VHL Family Forum

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Pheochromocytoma: Often Inherited

Adapted from an editorial by Robert G. Dluhy, M.D., Brigham & Women's Hospital[1]

Medical students often learn axioms in order to remember key features of a disorder. The "rule of 10" used to describe pheochromocytomas (pheos) is a good example: 10 percent occur outside the adrenal glands, and of those, 10 percent occur outside the abdomen, 10 percent are malignant, 10 percent are found in patients who do not have high blood pressure, and finally 10 percent are hereditary. The remainder are classified as sporadic, or random in the general population, not connected with a particular syndrome. When pheos are inherited, they are caused by a dominant gene, either alone or as part of the multiple endocrine neoplasia type 2 syndromes (MEN-2A and MEN-2B), von Hippel-Lindau disease (VHL), or in rare cases neurofibromatosis type 1 (NF-1).

An article in the *New England Journal of Medicine* by Dr. Hartmut Neumann and colleagues has dashed the rule of 10. In his study of a large group of patients with sporadic pheochromocytoma and no family history of pheos, approximately 25 percent had germline mutations of one of the four susceptibility genes for pheo. Now that we have tools to examine the gene itself, we get a very different picture than we had from clinical diagnosis alone.

This rare tumor of the adrenal gland is called a pheochromocytoma (or pheo). When pheos develop outside the adrenal gland they are called paragangliomas or extraadrenal pheochromocytomas. Patients with pheos may have high blood pressure that is difficult or impossible to control. In some rare cases the blood pressure can be low. Patients may have episodic headaches, excessive sweating, and palpitations as a result of the release of stored chemicals, called catecholamines, from the tumor. Left untreated, pheos may lead to a hypertensive crisis, heart arrhythmia, heart attack, or stroke.

When blood and urine tests reveal excess catecholamines, imaging studies of the abdomen usually reveal an adrenal pheochromocytoma. If the adrenal glands are normal, there is likely a paraganglioma among the structures of the sympathetic nervous system in the abdomen, chest, head, and neck. Similar tumors (called chemodectomas) that do not produce catecholamines may occur in the head and neck, including the carotid-body and glomus-jubulare tumors of the 9th and 10th cranial nerves. Even if these do not secrete chemicals, they usually cause a mass effect, such as cranial-nerve palsy and tinnitus or ringing in the ears. Paragangliomas below the head and neck usually present with signs of excess catecholamine production.

In the Neumann study, reviewed cases of a large group of patients from Freiburg, Germany, and Warsaw, Poland, with pheos that did not fit any of the syndromes. These patients were screened for inherited mutations[2] of four genes known to cause pheos: the RET proto-oncogene (MEN-2), the VHL tumor-suppressor gene (VHL), and two genes recently discovered to give rise to pheos and glomus tumors: SDHD and SDHB. Of the 271 patients studied, 66 (or 24 percent of the participants) had mutations in one of these genes.

30 (45%) VHL
12 (20%) RET
11 (17%) SDHD
12 (18%) SDHB

cont'd on page 2

Inside this issue!

Looking back, most of these patients with VHL or MEN2 had multiple tumors, often outside the adrenal glands, at a young age. 80% of the people with SDHD and SDHB had solitary pheos, and 40% were older than 30 years of age.

Even in the people who did not have a hereditary condition, one of these genes was found to be altered in the tumor tissue of these sporadic cases of pheo.

It is curious that so many factors can give rise to a pheo. We know that the VHL protein regulates the normal degradation of proteins such as hypoxia-inducible factor, which is implication in the response to conditions of low oxygen. SDHD and SDHB are part of the mitochondrial system in the cell which regulates oxygen sensing and signaling. Therefore the connection may be a defect in the oxygen-sensing system.

People with VHL have a 10-20% chance of having a pheo, with significant variations in families, depending where the mutation in the VHL gene occurs.

Neumann recommends that anyone with a seemingly sporadic case of pheochromocytoma be screened for mutations of the RET, VHL, SDHD, and SDHB genes. Earlier diagnosis of a condition like VHL or MEN will help to ensure better outcomes for these patients.

The Neumann study included people from two geographic regions, in order to reduce the effect of ethnic variation in the study. Nonetheless these two patient groups are both white Europeans within a distance of a thousand miles. We do not yet know whether these findings will be found to be consistent in all parts of the world. Hopefully this study will be repeated in other geographies and other ethnic groups.

Nonetheless, physicians should have a much higher degree of suspicion for familial syndromes in patients with apparently sporadic pheo. A young age and multiple extraadrenal pheos are clues that should prompt a physician to obtain a complete family history, a careful physical examination, and possible genetic analysis.

1. As printed in the *New England Journal of Medicine*, **346**:9, May 9, 2002. Reviewing the article by Neumann et all in the same issue, "Germ-Line Mutations in Nonsyndromic Pheochromocytoma," pages 1459-1466.
2. Germline mutations are mutations that occur in the genes of the egg or sperm and can therefore be passed to one's children.

The German team, in Padua: Dr. Hartmut Neumann, Sven Gläsker, Birke Bausch, and Dr. Klaus-Martin Kreusel.

Fruits Lower Blood Pressure

Most Americans and Britons do not consume the recommended five daily servings of fruits and vegetables. Many observational studies have reported health benefits of higher fruit and vegetable consumption. A new trial of increased fruit/vegetable intake on blood pressure finds that healthy British adults who added an average of one and a half servings per day decreased their blood pressure. About 345 people were in an intervention group, and an equal number were in the control group. After six months, blood pressure dropped by 3.4 mm for systolic and 1.4 mm for diastolic pressure compared to the controls. Five of 11 blood values of nutrients from plants were increased, confirming that subjects did eat more fruits and vegetables. The study appeared in the May 28, 2002 issue of the *Lancet*.

Here's what you need to know: these decreases in blood pressure may have public health significance. This change occurred without any drop in weight. Nurses spent 25 minutes with each person advocating greater fruit/vegetable consumption. Previous research in the U.S. found high intake of fruits and vegetables combined with low-fat dairy decreased blood pressure more than fruits and veggies alone. So, tell yourself that you should eat more fruits and vegetables. In the supermarket, it will help to keep you away from the processed snacks.

Nutrition News Focus, July 17, 2002 http://www.nutritionnewsfocus.com

Coming to Cleveland? Here's a comment from the last Conference ...

"Wow! I'm so glad we went! Next time we're going to bring our family doctor with us! We all feel much more confident now that we know what to watch for, and where to go for coaching in difficult times." -- see page 5 for details.

Banking Cord Blood

Stem cell research is leading us toward possible genetic therapies that may one day be able to help people with VHL. One concern will be ethical sources of stem cells. One possible source is cord blood, which may be frozen now in case it might be needed in the future by the individual, or shared with others in need.. Cord blood banks are giving hope to many people who would otherwise have spent months or years seeking a satisfactory match for a bone marrow transplant.

"Cord Blood" is the blood that remains in the umbilical cord and placenta following birth. Cord blood stem cells have the ability to treat the same diseases as bone marrow with significantly less rejection. Cord blood is collected after the baby is born and the umbilical cord has been clamped and cut. It is painless and safe for both baby and mother. When cord blood is collected and stored, the stem cells are immediately available for transplantation. Children make up a large portion of the 10,000 individuals each year who are unable to find a transplant in time.

Years of medical research have led to an amazing discovery: the blood in a baby's umbilical cord. First used in transplant in 1988, umbilical cord blood is a plentiful and rich source of stem cells -- the building blocks of the immune system -- that can be used to treat a variety of life-threatening diseases including leukemia, other cancers, and blood and immune disorders. In just the last few years, thousands of acutely ill patients have received treatment because of this tremendous medical advance.

Approximately 25% of these transplants have come from siblings, with the rest coming from donated cord blood samples. As more and more families save their cord blood, whether through donation or private storage, these numbers should increase dramatically. According to the *Journal of the American Medical Association*, "10,000 to 15,000 Americans each year who need a [bone marrow] transplant are unable to find suitable donors". Cord blood is an alternative transplant resource. As of the year 2000, more than 2,000 cord blood transplants have been performed worldwide.

According to the Cord Blood Registry, "It's kind of like having a "spare immune system" - just in case - and costs about the same as three months' worth of health insurance for your family." Doctors recommend that cord blood should be saved especially for children with unusual ethnic mixes, for whom donor stem cells would be very difficult to find.

There are many reputable cord blood banks where umbilical cords can be frozen. Obstetricians' offices usually have brochures and may have labs they prefer to work with. Alternatively, search on the internet for "cord blood banking." Prices vary considerably. Be sure that the cord blood bank you choose is accredited by the American Association of Blood Banks (AABB) and state licensed.

The Cord Blood Donor Foundation (CBDF) is a non-profit human health and welfare organization dedicated to providing educational and public awareness and promoting further research using umbilical cord blood stem cells from live birth for the treatment of disease. http://www.cordblooddonor.org/

The Cord Blood Registry (CBR), in San Bruno, California, is the oldest and largest of the cord blood banks and registries for sharing with families in need. 1-877-267-3256 or +1-650-635-1420 or http://www.cordblood.org

Out thanks to David T. Harris, M.D., Ph.D., Director of the Cord Blood Research foundation, for his assistance in preparing this article.

The Top 10 Ways to Love the Negativity in Your Life

By Christine Morris, Ph.D., Howard University.

Prosperity and abundance theories can backfire when our absorption with being positive causes us to resist and judge the very human experience of lack and limitation.

1. Remember the Chinese yin/yang symbol. Inevitably, we all experience the extremes of abundance and lack, freedom and limitation. Without the contrast, these concepts would be meaningless. In the Chinese symbol for yin and yang, there is always a speck of black in the white, and there is always a speck of white in the black.

2. Put out your welcome mat. In a calm, meditative state, imagine that your negative thoughts and emotions are strangers knocking at your door. Open the door and let them into your awareness — let yourself feel the intensity of your rage, sadness, fear, etc. — and then let that energy move through you. Ask your emotions what message they have for you.

3. Love the messengers. Your emotions — even the distasteful ones — carry messages for you about your authentic self and your true path in this life. No need to kill the messengers — love them! And ask them why they are there. What do you want but are afraid you can't have?

4. Fall apart if you need to. When we are experiencing significant stress, "holding it together" means we are trying to do things ourselves. When we give ourselves permission to fall apart — to stop looking and acting like we have it all together — we open ourselves to the possibility of receiving support.

5. Give in to your limitations. According to Rumi,[1] "the moment you accept what troubles you've been given, the door opens." When we resist or deny or avoid the inevitable limitations in our lives, their power over us grows. We open ourselves to greater possibilities when we welcome our limitations with gratitude and compassion.

6. Look to the turtle. No, your desire or dream isn't here… yet. Experiencing lack or negativity in your life doesn't mean your dream is pointless and futile. It just means you have to be patient and keep moving in small steps toward your desire.

7. Fan the flame. When you experience any type of negativity, you have the opportunity to clarify, focus, and strengthen your desire. Strong desire expressed through consistent action is what enables you to attract your dreams.

8. Pretend you're playing "hot and cold." Ever play "hot and cold" as a child? When you get farther away from the hidden object, someone says "you're getting colder." This is how lack and negativity in our lives help us — they give us valuable feedback about how to move away from what we don't want toward what we do want.

9. Cultivate inner peace. Inner peace does not come from avoiding negative experiences and emotions. Inner peace comes from having a spirit expansive enough to receive tumultuous emotions without losing our center. Who we really are is untouched by the emotions we experience.

10. Find any excuse to laugh. Here's a favorite cartoon from my husband's collection: "If you're having trouble laughing at yourself, put on some weight and let your children cut your hair!" Here's to us beautiful ones!

1. Jalal Al-Din Rumi was a 13th-century Persian philosopher best known for his love poetry. He was also a theologian, often mystical in the manner of St. John of the Cross. Kirkus Review (1999) feels that Rumi is now one of the best read poets in America. Deepak Chopra feels that Rumi's passionate poetry has "a healing quality."

ELST and VHL

At the Padua meeting Dr. Kathlyn Marsot-Dupuch of the French VHL Study Group reported a case history of a 38-year-old patient without family history of VHL who was discovered to have an ELST along with bilateral kidney tumors and pancreatic cysts. There were no retinal or central nervous system hemangioblastomas. Imaging studies showed a large vascular tumor of the temporal bone, but the inner ear appeared normal.

This case illustrates that ELST may develop with no change in hearing or balance. This patient did not show any of the more common symptoms of VHL, and a definitive diagnosis was only possible through DNA testing. The French Study Group recommends that ELST be considered as a major diagnostic clue of a diagnosis of VHL. They recommend that any patient with a vascular tumor of the temporal bone located in the area of the ELST should be checked for the VHL mutation.

From a paper presented at the VHL Symposium in Padua, Italy "Is endolymphatic sac tumor a major criterion for diagnosis of VHL?" by Kathlyn Marsot-Dupuch, Stéphane Richard, et al., French VHL Study Group.

Cleveland Meeting, October 11-13, 2002

Update on VHL: An Integrated Medical Approach to a Multisystem Disorder

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

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

The agenda includes a survey of the medical issues in VHL, diagnostic techniques and treatments, including legal and insurance issues and stress management. A special session is included on pre-implantation testing for VHL. Because of the expertise of the Cleveland Clinics in kidney issues, the program is particularly strong in kidney.

Dr. Andrew Novick

Families are welcome to attend this meeting. In the nine years since we began holding "patient/provider conferences," we and the physicians who have attended have come to believe strongly that it takes teamwork to manage VHL. The patient has an important role to play, and empowering the patient is good both for the patient and for the physician.

The conference is a wonderful opportunity to meet others with VHL. Andy noticed at his first meeting that other people also had "zippers" peeking out beneath the hairline. Tania noted that "all the women were wearing sensible shoes." Gale bumped into another person and both said in chorus, "Excuse me, that's my blind side." You will learn quickly that you are not alone, and that other people with VHL are just like you! -- nice, normal people with a health challenge to manage.

Dr. BrianClark

If you have not previously attended a VHLFA conference, you are encouraged to attend VHL 101, an introductory session with Joyce Graff on Friday morning (9:00 to 12) before the formal start of the conference at 1:00. You will have an opportunity to meet other attendees and VHLFA Board members, and gain an understanding of terms and concepts that may be new to you, especially in parts of the body with which you have not dealt before. The VHL 101 segment is free, but we ask you to register so that we know how many people to plan for.

Dr. Erick Remer

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

Highlights from the Agenda

Dr. Andrew Novick, Chairman of the Urological Institute, will talk about minimally invasive and nephron-sparing kidney surgery. He is one of the pioneers of kidney-sparing and adrenal-sparing surgery.

Dr. Brian Clark, Director of Reproductive Genetics, will talk about genetic testing for VHL, and preimplantation testing for genetic disorders like VHL. Dr. Clark is a specialist in prenatal diagnosis and medical genetics.

Dr. Erick Remer, a specialist in magnetic resonance imaging, ultrasound and computerized tomography, will talk about advances in diagnostic imaging.

Best-dressed!

Get your VHL Tee-shirt before the conference at special clearance rates! Damon G. models our Caring Tee. All our current stock of T-shirts has been reduced for summer clearance.

Shirts at $7 to $10, U.S. postage included, while supplies last. Call 617-277-5667 or see our online store at www.vhl.org

Our Experiences with DNA Testing

Frank & Paula C., Colorado

Obtaining the testing: Our doctors and local blood laboratories had several suggestions, but were not at all sure where to get DNA testing specifically for VHL. Of course, we immediately called upon the VHL hotline and spoke with Peggy Marshall, who provided a wealth of information. She helped us reach Dr. Catherine A. Stolle, of the Children's Hospital of Philadelphia.

Upon our contact with Dr. Stolle, forms were sent to us, then completed by us, as well as at the doctor's office.

Submitted the test: After Frank's blood was drawn at the doctor's office (in accordance with the directions of Dr. Stolle), Frank and I went directly to Federal Express asking for assistance in shipping the blood properly. Our doctor's office did not know how to ship the blood, and the specific directions (other than going through Federal Express) were not included in the information supplied.

Fed Ex needed the proper packaging for the blood. We stopped at an Urgent Care facility in the area to get the proper packing required for blood. The folks at the Urgent Care were nice enough to give us the packaging at no charge, and said it was how they shipped their blood samples. Fed Ex was in agreement with the packaging—we got lucky on our first try to find the packaging—not having to go to a laboratory or hospital.

The shipping fee was $25, out of our own pockets. We charged the DNA fees for the lab on a credit card (over $600), and when the results came in, requested reimbursement from our health insurance plan. We were able to be reimbursed for the cost of the testing. Upon renewal, we have noted our health insurance plan now excludes payment for DNA testing to determine a medical problem.

Who explained: Dr. Stolle was kind enough to talk to us about the testing, but most of what she said went way over our heads. The testing was not completed for 3-4 weeks, so when I called first to follow up, the testing was still incomplete.

Upon another call to Dr. Stolle, an assistant told me the results had been sent to our doctor's office just that morning. I immediately called, and was told the doctor would call me back. Dr. Ramos called and said Frank did test positive for VHL, and would send us the entire report, as it is complicated.

To this day, the only part of that report that means anything to us is the last paragraph that states in part, "This mutation has been reported previously in patients with VHL."

Points of confusion: Dr. Stolle said it was positive and we have reviewed the test results. Sure we are confused, but we are not scientists.

On the day we found out that Frank tested positive for the VHL gene, three hemangioblastomas were removed from inside his spinal cord, and we await the removal of a second brain tumor in his left, upper cerebellum. His first brain tumor was removed from the right, lower cerebellum.

Our experience with VHL began on March 20, 2001, and Frank was confirmed positive for the VHL gene on August 16, 2001.

I hope our information can help you and others. I cannot tell you how much we appreciate the help of Peggy Marshall in our desperation and confusion.

How to Avoid Confusion

Editor's note: In order to avoid the confusion expressed by Frank and Paula, it is preferable to order the test through a local geneticist or genetics professional. This will ensure that the local team knows how to ship the blood, and can assist in explaining the results to you in language you can understand. While everyone wants to speed through the process, this is one case where it is good to slow things down, talk through your fears and considerations before the testing, and have someone available to help you understand the full implications of the results, for yourself and others in your family. Optimally, schedule a 30-60 minute sessions with a genetic counselor to review the results. VHLFA is always willing to talk with you as well. 1-800-767-4VHL, info@vhl.org, or vhlfa@yahoogroups.com Discussion groups are also available in French, Spanish, and German.

DNA testing for differential diagnosis and for the first sample in a family are best performed by one of the "high hit rate" labs in the world. Dr. Stolle's lab has recently relocated to: Dr. Catherine A. Stolle, Dept of Pathology, Children's Hospital of Philadelphia, Abram Research Center 1106F, 34th and Civic Center Blvd, Philadelphia, PA 19104. Telephone 215-590-8736, Fax 215-590-2156, e-mail: stollec@email.chop.edu The complete list of DNA testing facilities for VHL is at http://www.vhl.org/healthcare/dna-src.htm

Who Explains the Results?

DNA-based diagnostic testing is considered to be "high complexity" testing by the agencies that certify clinical labs.* While every effort is made to describe test results in understandable language, the nature of the test and, therefore, the results are highly technical. Because of the nature of the clinical information in the report, test results must be communicated to the patient by the requesting physician or healthcare professional who is responsible for answering questions regarding the results and their implications for patient care.

It is a sign of our times that patients are being urged to be more pro-active in their own medical care. When they are, however, they may find the process confusing and the information difficult to interpret.

There have been discussions at the federal level regarding regulation of genetic testing. The conclusions of a panel of experts organized to advise the Surgeon General include the recommendation that genetic testing not be provided directly to the patient by the laboratories. Instead, testing must be requested by a physician or healthcare professional base on clinical necessity (as assessed by examination of the patient or the patient's records). The physician is then responsible for communicating the results, discussing the implications for the patient and the family, and coordinating patient care. In some cases, unfortunately, the local physicians are failing in their responsibility to their patients to serve this vital function. -- *Catherine Stolle, Ph.D., Children's Hospital of Philadelphia, Pennsylvania*

*In the United States, Clinical Labs are certified by State and federal governments under the Clinical Laboratories Improvement Amendments (CLIA) and the College of American Pathologists (CAP).

Ask the Family

Question: Has anyone with brain stem or cerebellum involvement experienced trouble traveling on winding, mountain roads? I know that I do. In fact, just this weekend I was on Emerald Bay Road in California, which actually has hairpin turns. That road can make me very dizzy and ill.

I was surprised that my companions in the car seemed irritated with me, when I asked them if they could please slow down. Perhaps my companions had a lack of understanding of my condition.

Is this a problem for anyone else. And, if so, what have you found that helps to relieve the dizziness? -- *R.M., California*

Answer(1): I have lived and worked in the Himalayas for 31 years and find that , although I have VHL, it is more a matter of acclimatisation. If you do travel on winding roads often and have to, the dizziness adjusts. Even people without VHL or cerebellar/spinal involvement react quite violently to our roads here-they're all hairpin in the Himalayas and the drops between hairpin and hairpin are astounding. I once had a friend from California stay and we took her to a small hill station -- small hills by our standards, anyway -- and she went into a substantial hysterical fit.

Depending upon the degree of the slope, the spatial variation between the point you last left and the one you're going to, the amount of terrain within your vision (which in any case has to adjust rather rapidly too to the ups and downs) and so on, giddiness and dizziness are not unusual in people not used to mountain roads. Even people who are react have become accustomed, may react badly again when there's a long gap between excursions. The person driving doesn't seem to react so strongly because their focus is adjusted to the stretch of road in front.

There's a big difference between being in a vehicle and walking, climbing: the latter being normal functions of a mammal's body, sitting on top of four wheels and rolling up and down is not. Lots of things happen physiologically when you move up and down mountain roads, and they're not all caused by VHL. The effects of your VHL issues might make it worse. She could try getting out of the car and walking up an incline and down again at her own pace. If that doesn't make her feel dizzy and giddy, then it's probably for other reasons. I'm not an expert, just learned a little by doing. Just thought this might be helpful. -- *Greta R., Nepal*

Answer(2): I have this problem too, resulting from brain stem surgery years ago. If you have the opportunity to be riding in a similar area again -- and obviously are a passenger and not a driver! -- see how you feel if you close your eyes before the winding starts. *Before* is a key word, as it may be beneficial if you do not even see the *beginning* of the winding. In my case, I am also bothered by the downward curve of even minor slopes in the road. If this trick eliminates the problem, you may be able to achieve at least some degree of relief (if not complete) with vestibular therapy vision and motion exercises.

I'm assuming you are up-to-date with all your scans. If you haven't already, talk to your neurosurgeon or neurologist or otolaryngologist about the problem and ask about vestibular testing to hopefully identify the particular spot(s) responsible for those miserable feelings. They may then be able to isolate some simple exercises to help you cope better with it. I know some VHL patients who have had total success overcoming this same problem and others, who along with me, have not.

We always marvel at the fact that so many folks go out and spend money in bars just so they can feel this way. (Smile) Others choose the amusement park rides. It just is not fun when you can't make it stop. A couple people have had varying degrees of success with different motion sickness medications, but you then have to wonder exactly what was the cause of those nasty feelings. I believe in always trying to find the explanation first, so no serious new or changing VHL problem (lesion) is overlooked. Then I personally try to adjust or improve the ways I handle the situation (i.e. movement tricks, eye focusing tricks) without chemical aids (and that includes the "natural" chemicals in herbs and health food stores products).

Always talk to your doctors about any new problems like this. Good luck...I suppose this means you won't be wanting to stop by Cedar Point Amusement Park in Ohio when you go to the conference this October? Me neither! -- *Fran Mott, Michigan Chapter VHLFA*

My Life with VHL

by Erika T., Switzerland

My name is Erika. I was born on 14 December 1958 and I grew up with my older sister Vreni in Arth-Goldau, Switzerland.

At the age of 15 I suffered sudden profound hearing loss in my left ear. At that time, my doctor was unable to determine the cause. He told me it was "a minor loss." He said that I still had one ear, and that should be enough. I can still remember how shocked I was that my problem was to him just a "minor" loss. For me there was nothing more to do at that time. I would not be handicapped by deafness on only one side. I could continue my life normally with it.

When I was 28, in 1986, I had the first of many disturbances in the hearing in my right ear. On my mother's side of the family there had been many kinds of tumors, but no case of hearing loss. One uncle in 1986 was told that he was being treated for von Hippel-Lindau (VHL), but there was no hint that it was a familial condition. My mother went blind at the age of 18, and at 35 was paralyzed on one side. She had brain tumors, but more is not known. She died at age 50. As children we knew nothing more than that our mother was ill, but she was strong and independent! To this day she is still a great example for me of humor, courage, and strength. But in those days there was practically no information available about VHL. No one imagined that we children could have this disease. It is amazing to me to see how much has been learned about VHL in the last two decades -- how much information is available to us today.

Health problems appeared for my sister Vreni and me about the same time. Vreni had a brain tumor, which could only be partially removed. For me, a tumor appeared in the petrous bone on the right side, near my hearing ear. In both our cases, the doctors rejected a diagnosis of VHL. Vreni was only 34 when she died, leaving three children behind. After that it was clear to me that there must be a relationship among our medical conditions, and I began my quest.

I got no help at all from the doctors. None of them had any ideas. One even said to me that any relationship was impossible and there were no similarities in my family -- that each occurrence was completely different and unrelated. In my case, my illness was taken care of, and I would be fine from here on. I truly wanted to believe that.

But I was now deaf.

Soon after I lost the last of my hearing, I was able to get very good lip reading lessons, and from the beginning I learned lip reading easily, so my communication is fairly smooth. However, I for a long time I did not accept being handicapped. On the outside I seemed content, but inwardly I was not. I even had thoughts that this is not the life I wanted to live. But I knew I had to come to terms with myself.

Erika T., Switzerland

I continue to cover it up, glad to be alive. But I have to come to terms with myself and my revised set of abilities.

I believe that each person who becomes deaf has different feelings about it. Everyone has a different reaction. It takes mental strength, an inborn optimism, a talent for lip reading, an ability to visualize, and hope, to deal with this fate.

Deafness may not come all at once. In the beginning it may improve. Then suddenly it becomes final. "Your hearing will never improve." It was again a shock for me. And who is responsible? Who is to blame? No one can be blamed for this. Today, with better understanding of these endolymphatic sac tumors and petrous bone tumors, and with early intervention, it is usually possible to save the hearing. But it is only in the last five years that this information has even existed, and then only if you look very hard to find the right expert.

I have VHL. A very good family friend finally helped me get a clear diagnosis in 1997. He learned about VHL on the internet and located Professor Neumann, and everything finally became clear. I finally learned that my illness did not occur by chance, and that is has a name. Moreover, great progress has taken place in its treatment: genetic tests, information about the changes in my gene, recommendations for screening and early intervention to prevent disabilities like mine.

Through DNA analysis we learned that my two children have VHL, and two of my sister's three children are affected as well. I know I have to learn as much as possible about it to help them. They should never have to go through the bitter experiences of my mother, my sister, and myself. Many mistakes were

made due to the inexperience and arrogance of the doctors. I can't help saying this frankly because it is so. My rage about this will take a long time to subside. And I know that I have to play an active role in the future.

For me, it is enormously important for all people affected by VHL to be located, so that they can get the advice and help they need to manage their health. Presently here in Switzerland the situation is not the best. We have good ordinary clinics and radiological institutes. Unfortunately, however, it is true here as everywhere in the world that we people with VHL must struggle for ourselves and must know more about VHL than most physicians. It is still very difficult to find the right clinic and the correct treatment. Even though Freiburg in Germany is only thirty miles from the Swiss border, treatment there is not covered by medical insurance. That is a serious problem for us. We are forced to put our trust in the physicians available to us, even though we do not know how much he knows about VHL. And we have to know when to press for better answers, and help the doctors get the advice they need to serve us better.

Many things have already happened to me, and I have had many operations. Many things still lie ahead for all of us. But who knows the what is going to happen? We need to know how to watch out for the possible issues in VHL, and to work with the doctors to find the best treatments and medicines. We should not allow ourselves to remain in a state of confusion -- by sharing information and working together we can help ourselves and all people with VHL.

We must try to live with it, adjusting to our fate. It is important to make adjustments as we go forward, and not give up. It is a great challenge, and all of us must be strong and courageous in the face of this and all the other challenges life will bring. I am very glad to have found so many caring people in this community. That is what makes up the VHL Family Alliance -- the many people with VHL, and the physicians and researchers who are working and studying to make the future for us as comfortable as possible. Many thanks to all who have contributed anything to the great success of the Alliance.

In closing, I thank the researchers and the many health care professionals and technicians; especially Professor Neumann who is always there and always has time for us; the physicians, and each and every one who has contributed to easing our lives. It would be wonderful if all mankind could have a better understanding of people who deal with illness and handicap, and who are trying to live out our lives happily and free from worry.

Erika is a member of the German-speaking VHLFA and our contact person in Switzerland. She is an active member of the German language online discussion group of the VHL Family Alliance, vhl-de@yahoogroups.com.

Endolymphatic Sac Tumor

Of all the people with VHL seen at the U.S. National Cancer Institute, roughly 10% develop an endolymphatic sac tumor (ELST). 10% of those (1% of people with VHL) develop tumors on both sides.

With early detection, hearing loss can be prevented in many people. There is a reasonable chance that a skilled surgeon can remove a small ELST and not damage the inner ear. Once an ear tumor gets large, though, it is much more likely that the inner ear will need to be sacrificed to remove the tumor completely, which is the only successful long-term treatment.

Awareness of VHL and ELST is still problematic, even though there are more case reports describing the relationship between them.

The most important thing for people with VHL to know is that routine screening for VHL will likely not find an ELST. While most individuals affected by VHL get routine brain MRIs (if they are in a good surveillance program), regular MRIs of the brain typically use a slice depth of 5 mm through the brain region. Slice depth or "cut" refers to the distance between the pictures. Images may be captured that include the inner ear region where an ELST can develop, but the small sac in which an ELST usually develops is only 1-2 mm in size, so there is a reasonable chance that these pictures, 5 mm apart, might "skip over" or not pick up the early ELST. If an ELST is suspected, dedicated MRIs of the internal auditory canal region with gadolinium contrast, usually with 1 mm "cuts" through the level of the inner ear, are the best way of detecting an early ELST.

Obviously, its not practical to order this detailed time-consuming and expensive scan on every person, so clinical suspicion and judgement have to be employed. If individuals complain to their doctor that they are experiencing episodes of vertigo, hearing loss, tinnitus (noises in their ear), a sense of fullness or pressure in their ear or perhaps facial weakness on one side, then a dedicated MRI of the internal auditory canal region is indicated. Even after such a scan is ordered, its necessary for the interpreting radiologist and other physicians to be aware of the possibility of an ELST and also to look for it on the scan. Sometimes a correlative CT scan of the same region can be helpful in confirming a possible ELST.

— Daniel Choo, M.D., Director, Center for Hearing and Deafness Research, Department of Otolaryngology, Children's Hospital Medical Center, University of Cincinnati College of Medicine, Cincinnati, Ohio

Family Fights Ignorance about VHL

By M. Scott Morris, staff writer, Northeast Mississippi Daily Journal, Tupelo, Mississippi

They say a lady isn't supposed to reveal her age, but Corinth, Mississippi, resident Peggy Marshall doesn't mind.

She has Von Hippel-Lindau disease, a hereditary condition that causes tumors to appear throughout the body. When she takes calls for the VHL Family Alliance's patient support number, people often can't help asking her age.

"They're amazed how old I am — that I've lived this long with VHL," 57-year-old Marshall said.

The sense of amazement is due to the disease. Since it's often hereditary, many patients have seen what it can do to their parents and family members.

"A lot of times their grandmother died at 29, their mom died in her late 20s and now they're in their middle 30s and everything is falling apart," Marshall said.

Chances are the person's grandmother and mother didn't know they had VHL and didn't get proper treatment.

Marshall, who has served in many leadership roles with the VHL Family Alliance since 1994, is committed to spreading the word about VHL so others can get an early diagnosis and a better chance of living a normal lifespan.

"If caught early, the tumors can be kept in check," she said. "If you don't get treatment, then the tumors are free to cause more damage."

Family history

The VHL Family Alliance estimates one in 32,000 people have the disease. Because of its hereditary nature, the disease usually affects whole families, not just individuals. However, not all cases of VHL are hereditary.

VHL holds few surprises for Marshall. Her mother died with the disease. Her brother, two sisters, two nephews, daughter and granddaughter have also been diagnosed.

"My mother knew she had VHL when she died. But when she died in 1982, we had not met anyone else with VHL," Marshall said. "Mother had a lot of guilt because she had four children with the disease."

Peggy's husband, Don Marshall, said a parent with VHL has a 50-50 chance of having a child with the disease.

"It's not like you have two children and one will have it while the other doesn't. It's like flipping a coin with each one," Don Marshall said. "In my (mother-in-law's) family, four out of five of her children got it. That's 80 percent of her children."

When a baby is conceived, the dad provides one set of genes and the mom provides another. Healthy people generally inherit two healthy copies of the VHL tumor-suppressor gene.

"Peggy is missing one. If something happens to the other gene, the tumors will grow," he said. "Most of us have two soldiers fighting a battle for us, but people with VHL only have one soldier."

Information "underload"

The Marshall family hasn't always been this informed. Like many people with VHL, the illness went undiagnosed for years.

Marshall's sister was diagnosed with glaucoma as a child and her eye was removed. The family now knows VHL caused a tumor in the eye.

"The eye does go into glaucoma when the pressure builds up," said Marshall, who also lost an eye due to complications of VHL. "It wasn't a total misdiagnosis with my sister. They just missed why it was glaucoma."

Don Marshall said misdiagnosis has been a common problem because VHL is not widely known.

"Peggy has seen specialists and said, 'I have Von Hippel-Lindau disease,'" he said. "They said, 'Excuse me' and left for a while. We know what they were doing. They were going to look it up."

Part of the VHL Family Alliance's mission is to educate family doctors about the illness.

"The biggest problem we have is underdiagnosis," Marshall said. "It often takes family members telling their doctors, 'My mother had brain tumors in her 30s and I have brain tumors in my 30s. Where's the connection?'"

New Director of Volunteer Services

In addition to the three new board members recently elected by the membership, the Board voted in July to elect Peggy Marshall to fill an unexpired term on the Board.

We had invited Peggy in the spring to run for re-election, but the timing was not right for her at that time. In July she agreed to take on a new challenge as Director of Volunteer services, overseeing all the U.S. volunteers who provide direct service to consumers. In this position she now oversees the Volunteer Service Programs:

- Hotline (which she has been chairing since 1994)
- Online Services (chaired by Maria Shipton)
- Chapters (chaired by Kathy Braden)
- Info line (chaired by Altheada Johnson)

Peggy is excited about this challenge, and we are delighted to have her back providing leadership for the services she has helped to pioneer. She is looking forward to supporting their activities and expanding our capacity to serve our ever-growing population with strength.

Peggy with her daughter Tammy

Hope for a cure

Treatment for VHL involves treating the individual tumors as they're discovered. That generally means surgery to remove or at least neutralize the tumors.

The Marshall's daughter, Tammy, has had five brain surgeries and still has tumors and cysts throughout her body.

"I have six brain tumors now," she said. "I still have little headaches."

Tammy also has a permanent partial disability in her right arm and hand that resulted from a 1991 brain surgery.

Families facing this disease hope for a drug that will destroy tumors without the need for surgery. Research in that area is ongoing, but there's no telling when such a beneficial drug could be available.

The good news about VHL is it has attracted a lot of attention from cancer specialists, according to Joyce Graff, a founding member of the VHL Family Alliance.

"People might wonder why we study a rare disease that affects a small number of people," Graff said. "I like to think the good Lord left us clues to solving the larger problems that we can find by studying these rare diseases."

Researchers are very interested in the tumor-suppressing gene that causes VHL.

"Research into VHL is helping to unlock the secrets of cancer for everyone," Graff said. "It turns out that 80 percent of all kidney cancer is caused by a change in the VHL gene, so researchers are interested."

Since it was founded in 1993, the Alliance has raised approximately $215,000 for research.

The Marshalls have been a part of that effort and are determined to press the issue and fight VHL the best they can.

"It's not so much for me, but for my granddaughter," Marshall said. "We want to stop this for her sake."

Beginning in the Eyes

For Peggy Marshall's family, the eyes have been the windows to uncovering tumors caused by Von Hippel-Lindau disease. "In my family it was particularly pronounced in the eyes," she said.

Marshall's diagnosis was too late to save her left eye. Early detection can check the growth of tumors while they're small.

Dr. Bernard Shipp, a Corinth ophthalmologist, said it's a relatively simple procedure to find tumors in the back of the eye but it involves a special test that some doctors might not regularly perform.

"It depends on the doctor you go to," Shipp said. "There's typically no symptom at all with the tumors. It's important to catch them early so they can be treated." He said someone with a history of VHL in their family should get their eyes checked once a year.

Marshall said that if tumors are found on the back of the eye, it's important not to stop there. "Tumors can show up in many parts of the body with this disease," she said. "It's important that you know what you have and do the proper screenings. If you let it get away from you, that's when you run into problems."

Magnetic resonance imaging and computed tomography scans are advised for people who suspect they have VHL.

Today, a DNA test is available to detect the presence of the gene that causes VHL. People who take the test and don't have the faulty gene don't have to worry about further screenings.

Joyce Graff, one of the founding members of the VHL Family Alliance, said people under 40 who develop tumors should consider the possibility of VHL.

"If you're under 40 and you have a tumor, there's usually some genetic cause at work," she said.

VHL is a hereditary disease but some people have gotten the illness without inheriting it. In those cases, something caused the tumor-suppressing gene to falter. Once someone has VHL, their children have a 50-50 chance of getting it.

According to the VHL Family Alliance, if you have two or more of the following symptoms, VHL could be a possible cause: vision disturbances; lack of balance; persistent vomiting, headaches, pain or weakness in arms or legs; uncontrolled high blood pressure; loss of hearing or Meniere's disease; any unexplained pain that stays in one place and lasts more than 1-2 days; tumors of the retina, brain, spinal cord, kidney, pancreas, adrenal glands, scrotum.

"These symptoms could also mean a lot of other things, so don't panic," Graff said.

For more information about Von Hippel-Lindau disease, contact the VHL Family Alliance, 171 Clinton Road, Brookline, Mass. 02445-5815. The patient support telephone number is 1-800-767-4845. The Web address is www.vhl.org, and the group's e-mail address is info@vhl.org.

Daily Vitamins Recommended

The *Journal of the American Medical Association* (JAMA) has issued a recommendation that all American adults take a multivitamin every day in order to supplement a healthy diet, keep their immune systems strong, and help stave off disease.

Since almost 80 per cent of Americans do not receive the recommended five daily servings of fruit and vegetables that would provide sufficient amounts of key vitamins and minerals needed for a healthy diet, supplementation has become extremely important, prompting the unusual move of the *Journal* recommending supplements on a daily basis.

In the article, the authors reviewed the association between inadequate intake of several vitamins and their association with a number of chronic illnesses. Their review of nine vitamins showed that elderly people, vegans (vegetarians who do not eat eggs), alcohol-dependent individuals, and patients with malabsorption are at higher risk of inadequate intake or absorption of several vitamins. Malabsorption, or an inability digestive tract of the body to derive all the nutrients from foods, is not uncommon in people with pancreatic cysts and tumors.

It is important to emphasize that the recommendation is to supplement to get to the optimal levels. Too much of a good thing is also not good. Excessive doses of vitamin A during early pregnancy, and too much of the fat-soluble vitamins taken anytime, may result in adverse outcomes. B vitamins are best taken as a group, in foods (liver, brewer's yeast, or wheat germ) or in a balance multivitamin.

Folic acid is essential for normal cell duplication, and may help prevent formation of tumors. Too little folic acid has been implicated as a cause of spinal bifida in babies whose mothers had insufficient levels of folic acid in the first months of pregnancy. Folic acid dissolves in water and is not stored in the body, so a sufficient quantity must be taken each day. Now that breakfast cereals and bread are supplemented with folic acid in the United States, the incidence of spina bifida has decreased significantly. Folic acid and vitamins B_6 and B_{12} are required for metabolism of homocysteine, and can the risk of cardio-vascular disease. Vitamin E and lycopene (the red pigment found in tomatoes, strawberries and watermelon) may decrease the risk of prostate cancer. Vitamin D is associated with decreased occurrence of fractures when taken with calcium. Calcium is essential to reduce the risk of osteoporosis.

Some groups of patients are at higher risk for vitamin deficiency and less than optimal vitamin status. Many physicians may be unaware of common food sources of vitamins or unsure which vitamins they should recommend for their patients. It is possible to get too much of a good thing, especially of the fat-soluble vitamins. But the greater risk is having too little of the essential vitamins needed to reduce the risk of chronic diseases, including coronary heart disease, cancer, and osteoporosis.

Best of all, be sure to eat your veggies! Include in your diet at least five services of fruits and vegetables every day.

Reporting "Vitamins for Chronic Disease Prevention in Adults," by Kathleen M. Fairfield, MD, DrPH; Robert H. Fletcher, MD, MSc, *JAMA* 2002, **287**:3116-3126

VHL Information grows in Chile

El Mercurio, the largest newspaper in Chile, recently published a Letter to the Editor from Pierre Jacomet, sharing his personal history with VHL and asking doctors to watch for evidence of VHL.

In June *El Mercurio* published another Letter to the Editor, from Dr. Gloria Valdes. Dr. Valdes praised Pierre's letter. "I wish to congratulate Pierre Jacomet for his efforts toward increasing our awareness of von Hippel-Lindau syndrome as well as other conditions involving pheochromocytomas … The thorough review he provided … will undoubtedly contribute to increasing an awareness of pheo-related conditions, not only in the general population, but also within the medical community."

Dr. Valdes also notes that a team has been assembled at the Clinical Hospital of the Catholic University of Chile (Santiago) where she works with a team focused on assessing and managing pheos. The team includes urologists, anesthesiologists and nephrologists specialized in pheochromocytoma. This pheo team has linked up with the Hospital Broussais in Paris (France) and the French VHL Study Group under Dr.Stéphane Richard, in order to carry out the genetic tests that will definitively diagnose the diseases that are revealed by the presence of pheochromocytomas. In closing her letter, she makes her pheo team available to Pierre, to the VHL Family Alliance and to the readership of *El Mercurio*.

This was not the result of a single letter, but of many conversations before the letter that Pierre conducted with members of the University medical community. It just shows what one committed individual can create!

Ask the Experts

Question: In discussing a forthcoming surgery, my neurosurgeon proposed to use a technique called embolization to block the blood vessels to the tumor and make it a little smaller. If that's the case, why not just do the embolization and not remove the tumor? Isn't that less dangerous? – *Altheada J., New York*

Answer: Embolization is the delivery of a material rather like glue into a blood vessel which supplies a tumor to block the blood supply to the tumor, permitting easier tumor removal with reduced blood loss. Embolization is commonly used for certain types of vascular tumors. Because hemangioblastomas are among the most vascular of all tumors, it is reasonable to think that it would be worthwhile with them.

Tumors that are ideally suited for embolization receive their blood supply from vessels that provide a significant blood supply to the tumor, and that *only* supply the tumor — not the brain, brainstem, cerebellum, or spinal cord.

Hemangioblastomas, however, rarely have a predominant source of blood from one or only a few vessels. Instead, the vessels feeding blood to hemangioblastomas are always a myriad of microscopic vessels that penetrate the tumor capsule, not a single dominant vessel, or even a few single vessels, that can be selectively blocked with benefit.

Thus, embolization has little to offer, is rarely indicated, and it may be associated with complications such as the emboli (the glue or particulate material) passing beyond the tumor and blocking a vessel that supplies the brain or other nerve tissue, causing a stroke. It can also cause the tumor to hemorrhage. The same is true for spinal tumors, although embolization may carry even more risk with spinal tumors because of the potential of blocking the anterior spinal artery and infarcting the spinal cord, causing damage to the cord due to interrupted blood supply.

In my experience with complete surgical removal of 329 hemangioblastomas in patients with VHL, we have never used embolization, or felt that it was needed.

The use of embolization alone for treatment would only provide incomplete treatment of the tumor being embolized, and would still be associated with the risks described above. In fact the embolization is only effective for the first 1-2 days because the body finds another route around the blockage.

-- Edward H. Oldfield, M.D., Chief of Surgery, National Institute for Neurological Disorders and Stroke, National Institutes of Health, Bethesda, Maryland

Coming to Cleveland?

Don't forget to come early for *VHL 101*, 9-12 on Friday morning. Come meet all the other newcomers, meet the VHLFA Directors, and get comfortable with the medical terms that will come up in the conference. Designed for family members -- doctors are welcome too! *-- see page 5 for more*

New VHL Clinic in Boston

We are pleased to announce the addition of another Clinical Care Center at the Brigham & Women's Hospital in Boston. To make appointments, please contact Dr. Peter Black's office, 617-732-6810, Fax: 617-734-8342, or Heather Galvin at hgalvin1@partners.org The Dr. Black is chairman of the Department of Neurosurgery. http://www.brighamandwomens.org/neurosurgery/Patient/VHLclinic.asp

Brigham and Women's Hospital has been ranked for several years as one of the best hospitals in the US with all the subspecialties needed to treat VHL patients. The Division of Neurosurgery has long traditions. Its first chairman, Dr. Harvey Cushing, was the pioneer of modern neurosurgery. The current chairman, Dr. Peter McLaren Black, is an authority in neuro-oncology and image-guided neurosurgery. He is joined in this initiative by Dr. Robert Friedlander, specialist in neurovascular surgery and by a group of physicians selected from the Boston community.

Participating in this clinic are physicians from the Beetham Eye Institute at the Joslin Diabetes Center, from Children's Hospital, and from the Dana Farber Cancer Research Institute. Dr. William Kaelin at Dana Farber is very active in basic research on the role of the VHL protein in the cell. Dr. Lloyd P. Aiello, ophthalmologist, is one of the pioneers of telemedicine for treatment of diabetic retinopathy worldwide.

This team has been involved for many years in the investigation of anti-angiogenic drugs for treatment of vascular tumors of the eye, and is nearing launch of an anti-angiogenesis study on VHL. VHL tumors are ideal targets for systemic antiangiogenic therapy because they are highly vascular, often multiple, and often located in brain areas with increased surgical risks. The VHL Clinic is collaborating with Dana Farber Cancer Institute (Dr. Kaelin) in a phase II clinical trial using a novel anti-angiogenic small molecule to treat hemangioblastomas of the central nervous system (CNS) and retina in VHL. This study has not yet opened to VHL patients. Further information will be posted on the VHL website when available.

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Teen Discussion Group Online

Heather C., Michigan, age 16

Teens face many challenges daily by friends and acquaintances. One thing that I face daily is my diagnosis with a disease called Von Hippel-Lindau. Many teens are diagnosed with this or they know someone who is. I did not feel that there was anything out there for us teens. That is why with the help of lots of people I created a Yahoo Group for teens with VHL.

This is a place where teens can share any questions they have on their experience with it and just talk. Right now we have about eight members. Hopefully this group will advance to the point that it is a top online support group. To join this group please send an e-mail to teenswithvhl-subscribe@yahoogroups.com.

Note to parents: The conversation is for and by teens. There are also two friendly resource people who listen from the sidelines to make sure that any misinformation is corrected: Maria Shipton from Philadelphia, and Matt N., an adult member who works professionally with teens in California.

European Leaders

Following the Padua meeting, the European leaders met Sunday to share ideas and learn from one another. The group included Luisa Guerra and four members of the Italian affiliate (two center front and three standing left), Florian Hofmann and Gerhard Alsmeier from Germany (standing in back), Joyce Graff from the U.S., two new members from Croatia interested in starting a group there (flanking Joyce), three members from Sweden (back right), Joke Jansen from the Netherlands (seated left), and three people from Spain including Karina Villar and Jesusa Martinez (standing right front), and (not pictured) Vibeke and Richard Harbud from Denmark and three genetic counselors from the U.K. willing to assist the U.K. group.

For support in Europe, see www.vhl-europa.org

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