



VHL Family Forum



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To Know, or Not to Know . . . That is *the* Question

It's one thing to choose for yourself whether you want to know if you carry the VHL gene. When Lisa Carcieri Tuttle surveyed 200 people with VHL in 1997, 45% had already been tested, another 33% said they would get tested.¹ For an adult, it represents fact rather than conjecture, facts on which you can build the rest of your life. If you don't carry the altered VHL gene, then you can stop worrying about VHL in yourself and your children. If you do carry the altered gene, you can use this early warning information to protect yourself.

Making the choice to do DNA testing for a child, however, is somewhat different. The professional associations of geneticists and genetic counselors, are firm in stating that DNA testing of a child should be for the benefit of the child, not the parent. Unless there is material benefit for the child, they prefer not to offer testing for young children. When the condition rarely affects very young children, as in VHL, the feeling is that the child should participate in the decision whether to test. However VHL can indeed occur in young children, and knowledge of which child is at risk allows you to focus watchfulness and attention on the children in the family who do have the altered gene.

At the VHL/MEN Symposium in the Netherlands in 1997, the attendees, predominantly internists, endocrinologists, and biomedical researchers, were asked at what age they would recommend DNA testing for VHL. 6% said prenatally, 15% between 1 and 5 years, 34% between 5 and 10 years, 16% when regular testing activities begin (about age 12-14), 17% said not until the child was old enough to choose, and 3% said only if symptoms are present.²

The opinion of the doctor is not necessarily the opinion of the parent, nor the child. There are clear issues of emotion and responsibility that emerge. Do you really want to know? Is it better not to know, and

to let the child have a normal childhood? If you know you have one child with VHL and one who does not, how will you react? Will you give one child more love than the other because of this information? or more attention?

"For me, I simply had to know. I knew that whatever the test results were, I was going to have this baby. My husband and I had talked about it, and decided that whatever God gave us, we would love and cherish our child. But I just had to know. I arranged for prenatal DNA testing. Putting it on my own health record didn't tell the insurance company anything they didn't already know about me. We learned that our daughter did carry the altered gene. It was difficult news, but in a way it gave us a chance to make peace with it before she was born, and strengthen our commitment to love and protect her." -- *Shelley, Maryland*

Not knowing is also difficult to live with. "The most terrifying thing my son could say to me was, "Mom, is there any aspirin? I have a headache." My

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heart would seize, and I would mentally go through my checklist:

- Take several deep breaths. don't panic. Easier said than done.
- Take it one step at a time, moving in an orderly manner through ordinary causes
- Check for fever. It could be a cold.
- Have others in the family, or in the child's close circle had similar symptoms recently? It could be the same cold.
- Give the child a standard headache remedy, like tylenol. Does the headache go away during the next hour?
- Watch for signs of heavy sweating, flushed face, panic or hyperactivity
- Over the next few days, is the headache still there? in the same place?
- Do some "casual" neuro testing — play ball, touch nose, walk straight. Do you see changes from the child's normal responses, balance, accuracy?
- "Where is the headache?" "If it were a fruit, what fruit would it be? What color? How big?" It helps to quantify the headache. Simply acknowledging an ordinary headache, and doing some relaxation breathing, can help it to go away. If it goes on longer, this game gives you a subjective measure to tell whether it's getting better or worse.

Any persistent headache, especially one that stays in one place for more than a day, should be checked with the doctor. Be sure to tell the pediatrician that there is VHL in the family. There are screening guidelines in the back of the Handbook. Ask that they do the screening routine appropriate to the child's age. Note that these are only guidelines — there are always exceptions.

Just because it says to begin testing at a particular age doesn't mean that an issue does not appear earlier, but it is rare at those earlier ages. If you suspect involvement, even before the age range begins, ask your doctor to check it out and hopefully rule it out. Headaches might be caused by brain tumors or pheos. In addition, there are many other perfectly ordinary and very treatable causes of headaches in children. Just take it one step at a time with a caring and cooperative physician who will take you seriously and walk you through the "trouble-shooting" to determine the most likely cause. It usually is a garden-variety headache.

With clear information on VHL status at younger ages, screening is finding small tumors at younger ages -- not so much because they are occurring at younger ages, but because we are looking. This raises new challenges for treatment -- what, if anything, should be done? Just because we can see a small tumor does not necessarily mean we need to act. New strategies must be developed for preventive maintenance to optimize quality of life.

DNA testing for young children

We heard from a number of members on this topic, but very few whose children turned out to have the VHL gene. We would appreciate hearing from others who have tested young children. How do you feel now about that decision? If you had it to do over again, would you make the same choice? Please provide feedback and information for others approaching that decision today.

Similarly, we would love to hear from young people whose parents made the decision for them. Are you content with your parents' decision?

Not at this Time

My genetic mutation has been genetically identified. I have an eight year old and a four year old. They have no symptoms. My daughter, the eight year old, has had one ophthalmological exam with a negative result. I don't wish to have them tested at this time because of the possible dangers of knowledge of a pre-existing condition as it pertains to insurability.

VHL is said to present most commonly in late teens and twenties.³ This diminishes my sense of urgency about having my children tested. I intend to be watchful with the children, as does my wife, and when they get older, I will discuss with them the nature of a genetic disorder and the risk that it poses to them. They can then have input about whether or not to be tested. Should any seemingly related symptom arise before they are tested, we would move to test the child to help determine the cause of the symptom. -- C.P., California

Gentle Bequest

One of the last things my late husband Eddie did before he died was to submit a sample of his blood and his mother's blood for DNA testing. Delayed diagnosis led to his death from metastatic kidney cancer. We found out on Tuesday that they found the mutation, and he passed away that Friday. I know it set his mind at ease to know that his children and the rest of his family could be tested to find out if they had VHL. He firmly believed it was better to know if you had VHL so you could be treated and followed properly.

We didn't know all the ramifications of being diagnosed with VHL. Since then four other people in our family have been diagnosed with the gene. Two had to have immediate surgery and two are being watched. I feel Eddie's insistence that everyone be tested once there was a match was a true gift to the four people who found out they have VHL. Now all four will have the best possible chance to be followed, and live as normal a life as possible. -- Linda G., Georgia

Not to be Missed

I am 42 years old and have been dealing with VHL since I was 15 years old. I have had bilateral pheochromocytomas, retinal hemangiomas, thyroid cancer

and kidney cancer. These of course have to be dealt with by surgery. If that was not enough, I have also had open heart surgery, appendectomy, C-section, and a complete hysterectomy, unrelated to VHL.

The reason for my letter is to report that we have finally gotten the genetic test back for my 6 year old daughter, and I am happy to report that she does not have the VHL mutation. YIPPEE!! I kept telling my husband that as long as she got my good looks and his good DNA I would be a happy woman. Well God has blessed us and we are so very thankful.

Having a child is a difficult and serious choice. I was born with a heart defect as well as VHL. So when I had my doubts and fears over the last 5 years if she had VHL and how I would handle it if she did, I would just remember what my mother would tell me growing up. She would say, "I wouldn't have missed this time with you, no matter how difficult and painful at times it seemed. I'm just glad that God gave you to me". Oh, I forgot to mention that my daughter has a heart defect also, and I'm just glad God gave her to me because our time together is wonderful. -- *Stephanie B., California*

Three for three

My husband comes from a family of four and two out of the four have VHL. We have three children, 4 yrs. 11, and 14. With the odds working in VHL's favor, we were sure at least one of the three would inherit the illness. Determined to stay ahead of the disease, we decided to go ahead with the DNA testing.

We received our phone call from the doctors office yesterday....all three children are negative! What are the odds of that? Thanks for letting me share my good news with you.... -- *Chris M.*

Best Decision

My mom was first diagnosed with VHL in 1996. She had her first VHL related surgery on her spine in 1979. She also had a brain tumor removed in 1985. It wasn't until another tumor was removed in 1995 that VHL was first mentioned. Our family had no idea. At the time of my mom's 1995 surgery, my grandpa was found to have a brain tumor. He had his removed in 1996, at the same time my mom had another brain tumor removed. It was at this time that her siblings were tested. It took much encouragement on the behalf of my father, myself and a couple of my mom's siblings to persuade the others to get tested so they could know for their children as well. My brother and I were tested and found to be negative, but we then learned our family had a deletion⁴ of the VHL gene, so we are awaiting insurance approval of another test to know for sure. At this time there is only a 20% chance we have VHL. I want to be tested to know for sure, so I don't worry if I have a headache or am clumsy one day that I have a tumor. I also want to be tested to I know that if I do have VHL, that I can get regular monitoring and appropriate treatment. My

mom went thirteen years before being adequately treated and I do not want this to happen to me, to any other family member nor to anyone else.

My mom had some serious side effects from her surgeries because her tumors grew so large and damaged so much of her brain before they were caught and surgically removed. My family sees what my mom went through and they don't want to end up as bad as she is. What I try to tell my family and want others to know, is that if you know you have VHL, you can get adequate testing, monitoring, and treatment for VHL related occurrences. You don't have to wait three months of being ill, with every other imaginable ailment being investigated before a tumor is found and VHL diagnosed. In my eyes, I see no option to the DNA testing. A negative test can ease a lot of worrying and heartache and at the same time discovering you have VHL is not a death sentence, it's the beginning of taking charge of your health. The wait for the results is an emotional roller coaster, but it is well worth it.

I'm 22, and it was one of the best decisions I ever made. -- *Jennifer O., Michigan*

1. VHLFF:5 (June 1997) 2:4-5.

2. Summary of the "Consensus Meeting June 28, 1997," from Dr. C.J.M. Lips, October 1997. His figures combine the electronic voting results with questionnaires turned in by attendees who left prior to the final session and are therefore slightly different from numbers reported previously in VHLFF which were based entirely on the electronic voting.

3. This is a myth. We have seen pheos in young children (age 8) as well as eye tumors (age 2), and even brain and spinal tumors (age 13). Eye tumors do not have obvious symptoms. If you are not going to do DNA testing, you should begin the gentle screening for children recommended in the handbook no later than age 6. -- *Peggy Marshall.*

4. Early tests using linkage analysis, especially in families with deletions, have been found to be unreliable. Please see the article on page 3 to determine whether you need to re-test to confirm the results.

Linkage Test Unreliable

Question: Audrey T. was tested by a reputable lab two years ago using linkage analysis. She was told she does have the altered VHL gene. She did comprehensive testing, and worried a lot both about herself and about her two adult children and potential grandchildren. Clinical testing revealed only one cyst on the kidney, which is not diagnostic of VHL. Subsequently, the actual alteration in the VHL gene in her family was located, making the direct test available. When her sample was re-tested using direct testing, they found that she does not have the altered gene! While we are thrilled with her results, we are shocked and angry about the two years lived with the wrong information. It raises the question: how many others received false-positive or worse, false-negative results from DNA testing in the past? Should people

get re-tested now that the tests have become so much more reliable? -- *Lois E., Minnesota*

Answer: As we all know, before direct mutation analysis became available, linkage analysis was offered to some families on a research basis and to some on a clinical basis by various centers. Linkage analysis is still used for analysis for some diseases when mutation analysis is not available or has a low rate of mutation detection. I.e., the technique is not obsolete in some circumstances for some diseases. However, for VHL, direct mutation analysis has largely replaced linkage as a preferable test in most cases. Linkage analysis has an inherent misdiagnosis rate for many reasons, that can include crossover events, errors in pedigree structure, incorrect assignment of paternity, low variability of markers, etc. It should be an individual/family decision on who might need retesting, made with the help of a knowledgeable physician or genetic counselor. For some families, all predictions may have already been proven correct and not need retesting.

For those given a normal result, retesting by mutation analysis would certainly be a reasonable option for an added measure of reassurance. It is important to point out that this applies to any lab that did linkage. I am aware of a research lab that gave a misdiagnosis based on linkage as well. Finally, it is important to remember that any lab test can be in error, for e.g. sample mix-up, regardless of the type of test. -- *Virginia V. Michels, M.D., Chief, Genetics, Mayo Clinic, Rochester, Minnesota*

DNA Testing finds 100%

-- *Berton Zbar, M.D., Maryland*

I just returned from a visit to Italy. While there I visited Alessandra Murgia and Dr. Opocher. Padua has considerable charm as a city that has crowds of people yet retains features of a medieval town. Padua would get my vote for an international VHL meeting.

With regard to the germline VHL mutation detection rate. First, Dr. Murgia has achieved a 100% germline VHL mutation rate (14/14 Italian VHL families). Dr. Murgia has been helped in her work by Dr. Catherine Stolle of the University of Pennsylvania.

Second, I recommended that DNA samples from VHL families for whom the mutations were not detected be sent to Dr. Stolle. This work was just completed and Stolle found germline mutations in 10 of the 11 families whose mutation had not previously been found. The sample in which Stolle did not find a germline VHL mutation detection is probably not VHL.

These results suggest that now a 100% germline VHL mutation rate is attainable by at least two labs: those in Philadelphia and Padua. Their methods are standard and published. Dr. Stolle is open to having other labs contact her for advice on mutation detection. Dept of Genetics, U. Pennsylvania School of Medicine, 415 Curie Blvd., Philadelphia PA 19104-6145; W: 215-573-9161; Fax: 215-573-7760.

Show Us Your Stuff!

Photos - Drawings - Paintings

We are seeking art work for the Year 2000 VHL Calendar! They can be any photograph, drawing, or painting done by a member of our community. Please send *only copies*, as the submission cannot be returned.

Please send by 15 June to: Pierre & Lisa Bonneau, 1761 W. Waterbuck Drive, Phoenix, Arizona 85742

Twelve winners will be chosen, and will be notified by September 1. Winners will be featured in the Year 2000 VHL Calendar.

Year 2000 calendars will be ready by early fall, and available for purchase at the pre-publication rate of \$3 each in quantities greater than ten.

Air Miles

Do you have air miles about to expire? Delta miles can be donated to VHLFA! If you wish to donate miles, please send your request in writing to Delta Airlines, Inc., Dept 745, Skymiles Award Travel, P.O. Box 20543, Atlanta, Georgia 30320-2543. VHLFA uses miles to send volunteer representatives to speak at large conferences, or to bring special speakers to VHLFA meetings.

See you in Atlanta!
See page 9 for details.

Most Important Person

by *Craig Lundsted, M.S.W.*

Who is the most important person in the medical management of a person with VHL? I asked this question while in the hospital for removal of my adrenal gland. The answers were usually the same. Doctors thought the doctors were. Nurses thought nurses were. Not so.

The most important person is YOU.

You need a primary doctor you have confidence in -- a doctor that knows about von Hippel-Lindau syndrome or is willing to learn about VHL. It is important to have as a gatekeeper a doctor willing to admit that he or she does not know all the answers and who will refer you to a doctor more knowledgeable in the area of concern.

Nurses who care are important. I had nurses who cared while in the intensive care unit. Knowing she cared minimized my anger at the clumsiness of one nurse who kept bumping into my bed.

Other hospital staff personnel are important if they give the feeling of concern and empathy. Family and friends are important for the support they give to the person with VHL. You are the most important person in the medical management of your VHL treatment for a variety of reasons.

- ♦ Nobody else can tell you how you are feeling. For example, I was the only person who could describe the symptoms of back pain and throbbing in my back invoked by even minimal exercise. Accurate descriptions can help the doctor pinpoint the source.

- ♦ Although the doctor can detect and identify serious problems like high blood pressure, prescribe medications, and make referrals; it is up to you to follow the doctor's advice.

- ♦ There are many unresolved feelings when you learn you have VHL and what it could mean to you. I needed to work through my feelings about an unknown future with VHL.

- ♦ You need to be your own advocate and take responsibility for learning about the disease and how you can help yourself. I had to be responsible for asking any questions I had and not be afraid of asking what seemed like dumb questions. The only dumb question is the question you don't ask.

- ♦ Because you are involved with other family members, you need to be aware and deal with their emotional feelings. Their support is important to your own emotional and physical health.

- ♦ Regardless of what your medical future is, you need to decide for yourself what gives you peace of mind, a sense of purpose for your life, and how to keep control of your life.

Although other people such as family members, friends, medical personnel, ministers and other

professionals are important, the person with VHL is the only one who must deal with all aspects in the treatment of the syndrome and of that individual. Quality of Life and Peace of Mind are slightly different for each person, and only YOU will know how to find Peace of Mind and Quality of Life for yourself.

Vitamins for Stamina

My brother Tony did the pre-surgery vitamin program which was presented at the Seattle conference and he is feeling marvelous after this surgery. I know it is early days yet but he says there is no comparison between how he feels this time and last year when he had partial removal of the other kidney - he reckons the vitamins etc. have a lot to do with how good he feels. Thanks for telling us about it.

-- *Maura H., Ireland*

Strength from Adversity

-- *Anonymous*

A man found a cocoon of a butterfly. One day a small opening appeared, he sat and watched the butterfly for several hours as it struggled to force its body through that little hole. Then it seemed to stop making any progress. It appeared as if it had gotten as far as it could and it could go no farther.

Then the man decided to help the butterfly, so he took a pair of scissors and snipped off the remaining bit of the cocoon. The butterfly emerged easily. But it had a swollen body and small, shriveled wings.

The man continued to watch the butterfly because he expected that, at any moment, the wings would enlarge and expand to be able to support the body, which would contract in time.

Neither happened! In fact, the butterfly spent the rest of its life crawling around with a swollen body and shriveled wings. It was never able to fly.

What the man in his kindness and haste did not understand was that the restricting cocoon and the struggle required for the butterfly to get through the tiny opening were God's way of forcing fluid from the body of the butterfly into its wings so that it would be ready for flight once it achieved its freedom from the cocoon.

Sometimes struggles are exactly what we need in our life. If God allowed us to go through our life without any obstacles it would cripple us. We would not be as strong as what we could have been. And we could never fly.

Osteoporosis

A side effect of steroid use

Altheada L. Johnson, MS, Registered Dietitian

You might wonder what an article on osteoporosis is doing in the *VHL Family Forum*. Isn't that the disease of old women? Yes, it affects 28 million Americans, 80% of whom are women. However, young people, men, and especially people with VHL need to be concerned too.

Osteoporosis is the loss of calcium from the bones causing them to become thin or porous, weak and easily broken. Calcium is laid down in our bones only during our growing periods when we are young. It is very important for pregnant or nursing women and youngsters to get the recommended amount of calcium in their diets each day. As we age, there is a normal, unavoidable loss of calcium from our bones. If, while we were young, we obtained the recommended amounts and have stayed active, this normal loss of calcium should have a minimal effect on our bones.¹ It is important that everyone maintain a good calcium intake throughout life. (See Table 1).

Along with the normal loss that occurs as a result of aging, a number of other things can contribute to the development of osteoporosis: inactivity, poor diet and prolonged steroid use. Exercise not only keeps the muscles strong, but also helps the bones maintain their strength. Weight bearing is the key.²

Prolonged use of steroid drugs has been shown to destroy bone.⁴ It is very likely that if you have VHL you will, at some time in your life, have a procedure which will involve the use of some steroids. Steroids may be used before, during or after brain or spinal surgery and some eye procedures to control swelling around the brain, spinal cord, and optic nerve. While these drugs are important tools in successfully recovering from these procedures, care must be taken to ensure against unwanted side effects later in life. Steroids are still the basis of anti-rejection treatment after kidney transplantation, and there is an enormous problem with osteoporosis worldwide among transplant recipients.

What constitutes "prolonged use"?

Five milligrams or more, per day, of a glucocorticoid steroid (Prednisone) for six months or more presents a big risk factor for steroid induced osteoporosis. Decadron, usually used during brain surgery, is 5 times stronger than prednisone (deltasone). Therefore, 1 mg of Decadron for one month or 5mg of prednisone for six months requires preventive therapy to avoid bone loss.

The symptoms of osteoporosis are often silent in the early stages but may include back pain, compression of the spinal column (loss of height) and easy fracture of bone, especially the wrist, shoulder or hip.

The U.S. National Institutes of Health Recommend:

Age Group	Milligrams Calcium per Day
Infants: Birth to 6 months 6 months to 1 year	400 600
Children: 1 to 5 years 6 to 10 years 11 to 24 years	800 800-1200 1,200-1,500
Men: 25 to 65 years Over 65 years	1,000 1,500
Women: 25 to 50 years Postmenopausal Taking HRT Not taking HRT Over 65 years Pregnant/nursing	1,000 1,000 1,500 1,500 1,200-1,500

Table 1. Source: N.I.H. Consensus Development Panel on Optimal Calcium Intake

Prevention and treatment would include a baseline bone density test, general risk factor reduction: no smoking or alcohol, increased calcium and vitamin D intakes and increased activity. In addition, if you are a postmenopausal woman then you will want to speak to your doctor about starting hormone replacement therapy (HRT).

Studies have proven that the use of 5-10 mg per day of a bisphosphonate called Fosamax, along with increased calcium intake, can prevent further bone loss.⁴ Actinol will soon be approved by the FDA. Other treatments include new designer female hormones and a nasal spray. Ipriflavone, a non hormonal nutrient, has also been shown to inhibit the bone destroying actions of osteoclast and stimulate the activity of bone building cells, the osteoblast.⁵ Bone loss cannot be regained to previous, non osteoporosis levels but some bone can be regained.⁴ Treatment means to stop further loss. Short term steroid use is generally not a concern.

Aseptic necrosis is, however, a rare result of short term steroid use where blood vessels feeding bone are destroyed and as a result, the bone dies. This usually occurs in the hip resulting in the need for hip replacement. Unfortunately, there is no way to know who is susceptible to aseptic necrosis and preventive measures are not useful here.

So, what does all this mean? It means that osteoporosis is preventable and treatable. It means that we all must get the recommended allowances of calcium in our diets each and every day. Doing this is

easier than you think. There are many supplements on the market that contain calcium with other minerals and vitamins. I think foods high in calcium are a better way to go. All milk products are an excellent source of bone-building calcium—stick with the low fat or fat-free versions (see Table 2).⁶ Canned salmon (with the bones), dark green leafy vegetables like collard greens, kale and broccoli are good non-dairy sources of calcium. We can all do with more vegetables of every kind in our diets, as they can provide not only calcium but other minerals, fiber, vitamins, and phytochemicals. Finally, our bones, teeth and waistlines would thank us for replacing the soda in our diets with skim milk or juice fortified with calcium.

One Calcium-rich Serving of each of these foods provides about 250-400 mg Calcium:

- ♦ 1 c. skim or low-fat milk, or low-fat or nonfat yogurt
- ♦ 1-1/2 ounces cheese
- ♦ 1 cup calcium fortified juice
- ♦ 3 ounces canned sardines with bones

Table 2. From a fact sheet supported by a grant from the National Osteoporosis Foundation, 1150 17th St., N.W., Washington, D.C. 20036-4603, 800-223-9994.

1. Jane E. Brody, *The NY Times*, "Guidelines Offer an Earlier Blueprint for the Battle Against Bone Loss," 10/20/98
 2. Susan Gilbert, *The NY Times*, "Weak Bones Among Men Are Linked to Estrogen," 12/08/98
 3. Jane E. Brody, *The NY Times*, "Calcium Takes Its Place as a Superstar of Nutrients," 10/13/98
 4. K.G. Saag et al (1998) *N Engl J Med* 339:292-9
 5. Steve Love, *Healthy Talk* (a publication of The Vitamin Shoppe) Jan/Feb. 1999 "Ipriflavone: A New Non Hormonal Nutrient for Osteoporosis"
 6. National Center for Nutrition and Dietetics Nutrition Fact Sheet- "Choose Calcium-Rich Foods for Strong Bones"
- Special thanks to Dr. Stanley Wallach, Endocrinologist at The Hospital for Joint Diseases, New York, and Dr. Hartmut P.H. Neumann, Nephrologist, Albert-Ludwigs University, Freiburg, Germany, for their assistance in writing this article.

Dear Ones - Just have to tell you - Tim is our son and his wife Heidi is one of the most beautiful persons I know. She is mother of Rachel (Rachel on the beach is one of the five winners for the calendar -- she is on the front). They are such a loving, caring family and so much support for Tim. I thank God for your research in this difficult illness. We wish we could send you a million dollars. -- Jennifer C., Florida

Thank you very much for all the information you sent for my sister. I have sent a blood sample for testing on myself. The truth of the matter is that the calendar you sent out this year is the reason I had the blood test done. I noticed some of the symptoms in myself, although my doctors say it is probably just a pinched nerve and reflux disease. I'll know in a few weeks. -- Susan L., New York

Ask the Experts

Pregnancy and VHL

I am pregnant, and I have some very small cerebellar hemangioblastomas. My doctor is wondering if we should schedule a Caesarean section, to minimize straining and prevent rupture of the brain tumors. Also, is it okay to have an epidural and a relatively natural delivery? What do you think? -- Hélène P., Québec, Canada

There is no reason that you cannot have an epidural, and there is no need to schedule a Caesarean Section if you have small, quiescent brain tumors. However, if the tumors are large, or have a cyst, and are exerting intracranial pressure, then it would be best if the labor and delivery were as short as possible, and a Caesarean would be indicated.

As you undoubtedly know, during pregnancy tumors may grow or a cyst may appear. We do not know whether this is in fact caused by the pregnancy, or would have occurred even if you were not pregnant, but certainly the disease does not stop during pregnancy. It will be up to your neurosurgeon to evaluate the situation with an MRI and clinical examination and make the best recommendation in your situation.

See also Grimbirt, Richard, et al., "Pregnancy in VHL disease." *Am. J. Obstet. Gynecol.* 1998

- Stéphane Richard, M.D., Président of the VHL Study Group, Hôpital de Bicêtre

'Birth' Announcement

Hello everyone!! Just wanted to bring everyone up to date on Cary. He had his surgery today, and he is doing well!! The doctor removed a tumor on Cary's one small "Kidney McNugget" that was the size of a "Ping-Pong ball" and fortunately he was able to remove it without damaging or removing the kidney.

Cary and I were joking this weekend that we wanted to send everyone an "announcement" to notify all that he was fine, the tumor weighed in at .2 ounces, was 3.2cm, was extracted at approximately 2:30pm, and we named it "Kidd" (short for Kidney). (Humor is the best medicine!!!)

The surgery took about 4 hours. Cary is doing well tonight, very tired. Feeling very little pain at this point (epidurals work wonders!!!!) I will be back at the hospital tomorrow and the doctor wants to get him up and walking around already. Thanks to Michelle, Laverne, and family for watching the boys for me so I can spend time at the hospital!! Thanks so much for everyone's support, thoughts, and for caring!!! Take care! -- Gail S., New Mexico

Paralyzed? Don't give up too soon!

by Gale L., Florida

I just want to set the record straight. Many of us VHL folks that have had spinal cord surgery go through a state of paralysis because of trauma to the spinal cord. Some may have permanent damage because of injury to the spinal cord during surgery and others may have a temporary period of paralysis. Only through dedication and hard work can these people expect to regain any mobility.

Only through dedication and perseverance was I able to walk out of the hospital about five weeks after cervical spinal cord surgery (utilizing a cane). I feel this is only because I had physical therapy five days a week, 6-8 hours a day, while I was in the rehabilitation unit in the hospital.

Although I was able to walk out of the hospital, I had total paralysis of my left arm with shoulder subluxation. This is because the trauma to the cord happened mostly on the left side. It took nearly three additional years of occupational and physical therapy to get where I am today. Even if the feeling doesn't come back, you can often learn to use your limbs in a different way and get back on your feet.

No one can guarantee that function will return, but if you don't try I can guarantee you that it won't. If the people that are paralyzed today do not work hard to improve their condition, the only thing I can tell them is to get a comfortable pillow for their wheelchair. If the doctor has not prescribed physical therapy -- right away -- then the family needs to make noise and get the therapy needed. If nobody will pay for a therapist, the family can learn to do the exercises and do it at home. Don't give up too soon, don't assume that damage is permanent until you have tried hard for months. It is not easy. It takes initiative and persistence and a desire to accomplish what they are trying to do. Even after all that hard work it still may not work, but you won't know unless you try, and then you will not be left to wonder what might have been if you had worked harder.

Do whatever it takes to get the strength and courage to do what you must. This is one thing that no one can do for you. You must do this for yourself. The surgery is the easy part, you were sleeping. The doctors and surgeons did their part you are alive, but it doesn't end there. This is just the beginning, the beginning of a long arduous process. Rehabilitation is

probably one of the hardest things you will ever do in your life, but it will probably be the most memorable. In 1987 when I was going through my rehabilitation, the Rocky movies were very popular. Before and/or after my grueling therapy, I would put on my headphones and listen to "Eye of a Tiger". These were the last words I heard every night before going to sleep. Sometimes the therapy was painful, but it was more frustrating than painful. The real pain begins when the wounds start to heal and you realize you can't do the things you used to. What really hurts even more is knowing that you didn't even try. I remember when getting out of bed was a chore. I also remember showing up to therapy with my shoelaces untied. I especially have fond memories of finally tying those shoelaces. Even to this day when I feel like I need some form of formal therapy, I ask my general practitioner for a prescription and off I go again.

Muscles atrophy very quickly if you don't use them on a continuous basis. Therapy should never stop-- formal or otherwise. Walking is therapy, cooking is therapy, even folding laundry is therapy. When you move any muscle in your body (especially the ones that are difficult to move) is all considered a form of rehabilitation. Remember the old adage--if you don't use it, you will lose it? Doing physical and occupational therapy may not lead you in a so called state of "normal," but there is only one direction to go, and that is doing better than you were doing. The sooner you start your therapy after surgery the easier it will be. This is also the time you gain the most function.

If it was your child that went through what you have gone through, would you let them give up?

Chat with Jay Platt

We are inaugurating a series of online "chat" sessions (a group typing fest) under the sponsorship of two health network sites. We will appreciate your feedback on the mechanism, and the kinds of topics you would like to see addressed.

The first two are chats with Jay Platt, who recently completed the Appalachian Trail, and who has led our fund-raising effort this year.

March 17, Wednesday, 7 pm Pacific US time (10 pm Eastern US time) at www.worldwithoutborders.com

March 25, Thursday, 8-9 pm Eastern US time. Free registration at Dr. Koop's site is required, to give us a level of privacy. Please see www.drkoop.com or www.vhl.org for more information.

Watch the VHL website for announcement of additional chat session. Please submit your ideas for future chats.

**See you in Atlanta!
See page 9 for details.**

Atlanta Meeting

June 4-6, 1999

"This educational conference brings together consumers and health care providers as well as educators and the latest research information. Such a consortium is a paradigm for sharing information," says Dr. Louis J. Elsas II, Professor of Pediatrics and Biochemistry at Emory University School of Medicine. "The Division of Medical Genetics, Emory University, is proud to co-sponsor with the VHL Family Alliance the Sixth International Patient/Provider Conference on von Hippel-Lindau." Dr. Elsas will co-chair the meeting with Eva Logan, chair of the Georgia Chapter of the VHLFA. Eva welcomes y'all to Atlanta and invites you to join us at the Sheraton Colony Square Hotel, 188 14th Street, N.E., Atlanta, Georgia 30361.

Families gain a once-in-a-lifetime experience of meeting others with this condition, and the leading physicians and researchers with expertise on this disease. Physicians gain insight into living with a rare disorder, and the diagnostic techniques and treatment options that can make a world of difference in a patient's quality of life.

Emory University will award up to **14.5 hours of continuing medical education (CME) credits** for doctors who attend this meeting. Final registration is not yet set, but will be approximately \$125 for most attendees, and \$180 for those obtaining CME credits. Credits for nurses, dietitians and genetic counselors can also be arranged. Please inquire.

We need you to make the experience complete! Start shopping now for inexpensive fares, or saving up those frequent flyer miles! Nearby businesses may also be willing to donate frequent flyer miles to help with your transportation. Sessions are planned from noon on Friday the 4th through noon on Sunday the 6th. The hotel is about 30 minutes from the airport, and shuttle service for \$10 per person one-way is easily available. Advance reservations are recommended for wheelchair service. Call 404-524-3400.

Reservations must be made directly with the hotel before May 5 to qualify for the VHLFA conference rate of \$99 for up to four adults in one room. Call Sheraton at +1-404-892-6000, or 1-800-422-7895 (U.S. outside Georgia), or fax to 404-733-6997.

This activity has been planned and implemented in accordance with the Essentials and Standards of the Accreditation Council for Continuing Medical Education through the joint sponsorship of the Emory University School of Medicine and the VHL Family Alliance.

The Emory University School of Medicine is accredited by the ACCME to provide continuing medical education for physicians. Emory University School of Medicine designates this continuing medical education activity for up to 14.5 hours in Category 1 credit toward the PRA of the AMA. Each physician should claim only those hours of credit that s/he actually spent in the educational activity.



Too Expensive

and other reasons for not going to the meeting

We all have our reasons for not going to a VHL meeting. Maybe it's really too much airfare this year, but the meetings move to different regions in hopes of being close to you at some point. If the meeting is within 500 miles of you, please take a hard look at your thoughts about not coming. I could tell you about hotel food charges and the high cost of renting projectors. I could tell you about all the speakers who come at their own expense to keep the registration low for attendees.

Instead, let me ask you to ask yourself whether your reasons are really valid, or whether it's really your own avoidance of coming. This is a chance-of-a-lifetime to have this meeting closer to you than it may ever be again. When the meeting was in Hawaii, one lady who lived in Hawaii said to me that she could commute from home, but the registration fee was too expensive. "Well," I said to her, "it will never be cheaper." With fifty states, how often will we hold the Annual Meeting in her state? No airfare (she could drive), no hotel (she could stay at home). There were people who paid thousands of dollars to be at the Hawaii meeting, and she was balking at \$150? At that point, you need to inspect your reasons and see what it is that is really keeping you away.

We all have things that stop us. Just ask yourself if you are letting yourself be stopped when you could push forward and have some real breakthroughs. You might want to call the hotline and talk about your fears and concerns with someone who has been there.

You are the only one who can make or break your own success. You can let VHL stop you, or you can break through that barrier and have a Happy Life in spite of it all. We all have limitations, and we all have more power than we exercise. It's not about "dwelling on it," it's about acknowledging this part of our real lives and learning to manage it well, keep it in its place.

There will be audio tapes for the information, but it's not half as valuable as being there, meeting people, asking your very own questions, hugging people, being hugged, having someone truly understand what you're going through in a way you have never experienced before. Do you really want to deprive yourself of that?

“

Paris was my second VHL conference, and I learned a lot again.. Not only through the speakers, but also by talking to several people, and just by being there. -- Chris H., Belgium

”

Introducing New Board Members

Kathy Braden, Indiana

Kathy Braden joined the Board this year as Director of Professional Education.



I joined the VHL Family Alliance in 1995 after my husband, Andy, and his sister were diagnosed with VHL. We live in Indianapolis, Indiana with our 13-year-old daughter and 10-year-old son. We are co-chairs of the Indiana Chapter of the Family Alliance.

We got involved because of our experiences with misdiagnosis and delayed treatment.

Andy's sister was diagnosed as having kidney cancer with a metastasis to the brain. She underwent full brain radiation to kill the cancer site in her brain, and was told she had six months to live. Their mother had died of kidney cancer, so Andy went for a checkup himself. Andy was found to have a kidney cyst, which they biopsied in an open operation and determined it was only a cyst. He told the doctors he wasn't feeling right, had a lot of headaches, and they told him it was stress. They sent him to a psychiatrist, and told him it was "all in his head." They proved to be right, but not quite as they meant it -- three months later it was clear that it was a brain tumor. That was July of 1995.

Andy was also told that the brain tumor was a metastasis from renal cell carcinoma, even though they couldn't find a primary kidney tumor, so they sent him to radiation oncology for a consultation. Thankfully, the neurosurgeon did not believe the pathology report and sent it for a second opinion prior to ordering full-brain radiation. It was not a met, it was a hemangioblastoma. The correct diagnosis was VHL.

We attended the Hawaii meeting. It was a long way to go, but we thought if the meeting is terrible, what a place to go lay on the beach! We went and only went to the beach one time. We were overwhelmed and overjoyed. The contacts were incredible -- NIH, the family members, the doctors -- people who knew what you were talking about as opposed to "V.H.What?"

Four years later, Andy's sister is doing fine. After Andy's diagnosis they looked again at her case and realized it was VHL, not metastatic cancer.

Our daughter did a science project on vhl, won an award. It was hard to start with, sharing with her

friends. Some kids really paid attention, and realized she too is at risk, but it has not been an issue, and was a positive for her. We have chosen not to do DNA testing yet. I don't want to give them an excuse to do stupid things. They go for screenings and it's no big deal yet.

I have a bachelors degree from Ball State University and work part-time at St. Francis Hospital and Health Centers as a Functional Outcomes Coordinator, looking at the patient's perspective of how they think they feel and how they are doing. We surveyed cardiac patients, and are now following breast cancer patients, doing a survey prior to intervention and then quarterly for a year to determine what side effects, problems, etc. may have come up, and if there is something the hospital can do to help.

I volunteer at my son's school on a regular basis and spends the majority of her time driving car pools and assisting with 4-H projects (sheep and goats). We live on the edge of Indianapolis. My parents have two acres. The sheep winter at my sister's farm and summer at my parents', so the kids get to help care for the animals, walk and wash and shear them and get them ready to show at the Fair. Chris also competes in Legos, and Andrea has entered in cake decorating and gift wrapping.

Susan McGuire, North Carolina

Susan joined us as Director of Public Relations.



I am an office administrator for a general dentist and have worked 17 years for the practice. My husband Brian and I have been married for 26 years and we have two grown children, Patrick and Justin. I enjoy travel, reading, sewing and music. I play in our church's adult handbell choir and direct a youth handbell choir.

I became involved with VHL Family Alliance after watching the dedication of my sister-in-law Peggy Marshall and brother Don to the organization. I felt there was a vast amount of beneficial information and work being done by the folks in the VHLFA. I had a sketchy knowledge of VHL as our family has known of the disease since Peggy's diagnosis in the 1960's. The more I learned about VHL the more questions I had about its diagnosis, prognosis and treatment. VHL has impacted several people very special to me, and as I met other people with VHL and their family members I knew I would like to help out in anyway VHLFA could use me.

I graduated in 1991 from NC State University with a Bachelors Degree in Communications and Public Relations. I always thought I could use my education to volunteer with a non-profit organization, as I was not prepared to make a career change after working several years in the dental field.

When Brian and I visited Don and Peggy we were amazed at the amount of materials and resources that were available to people concerned with VHL. We talked about different public relations programs and what might be done in the future. If any of you know Peggy, she never seizes to grab an opportunity to recruit new volunteers and I was asked if I would consider helping on the Public Relations committee. I have not regretted for one moment saying yes.

Last year was my first attendance at a National VHL meeting and I met many new friends and learned new information about VHL and its manifestations. As a lay person, I had a crash course in medical terminology. In the past year I attended the National Organization for Rare Diseases (NORD) conference and Alliance for Genetic Support Groups (AGSG) conference. I have compared and shared information with other non-profit groups and have in returned learned ways to help us get our message out to the public. I currently chair the Public Relations committee and I hope to help VHLFA make great strides in reaching the print and broadcast media. Jay Platt's hike was a national media event and with the help of Maria from PA, her friend Lou, and many others Jay was able to get local and national press. Joyce Graff was our eyes and ears as we followed Jay on the Appalachian Trail from our computers. It has been both challenging and exciting to work this year in on projects for VHLFA.

I feel very privileged to be a part of the VHLFA and its wonderful group of caring and giving people. We are all working toward a very informative and successful National VHL meeting June 4-6 in Atlanta. I hope to meet many of you there.

New Zealand

-- Jon Johnson, Auckland, New Zealand

We are very sorry that Val and I couldn't make the conference in Paris this year, however we are pleased to report that our group in New Zealand is slowly growing as more and more individuals become aware of our activities. It has certainly surprised us how many patients and supporters have come forward for information and help since we returned from the conference in Hawaii two years ago, and it is particularly pleasing to be able to offer them assistance based on Family Alliance material, and be able to tell them that they certainly are not alone and many individuals

in the Family Alliance and medical personal throughout the world are making inroads into research and patient treatment and care.

Progress here has been made with us gaining the support and help from the Northern Regional Genetic Service, which covers a large section of New Zealand and with the help of Ingrid Winship and her staff we are in the process of gaining support from various medical specialists who have an interest in VHL.

We have also managed to receive some financial assistance from our Lottery Commission to offset initial setting up costs and hopefully arranging our first meeting (mini conference) involving medical staff and families. It is hoped to arrange this in Auckland in 1999.

We would like to wish all those attending this years conference the best, and let you know that even down here on the other side of the world we are thinking of you all and would like to express our gratitude and appreciation for the many hours many of you put into trying to beat and treat VHL.

We are looking forward to following the progress made since Hawaii.

Regards, Jon & Valerie Johnson

News from Scandinavia

-- Richard Harbud, Denmark

We have some exciting news regarding the Scandinavian Knowledge Base Working Group. Vibeke will be attending part of the 7th meeting of the Scandinavian neuro-oncology group, in Lillehammer, Norway, on the 18-21 March. One full day of the meeting is to devoted to VHL. VHLFA Denmark is sponsoring the participation of Dr Thomas Rosenberg, an eye specialist from Denmark. Dr. Rosenberg, a major asset to the Danish VHL association, was unable to attend the Paris meeting because of a prior engagement. He is very keen to make international contacts within the field of VHL.

Also attending the meeting is Dr. Mika Niemelä, neurosurgeon from Finland, who as you will recall was at the Paris meeting. Most importantly, doctors from all Scandinavian countries will be there. A major breakthrough in international cooperation is at hand.

We have also just had two articles published in the Danish Weekly Medical Journal. Both articles are written by doctors who help us with their support and knowledge in the association. The first article is written by Dr. Hans Ulrik Møller, whom you met in Paris. He wrote a detailed report of the VHL Paris meeting. The second article was written by Dr. Marie Louise Bisgaard, a clinical geneticist, who is now doing DNA testing in Denmark.

Clinical Studies -- Can You Help?

Hearing issues? Vertigo? Tinnitus?

NIH looking for participants for ELST Study

Dr. Edward H. Oldfield of the National Institute for Neurological Disorders and Stroke, Bethesda, Maryland, USA, is heading a study of endolymphatic sac tumors (ELST).

In addition to being associated with tumors of the kidneys, brain, spinal cord, adrenal glands and retinas, VHL has recently been associated with tumors of the inner ear. More specifically, the endolymphatic sac (ELS) has been found to be the site of origin for specific VHL-associated inner ear tumors. The ELS is thought to be a site of regulation of inner ear fluids. Tumors of this sac (ELST) have been associated with hearing loss (either sudden or gradual) as well as vertigo (balance disorder) and tinnitus (noise in the ear). In the limited cases that have been described in the literature, the recommendation has been for immediate surgical removal of the tumor. However, the rarity of this tumor has resulted in most tumors only being recognized after they have grown quite large and already caused deafness and balance dysfunction.

At NIH, one of the largest groups of VHL patients has been studied and followed on a regular basis. This has provided the unique opportunity to closely examine patients with this disease and detect these inner ear tumors very early in their development. The advantage is that ELST's can be removed before they can cause permanent hearing loss and vertigo. Since there have been no reports on early intervention for early stage ELST's, the basis for recommending this surgery for early tumors is preliminary data from the NIH experience. Of the 13 patients with late stage ELST's diagnosed at the NIH, all 13 have shown severe deafness and balance dysfunction. A review of the case reports in the literature seem to support this observation. Therefore it seems that untreated ELST's will eventually cause hearing loss and/or balance dysfunction. The exact timeframe for development of these problems is not known.

Candidates for this study will have confirmed or suspected ELST based on audiologic or vestibular testing. Candidates may choose surgery, or non-surgical follow-up at 6-12 month intervals for five years, with neurologic, audologic testing, and MRI scanning of the inner ear region. If necessary, you and your doctors could elect to change your management to one involving surgery. The goals of the study are to find better ways of diagnosing ELSTs early, and

finding optimal treatments that preserve hearing and balance.

Eligible patients who have already been seen at NIH will be invited to participate. If you believe that you may also be a candidate for this study, you are welcome to contact Suzanne Lischynsky, R.N., of the office of Dr. Edward H. Oldfield, 301-496-5728, Fax: 301-402-0380

People will be excluded from the study if they are: pregnant or lactating; have multiple abnormalities of the middle and inner ear that would complicate analysis of the cause of hearing loss; patients undergoing chemotherapy with ototoxic agents. Additional inclusion and exclusion criteria are available from Ms. Lischynsky.

Smoking and Tumor Growth

The U.S. National Institutes of Health (NIH) is performing research to understand the genetic abnormalities that lead to the development of tumors in people with VHL. This work has led to the identification of the VHL gene, as well as an understanding of which types of mutations lead to the growth of different tumors in people with VHL. Ongoing natural history studies are describing the growth of different tumors over time, the correlation with VHL gene mutation type, and the effect of smoking on tumor growth.

The NIH has continued interest in evaluating the natural history of tumors found in people with VHL, genetic abnormalities associated with these tumors, and the role of environmental exposure to the growth of these tumors. Medical care and travel expenses are provided by the U.S. Government.

Interested people with VHL should call +1 (301) 496-6353.

Angiogenesis Inhibitor

The clinical trial of an angiogenesis inhibitor starting in Boston this year is still looking for subjects. Details are in the December 1998 issue of VHLFF, or at www.vhl.org. For more information, please contact Sarah Mott at the office of Dr. William Kaelin, 617-632-4747, Fax 617-632-4381, or e-mail: sarah_mott@dfci.harvard.edu

*For the latest in research results and clinical trials information,
come to the Atlanta meeting!
See page 9 for details.*

***This issue is dedicated to Jay Platt, honoring his 4,000,000 steps for VHL!
We are grateful for the many gifts from...***

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 Neville Broadbudd, by Virginia Hufford
 Deborah Bruff by John & Edna Benz, Tamara
 Bentz
 Paul Buchanan, by Harry & June Wilcox
 Don Chew, from USAG-Scott AFB, Illinois;
 Rapid Delivery Service, Marris Cole, J.D. &
 Janice Duff, Joseph & Theresa Fischer,
 Mary Lax, Brenda McLeod, Albert Meena,
 Eileen & Scott Merrill, Davenport Mosby,
 Michael Ozbourn, Margery Render, Leroy
 Stram
 Bernard Christensen by Theresa Blazich
 Brad Cox by Tania 'Durand
 her Dad, by Cathy Clifford
 Marvin Dankenbring, by Vicky & Stanley
 Palmer
 Carol Darmofal, by David Darmofal

Mrs. Ora York Dixon, by Harry & June
 Wilcox
 Mrs Nancy Dyches, by Virginia Hufford
 Carolyn Flanagan, by Wilma Elkins, Carl
 Hedberg
 Naomi Flicke, by Evabeth Thomas
 Thea Franzini by Donna Bauwens
 Frank Graff, by Walter & Sandra LaFeber
 Dr. John Graham by Robert John Bayer
 Lois Grisson, by Deirdre O'Connell
 Charles M. Hall Sr, by Tondra & Michael
 Bolinger, Louise Browning, Tondra Brunk,
 Epsilon Lambda Chapter, Mark & Aileen
 Friedlein, Diana Halberko, Martha Miley
 Madge Hall by Louis & Wilma Briscoe,
 Kathleen Neary
 Lawrence Harper, by Linda Smiley
 Orville Kendrick, by Betty & Emily Heide
 Mrs. Edith R. Larkman, by Harry & June
 Wilcox
 Caren Lewis by Patricia & Vernon Fruehauf
 Pearl Mathewson by Brenda Nelson
 Meg Moody by Mary McCabe
 Wilma Mullins by Joyce Graff
 Frieda Nierman, by Vicky & Stanley Palmer
 Raymond Pulley, Jr & Sr, by Marguerite
 Golding
 Matthew Rider by Rita Miller
 Skeezix, by Donna Bauwens

Are you coming to Atlanta?

Call the hotline for a complete brochure, or get one from our website, www.vhl.org

Early birds who register before May 5 will be eligible for a discount.

Hotel reservations must be made by May 5 to qualify for the conference rate.

See you in Atlanta!

DONATION & MEMBERSHIP INFORMATION

Please check one: I am a ☐ Person with VHL ☐ VHL Family Member ☐ Supporting Friend
☐ Health Care Professional (physicians nurse, social worker, etc.)

Occupation, or medical specialty: _____

All members receive the VHL Family Forum. Check here ☐ if audio version is needed

Name: _____

Address: _____

City: _____ State: _____ Zip/Postcode: _____

Country (if not USA): _____ Fax: _____

Phone (home): _____ Phone (work): _____

Membership:

☐ New member ☐ Renewal (Dues \$25 per household, \$35 per professional) \$ _____

☐ I cannot afford dues at this time, but please accept my donation (any amount)

Yes! I want to join Jay's team!

☐ Please send me _____ brochures about Jay's hike for me to share with others.

Donation:

Tax deductible donation of . . . (amount) \$ _____

☐ In Honor of _____

☐ In Memory of _____

☐ Please send a card to: _____

Purchase merchandise: (proceeds help the VHLFA!)

☐ # _____ Copies of the 1999 VHL Calendar @ \$10 each (call for quantity prices) \$ _____

only while supplies last -- order larger quantities now for 2000!

☐ # _____ Copies of Pierre Jacomet's CD of Bach's Goldberg Variations @ \$12 each \$ _____

☐ # _____ Copies of *How Are You?* by Patricia Foote @ \$17 each \$ _____

☐ # _____ Copies of *Sacred Process* by Karen Koenig @ \$15 each \$ _____

☐ # _____ Copies of the VHL Video @ \$20 each, (specify _____ US or _____ PAL format) \$ _____

Payment Method:

☐ Enclosed check, payable to the VHL Family Alliance **TOTAL:** \$ _____

☐ Master Card/Visa Card # _____

Expiration date: _____ Name as it appears on the card: _____

Signature _____

Send this form to VHL Family Alliance, 171 Clinton Road, Brookline, MA 02445 USA

4,000,000 steps for VHL!

Jay has now gone the distance -- and we're catching up! \$92,580 as of 5 March

"Everyone who showed up to celebrate with Jay went an extra mile or two to do so -- it was not a pretty sunshine day; it was not an easy drive; it was not a pleasant walk in the woods. Everyone made a special effort and stretched outside their comfort zone and persevered -- and we did it because of Jay. We hope that everyone will keep Jay's model and inspiration in their hearts when they need to remind themselves to hang in there, keep going, and just do it." -- *Joyce Graff, Dahlonega, Georgia, January 23, 1999*

As a vertical line of tornadoes marched eastward across the map of Georgia, a stalwart group of family and friends gathered in Dahlonega, Georgia, to walk the last mile of the Appalachian Trail with Jay and celebrate with him.

Even if you figure a 34-inch pace, which would be longer than most paces on the Trail, Jay walked 4 million paces from Maine to Georgia. It's that kind of extra-ordinary effort it will take to cure VHL. We're on the trail of a cure. With teamwork and persistence, we will do it. *Thanks to all who are helping!*

"I am sure Georgia has been on your mind since you began this journey last August, and as one who has had the privilege of following your progress, it is my distinct pleasure to say "Welcome Home, Patch!"

Courage against great odds is the essence of our American Spirit, and your perseverance in completing this goal stretches the boundaries of our potential as a people. As the trail ends, and you continue your most difficult challenge, I am confident you will remain a role model for individuals living with von Hippel-Lindau syndrome.

Your remarkable Maine to Georgia expedition is now part of the rich history and lore of the Appalachian Trail, and for those striving to overcome von Hippel-Lindau Syndrome, you are a true folk hero. Again, congratulations on your thru-hike and best wishes to you and Paz for a long and happy marriage." -- *Arlen Spector, U.S. Senate, Pennsylvania*



Meet Jay! In Atlanta, or online. See page 9.

VHL Family Forum

Newsletter of the VHL Family Alliance
171 Clinton Road
Brookline, MA 02445-5815

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