.

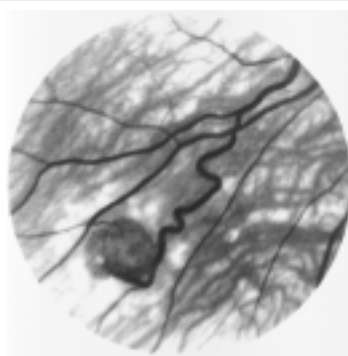


Figure 1: Retinal angioma in the lower left quadrant of this photograph. Darkened areas show the accumulation of fluid under the retina. Photo courtesy of Joslin Diabetes Center.

Let me describe the process of photocoagulation, the most common treatment currently available to people with VHL. The goal is to shrink or ablate the vascular tumor so that once a tumor like this is discovered we want to deliver hot energy or a hot light to this tumor. The pigment within the tumor absorbs the light. It works best in tumors that are a little darker, but it works well in VHL tumors as well. We want to deliver the energy, and have the energy absorbed by the tumor. The ophthalmologist delivers points of light, making big circles. If you hit it too hard with a narrow beam, the tumor can bleed and the blood can go into the vitreous cavity and then that begins to set up a connection between the vitreous gel in the center of the eye and the tumor. Those kinds of connections can lead to problems in the future.

We apply fairly broad laser doses gingerly at first. A small tumor might receive ten doses, then the patient will go home. About four weeks later we may deliver another ten. Gradually the intensity of the light beam is increased a little more until we are pretty sure that there is enough of a fibrous coating over the lesion so that it will not spontaneously bleed. After a while then the lesion will acquire a little fibrous tissue cap, and the tumor begins to shrink beneath this cap. As long as we can see little portions of the tumor from the side we will continue to deliver laser therapy to the tumor. The objective is to be able to shrink the tumor until it is just a little nubbin in the retina. At that point we can really apply energy to it and ablate it.

Occasionally the tumor will shrink. It will almost always develop this little white fibrous tissue cap, making it difficult now to deliver additional energy if the edges of the tumor are not showing. So our next strategy then is to try to strangle the tumor, to decrease the blood supply to the tumor by applying at first gentle and then more strong laser therapy along the sides of the artery supplying the tumor. The laser treatment then stimulates fibrous tissue growth around the edges of the artery, and that fibrous tissue then begins to gradually constrict the lumen, the opening down the middle of the artery. And that is a good strategy.

If laser treatment doesn't work, the other strategy we invoke then is cryosurgery. We bring a cryosurgical probe around the outside of the eye, advance it until it is right beneath the tumor, and then apply

freezing energy which penetrates through the white coat, through the vascular coat, and finally through to the retina. You can apply enough energy so that this entire lesion can be turned frosty white and using a freeze-thaw technique you can achieve very good damage to the lesion.

The main goal is the shrinkage or stabilization of the lesion. We want to decrease the leakage so that the amount of fluid beneath the retina decreases, and we want to improve central vision. In this eye (Figure 1) there was reduced vision, down to the level of 20/30, caused by the accumulation of material beneath the macula. We can see that the fluid action and the exudate action extends downward, and the lesion is out to the left. There are abnormal vessels leading up to the lesion, the artery and the vein.

The lesion was exposed to laser therapy. Six months later we began to see a decrease in the amount of exudation and the vision improved to 20/20. Seventeen months after the treatment the vision came back to 20/10 and we can only see a very small fleck of protein beneath the retina. This is a good response.

Occasionally a lesion will develop in a position near the optic nerve which can be very detrimental to vision. When the lesion develops in this position there is not much we can do to improve vision, but there is still a lot we can do to prevent the lesion from going down the cascade where we will lose the eye.

In summary, this condition can be managed. It is managed very well, especially if we get to the lesion early. Periodic examination is extremely important, especially if there is a family history. With the onset of reduced central vision everybody — whether VHL or not — should consult an ophthalmologist to find out why the central vision is degraded, and then we discussed the treatment options, namely laser and cryosurgery.

Questions:

If there is a lesion on the optic nerve, what would you do? If it is not growing and not exuding material that travels beneath the macula, just watch it. We have excellent ways of watching it now, one of which is photography. But if all of a sudden it begins to grow, what can you do then? Well, you would hesitate to use hot laser unless you were pretty sure that the nerve fiber bundle (the bundle of nerves coming from the macula to the optic nerve) was actually beneath the lesion and not draped over the top. If they are draped over the top of the lesion then you would be excluded from using hot laser. Could you do cryosurgery? If this lesion is sitting right on top of the nerve there is no way that you are going to get the cryo probe beneath the nerve and apply the energy, because cold energy is very destructive to nervous tissue. That's when you would think about using PDT.

What is Photodynamic therapy? PDT differs from laser therapy in that we take advantage of the fact that the lining of newly growing capillaries tend to attract lipid

molecules (fats). There is a dye out there now call visio-dye that when injected into a person's vein combines itself with molecules of low density fat. This combination molecule then circulates throughout the body. Normal blood vessels will not attract low-density fat, but the lining of new blood capillaries or growing capillaries will. So ten minutes after infusion the dye along with the low-density fat will have saturated the lining of the abnormal new blood vessels in the VHL lesion. That target, then, is irradiated with a cold laser, a diode laser that does not cause any formal effect, but at the wave length of 680 nanometers it will excite the dye. The dye then produces single oxygen molecules which are very toxic to the lining of these capillaries. That then coagulates or thromboses the capillaries and turns them off. So that is probably the modality I would favor.

What is your experience with that? Is PDT successful in treating VHL lesions and maintaining vision?

So far we have little experience with PDT and VHL. The results from PDT with age-related macular degeneration are approximately 60% stabilization and/or improvement of vision. So compared to what we had before, that's super. But there are treatments now in clinical trial involving anti-VEGF molecules. We want to discourage the growth of new blood vessels beneath the macula. Presumably these anti-VEGF treatments might be useful also for VHL.

My daughter's two angiomas were discovered in 1998, and they are just watching then, they haven't done anything. I feel like I'm just waiting for something to happen. It all depends on the location of the tumors, if they are too close to the nerve, or too close to the macula. That might be a reason for temporizing.

They say they are in the periphery. What strategy would you follow in a case like that? Because I have seen patients develop multiple tumors over the years, I prefer keeping up with them so that I know that I have handled this one. I keep watching, and if another one pops up I handle that one. It's just another philosophy.

At what age do you recommend beginning eye exams? If there is a family history, I would recommend beginning at age 3-5. It is easy to do a good exam on a child of five. If there seems to be a high penetrance in the family, I would even recommend doing an exam under anesthesia as early as age 1.

In our family there is a lot of involvement and most people have multiple tumors by age 9. We also see a lot of growth in puberty. Under those circumstances would you not recommend that they be checked every six months? Yes. Once a tumor is seen, then I think a schedule of every six months is a very good schedule to follow.

In our family it seems that when you treat a lesion it only gets worse. Sometimes when you treat a lesion you will actually stimulate that lesion to begin to exude fluid into the subretinal space. Not every lesion responds the same way, and therefore you have to tailor your treatment to the particular case, go slowly, and assess the response.

Based on his talk at the VHL Conference, Palo Alto, California, June 2001.

Can One Person Really Make a Difference? You Bet!!

by Alice M. Coday, Washington

When I was a high school student I went to hear Margaret Mead speak at the University of Delaware. I realized that day, my single voice could make a difference. Her quote is on my bulletin board today; "never doubt that a small group of thoughtful, committed citizens can change the world: indeed, it's the only thing that ever has." Relationships with lawmakers can play a critical role in supporting issues that you care about. Each and every one of us can make a difference.

The events of September 11th and the war on terrorism present new fiscal challenges for lawmakers. Congress now faces a budget battle on all fronts. It is imperative once again that the United States not retreat from its role as the world leader in cancer research and treatment.

February 6, 2002, was Nationwide Lobby Day for the American Cancer Society. A representative from each of the 50 states met in Washington, DC, to meet with lawmakers. I was one of 3 volunteers representing Washington State. I have been part of the Von Hippel-Lindau research protocol at the NIH for the past 9 years. Three surgeries have been performed at the NIH. The fact is, the NIH has been my life line. I truly believe without the NIH I wouldn't be here today; therefore I feel it is important to do everything within my power to support the war against cancer.

Can you make a difference? You bet! These are issues that affect us all, and are supported by both major political parties. Read up on the following and make your own voice heard! What I have chosen to do is to ask my legislators to take the following actions:

Increase investment for cancer research and application programs.

Complete the five-year doubling of the NIH budget by providing \$27.3 billion to NIH in FY 2003, and continue expanding the investment in conquering cancer when that doubling is complete.

Fund NCI at \$5.69 billion, the level proposed by its Director in the NCI's Bypass Budget.

Increase access to cancer care, prevention and awareness programs.

Enact legislation that assures access to clinical trials; advance legislation that eliminates barriers to cancer screening tests and support programs that increase awareness and prevention of cancer.

Co-sponsor legislation (HR 1520 or S 710) introduced by Senators Jesse Helms (R-NC) and Edward Kennedy (D-MA) that would ensure private insurance coverage of the full range of colorectal cancer screen-

ing options, according to ACS guidelines.

Sustain increases for vital cancer outreach, awareness and screening programs at the Centers for Disease Control and Prevention (CDC).

Reduce health disparities among minorities and the medically underserved.

Ethnic minority and medically underserved communities are disproportionately affected by cancer. They are more likely to develop and die of cancer than any other group. Fund the National Center for Minority Health and Health Disparities (NCMHD) at the NIH at \$199.6 million.

And last, let's see the Patient's Bill of Right's enacted this Congress.

Cancer is the second leading cause of death in the United States, claiming the lives of one in every 4 Americans. The good news is that more Americans are surviving the disease more than ever before. Almost nine million Americans alive today have a history of cancer!! This is twice the number of survivors compared with 30 years ago. We can win the war and you can make a difference!! Contact your lawmakers today.

Audrey on the Line

Audrey Clifton and her husband Don had a quick initiation to staffing the 800 line. It was feast or famine with the calls! They enjoyed talking with everyone who called in January and February.

Audrey has had a diagnosis of VHL since age 32. She is now 64, a mother of three and a grandmother many times over. She has had numerous operations on eye, kidney, brain, and spine, and an ELST in the ear. She also had gamma knife surgery at the University of Virginia for a brain tumor. She has been the Chairman for North Carolina for several years.

Audrey's advice? Try to find a doctor who knows something about VHL. Do not hesitate to get a second opinion or if necessary a third opinion prior to making a decision where surgery is concerned.



Audrey and Don love to help people. They live in Fayetteville, North Carolina, where Audrey is very active in the Outreach program of her church.

Audrey (left) at a North Carolina chapter meeting, 1998.

Valentine Pageant seeks cure for VHL

By Kim P., Illinois

I was almost fourteen years old when I had my last Valentine's Day with my father. When I received the last newsletter from the Alliance and read about David running a marathon, I decided it was time I did something to help support the VHLFA. My husband Darrin and I have two daughters, Danielle and Rebecca, and I love doing pageants with my girls. So 18 years after my father's death from kidney cancer, I decided to sponsor a pageant in his memory.

At the time of his death in 1984, a physician recommended that we children be screened for possible problems. A scan revealed that I had a small tumor on my pancreas, but my mother was told not to worry about it. Even earlier, at age 5, doctors had discovered a tumor in one of my eyes. They told my parents it had scarred over and not to be concerned.

When I was 18 I began having dizzy spells. Doctors discovered and removed a tumor from my ear, and later another from my brain. In 1992 I was diagnosed with von Hippel-Lindau. One of my sisters also has the disease. We knew nothing about it, so I began doing research.

It quickly became clear that VHL is what had killed my father. Maybe if we had known more than about treating it, he might still be alive.

Six years ago when I was pregnant with my first child I had severe back pain and numbness in my left leg. Tests revealed a tumor on my spine. After the birth of Danielle, followed swiftly by spinal surgery, I had to undergo physical therapy to regain the use of my paralyzed leg. I felt a lump in my abdomen, but the doctors told me it was nothing to worry about.

Six weeks after my second daughter's birth, the lump was still there. I would lie down, and it would feel like I was lying on a tennis ball. Tests revealed a malignant tumor the size of an orange on my pancreas. Doctors were planning to remove my pancreas, when I read an article in the VHL newsletter about pancreatic tumors. I realized that radical surgery was not my only option. I went to the U.S. National Institutes of Health (NIH) where they removed 80% of my pancreas, part of my stomach and small intestine. But they saved enough of my pancreas that I am not diabetic! They also found an endolymphatic sac tumor and successfully removed it. My doctors at home had thought it was only scar tissue.

So you can understand my gratitude to God for the VHLFA newsletter! It has led to better medical care, and truly saved my life. I still have tumors in my neck, cysts on my kidneys, and a few brain tumors, but I'm not giving up. VHL is slow growing. I'm like everybody else, I just have more scars.

The pageant came together in a month. It was held at the Illinois Centre Mall. We had a total of 73

entrants — way beyond my hopes. We got a lot of wonderful media coverage which helped tremendously. We were also blessed with donations of money for trophies and gifts for the children. Each child received a crazy straw and a coupon for a cheeseburger from McDonald's, candy hearts from Target, and Valentine candy from Schnuck's grocery store. The overall boy and girl winners each received a \$50 savings bond from Old National Bank and Banterra Bank, a 2-hour limo ride from P&B Limo, a plastic picnic table from Wal-Mart, a teddy bear from Kroger, and a portrait package from J.C.Penney and Don Barden Photography. We gave out 42 trophies: first, second, and third place in each age group for both boys and girls. Miss & Mr. Sweetheart were chosen in each of seven age groups, from 0-6 months to 5-6 years. It was such a success, I hope to make it an annual event!

Several local newspapers printed my VHL story and the details of the contest, encouraging people to register in advance for \$20, or on-site for \$25. The Mall provided advertising help — they even ran TV ads! They set up the stage, chairs, PA system and everything. Two TV channels covered the event.

We are very grateful to the many volunteers who donated their time, and the many who gave extra donations. My Mom, Marsha, and Darrin's Mom, Cheryl, were super help registering contestants. Danielle, Rebecca and Crysta passed out candy. Nikki helped keep them supplied, and did a super job getting the trophies passed out. Darrin was the official money taker (he was really good at that), and he was also the official vote counter and judge harrasser!

My sister Krystal got several donations that helped out tremendously and my sister Wendy rounded up several contestants. Kay passed out many forms that attracted contestants. I could just go on and on and on. I'm grateful to all!

Someday we won't have to do fundraisers any more for VHL because they'll find a cure. But that's why this is so great because we're one step closer, one pageant at a time! I know my Daddy would be so proud!

Kim presents trophies to Miss and Mr. Valentine, Miss Sweetheart, and Tiny Miss Sweetheart.



Women Respond Differently to Stress

A landmark UCLA study suggests that women respond to stress with a cascade of brain chemicals that cause us to make and maintain friendships with other women. It's a stunning finding that has turned five decades of stress research — most of it on men — upside down. "Until this study was published, scientists generally believed that when people experience stress, they trigger a hormonal cascade that revs the body to either stand and fight or flee as fast as possible," explains Laura Cousino Klein, PhD, now an assistant professor of biobehavioral health at Pennsylvania State University in State College and one of the study's authors. It's an ancient survival mechanism left over from the time we were chased across the planet by saber-toothed tigers. Now the researchers suspect that women have a larger behavioral repertoire than just "fight or flight." In fact, says Dr. Klein, it seems that when the hormone oxytocin is released as part of the stress response in a woman, it buffers the fight or flight response and encourages her to tend children and gather with other women instead. When she actually engages in this tending or befriending, studies suggest that more oxytocin is released, which further counters stress and produces a calming effect.

This calming response does not occur in men, says Dr. Klein, because testosterone — which men produce in high levels when they're under stress — seems to reduce the effects of oxytocin. Estrogen, she adds, seems to enhance it.

The discovery that women respond to stress differently than men was made in a classic "Aha!" moment shared by two women scientists who were talking one day in a lab at UCLA. "There was this joke that when the women who worked in the lab were stressed, they came in, cleaned the lab, had coffee, and bonded," says Dr. Klein. "When the men were stressed, they holed up somewhere on their own."

"I commented one day to fellow researcher Shelley Taylor that nearly 90% of the stress research is on males. I showed her the data from my lab, and the two of us knew instantly that we were on to something."

The women cleared their schedules and started meeting with one scientist after another from various research specialties. Very quickly, Drs. Klein and Taylor discovered that by not including women in stress research, scientists had made a huge mistake: The fact that women respond to stress differently than men has significant implications for our health.

It may take some time for new studies to reveal all the ways that oxytocin encourages us to care for children and hang out with other women, but the

"tend and befriend" notion developed by Drs. Klein and Taylor may explain why women consistently outlive men. Study after study has found that social ties reduce our risk of disease by lowering blood pressure, heart rate, and cholesterol.

"There's no doubt," says Dr. Klein, "that friends are helping us live longer." In one study, for example, researchers found that people who had no friends increased their risk of death over a 6-month period. In another study, those who had the most friends over a 9-year period cut their risk of death by more than 60%.

Friends are also helping us live better. The famed Nurses' Health Study from Harvard Medical School found that the more friends women had, the less likely they were to develop physical impairments as they aged, and the more likely they were to be leading a joyful life. In fact, the results were so significant, the researchers concluded, that not having close friend or confidante was as detrimental to your health as smoking or carrying extra weight!

And that's not all: When the researchers looked at how well the women functioned after the death of their spouse, they found that even in the face of this biggest stressor of all, those women who had close friend and confidante were more likely to survive the experience without any new physical impairment or permanent loss of vitality. Those without friends were not always so fortunate.

Yet if friends counter the stress that seems to swallow up so much of our life these days, if they keep us healthy and even add years to our life, why is it so hard to find time to be with them? That's a question that also troubles researcher Ruthellen Josselson, PhD, coauthor of *Best Friends: The Pleasures and Perils of Girls' and Women's Friendships* (Three Rivers Press, 1998). "Every time we get overly busy with work and family, the first thing we do is let go of friendships with other women," explains Dr. Josselson. "We push them right to the back burner. That's really a mistake, because women are such a source of strength to each other. We nurture one another. And we need to have unpressured space in which we can do the special kind of talk that women do when they're with other women. It's a very healing experience."

Summary of an article published in *Psychological Review* 2000 Jul;107(3):411-29

Let's cure VHL in this decade!

Write us in on your United Way campaign!

U.S. Federal workers, choose # 1098
in the Combined Federal Campaign!

June 6-8: Symposium in Padua

Due to the success of this worldwide Symposium on VHL, we have moved the venue from the charming (but somewhat uncomfortable) old medical school to Praglia Abbey in the suburb of Bressio just outside Padua, renovated to be a modern conference center. This facility is wheelchair accessible.

Experts on VHL from all over the world will gather in this beautiful setting to share their latest findings, advance the management of VHL, and facilitate collaboration on research toward a cure.

Abstracts on a wide variety of important topics have been received, and the Committee is finalizing the agenda. Current information can always be found at www.vhl.org/conf2002, or at www.vhl.it.

Registration is \$264 (Euro 250) for the three-day event, including all lectures, lunches, coffee breaks, and an informal get-together with drinks and a Congress Kit of handouts. There are discounts for students and families, and one-day registration fees. Discounts for registration prior to May 1.

Family members will be most comfortable on the last day of the conference, where topics will be more clinical in nature. A VHLFA leaders' meeting will be held on Sunday. All volunteers are welcome.

There are discounts for registration prior to May 1.
We look forward to seeing you!

Note: the meeting site in Padua has been changed.
Please review your hotel choice on the website.

Register on the website <http://www.vhl.org>
or send a fax requesting the registration form to:
Italy: +39 049 657391; US: +1 617 734 8233

Where to Stay

Praglia Abbey is a famous Benedictine abbey, founded in the 11th century, but almost entirely rebuilt in the 15th and 16th centuries. Its main features are the typically Tuscan elegance and lightness, with a Romanesque bell tower. There are four 15th century cloisters and a large number of works of art. It is a center for the recovery and restoration of ancient and damaged books.

The Abbey is located near Padua (Padova) and the warm springs spa town of Abano Terme. Transportation will be provided from either of these towns.

The official language of the symposium is English.

The average temperature in Padua in June is expected to be 15-20 degrees Celsius, or 65-75 Fahrenheit.

Rooms can be reserved through the tourist bureaus of Padua (fax: +39-049-650794) or Abano Terme, or through your travel agent. See the website www.vhl.org/conf2002 for details.

If you decide to add a vacation in Italy, your options could be endless! -- *Jacki Hunsberger, Odyssey Travel*
Tel: +1 (610) 222-0550 or (US/Canada) 800-972-3178
Fax: +1 (610) 222-0520

October 11-13: Update on VHL

An Integrated Medical Approach to a Multisystem Disorder

The Cleveland Clinic Foundation and the VHL Family Alliance will sponsor a continuing medical education event for primary care physicians, nurses, and genetic counselors to be held at the Marriott Airport Hotel, October 11-13, 2002. The meeting will be chaired by Dr. Robert Uzzo of the Fox Chase Cancer Center and Dr. David Goldfarb of the Cleveland Clinics. Continuing Medical Education credits will be awarded by Cleveland Clinic to physicians and genetic counselors for the sessions they attend.

Because VHL crosses so very many medical specialties, it is a challenge for any health care professional to follow a patient with VHL. Without the cooperation and close collaboration of the patient and the family, it is practically impossible. For this reason we always work to engender a partnership between the families and their health care professionals.

The agenda includes a survey of the medical issues in VHL, diagnostic techniques and treatments, including legal and insurance issues and stress management.

Families are welcome to attend this meeting. In the nine years since we began holding "patient/provider conferences," we and the physicians who

have attended have come to believe strongly that it takes teamwork to manage VHL. The patient has an important role to play, and empowering the patient is good both for the patient and for the physician.

The conference is a wonderful opportunity to meet others with VHL. Andy noticed at his first meeting that other people also had "zippers" peeking out beneath the hairline. Tania noted that "all the women were wearing sensible shoes." Gale bumped into another person and both said in chorus, "Excuse me, that's my blind side."

If you have not previously attended a VHLFA conference, you are encouraged to attend "VHL 101", an introductory session with Joyce Graff on Friday morning before the formal start of the conference. You will have an opportunity to meet other attendees and VHLFA Board members, and gain an understanding of terms and concepts that may be new to you, especially in parts of the body with which you have not dealt before.

For registration information, see <http://www.vhl.org/conf2002> For hotel reservations, call Marriott at 1-800-228-9290 or (216) 252-5333

Eva Logan's voice

By Joyce Graff

When I first met Eva on the telephone, I was immediately charmed by her warm Southern tones, her delicious sense of humor, and her optimistic view of the world. On a subsequent visit to Atlanta, she and I were interviewed by CNN. She asked the camera man whether her eyes looked okay in the sunlight. When he said yes she was relieved, and she shared that she had one prosthetic eye. Years after she lost vision in that eye, it became increasingly painful and the doctor recommended removing the eye. She was fearful that it would degrade her appearance. The surgery went well, and she found an excellent ocularist who made her a perfect match for her seeing eye. On the airplane returning from picking up her prosthesis, she noticed a man across the aisle staring at her. Feeling self-conscious, she asked if anything were wrong. "Not at all," he said. "Sorry to be staring, but you have the most beautiful eyes." "No problem," she said smiling, "you have no idea how much that means to me."

“Eva was the first person I spoke with when I called the hotline and she even took the time to call me back a few times to see how I was doing. She was precious. — Tara E., Colorado”

Her willingness to share incidents like that one, and to encourage, to educate, and to empower others was a hallmark of her life, not only in the VHL community. For eight years Eva Logan was a member of the committee of volunteers who answer the VHL Family Alliance hotline. Sweet and sassy, with a delicious sense of humor and an instinct for empathy, she spent her life as a "Tennessee Volunteer" in the truest sense.

Eva Hayes Logan was born and raised in Memphis, Tennessee. The youngest of five children, she was the impish one, always pestering and delighting her three older sisters and one older brother. She was friends with the two daughters of Dr. Long, a Protestant minister on their street. When she was staying over with them one night, Dr. Long asked the girls to quiet down and go to sleep. Eva matter-of-factly informed him that when she was staying over, the house rules did not apply.

Eva and Chris Logan were high school sweethearts who married and enjoyed twenty-eight years together as husband and wife. Chris attended the University of Tennessee in Knoxville, and he and Eva traveled extensively before settling in the Atlanta area in 1989. Chris was in the U.S. Air Force and later became a commercial pilot. In each location where they established their home – Germany, Oklahoma, Nevada, Louisiana, Atlanta – Eva adapted and developed friendships that spanned the test of time.

Chris and Eva Logan



From an early age she enjoyed volunteer service. She served in the Rainbow Girls as a teenager. In her adult life she was a preschool teacher, a physical therapist aide at the Veterans' Hospital. Her greatest effort was in preventing domestic violence and protecting battered women. She served as a coordinator and speaker for the Fayette County Council for Battered Spouses. She was instrumental in the creation of Lighthouse, a transitional home for battered women and their children. She helped to start the Teen Dating Violence Program, educating teens in avoiding and stopping date abuse, and counseled with adult women to empower them to break the cycle of abuse.

In addition to her work with the VHL hotline, she spoke before groups of medical students and personnel at Emory University in Atlanta and at the National Cancer Institute. She spoke before committees of Congress on the subject of VHL, encouraging them to increase funding for VHL research at the National Cancer Institute.

When I was feeling anxious about VHL issues in my own family, I always knew I could phone Eva for a dose of warm down-home wisdom and perspective. Eva passed away January 25, 2002, at the age of 49. We celebrate her life and forever cherish her memory.

My Aunt Eva, by Kari Beth N., Mississippi

You left this world just yesterday,
To be in a better place.
I already long to hear your voice,
And see your smiling face.
There is so much I wish I could have said to you,
So many thank you's for your love and care.
But your time on earth is done,
And I will have to wait to get to heaven and tell you there.
I prayed for God to tell you,
How dearly I loved you and how much you meant to me,
I tried my best not to cry,
Because I did not want you to see.
You will never know how much you inspired me,
And how your life impacted mine.
Your bright light of love for all of life,
Will continue to shine.
I thank God for the Memory of you,
That I will always keep with me.
And I will push myself to make you proud,
Just you wait and see.

How could this happen?

Eva's death was caused by metastatic cancer throughout her body. One kidney tumor grew to 5 centimeters, moved outside the kidney capsule, and metastasized.

Nearly every day for the past month people have called the hotline to ask, "How could this happen? And to Eva, of all people? Eva is the one who has been coaching me in managing my health!"

We have seen it before — Adele Davis was one of the first proponents of nutrition for cancer prevention. When her husband died of cancer in his 70's many people took that as an opportunity to question her entire philosophy. Dr. Jim Fixx, the first big advocate of running and aerobic exercise to take care of your heart, died of a heart attack. Such things call the strategy into question, and it is always good to examine and learn from such sad events. But just as their strategies proved to be good ones, the strategies that Eva taught — those which are to be found in the *VHL Handbook* — are still what we need to follow. And we definitely need to learn more!

In the spirit of learning, we posed some key questions to our medical advisors:

Question: We have a "guideline" that says we can wait until the tumor reaches 3 cm before taking action. We know that's not foolproof, but is a rough measure of tumor activity. The kidney tumor in question seemed to grow more quickly than expected — growing in six months from 2.5 cm to 5 cm. We know that size is only a crude estimate of the danger level of a kidney tumor — what we really want to measure is its activity level. Are there any better ways to measure the activity level of a tumor?

Answer: Not yet, but this is an active area of research for all cancers. It's the same question we have in the pancreas, the breast — everywhere that cancers can grow, there can also be benign growths, or early growths, that require more or less action depending on their levels of activity. When activity levels are low, less invasive actions can be taken that will solve the problem with little impact on the patient. When activity levels are high, more aggressive action should be taken even if there is more surgical impact, because the impact of not acting would be more destructive than the impact of the surgery.

My sister Eva and I were closer than twins, though I was eight years older. I can't find the words to describe how much I miss her voice and her presence in my daily life. We shared our deepest feelings, our impossible dreams, our worries for the family and their health problems. We loved and encouraged each other endlessly.

Thank you for honoring Eva in this way.
— Peggy Marshall, Mississippi

There are modalities currently in clinical trials that should help us — for example, PET markers to show on a scan the level of VEGF production surrounding the tumor. Once we have this kind of tool available, we will be able to monitor a tumor more closely and more often with less cost, less radiation exposure, and more helpful information.

Question: When the 5 cm tumor was removed, they had to remove some back muscle to get a clean margin. Clearly at this point it was known that the tumor was outside the kidney capsule, and the pathology report showed that it was a grade 3-4 tumor (with high metastatic potential). In a case like this should there be follow-up radiation or chemotherapy? How frequently should you have check-ups to check for metastasis?

Answer: The situation you describe is already dangerous. These are good questions that can only be answered in conference with your own medical team, with all the scans, pathology information, and the total medical picture at hand. If you are not comfortable or confident with the answers being given locally, do not hesitate to seek a second opinion, especially from a team with greater experience in VHL. Chemotherapy itself can be a difficult course, has a low chance of success, and is not for everyone. Further surgery may or may not be helpful.

Question: Eva had had a difficult recovery from a prior kidney surgery, and dreaded having open surgery again. She wanted to have Radio Frequency Ablation, not open surgery. This relatively large tumor did not respond to RFA treatment, but continued to grow. What should we know about qualifying a kidney tumor for RFA treatment versus open surgery?

Answer: It is still early in our learning about radiofrequency thermal ablation (RFA). This minimally invasive, image-guided therapy may now provide effective local treatment of VHL tumors, and can also be used in other cancer tumors as an adjunct to conventional surgery, systemic chemotherapy, or radiation. We have the greatest amount of data for liver cancer, and have treated 24 kidney tumors including 19 patients with VHL (see Note 1). While nearly all patients came through the treatment itself very well, we do not yet know how well the tumor will be inactivated over the long term. This treatment will remain experimental until we have a better grasp on ways to measure tumor destruction.

If a target can be seen with CT, MR, or ultrasound, then a needle can be placed into it. If a needle can be placed, then the target tissue or tumor can be ablated and destroyed. If a clean margin is created, then the tumor should not recur at that site. Recent developments in RFA allow this treatment process to be done in a safe, predictable, and cheap fashion with low

complication rates and minimal discomfort, on an outpatient basis. Further study is required to assess which patients will benefit from this new treatment, and most patients will not be candidates due to the size or location of the tumor. Although long-term data have yet to be reported, early results suggest that RFA may prove to be an effective treatment option for small kidney tumors (under 3 cm), disabling them before they grow to larger sizes. The standard of care for tumors larger than 3 cm continues to be surgical removal of the tumor.

As Eva's husband Chris said, "In 20/20 hindsight, I feel our medical system has become so specialized in their fields of expertise that the patient has to now act as their own general health coordinator. In too many cases the general practitioner just shuffles you off to the various specialists and doesn't fulfill the role of coordinator needed to protect the patient." Get copies and read your radiology reports and pathology reports. Take someone with you to appointments. Don't depend on understanding everything the doctor tells you after surgery while you are still in pain and on drugs. Read the reports again a few weeks later, and ask all your questions. Don't be shy. The doctor has many patients, but you have only one body. It's your responsibility, even more so that the doctor, to take charge of your own health.

"Even though Eva wasn't given the opportunity to grow old as gracefully as I would have liked," says Chris, "she did receive better healthcare than the generation before her. Her ability to get more and more information concerning VHL did permit her to minimize or alleviate the affects this disease has the potential to inflict on the individual. Unfortunately, she didn't beat the disease, but she certainly made her mark on improving people's lives. She left us much to treasure."

Note 1: B. Wood et al., "Percutaneous tumor ablation", *American Cancer Society* (2002). C. Pavlovich et al, "Percutaneous tumor ablation of small renal tumors: Initial Results", *Journal of Urology* 167:10-15, January 2002.

See also <http://www.cc.nih.gov/drd/rfa>

Our thanks to Chris Logan, Peggy Marshall, Dr. Peter Choyke, and Dr. Bradford Wood of the NIH for their assistance in preparing this article.

Gale Lugo and the Florida Chapter, January 2002

Thank you for a wonderful experience at our meeting in Orlando. We learned a lot, but, we had so much fun, too. The remarkable thing is the attitude of all the guests. We all knew the incredible problems experienced by all, but, no one wore their problems on their sleeve. They just shared what might be of benefit to all. We had a lot of fun learning, too. Thanks again, Gale, you and your husband are great hosts. -- Alva L., Florida, a VHL Veteran, age 78

Ask the Experts

Skin Involvement?

Question: I recently visited my family doctor concerning strange bruising or rash on my feet and legs. I am wondering if there is anything in VHL that causes this, or not. My family doctor is referring me to a dermatologist to see if there is some other reason, or it may be nothing. I have had these marks for almost a year now. One thing my doctor did say that is a possibility is that it could be blood vessels growing close to the top layer of skin.

Answer: I have not seen bruising or rash on the feet and legs in any of the hundreds of patients affected by VHL whom I have screened. Other causes should be pursued in the differential diagnoses, including localized vascular insufficiency or peripheral neuropathies. If bruising is generalized (occurring randomly on various part os the body) see a Hematologist who will order blood tests that may diagnose a treatable problem.

Regarding VHL, I would also check for an elevated red blood cell (RBC) count and elevated erythropoietin levels, causing a reddened or flushed appearance, although this does not tend to be localized as described in this case. There have been a few individuals with VHL who have had secondary polycythemia (abnormally high RBCs). If the RBCs are abnormally high, a hematologist will advise whether the level is high enough to require therapeutic phlebotomy (drawing off of some blood) to relieve vascular congestion.

-- Gladys Glenn, M.D., Ph.D., Medical Officer, Genetic Epidemiology Branch, National Cancer Institute, Bethesda, Maryland

Standing MRI?

Question: What can you tell me about the open standing MRI? Are the pictures as good as with the tunnel kind? So many people are claustrophobic that this would be an interesting option as long as the definition is good. Are there a couple of key questions that people could ask to determine whether it would be a wise decision to use the open MRI or not?



Answer: Experience with this type of magnet is still limited. The images are not the same quality as with the enclosed magnets which are also higher in field strength. Moreover, the field of view is smaller, so each image covers a smaller area of the body and more pictures are required.

My first choice for claustrophobic patients who need an MRI would be some light sedation and scanning in a good enclosed high field magnet. Failing that, some of the newer open horizontal magnets are getting pretty good. A Standing is my last choice because of the lack of experience with it and the problem of motion -- imagine standing perfectly still for 5-10 minutes! However, I agree completely that the manufacturers should come up with better designs for their magnets, and there are some under development. But meanwhile we shouldn't compromise on image quality, since missing those little tumors can be a big problem

Here are the question I would ask myself when considering an MRI:

1. Can I tolerate (even with some mild sedation) a closed MRI scan? If yes, go for a 1.5 Tesla magnet, (preferably one of the major manufacturers like Siemens, Philips or GE). (Tesla is a measure of the strength of the magnet).

2. If no, what are the available open MR scans in my area? Pick the newest one in the area in a radiology group with a good reputation, (latest models of Toshiba, GE, and Hitachi are good). The early ones were barely satisfactory. The standing open is another option but is not superior to the others for VHL screening. We recently saw a case in which a large mass was missed on an open magnet but was picked up on the closed one. They are not yet equal.

3. Make sure the radiology group has a knowledgeable neuroradiologist familiar with VHL! People are as important as machines! If this is not available locally, ask another radiologist with greater VHL experience for a second opinion on the scans.

-- Peter Choyke, M.D., Chief of MRI, Diagnostic Radiology Branch, Clinical Center, National Cancer Institute, Bethesda, Maryland

Cleveland meeting: Moved to Oct. 11-13

We apologize for any inconvenience.

Please change your hotel booking

For details, please see page 7

Health Care Proxy

A durable power of attorney for health care is a signed, dated, and witnessed legal document commonly called a "health care proxy" or "health care power of attorney." This document differs from other durable powers of attorney in that it is specifically limited to health care and medically related decisions.

For anyone facing surgery, this document can be one way to ensure that he or she stays in charge of his or her own destiny. The patient (known as "the principal") appoints an individual (called "agent," or "proxy") to make medical decisions in his or her behalf if (and only if) the patient becomes incapacitated and unable to make such decisions. The agent can be any competent adult, and is usually a family member or close friend. The appointed agent will take over medical affairs and act within the guidelines and restrictions should the patient become incapacitated due to illness or injury.

A durable power of attorney for health care should:

- Adhere to the rules of the state or country in which it is created (local laws vary)
- Include a backup if the original agent is not available
- Spell out clearly what matters the agent can and cannot handle

“ This can be one way to ensure that you stay in charge of your own destiny. ”

Prior to signing a durable power of attorney for health care, the principal and agent must designate under what conditions the document becomes effective. The agent would then be responsible for all medical matters until the principal's recovery or death.

Most states also allow the principal to request that he or she not be given "life sustaining procedures" in cases of terminal illness. The agent will be able to follow the principal's wishes to a greater extent if the document clearly defines and specifies the powers being granted. This should also be discussed openly among family members, as it presents a problem for doctors if the whole family is not in agreement.

If your loved one signs a durable power of attorney for health care, make sure he or she communicates all wishes, values, and preferences to his or her agent. A lawyer can help you understand how to tailor the durable power of attorney for health care to fit your loved one's wishes.

Hospitals often have forms available that follow the rules in your state or country, or you may wish to contact an attorney.

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Ask the Family: DNA Testing

A conversation from our online discussion group, vhlfa@yahoogroups.com

Question: What benefit would there be to those of us who already know we are affected, to have a DNA test to confirm? Is there any information that a blood test could give us? I already know I have VHL.

— Brian N., Massachusetts

Answer (1): Genetic testing is needed for the parents, in order to be able to test the kids. It helps to find their mutation much faster if they have already diagnosed where the mutation is in the VHL parent. I am not sure if finding out your mutation has any bearing, unless you need it for other family members. Maybe somebody else has more information about that?? My sister had hers done, even though we knew she had VHL, so that both of her sons could have the genetic testing. Even in the last year or so, testing has become even more reliable than before. Her boys were 7 and 9. One has VHL and one does not.

— Lori M., Wisconsin

Answer (2): Aside from providing a match for other family members to be compared to if you were the only diagnosed member, the results from your positive DNA test would be added to the growing database of info on VHL. The greater the numbers of cases studied, the better the chances are of finding and beginning to understand the many variations of VHL... some of this might not become useful for years yet, but each tiny data point is part of the puzzle. It isn't of direct value, but could well come back to serve you or your descendants some day. A current "benefit" in some cases is being able to distinguish between VHL type I, and type IIa and IIb, which helps anticipate which tumor types are most likely.

— Elaine F., New Hampshire

Question: What is the common wisdom about testing children? My niece is 6 now, and she has never been tested. As much as my mother and I push for it, he would rather not have her tested so young. His physician believes that a positive result will cause him to blame every little snuffle on VHL. So, what is the consensus — panic versus knowledge? Seems easy to me, what do you all think? (she is asymptomatic). — Brian N., Massachusetts

Answer (1): We believe that the genetic testing is so much better, simply because then you know. Others, however, feel that parents or other children may behave differently toward the one with VHL versus the ones without it. If I found out that my parents had not tested me genetically, and it was available, I would be angry at having to go through awful tests year after year for *nothing*. Why put your children through it? Of course, some people just

don't want to know. There is one downside to genetic testing, insurance could be harder to get, once a positive test comes back indicating they have VHL. It is recommended if you don't want to know that at least you do some of the general tests yearly to check on the health of the child. When they are this young, they start out with only minor tests.

My sister was asymptomatic for a very long time. They were sure that she did not carry the VHL gene. Nothing was showing up to indicate VHL. (DNA testing was not available then.) However, on her 21st birthday she was released from the hospital after having a hemangioblastoma removed from her cerebellum. I was released from the hospital only two days before her, after having the same surgery. Her tumor was twice the size of mine. We had known I had VHL, but hers was a very big surprise! My point is that symptoms don't always show up to let you know something is going on.

My husband and I have adopted, so we have not had to face this issue. We would definitely test, if we ever went down that path, as soon as our doctor recommended it. Of course there is also testing available while you are pregnant, in case you would want to know that early.

Take care, Brian, and good luck with your niece! Be patient and persistent all at the same time!

— Lori M., Wisconsin

Answer (2): I'd like to add my two cents worth. When you begin to think about insurance companies and the impact of genetic testing on acceptability, it is pretty easy to see how one could think that they were "keeping the secret" by not getting tested. In reality, I cannot imagine that there are any insurance companies that could not view one's medical files and be stunned at the number of screenings that are part of our lives. I think it would be much better to have the DNA test and hopefully make all the screening unnecessary with a negative test. Who needs the expense and trauma? Even if you test positive, knowing is the first step in taking control of your life and taking better care of yourself. — Deb C., Michigan

Answer (3): This is a personal opinion and only one, but when I had my spinal tumors removed in 1998, we were sure that I had VHL. Our first step after me was to determine if my two children also had the VHL gene. Neither I, nor my husband, ever wanted our children to go through all of the tests if they were unwarranted. Much to our sorrow, both of the boys have the gene. It is now up to us, as parents, to follow through with their tests until they become of age. The only medicine we have at this point is prevention and screening. Wouldn't you want to know one way or another before a disaster strikes?

Whatever decision you make will be up to you and your family, good luck on your choice.

— Rhonda S., Malaysia

Answer (4): I don't know about common wisdom, but we've decided that there is little to be gained from testing before the "heavy-duty" scanning starts at puberty. Our kids are 6 and 9, and haven't been tested yet. It was pointed out by a genetics counselor, that once you have the results, a positive result will alter a family's dynamics, even by a wee bit, no matter how hard you try (or maybe *because* you try so hard) — it's inescapable. Delaying that day isn't necessarily a bad thing, so long as it doesn't increase the risk for the child. It is an easy thing for me to rationalize about testing, but on a purely emotional level, the simple hint of the thought that this could affect either of my children directly just turns me to ice. I cannot be the least bit confident that I'd deal with the news half as well as I'd wish to. Knowledge can bear a grain of panic, and we may find temporary security in putting off testing as long as prudently possible.

There is very little difference in how a pediatrician would examine a pre-teen that tested positive vs. negative. Other than retinal checks that are done by my husband's ophthalmologist, pho testing, and close attention to neurological development, our yearly checkup routine is quite ordinary. Our pediatrician has had pediatric VHL experience, and I trust his judgement. Once the intensive screening begins, I can't see putting a child through it if it isn't necessary, and I'd push for testing. I have to admit though, I'm starting to wonder if the concept of having inherited this disease isn't easier to absorb for a younger kid (ages 10-11) than for a teen. A young child's grasp of the ramifications might not be quite as wide, and the future beyond a month or two away is generally fuzzier. Being a teen is such an emotional time, that being hit with this scary bombshell, and then an arsenal of scans, is an awful lot to cope with all at once. I think I'd like to see the "idea" of a positive result being digested, before having to deal with the "reality" of it. We're considering DNA testing in the next year or so, to give us all time to come to terms with any fallout. — Elaine F., New Hampshire

VHL Stands for . . .

The Online discussion group has been having a half-teasing conversation about coming up with an alternate resolution for the acronym VHL: Marc suggested Very Handsome and Looking (just kidding). Loreen suggested that VHLFA helps us move from Very Heavy Load to Very Healing Love. Deb C. suggested that we are all living Very Heroic Lives. Our transformational statement is that we want to find a cure and make VHL stand for Very Happy Life.

What do you think? Please send your suggestions to editor@vhl.org or 1-617-277-5667

Volunteering for Health

from Medscape Health for Consumers, Beth Israel Deaconess Hospital, Boston, www.medscape.com

In 1974, when Louise Jackson was in her late 50s, her knees had become so painful that doctors prescribed a cane to help her walk. But as Jackson became more and more involved with the foster-grandparent program of the national Senior Service Corps, she found her days getting busier and busier.

One day, she lost the cane—and never bothered getting another one. Today, at the age of 84, Louise Jackson puts in full days teaching troubled parents—many of them drug addicts—how to parent their children, escorting them from the grocery store, to their children's schools, to the laundromat and back home. "It's like the whole family's my children," she says.

And those bad knees? Oh, they still bother her from time to time. "Sometimes I get up and my knee's hurting me and I think, 'I don't want to get out of here.' But I get up and go and a little while later, I don't think about my knees any more," she says. "I know my health is better. If people would just get up out of their house and do something for other people, they'd feel better about themselves."

More and more, research is telling us what Louise Jackson and other senior volunteers already know: Volunteering not only makes people feel better about themselves, it actually lengthens their lives and improves their health. A 1999 study from the University of Michigan found that seniors who spent less than an hour a week volunteering were less likely to die over the period of the study (7 1/2 years) than those who didn't volunteer at all.

By carefully controlling for factors like existing health conditions and other physical activities, the researchers also documented that people live longer because they volunteer—rather than people volunteer because they're healthier and thus likelier to live longer. "Quite a few people assume that older volunteers should benefit in terms of better health and well-being," says Marc Musick, PhD, an author of the study who's now affiliated with the University of Texas. "This study is one of the first to document that is true in a nationally representative sample of older Americans."

This community is strong because of its many willing hands and hearts

Please call to offer your help

VHL Family Alliance

info@vhl.org, +1 617 277-5667

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Carlos Casal, by Joyce Graff
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 Exclusives
 Audrey Tobin, by Susie Clark-Tenney
 All the VHL volunteers, by Carol Karal, John & Mary Jo Libertino
 Timothy Walter, by Joseph & Jenifer Chapko
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In Memory of . . .

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Carrying the Torch

The Olympic Flame, brought by air from Greece, traveled 13,500 miles through 46 U.S. states on its way to commencing the Salt Lake Games. Over 11,500 torchbearers and countless others participated, making this Torch Relay the largest in the history of the Olympic Games.

In Olympia, Washington, one of the torchbearers was Larry Bennett, 39, who has VHL. Ten years ago a number of doctors were removing whole kidneys. Larry lost the last of his own kidney power in 1993, his second nephrectomy. After a difficult year on dialysis, he received his new kidney from his brother Darl. "Dialysis was life-preserving," his wife Debbie says, "but the quality of life was one of mere survival — the transplant literally gave us our family back. Larry is doing great now. We don't take a single day of healthy living for granted."

In her nomination letter, Debbie wrote, "He is an inspiration to many because of the numerous trials he



has been through. He has had each kidney removed due to cancer, had a kidney transplant and 2 brain surgeries in the 18 years of our marriage. This would devastate many, but he truly looks at these as part of life — bumps in the road to deal with and move on. He truly is an inspiration."

When he ran near an elementary school, a group of first graders wanted to touch the torch. They asked Larry why he got to carry the torch. "Instead of giving them a long talk on genetics, I just told them with the help of my family I survived being very sick."

We cannot control everything that happens to us in life — accidents, layoffs, and health issues — we can only control how we react to these events and how we deal with them. We celebrate the Olympics by honoring each person with VHL for the way they manage their own "marathons" with this chronic illness.

Photo caption: Larry with his family: L to R, Jordan (13), wife Debbie, Nicole (15), and Andrew in front (11).



VHL Family Forum

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