VHL FAMILY FORUM

Family, Friends, Physicians, & Researchers dedicated to improving diagnosis, treatment, and quality of life for people affected by von Hippel-Lindau.

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Improving Diagnosis of VHL

With the September 2004 issue of the *VHL Family Forum* we included a survey, asking people to share some information about their VHL histories that might help researchers. We had a wonderful response! Altogether 172 people reported information on 290 cases.

36 (20%) of the surveys were received from outside the United States — from Canada, England, Scotland, Ireland, Europe, Australia, New Zealand, and South Africa — a total of 15 countries were represented in this survey.

73 people (25%) were diagnosed through DNA testing. This is a sharp increase from surveys we did five years ago, which is to be expected, since the technology has improved so much since then. Fifty-six people (19% of the total) had a diagnosis of VHL before they had their first symptom. This is a wonderful advancement, allowing those people to begin screening early, and catch issues at very early stages.

We asked people at what age they were diagnosed. Ages ranged from zero to 84, with a median age at diagnosis of 25 years. Age at first symptom ranged from one to 71, with a median age of 20 years. So the ranges were very broad, with the highest probability in the young 20's. Fifty-five (19%) of these people had their first symptom before the age of 15.

Of the 243 people who provided both an age at diagnosis and an age at first symptom, 56 (23%) were diagnosed before the first symptom, through DNA testing. Of these, 24 (40%) were under the age of 12. Eleven still have no symptoms, and a total of 124 were diagnosed within the first 1-2 years. The physicians working with these patients achieved a diagnosis within a reasonable timeframe. In other words, the system worked well for 72% of the group. Others were not so fortunate.

Thirty (12%) of these people waited 2-5 years for a diagnosis.

Twenty (8%) of these people waited 6-12 years for a diagnosis.

Seventeen (7%) of these people waited more than 14-43 years for a diagnosis.

During that time, minor issues became larger issues. In some cases a small problem treated as an isolated incident could have served as an early warning signal if it had been properly interpreted. One man, diagnosed with metastatic kidney cancer at age 66, realized in retrospect that he had been coping with the effects of an undiagnosed pheochromocytoma for 43 years. That means he spent 4 decades with an undiagnosed set of symptoms that were significantly impacting his cardiovascular system and his emotions. At any point in those four decades, if the diagnosis had been made, he might have been spared the very heavy toll of metastatic kidney cancer.

Ten years ago we did an on-site survey at the Boston meeting, where we asked some of the same questions. There were only 37 people responding to that survey, so it is not altogether fair to compare the two. But if it were, this comparison shows that there is at least a 5% improvement in the rate of diagnosis. Our sense of the situation is that it is even better than that. Both studies include history, and of course history has not changed. But the outlook for our young people is significantly brighter.

DNA diagnosis is available as a tool to determine which members of VHL families are in fact at risk. In addition, DNA diagnosis is now available for use in differential diagnosis. When a doctor is trying to decide whether you have disease A or B, the process of differentiation is often unclear. It is very helpful to have a test, like the DNA test for VHL, that can help make that decision clear.

cont'd on page 2

Inside this issue!

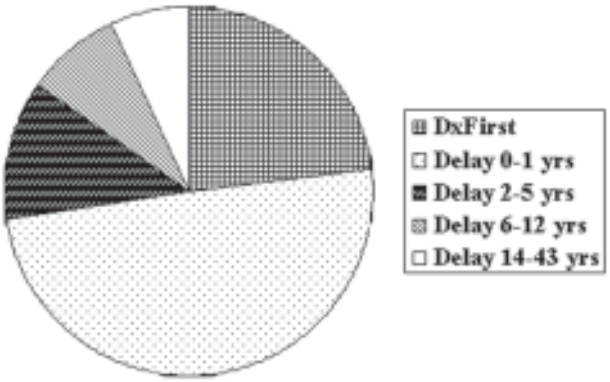


Figure 1: Timely Diagnosis: *Of the 233 respondents providing sufficient data for analysis, 23% were diagnosed before the first symptom, and another 49% were diagnosed within one year of the first symptom. The remaining 28% waited up to 43 years for a diagnosis of VHL.*

The key is to ensure that doctors will consider VHL as a possible diagnosis. That is something we can all help with — raising the visibility of VHL, helping doctors and the general public think of it more readily.

We asked people what that first symptom was. Among the people who had a delayed diagnosis, their first symptom — the one NOT diagnosed as VHL — was reported as shown in Figure 2.

We have felt for quite a long time that the physicians most likely to make a timely diagnosis of VHL are ophthalmologists and neurosurgeons. Brain and eye tumors are most prevalent, with about 60% of people with VHL likely to have one or both of these tumors. We see that clearly shown in this survey, but we also see that there is still room for improvement. Only about 60% of eye, brain, or adrenal tumors were diagnosed within the first year.

While eye and brain tumors were likely treated as single tumors, pheos were often totally misdiagnosed as stress, mental illness, or heart problems. Cardiac issues are easier to diagnose these days, but the distinction between emotional and adrenal issues remains very difficult without testing. In your own family, if you suspect that a pheo diagnosis is being missed, do not hesitate to press for additional testing and get another opinion.

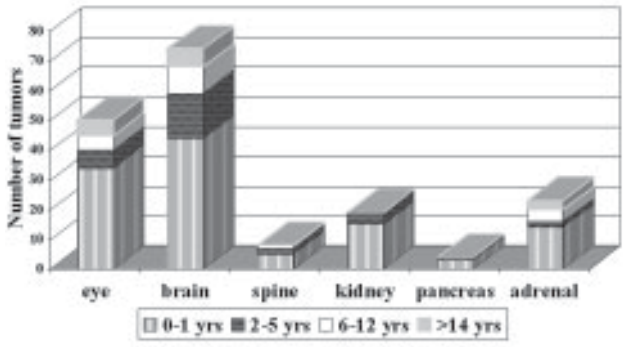


Figure 2: First Symptoms: Timeliness of Diagnosis by Tumor Type. While the majority of tumors in each category were properly identified as being VHL, there is still room for improvement, especially among ophthalmologists, neurosurgeons, and in identification and localization of pheochromocytomas and paragangliomas.

We have learned a great deal about the natural history of these tumors over the past dozen years. At this point, the VHL Family Alliance and its advisors are working to turn this information around and look at it more broadly — in the general population, what is the likelihood that a particular tumor might be VHL? How worthwhile is it for the physician to do the additional testing it takes to determine whether this person has VHL?

Here are a few of the lessons we have learned:

1. Bilateral epididymal cysts are considered enough to diagnose VHL.

2. Endolymphatic sac tumors rarely occur outside VHL.[1]

3. People with hemangioblastomas in the retina or central nervous system are highly likely to have VHL, especially if the tumor occurs for the first time in someone under the age of 50, or if there are multiple tumors.[2]

4. Where there are multiple tumors, on both kidneys or adrenals, there is almost certainly a genetic cause, especially in younger people (under 50).[3]

5. 24% of adrenal tumors in one large study had a genetic origin; 30% of those were found to be VHL.[4]

6. VHL is the leading hereditary cause of clear cell kidney cancer.[3]

7. 20% of people with VHL are the first in their family to have VHL.[3]

Please help us raise the visibility of VHL, to make it ever more probable that the physician will consider a diagnosis of VHL, and do the tests needed to rule it in or out. An early diagnosis will help to avoid the worst consequences of some of the later-onset issues like metastatic kidney cancer.

Many thanks to all who participated in this survey, and to Robin Cochrane for the data entry and assistance with analysis. Thanks too to all the many physicians whose research on VHL has moved our knowledge forward in the last decade.

1. Russell R. Lonser et al, "Tumors of the endolymphatic sac in von Hippel-Lindau disease." *N Engl J Med* 2004, 350:2481-2486

2. Stéphane Richard et al, "Central nervous system hemangioblastomas, endolymphatic sac tumors, and von Hippel-Lindau disease." *Neurosurg Rev* 2000, 23:1-22; discussion 23.

3. Russell R. Lonser, Gladys M. Glenn, et al., "von Hippel-Lindau disease." *Lancet* 2003, 361: 2059-2067.

4. Hartmut P.H. Neumann et al, "Germ-line mutations in nonsyndromic pheochromocytoma" *N Engl J Med* 2002, 346: 1459-1466.

New Handbook

By Joyce Graff, Editor

Along with this newsletter we are sending a new edition of the *VHL Handbook*. The online version has been updated to include this new text, and a downloadable file has been put on the internet. Additional paper copies may be purchased (see page 15).

This booklet is designed for use by patients and their local doctors, helping you build your own healthcare team. It provides a practical summary of the scientific literature and clinical studies on VHL. In the back of the book is a bibliography of leading articles about VHL, to give the physicians a quick guide to the most important articles. There is also a glossary of the medical terms to help patients learn the language that their medical teams will use.

The 2005 Edition of the *VHL Handbook, What You Need to Know about VHL*, has been significantly updated, incorporating what has been learned about VHL since the previous update in 1999. It also includes what this community has learned — the experiences you have shared with us have significantly contributed to our collective learning! Thank you!

The information about endolymphatic sac tumors has been completely changed.

The section on Reproductive Health has been significantly updated, especially the part for women. There is still no practical way to distinguish for sure before surgery between a benign cystadenoma and a cancerous tumor, except through close observation of its behavior. We are working with the ovarian cancer research community to find a better test. Even in the general population, 90% of the tumors in this region removed from pre-menopausal women are benign. That is good news — but only after surgery! Everyone needs a better way to tell for sure whether surgery is really needed.

There is a new section on Pregnancy and VHL. We still do not have data to show whether tumor growth is spurred by pregnancy, but some of the bodily changes that occur during pregnancy can certainly make things worse, at least for the duration of the pregnancy. It is very important to have a thorough checkup before and after, so that you and your medical team have the information you need to protect you. If you are already pregnant, read this section and get a check-up right now.

Recommendations for in-depth discussion of stereotactic radiation have not changed, but have moved into the body of the text, since some people were not finding them in the back of the booklet. Anyone considering stereotactic radiation should definitely read this section and have the suggested conversation with the medical team. If you don't, you are at significant risk that the treatment may go wrong. Once they give you the radiation, they can't take it back, so you really MUST have this conversation BEFORE treatment.

Recommendations for treating adrenal tumors stress the preference for laparascopic partial adrenalectomy wherever this approach is available and appropriate. Even if this is the first adrenal gland affected, there is significant risk of an adrenal tumor on the other side in future, so care should be taken to conserve as much functioning cortex as possible.

We have a better understanding today of paragangliomas (pheos that occur in other parts of the body) and better ways to test for them. This is still a difficult area, and a new international consortium is being formed, scheduled to meet for the first time in fall 2005, to make better progress in standardizing the testing and interpretation of the results, and in finding the location of these tumors.

Experience with kidney tumors over the last several years has confirmed the reliance on the guideline of leaving kidney tumors until the largest tumor approaches a size of 3 cm. Radio Frequency Ablation has proven effective in treating tumors that qualify for this less invasive approach.

Suggestions for screening children have changed considerably as we have identified more young children with VHL through DNA testing, and are having more experiences with early affects. Issues do crop up in children under the age of 12, and some level of screening is advised beginning at ages 1-2, to catch and treat or monitor early issues as appropriate.

The section on Nutrition has been enriched with expertise from dietitian Altheada Johnson, Gale Lugo, and the Harvard School of Public Health. Living Well With VHL has been enriched by Dr. Gary L. Wood of Tampa, Florida, a clinical psychologist who has VHL.

We are very excited about the new Handbook, and hope that you will find it useful. Your feedback is always important in shaping the next handbook.

Special thanks to our Medical Advisory Board members, listed on page 14 of this issue, and to the many additional physicians and researchers who provided key information to enrich this booklet. Thanks too to the many VHLFA members who contributed their stories and reviewed sections to add their important perspective.

Seven volunteer translation teams are working to create local versions of the Handbook in French, Spanish, Dutch, Italian, Japanese, Chinese, and Ukrainian. Announcements will be made on the language-specific websites as soon as these versions are ready. If you are interested in helping to create another language edition, please contact editor@vhl.org Thank you!

Allergies to Contrast Media

Peter L. Choyke, M.D., U.S. National Cancer Institute

Dr. Choyke

VHL patients undergo many imaging studies requiring injected and oral contrast media, and some patients develop allergies to these contrast agents. There are many myths and misunderstandings surrounding contrast allergies leading to wide variations in policies among different imaging centers and hospitals. The goal of this short article is to dispel some of these myths.

Allergies can occur with any contrast agent, whether injected into a vein or ingested orally, whether for CT scanning or MRI. Allergies are more common with the CT contrast agents ("iodinated" contrast) than with MR contrast agents ("gadolinium" contrast) but they occur in both. There are different manufacturers of both iodinated and gadolinium contrast. Some people are allergic to one particular brand of contrast agent and not to another in the same class. Some people are born with allergies to contrast agents and some develop them over time, even after many years of reaction-free exposure to contrast media.

Question: You say that allergies to shellfish are not related, yet at every scan I am asked about allergies to shellfish. Why is this?

Answer: This is an urban legend that will not go away.

Not all allergies are the same. There are five basic types of allergies and some very rare additional types that will not be discussed. Nor will I discuss medical treatment of these reactions. A reference is provided for this purpose.

The five basic types of contrast reactions are:

- Skin hives: reddened splotchy areas on the skin (face, trunk, arms) that develop soon after injection. They can be very itchy and uncomfortable and take several days to go away.

- Throat tightening or laryngeal edema: This is an internal hive that affects the breathing tube causing difficulty breathing. This can range from mild throat "tightening" to a severe "choking" sensation. It is often accompanied by swelling of the face.

- Asthma: This reaction involves the airways in the lungs and results in wheezing. Asthmatics are more prone to this reaction.

- Shock: This is a systemic reaction to contrast media where the blood pressure decreases suddenly and other serious symptoms occur. This is a medical emergency

- Delayed: Some patients experience headaches, muscle pains and flu-like symptoms up to 48 hours after contrast media. This is a self limited reaction.

Skin hives and minimal throat tightening are the most common reactions and are considered mild reactions. They can usually be prevented with oral steroids and an antihistamine (benadryl) taken beforehand. Even if there is a "breakthrough" reaction it will be mild and therefore the risks associated with contrast exposure are very low. To put this in perspective approximately 2% of patients getting iodinated contrast media and fewer than 1% of patients getting gadolinium contrast media will develop this type of reaction.

The other reactions are considered more serious but fortunately are much less common (by a factor of 10). Reactions leading to death occur in about 1:250,000 patients. However, if you have had a moderate or severe reaction you obviously do not want to experience it again. So, most patients opt for another kind of study. For instance, if you are allergic to CT contrast then you would switch to MRI for your studies and vice versa.

Most adverse events occurring after contrast media are NOT allergies. Feeling warm, flushed, nauseous and even vomiting are side effects of the drug, not allergic reactions. Some people are very sensitive to contrast, but not allergic. It usually helps to slow the rate of administration of the contrast media; this will usually reduce symptoms. Also, contrast media, especially iodinated contrast media can damage the kidneys, a process known as "contrast nephrotoxicity". Predisposing risk factors for contrast nephrotoxicity are previous renal diseases, diabetes and hypertension. If you have had much of your kidneys removed surgically, your doctors will often change from CT to MRI to reduce the risk of contrast nephrotoxicity. Finally, allergies to "iodine" or shellfish are not related to allergy to contrast agents.

If you have developed an allergy to contrast media then there are some simple things you can do:

- Always find out and write down what type of contrast and what brand name you were allergic to. It may be possible to avoid that brand in the future. I know patients who are allergic to one brand of gadolinium or iodinated contrast media but not another brand.

- If the reaction was mild you can take a simple "pretreatment" or "prophylaxis" consisting of several pills of steroids and benadryl before the exam to prevent reactions.

- If the reaction was moderate or severe you can change to another kind of scan using another class of contrast agents.

- If the reaction was severe but your doctors still feel you need the injection of that kind of contrast then you should be pretreated but also have anesthesiologists or other emergency health personnel "standing by" to treat the reaction if it occurs. However, in reality, this is hardly ever done. In my career of 20 years I have seen it done only once. There are almost always safer alternatives.

It is still unknown why reactions occur in some people and not in others. Some doctors have devoted their whole careers to trying to understand the answer to this question. Indeed, there are some reactions that I do not discuss here that are very difficult to explain. Fortunately, modern contrast agents have very low reaction rates, and this article will not be relevant to most patients with VHL. If you do have a contrast reaction I hope these comments have shed some light on the subject.

Question: We have heard you say that oral contrast should be used for abdominal CT's along with the intravenous dye. Yet my hospital declines to administer the oral contrast, saying they feel it is not necessary. What do you recommend and why?

Answer: Oral contrast is important for identifying the bowel and differentiating it from adjacent structures. If the bowel is not opacified by oral contrast it can be misinterpreted as a mass. One added benefit of oral contrast is that it hydrates the patient and this lessens the chance of contrast nephrotoxicity and rids the body of the contrast media faster. Some centers use water as an oral contrast agent instead of conventional oral contrast. This is an acceptable alternative to oral contrast media since it hydrates the patient and also provides a "negative" contrast. I do not feel strongly about what kind of oral contrast you take, just as long as you take something.

References:

1. Thomsen HS. "Guidelines for contrast media from the European society of urogenital radiology." *AJR* 2003; 181:1463-71

2. www.chestx-ray.com/Practice/Contrast.html

Help Melissa do her Personal Best for YOU!
Help her meet the Janus Challenge
and win more money for VHL research!
See page 16 for details.

John, Missy, Adam, and Taylor

His Valiant Battle

by Ellen R., Indiana

Please accept this donation in memory of my brother, John Rainier, who recently lost a valiant battle with VHL. John passed away on May 24, 2003, after a severe infection. He was 53 years old. John fought his battle with VHL for about 24 years. Through the many surgeries and eventual dialysis, he persevered – never giving up hope or a desire to be as strong as he could be and give as much as he could to others.

John was one of "America's brightest and best." He graduated near the top of his class in 1972 from the United States Military Academy at West Point. Later he served as a faculty member at West Point. He rose to the rank of Lt. Colonel.

I learned of my brother's death on Memorial Day and I thought ... how appropriate. Even though John did not serve in an active-duty war setting, he certainly fought VHL with a great deal of courage.

Thank you for all of your efforts to fight this disease. John benefited greatly from consultations with VHL experts whom he found through your organization.

Hope and Life-Saving Action

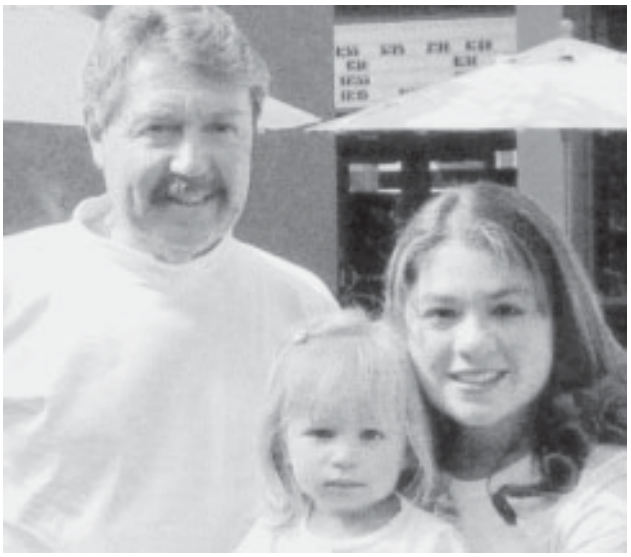
By Melissa K., California

Hello, my name is Melissa. I want to briefly write to you about my family who inspired me to do a fundraiser for VHLFA. My father, Ron P., age 62, was diagnosed with VHL this past February. He had his kidney removed last year due to renal cell carcinoma. His remaining kidney still has cancerous tumors on it, but luckily it is still functioning. My Dad had no symptoms at all until a simple blood panel revealed he had a high level of creatinine in his blood, which prompted his doctors to look at his kidneys. Luckily he has no other signs of VHL at this point, but needs to have further testing done. My Dad's sister, my Aunt Susan (age 42) also has tumors on both of her kidneys, her pancreas, and some on her liver. She too has had no symptoms. She tested positive for the VHL mutation as well, but because she just learned about this in August she has not had any medical intervention yet. Since then, my daughter, Keri (age 2) and I (age 29) have both tested positive for the VHL mutation too. Luckily, we do not have any signs of VHL after numerous tests. We also learned that my grandmother, Gertrude P. (age 84) has the VHL mutation, but she has never had any symptoms or complications her whole life!

We still have a couple of family members that need to be tested. I have 2 brothers, Michael (age 27) and Ryan (age 25) and a nephew Mickey (age 4) who will be tested soon. My grandmother comes from a family of nine children. She only has 2 sisters that are still living. But 43 other family members could potentially have VHL! My grandmother has many nieces and nephews. We are not sure if anyone else will decide to be tested, but they have all been informed. It is unusual that no one else has shown signs of VHL, so it may be possible that the mutation began with my grandmother.

It has been a difficult year for our family finding out that we have this rare inherited disorder, but when we found the VHLFA website it gave us hope. We were able to learn so much from the site and we still learn something new every time we visit it. We are also grateful for all the publications and for the Handbooks that your organization has sent to us. It is scary when our doctors do not know anything about VHL or they've never heard of it! But through the VHLFA we have been able to educate ourselves, doctors, family and friends. I truly feel VHLFA has given hope to those who don't know where to go. I don't know what we would have done if it were not for VHLFA!

Through the VHLFA website we were also able to learn about the U.S. National Institutes of Health and their fabulous research studies. Thankfully, my Dad and my aunt were just accepted into a research study. We feel blessed that they will be able to see some of the best doctors in the world. We also learned from the NIH that my Dad's type of kidney tumors have never been seen before with anyone else they have studied. It will be interesting as they learn more about it.



Melissa K with her Dad, Ron, and her daughter, Keri.

On behalf of my family, I decided to do a fundraiser for VHLFA because of its help and dedication to those with VHL. I didn't do anything spectacular, I just sent out letters to family and friends, but I found that everyone who responded was very supportive and generous. I just felt that I had to do something for VHLFA in honor of my grandma, my Dad, my aunt, my daughter, and to all those who are dealing with VHL. We have been blessed in that VHL has not been as cruel to us as it has to other families, however I understand how many families feel with this diagnosis. I know it is difficult to live with. I felt that doing something to find a cure for VHL was the right thing to do. I pray that a cure can be found in my daughter's lifetime so that she will never have to worry about VHL. I also hope that my Dad and my aunt can live long lives despite this new diagnosis. My Dad and my aunt have had a hard time learning about this, just as I have. It was so unbelievable to think that our family had this rare disease. Here I thought we were pretty

Has the VHL Family Alliance made a difference in your life? Please help to keep our services strong, and fund research. **Thank you!**

normal and to find out that there aren't many people in the world with VHL was shocking! I remember talking to a few geneticists that were in "awe" to talk to someone with VHL because they have never encountered a VHL patient in their practice! It was also scary telling my doctors about this, especially when they've never heard of it! Luckily, I had my VHL Handbook for my doctors to refer to.

Another reason I decided to do this fundraiser was to raise awareness of VHL. Since VHL is so rare, I felt people needed to know about it. I also wanted to emphasize the importance of getting a regular physical and blood panel done no matter what your health status is. I find that everyone talks about being healthy, eating right, and exercising, but I never hear people talking about the importance of regular doctor check-ups. I truly believe that my Dad's routine physical and blood panel are what saved his life and that of all of our family members with VHL. My Dad actually had high blood pressure, and with my Mom's help she prompted him to see his doctor. Instead of his doctor just giving him blood pressure medicine and sending him out the door, she decided to give him a physical and to do a blood panel. When his doctor found high levels of creatinine in his blood, she knew his kidneys were not working properly. An ultrasound, CT scan, and MRI eventually led to surgery, which led the doctors to believe this was VHL. I never realized how much your blood can tell about what is going on inside your body, so I felt that it was important for people to realize this just as I did. (*See Note below.)*

I began the fundraiser on September 20, 2004, and ended it on October 20. Thus far my family, friends, and I have raised $3500. I have enclosed checks along with the VHLFA forms I downloaded from the website. I was truly amazed that we were able to raise that amount of money by just sending our simple letters! I hope this money can help out in many ways, and hopefully a cure can be found one day.

Grandmother Gertrude and Aunt Susan.

United Way? write us in!
CFC? #0242
Please print out one or more posters from our website to hang in your office for workplace charity campaigns.

Thank you again for all of your help to our family and to everyone with VHL. We truly appreciate the VHL Family Alliance!

Editor's Note: We are grateful to Melissa and all her family and friends! We should also note that VHL does not always create a disturbance in the function of the kidney, so a blood panel is not sufficient to check for VHL. It is certainly fortunate that in this case it showed up in the blood work and the doctor did such conscientious follow-up. People who know they may be at risk for VHL should follow the screening guidelines in the Handbook to check for possible kidney problems.

Healthy Kidneys

By Tammy M., Pennsylvania

I have had VHL for approximately 12 years now, although it was not diagnosed until I had a carotid body tumor *(see note)* removed, back surgery, and discovered angiomas in my eye. It was my eye doctor who diagnosed the possibility of VHL and told me to get my kidneys scanned immediately. At that time I looked up VHL on the internet to discover the VHLFA website and information on what VHL kidney cancer was all about.

You will never know how helpful this website has been!!! At that time, a lot of places were doing renal-sparing surgery, so that was some hope for me. My local urologist diagnosed the kidney cancer and said he didn't know much about VHL but he thought it was a situation where the tumors would keep recurring so "we might have to take both of your kidneys out and put you straight on dialysis." Boy, was I glad I found your website!!!

That was eight years ago and I have yet to have any invasive kidney surgery. I was fortunate to be involved with a kidney study through NIH and have had radio-frequency ablation done successfully on three separate kidney tumors over the past four years.

I didn't think I was the type of person who would ever take time to type a letter to VHLFA but I had to just say thanks to all of the people who keep the website going and the research monies and the many other events to help VHL patients.

Note: a carotid body tumor is an extra-adrenal pheochromocytoma (also called a paraganglioma) near the carotid artery in the neck, below the earlobe.

My Miracle Baby

by Amy W., R.N., Kansas

I am a third generation VHL survivor. My maternal grandmother died from metastatic renal cell carcinoma when I was 12 years old. She had already lost her vision due to a tumor on her optic nerve and also had a craniotomy for a brain tumor. At the time, we didn't know that it was VHL.

My mom also has VHL. She has had two craniotomies (for brain tumors), two laminectomies (for spinal tumors), three partial nephrectomies, multiple eye surgeries that have resulted in loss of vision in her right eye and she still has a spinal cord full of tumors that have left her with extremely decreased sensation and strength below her neck.

I have a little brother who is 15. He has made the decision not to have the blood test done, so instead my mom makes sure that he gets his screening done. So far he hasn't shown any signs of VHL. Of my mom's siblings, one of her brothers has been tested and is negative, her sister passed away from a brain aneurism, and was never tested (however she and her husband didn't have any kids) and mom's youngest brother hasn't been tested.

I am 27 years old and was diagnosed with VHL when I was about 13. My mom had only been diagnosed for about a year or so before me. My diagnosis came after finding retinal hemangiomas. I have had multiple eye procedures, but because of the location of the tumors and the wonderful care I have received, I still have 20/20 vision.

My mom has always been as honest with me about VHL as possible, however when I was diagnosed we were still extremely new to the disease. She told me at the time that the theory was that VHL could be "triggered" by hormonal changes, but I didn't really get it. When I was 19, I went on birth control and four months later I was having a craniotomy.

My only symptom was having the hiccups between 20 and 40 times a day. I had seen a wide range of different doctors, and all any of them could offer was a pill that would make my hiccups go away. To me that just didn't sound right, surely there was a reason for them and I wanted to know what it was. I convinced my primary doctor at the time to order an MRI of my head and a CT of my kidneys, as Mom and I felt I should probably have some baseline scans done. After having my head MRI reviewed, it was discovered that I had a tumor in the brain stem and it was pushing on the area that controls hiccups. After a few different opinions, I told my Mom that I only wanted her neurosurgeon to do the removal, as I had seen firsthand the work he could do from her previous surgery. My surgery was a complete success.

At that point I quickly learned that VHL wasn't anything to play with and it was going to be a big factor in my life decisions forever. I also came to the conclusion at that point that, at least for me, hormonal changes were going to be tough.

In May of 2002 I graduated from nursing school. I married my wonderful husband Lee in August of 2002. He was aware of my VHL long before the wedding, but he had only experienced my Mom's recovery from one of her partial nephrectomies. It was still pretty foreign to him. We had spent many hours talking about children. We both knew that we wanted a family, but the means of attaining that family wasn't clear. He was extremely supportive and told me that he would be happy adopting or having our own children, if I thought it would be safe for me.

A couple of months after the wedding, baby fever really hit hard. I wanted a child, and I wanted to have my own child. So I started having all of my screening done. I had multiple kidney cysts that had been there and remained unchanged since my very first scan six years earlier. I also had multiple pancreatic cysts (one large enough to measure) that were again unchanged from the beginning. I had a spinal tumor that was found six years earlier that also had never changed and I had a cerebellar brain tumor that had been found two years earlier, but had remained stable for those two years.

I talked with my neurosurgeon and my urologist about my thoughts of getting pregnant, and both felt this was as good a time as any. I consulted with a perinatologist (a specialist in high-risk pregnancies) and we discussed VHL and my manifestations. She was very honest and told me that she didn't have personal experience with VHL, but she would do her homework. She sent multiple e-mails to other perinatologists and copied me on all of them, as well as their answers. We met again, and she agreed to take me as a patient. She said we would develop a plan for delivery that would be safe for me and the baby.

Learn from each other!
Support one another!

Attend a meeting near you.
See the roster of 2005 meetings
on page 12.

I got pregnant in January of 2003. Lee and I were so happy. We had agreed not to tell anyone, except our parents, until we were through the first trimester. However, that plan didn't work. I began having terrible headaches during my 8th week of pregnancy. I called my doctor and we tried everything. I had given up caffeine cold turkey, so I started drinking some caffeinated things to see if that would help. It didn't. By now, I was having trouble standing up from a sitting position without feeling sick to my stomach and having terrible pain. Finally, after about a week of this, my perinatologist sent me for an MRI of my head (which is perfectly safe during pregnancy due to the lack of radiation, and the contrast is also safe). The tumor that had been stable in my cerebellum for two years had taken on a life of its own and was now blocking my cerebrospinal fluid from draining into my spinal cord to maintain the fluid balance in my brain.

My perinatologist took it upon herself to consult my neurosurgeon, and together they agreed that the best plan was to wait until after my 12th week of pregnancy, when the baby is formed, to do surgery. Unfortunately, I couldn't make it that long. My neurosurgeon was afraid that the pressure in my head was going to cause me permanent damage if it wasn't relieved immediately, so he personally called my house, told me to pack a bag and get to the hospital immediately. He scheduled surgery for the next day, but wanted me in the hospital in case something happened during the night. I was only in the beginning of my 10th week of pregnancy.

I had surgery on April 7, 2003, to remove the tumor. After being anesthetized, my surgeon had to put a ventriculostomy (external brain drain) in to relieve the pressure before he could go after the tumor. He was forced to leave me on the table under anesthesia (and pregnant) until enough fluid had drained to allow him to proceed safely. My whole surgery took about six hours, but it was a complete success. After surgery all anyone could tell me about my baby was that there was still a heartbeat.

By now you can image the crash course that my husband was receiving on VHL. He was worried about me and worried about the baby. We spent the next seven months having multiple sonograms done. Each one assured us that the baby was fine, normal, whatever word you want to use, but nothing was going to convince me until I could see this child for myself.

Two weeks before I was to be induced, my perinatologist sent me for the whole series of head and spinal MRIs to make sure there were no changes. All that was found was the spinal tumor that had thankfully remained stable for my whole pregnancy. I also went to see my eye doctor before

Lee, Amy, and Kaytlynn at nine months.

delivery and he found a small retinal lesion. Armed with this information, my doctor consulted with the anesthesiology team at the hospital, and presented my case to them and told them what she wanted to do and why. With everyone on board, we proceeded.

I was induced on the Friday of a weekend that my doctor was on call. While I had met with all of her partners and they all agreed on the same delivery plan, I wanted her to be the one handling the situation. We got to the hospital at 8:00, they started Pitocin at 9:00, and broke my water at 11:00. I had an epidural placed (thus one of the reasons for the spinal scans pre-delivery). My doctor was adamant that I not push (she was afraid of elevating the pressure in my spine and head and causing problems with my eye tumor or my spinal tumor) so we let the contractions work the baby through the birth canal, and then I played tug-of-war with the nurse. She wrapped a bed sheet around her waist and whenever there was a contraction, I would pull against her, helping the baby get through the birth canal. Finally the baby was far enough along that my doctor (who had an extremely high level of expertise) was able to use forceps to get the baby all the way out. At 3:10 on October 24, 2003, a perfectly healthy, 7lb. 5oz. Kaytlynn was born. She is my miracle baby.

Even with everything that we went through, I have never regretted the decision that we made to

cont'd on page 10

have our own child. Yes, I worry that she might have VHL, but I also realize that the advances we have made and continue to make will be at her disposal. We are going to have her genetic testing done when she is a little older, when she is a little slimmer and has easier veins to stick and when we are used to having a child who bumps her head and has a cold, things that all kids do. We need to know that if she does have VHL, everything that happens isn't going to be related.

We have already had a complete eye exam, dilatio and all, done on Kaytlynn. She has also had an ultrasound of her kidneys done. Both of these were done at the request of the doctors that I want to follow her if she does indeed have VHL. While some would argue that it could be a waste of my time and money to start developing her team of pediatric specialists now, before I know her genetic results, I don't agree. Part of it is being a mom, wanting only the best of your child and knowing that you have done all that you can to provide for them, in all areas of life. The other part is being a nurse and seeing too many people who aren't proactive and settle for whatever care falls into their lap. I refuse to settle for anything other than the best for myself, so to take care of my child, you better believe that the doctor is going to be top notch.

I don't spend time wishing it wasn't my grandma, or my mom or myself. It is a fact of life that can't be changed, but must be dealt with. We have a rule in our family that you never go to the doctor alone, even if you don't expect to hear anything new. Everyone needs support, and two sets of ears are always better than one. My mom and I have our CTs done together so we can make faces at one another while we drink the terrible contrast. When the time comes for a change in the news, I allow myself one good cry, and then it is nose to the grindstone to figure out what needs to be done and to get myself physically and mentally prepared to deal with the newest challenge. That doesn't mean that there aren't days that I get tired of dealing with it, but the fact remains that I can't change it and I am not willing to let VHL define me.

We live with the motto that "God will never give you more than you can handle". There are definitely days that I think He is pushing His luck, but then I hear a sweet little voice say "mama" and I know it is all worth the effort.

For VHL women who are considering pregnancy, I have the following recommendations:

- Consult with a perinatologist before you become pregnant (some will even agree to just oversee things if you already have an obstetrician that you feel comfortable using).

- Discuss delivery day, long before the time comes. Make sure that everyone agrees on the plan, including any partners of your doctor who could potentially deliver your baby. Make sure that you are 100% on board with the plan. If your doctor suggests forceps or a vacuum birth, make sure they are fully trained, use the techniques often, and that the hospital has supportive services available should the baby need them.

- Avoid a Caesarean-section whenever possible. While the scar from a C-section should be well below the area for entrance when doing anything to the kidneys, why risk adding scar tissue to your abdomen when renal cell carcinoma could realistically be in your future?

- Be sure to have thorough and complete scanning and an eye exam done before you become pregnant and shortly after the baby is born to know what you are dealing with at all times.

- Don't be afraid to ask questions. Make sure your doctors know about VHL, and if not, that they are willing to learn about it.

- Talk to other women who have made the decision to have or not have their own children. No one else can make the decision for you, but knowing someone else had the same hopes and fears can be reassuring.

Editor's Note: *This is Amy's story. There is still no data to prove whether hormones definitely trigger tumor growth, or whether the tumor was just ready to make its move when the hormones changed, but times of hormonal change are definitely a time for caution and careful monitoring. As described in the new Handbook, the normal changes of pregnancy can certainly inflate existing tumors. If you have had experiences with pregnancy — both positive and negative — and have good documentation of your case, with good before and after pictures, and would be willing to participate in a study of pregnancy and VHL, please contact the VHLFA at 1-800-767-4VHL.*

Looking for a special gift?

Something unique and inspirational?

See page 15 of this issue for some very special CD's and books, from artists with VHL in the family.

Proceeds of these purchases go to VHL research.

Giving $150 or more? claim one of these gifts free with our thanks!

All donations in January thru March will count toward Melissa's Triathlon.

Memphis Meeting

Fifteen people gathered in Memphis November 20 for the 2004 Mid-South meeting. People came from Arkansas and Mississippi to learn about new sources of treatment for people with VHL. Speakers were Selvi Palaniappan, a genetic counselor from Vanderbilt in Nashville; Dr. Chris Friedrich, a geneticist from University of Mississippi in Jackson; and Dr. Jose Claudio Rocha from Brazil who is in Memphis on a post-doctoral research appointment at St. Jude's Children's Cancer Research Center in Memphis. Ms. Palaniappan and Dr. Friedrich spoke about their practices, and about services for VHL available in Jackson and Nashville.

Dr. Rocha told us the story of his research project in Brazil. He diagnosed a patient with VHL and checked the records at his very large, very fine hospital in Saõ Paolo. There were no other VHL patients listed in their database! Finding this impossible to believe, he worked to spread word of his research project among physicians, and on radio and television. Over the next five years he identified 80 people with VHL in 27 families in Brazil. He has begun a family support organization there, in cooperation with VHLFA, and is offering free DNA testing for people with VHL in Central and South America.

Dr. Rocha told us about the state of research toward drug therapies for VHL, and the work he has been doing at St. Jude's. While there is good progress, we should not pin all our hopes on drug therapies. The most successful approach today is prevention — early diagnosis, and appropriate treatment. Remember that for breast and prostate cancer the greatest advances in survival have come about because of early detection. If tumors are found earlier, they are much more successfully treated. In the near term, the guidance provided by the VHL Handbook and the partnership of your medical care team are the best way to maintain your health.

The Mid-South is a heavily rural area, with fewer services than many more populated regions of the United States. Dr. Rocha mentioned that in his home city of Saõ Paolo there are 18 million people. There are only 12 million people in all three states of Tennessee, Mississippi, and Arkansas — only two-thirds the population of the city of Saõ Paolo!

There are certainly some talented physicians in the Mid-South area, but it has been difficult to find people who are familiar with VHL. People with VHL have had to shop one by one for specialists in a particular field, with little help and mixed success.

Three major improvements have come about in the last few years.

First, some people with expertise in VHL have moved into the region, notably:

- Dr. Lewis Blevins (endocrinology) and Selvi Palaniappan (genetic counselor) moved from Atlanta to Nashville. They have set up a VHL Clinical Care Center at Vanderbilt in cooperation with the VHLFA. +1 (615) 322-8960

- Dr. Mary Curtis (genetics) moved from Iowa to the University of Arkansas in Little Rock. +1 (501) 320-2966

- Dr. Chris Friedrich (genetics) moved from University of Pennsylvania to University of Mississippi, Jackson. +1 (601) 984-1900

- Dr. Bruce Korf (genetics) moved from Boston to University of Alabama, Birmingham. +1 (205) 934-9411

Second, these genetics professionals have been building teams around themselves to assist people with a number of genetic diseases, including VHL. They are willing to assist patients with VHL to find an expert who can assist with a particular problem.

Third, there are a number of new departments of Cancer Risk Analysis. For example, at Baptist Hospital in Memphis there is a new division, Baptist Centers for Cancer Care. The genetic counselors there, Eric Fowler and Ellen Neese, do not have expertise in VHL. However, they are very willing to assist families with DNA diagnosis and with obtaining appropriate care.

We often tease that if you can't find an expert, you can help to grow one. Doctors cannot be expected to be experts in every rare disease there is. But a doctor with good skills in his or her field, and a willingness to learn, can be an excellent team member. Consider yourself the "owner" of your healthcare team, as if you were the owner of a sports team. You may not know how to do the job yourself, but you know the qualities you need in your players, the talents you need to hire, and the attitudes that will make the team work well together.

You also need a good quarterback — a good leader within the team who makes sure the ball is passed to the person who can lead the team to victory. Your general practitioner may be the person to play this position on your team, or a genetics professional in your area may be able to assist.

And always, keep your own good common sense turned on. No one knows all the answers in VHL. No matter what the level of expertise on your team, your own opinion also counts. Using the Handbook and second or third opinions when necessary, you can create good care for yourself.

We had a great time visiting together, and sharing ideas with some attendees who were new to VHL. We decided to set a date for next year's meeting and work to make it even better! Mark your calendar and be sure to join us next year, Saturday, November 19, 2005.

Educating Congress

Washington, D.C., Educating our Members of Congress. Early February, 2005. VHLFA members are invited to join us for a day of meetings with Senators and Representatives to discuss health concerns. If you are interested, please call 1-617-277-5667 and talk with Joyce.

Calendar for 2005:

Manchester, England, UK meeting, April 9, 2005. For details contact Mary Weetman, uk@vhl.org +44 (0)170 635-8055

Copenhagen, Denmark, Nordic Countries Regional Meeting, April 15, 2005. For details, contact Vibeke and Richard Harbud, harbud@post5.tele.dk +45 4676 7033

San Antonio, Texas, Regional Meeting, May 21, 2005. For details, see www.vhl.org/meetings or call 1-800-767-4VHL.

Binghamton, New York, Regional Meeting, June 4, 2005. For details, see www.vhl.org/meetings or call 1-800-767-4VHL.

Chicago, Illinois, U.S. Annual Meeting, June 25, 2005. For details, see www.vhl.org/meetings or call 1-800-767-4VHL.

Memphis, Tennessee, Regional Meeting, November 19, 2005. For details, see www.vhl.org/meetings or call 1-800-767-4VHL.

2006:

Salt Lake City, Utah, U.S. Annual Meeting, June 10, 2006. For details, see www.vhl.org/meetings or call 1-800-767-4VHL.

Toronto, Canada, Worldwide Medical Symposium, May/June 2006. Details will be posted at www.vhl.org/meetings as they become available.

The Nordic Workshop

15 April 2005, 11 am to 6 pm
Hotel Sct Petri, Copenhagen

In 2008 the 10th international symposium on Von Hippel-Lindau Disease (VHL) will be held in Copenhagen. At this symposium we want to have a strong Nordic representation. We therefore invite physicians from all the Nordic countries to participate in the Nordic workshop next spring.

The following issues will be covered:
- Sharing of experiences from the different countries
- Idea for joint Nordic clinical trials and joint patient surveys
- Creation of a Nordic coordination/steering group

The final program will follow later. The program will start at 11 am and finish at 6 pm

The symposium will have free admission and will be sponsored by the Danish patients' organization. However, our funds are not unlimited, therefore we kindly ask speakers from the different countries to volunteer.

We are currently trying to generate a list of VHL-interested doctors in the Nordic countries. We would very much appreciate it if you would send us your contact information (e-mail and postal address) and the same information from any of your colleagues who could be interested.

On behalf of the Danish Medical VHL-doctor group and the Danish patients' organization:

Thomas Rosenberg, Staten Øjenklinik Rymarksvej, 12900 Hellerup, Denmark, E-mail: roseeye@visaid.dk

Vibeke Harbud, Foreningen af Von Hippel-Lindau patienter, Fiskervejen 10, 4000 Roskilde, Denmark, E-mail: harbud@post5.tele.dk

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Gift Books:

❑ *A Time to Walk, lessons learned on the Appalachian Trail, by Jay Platt* @$15 each $ _____

❑ *Sacred Process, the inspirational story in poetry of Karen Koenig @$15 each* $ _____

❑ *How Are You?, Manage your own Medical Journey, by Patricia Foote @$15 each* $ _____

All prices include U.S. domestic postage. For mailing to destinations outside the U.S., please add $5 $ _____

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
Canadians, please send donations to: Canadian VHL Family Alliance,
4227 Hamilton Road, Dorchester, Ontario, N0L 1G3 Canada

Going for the Gold!

by Melissa Thomas, Texas

On July 24, 2005 I am going to set to achieve the greatest athletic goal of my life by competing in the 7th annual USA Ironman Triathlon in Lake Placid, New York. In my efforts to complete this 2.4 mile swim, 112 mile bike ride, and 26.2 marathon run in 17 hours, I am also running this race in honor of my father, who has Von Hippel-Lindau (VHL) disease. My goal is to raise money for VHL research so that a cure can soon be found.

My dad first became aware that something was wrong in 1955 when floaters showed up in his eyes. In 1963 the problem was diagnosed as VHL.

From 1963 until 1986 he underwent numerous surgeries treating the eyes. Unfortunately in 1986 one of the eyes had to be removed. CT's and MRI's revealed that my dad had many other tumors in his body including his brain, spinal cord and cancer in his kidneys. My father has been through several surgeries over the years including 4 brain tumor surgeries and 3 kidney surgeries.

Throughout his battle with VHL he has remained more positive than one can imagine. My dad always looks at the glass as half full. He exercises everyday, follows a strict diet, and has an amazing outlook on life. He has out-survived what doctors had told him in the past because of his sheer determination. He is my inspiration behind the Ironman.

Please support my dream of crossing the finish line of the Ironman Triathlon by helping me raise money for VHL research. Even the smallest donation will get me closer to my goal of $100,000. You can donate online at www.vhl.org/ironwoman or on page 15. Thank you for your support!

Melissa Thomas (center) with her parents, Todd and Mary.

IMPORTANT UPDATE:

Janus Charity Challenge Participant

In conjunction with my fundraising efforts I am taking part in the Janus Charity Challenge. The Janus Capital Group is making additional contributions to the athlete's non-profit organization to the top 30 fundraisers at Ironman USA Lake Placid in the following amounts:

1st Place- $10,000 *bonus from Janus Fund!*
2nd Place- $8,000
3rd Place- $6,000
4th Place- $4,000
5th Place- $2,000

*The next 25 receive a $750 contribution

Please help me "go for the gold" by becoming the top fundraiser.

Support Melissa's Triathlon for VHL, and meet the Janus Challenge!

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

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