



VHL Family Forum



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Finding Elusive Pheochromocytomas

Review of studies by Graeme Eisenhofer, Ph.D.¹

A newly developed blood test to detect potentially deadly tumors that form in the adrenal glands has been shown to be significantly more sensitive than traditional diagnostic tests. The new test provides earlier and more accurate diagnoses of these tumors in patients with an inherited predisposition to develop them, possibly preventing complications or death. The study, led by researchers at the U.S. National Institute of Neurological Disorders and Stroke (NINDS) and the National Cancer Institute (NCI), appeared in the June 17, 1999, issue of *The New England Journal of Medicine*.¹

"Although rare, these tumors are clinically important because they must be excluded as a surgically curable cause of hypertension in many of the people who develop high blood pressure," says Graeme Eisenhofer, Ph.D., a researcher in the Clinical Neurocardiology Section of the NINDS, and lead author of the study. "If the tumors are not diagnosed and removed, they can have potentially catastrophic consequences for the patient."

The mainly benign tumors, called pheochromocytomas (pheos), are sometimes found in patients with von Hippel-Lindau (VHL), a familial cancer syndrome with neurologic complications in which affected individuals inherit a predisposition to develop tumors in a number of organs, including the adrenal glands that sit atop the kidneys.

"Pheochromocytoma can occur in VHL patients as young as 4 years of age. Deaths from unsuspected adrenal gland tumors have been reported in young children in these families and it is important to make the diagnosis early so that surgical intervention can be performed," said W. Marston Linehan, M.D., NCI's Chief of Urologic Surgery.

A patient who develops pheos for any reason is at risk for dangerous and unpredictable surges in blood levels of certain adrenal gland hormones that regulate blood pressure and which are responsible for the so-called "fight or flight" responses to stress. The surges

in hormones and resulting spikes in blood pressure put the patient at risk for heart attack, stroke, hemorrhage, or sudden death.

Currently, the most reliable tests for pheos use imaging technologies, such as CT (computed tomography) or MRI (magnetic resonance imaging), which can be time-consuming and expensive and which do not necessarily identify a tumor as a pheo. Confirmatory biochemical tests are required for accurate diagnosis. Several biochemical tests are available which measure blood and urine levels of the adrenal gland hormones. But in many cases these tests are not accurate, because some pheos do not release the adrenal hormones regularly or in significant amounts. These tests depend upon catching the pheo during an active episode.

Drs. Eisenhofer, Linehan, and their colleagues studied enzymes important in catecholamine production that may have altered activity in pheos in contrast to normal adrenal tissue. Metanephrines are produced mostly by pheos, not normal tissue. Dr. Eisenhofer and colleagues found that measurements of blood levels of these chemicals makes it easier to differentiate secretion of normal tissue versus pheo. Thus these measurements give a more accurate diagnosis of pheos. A person with a normal plasma concentration of metanephrine and normetanephrine can be fairly confident of not having a pheo. Because of the high

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reliability of these tests, additional tests are not needed to rule out a pheo, significantly reducing costs.²

Altogether, they measured the amounts of normetanephrine and metanephrine in 26 patients with VHL and 9 patients with multiple endocrine neoplasia type 2 (MEN2), another rare genetic disease characterized by pheochromocytomas. Use of the new test detected 97 percent of the tumors, whereas the other tests detected only 47 to 74 percent of tumors. Although particularly useful in diagnosis of tumors in VHL and MEN2, the test also shows promise for improved diagnosis of pheos in the much larger population of patients with high blood pressure where the tumor needs to be excluded.

Symptoms. Pheochromocytomas are usually benign. They may occur in or near the adrenal glands, or anywhere along the sympathetic nervous system roughly from the base of the skull to the bladder. The most apparent symptom, caused by the increased secretion of epinephrine and norepinephrine, is hypertension, or high blood pressure. This hypertension may be constant or intermittent. Attacks may occur every few months or several times daily, and typically last less than five minutes. Physical and emotional stresses can initiate an attack. During severe attacks, patients may experience headache, sweating, apprehension, palpation, tremor, pallor or flushing of the face, nausea and vomiting, pain in the chest and abdomen. There may be a tingling, burning, or crawling sensation on the skin of arms or legs or urinary difficulties.

Testing options. The most commonly used test for a pheo is a 24-hour urine collection. All the urine is collected for a 24-hour period, kept refrigerated, and then analyzed for levels of catecholamines and epinephrine. Patients are asked to avoid caffeine, bananas, vanilla, chocolate and a lengthy list of other foods for two days before the test. Many foods can cause false positives, but caffeine is the most frequent cause of false negative results. The test is somewhat inconvenient, as you have to keep a jug of urine in the refrigerator, and you have to remember to save all urine for this period, even if you wake in the middle of the night. This test is even more difficult to perform reliably with a small child.

Pheos that occur in the adrenal glands themselves are usually the easiest to find. They usually appear quite clearly on a CT or MRI, even when they are quite small. CT and MRI are equally good at showing them. The hardest ones to find are those which occur outside the adrenal glands, in the tissue of the sympathetic nervous system, anywhere from the base of the skull to the bladder.

Dr. Eisenhofer and his colleagues compared the normetanephrine and metanephrine levels in the blood against the blood levels of catecholamines (epinephrine and norepinephrine) and the levels of

these and other chemicals in the urine. They found that the blood test was 97% accurate in detecting the presence of pheo tumors, while the other biochemical tests were only 47% to 74% accurate. All patients with MEN-2 had high blood concentrations of metanephrine, while the patients with VHL had almost exclusively high blood plasma concentrations of only normetanephrine. One person with VHL had a normal plasma level of normetanephrine. This patient had a very small adrenal tumor (less than 1 cm.) The higher the sensitivity of measurements of plasma normetanephrine and metanephrine, the more accurate the test in finding pheos.

The study recommends use of HPLC measurements of plasma free normetanephrine and metanephrine as the initial biochemical test of choice.³ To avoid false-positive results, a list of any drugs the patient may be taking should also be considered, and the patient must be cautioned not to take acetaminophen in any form (e.g. Tylenol, Excedrin, or as an ingredient in cold medications) for at least five days before the sample is drawn. It is best if the sample is obtained in the morning after an overnight fast (water and non-caffeinated soft drinks are permissible). Caffeinated or even decaffeinated coffee should be avoided for at least 24 hours before the test, and the doctor should be told if these have been taken, as they can cause higher levels of dihydrocaffeic acid in the bloodstream and reduce the accuracy of the test.

If plasma free metanephrines have been run, and they are well within the normal range, then it is highly unlikely that the patient has a pheo and there is little need for further tests. On the other hand, blood or urine catecholamines, even when performed in combination, may yield normal results when there is in fact a pheo present.

Locating the tumor. In most cases of positive biochemical results, CT or MRI scan of the entire abdomen will usually locate the tumor. However, in many cases it is also appropriate to follow up with MIBG scintigraphy -- preferably using the 123-iodine labeled compound rather than the 131-iodine labeled compound -- to establish more reliably that a located mass is a pheochromocytoma, or to locate an extra-adrenal pheo. 123-iodine is ten times as sensitive as 131-iodine. It tends to be less available because it has a much shorter half-life and therefore has to be used within 24 hours of preparation. It is most available near large university centers where they are able to do the "labeling" process in their own research facilities. MIBG-131 finds only 60% of pheos in VHL; MIBG-123 finds in the range of 95%.

Treatment. The treatment of choice whenever possible is laparoscopic adrenal sparing surgery. Since VHL patients often have bilateral pheos in the course of their lifetime, it is important to retain as much adrenal function as possible even when dealing with a single pheo.

1. Eisenhofer, G.; Lenders, J.W.M.; Linehan, W.M.; Walther, M.M.; Goldstein, D.S.; Keiser, H.R. "Plasma normetanephrine and metanephrine for detecting pheochromocytoma in Von Hippel-Lindau disease and multiple endocrine neoplasia type 2." *N.E.J.M.* 340:24 (1999) 1872-1879.
2. Eisenhofer, G; Walther, W.M., et al, "Plasma Metanephrines: Novel and Cost Effective Test for Pheochromocytoma," Proceedings of the 1st International Meeting on Adrenal Diseases, *Brazilian Journal of Medical and Biological Research*, September 7, 1999.
3. Sources of testing for plasma metanephrines:
(1) NIH will do the test without charge in the research lab, but it can take up to six weeks. For instructions and consent forms, see

<http://www.vhl.org/profinfo/pheo>
(2) Clinical testing with a 2-day turn-around is available from the Mayo Clinic by calling 1-800-533-1510 or +1 (507) 266-5700. The published fee as of 8 Nov 1999 was \$122.50. CPT code 82491. Please note that the information faxed to us by Mayo consisted of blood drawing and shipping instructions only and did not include patient instructions. You should also read the information posted at <http://www.vhl.org/profinfo/pheo>
Our thanks to Dr. Walther, Dr. Eisenhofer, Debra Harlander and Mary Peebles and the Pheochromocytoma Support Group for their assistance with this article and for their ongoing partnership in support of people with pheos. See <http://members.aol.com/ThreePeb/indexpheo1.html>

Coping with Neck and Back Pain

by Deb Hogan, P.T., Massachusetts

Editor's Note: Please note that unexplained neck and back pain should first be checked with your neurologist or neurosurgeon, to determine that there is no tumor that could be causing the pain, and that might be worsened with manipulation. Always check with a neurologist before undertaking any chiropractic treatment or physical therapy on the neck or spine.

Eight out of ten people suffer from neck and back pain. It is the second highest reason for a doctor's visit, behind respiratory infections, and the leading cause of worker absenteeism. I am a Physical Therapist (P.T.) specializing in the care of neck and back injuries and I also have VHL. Having worked as a P.T. for over 10 years and having just been diagnosed with VHL, I laugh at the irony of now suffering from neck and low back pain! I certainly have empathy for my patients!

Neck and Low Back Pain (LBP) is common among patients with VHL because of the central nervous system (CNS) tumors, scar tissue from spinal surgeries and the stress that comes with a chronic illness. I remember as a child, watching my mother apply BenGay¹ to her aching back. She wore a brace with metal stays for years. She would awaken early in the morning, take her pain medicine, and then go back to bed to wait for its effects. This is the way she managed her pain so that she could support me, as she was a single parent.

How do you manage your pain? Do you use heat or ice? Is stretching helpful? How about sleeping? Is your pillow a problem?

As a Physical Therapist and as one who suffers from VHL, I would like to provide some helpful hints for managing neck and LBP.

1. Once your pain becomes chronic, using heat or ice is a matter of personal preference. I use a hot water bottle 1-2 times per day at the base of my neck. Heat relaxes tissues and increases blood flow. Ice, on the other hand, is a fantastic analgesic, if you can tolerate the cold!! Ice decreases circulation and keeps swelling down. Try heat or ice instead of pain medicine to manage your pain.

2. I teach all of my patients proper posture and body mechanics. Your head position is very important. Your head weighs 10-12 lbs. If your head is in front of your shoulders, during computer work for instance, the weight of your head increases 3-5 times (as far as your muscles are concerned). So the muscles that support your head are supporting the weight of a 3 year old child instead of supporting an infant. This will cause fatigue and pain. Keep your head back so that the hole in your ear is over your shoulder. Practice an exercise called a "chin tuck". Think of the chickens in the farmyard, clucking and moving their heads back and forth. They are doing chin tucks ... tucking the chin backward (not looking up). This is a helpful exercise to alleviate neck pain and normalize posture.

3. Over half of my patients complain of pain in the morning. Some of this is related to sleeping posture --sleeping on the stomach or sleeping in a 3/4 turn that twists the low back all night. How does this cause low back pain? Try putting your leg up on a chair to stretch the back of your thigh. Stay that way for several minutes and then try to move your leg! This pain is similar to the pain we experience from poor sleeping positions held over many hours. Lying on your side with a pillow between the knees, or on your back with a pillow under your lower legs, are two great sleeping postures. Avoid reading in bed or using too many pillows to sleep. Try rolling up a towel lengthwise and placing it inside your pillow case. This adds extra support for your neck.

4. A stretching program is important, as is regular exercise. This program should be given to you after a thorough evaluation to see what muscles are weak and where you are lacking in Range of Motion.

Those of us with VHL have chronic issues that have no easy answers. I find that as I take my mind off myself and focus on others, I am helped; As I keep a positive attitude, I am better able to cope. Prayer and meditation help me relieve stress. Pain does not have to keep you from living and contributing. The choice is yours. Just remember that you are not alone.

1. A U.S. brand of mentholated cream for muscle aches

One More Round

by Peggy Marshall, Chairman of the Alliance

Little did I know when I wrote my last article for the *VHL Family Forum*, "Up For the Fight", that I would have brain surgery in such a short time. On September 9, 1999, I had bilateral brain surgery on the right cerebellum to remove a hemangioblastoma and on the left side to repair a spinal fluid leak from a previous surgery.

The symptoms were sudden and distinct and I knew there was increasing pressure. We know that each manifestation of VHL is different and symptoms vary. But I believe each of us knows our own body and its functions better than anyone. If something is not right, we know.

Do I practice what I preach (as the old saying goes)? You bet I do! I called and made an appointment immediately. Is it ever easy to hear you need surgery? Although I dread those words as so many of you do, once we hear it is necessary, the choice becomes easier. Early intervention is important many times in determining the outcome.

I want every one in my VHL Family to know that my surgery was very successful, recovery has been quicker than one could hope for and physically I have more energy than I have had for years. My neurosurgeon remarked that my balance is better than it has been for years. We agreed that the spinal fluid leak had caused an accumulation of spinal fluid that put pressure on my brain stem resulting in various symptoms, such as difficulty with balance, weakness in right arm and leg, and intense tiredness.

I learned some interesting things from this surgery. When I called to make an appointment the nurse remarked that my symptoms probably were not coming from the tumor. I was not sure what the cause was, but knowing there was increased pressure, I pushed for a MRI. I also know from experience that symptoms may not necessarily match what the medical professionals feel is causing the problem. There is much to learn about the body's neurological functions. I think we have to be strong participants in our health care and not dismiss possibilities too quickly. My neurosurgeon was not available to review the scans with me the day of the MRI. The radiologist who reviewed the scans did not see the accumulation of spinal fluid. I insisted that my neurosurgeon review the scans later that week. The neurosurgeon recognized the problem and worked with me to schedule surgery. I also had a relatively small tumor that he felt could be easily removed at the same time.

After my earlier brain surgery in January 1996, I had ongoing difficulty with balance and fatigue along with severe spinal headaches. It was four months before I could even sleep in a fully reclined position.



My recovery had been long and difficult due to brain swelling before and after the surgery. Follow-up MRI's indicated the leaking spinal fluid was decreasing and it was decided that nothing further was indicated. My neurosurgeon told me my balance was as good as it was going to ever be. I accepted his opinion and learned to compensate in many ways, pacing my activities during the day so I would not be overly fatigued in the evenings. I remember hearing comments from friends and family such as "You are not as young as you used to be" and "If I did what you do, I would be tired, too". I have taken care of children for over 20 years and rarely sat down. I did not understand why, all of a sudden, I was so very tired.

After the surgery this past September and the repair of the spinal leak, I was given a new lease on life. It was not my age or my job. It was displaced, trapped, leaking spinal fluid. Now I am the person I was before my surgery in 1996. I learned that if deficits occur, it does not necessarily mean they will be permanent. And it is perfectly within reason to question your medical professionals about treatments. Do not ever give up!

After three and one half weeks from the date of surgery my neurosurgeon cleared me to go back to work. I think even he was surprised. It has been truly remarkable!

Even though VHL can manifest itself at any time, I feel very strongly that a well informed patient and medical team can help us through each episode with a better outcome.

Don and I are counting our "Blessings" and God has answered a multitude of prayers on my behalf. We want to thank each of you who offered your love, support, and prayers the past few weeks. God has also blessed us with a loving; caring "family" that truly understands what we are going through.

Special Children, Challenged Parents

"I wrote *Special Children, Challenged Parents* to deal with issues that no other book about disability deals with, especially a major male dilemma: Men, including myself, have a hard time facing things they can't fix," says Dr. Robert A. Naseef. "Men feel powerless, inept, sterile. We can't simply work harder to fix our child's disability. Instead, as we struggle with the grief and darker side of what it takes to face our broken dreams about our child, we must deal with our powerlessness." According to Dr. Naseef, "Fathers as well as mothers can go through a process that leads from grief to hope, from isolation to relatedness, from emptiness to fullness, and from shame to pride. This process can be long and arduous, but it can also be heartfelt and inspiring."

Special Children, Challenged Parents is a singular book because it offers the views of a man who is a psychologist and the father of a child with autism. This volume addresses the issues of all parents with special-needs children, and it focuses specifically on the issues and needs of fathers. *Special Children, Challenged Parents* not only helps men to open up in their own way, it also helps women to get a glimpse inside the psyche of men. Couples can also use this book to connect and communicate better when their marriages seem ready to crumble from strain.

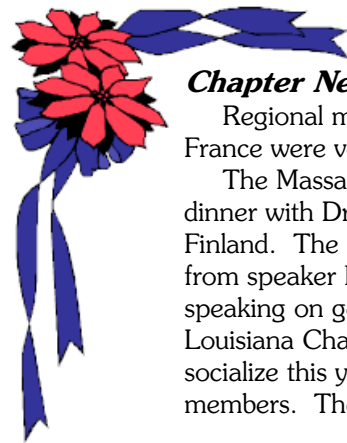
“ Men, including myself, have a hard time facing things they can't fix. We can't simply work harder to fix it ... we must deal with our powerlessness. -- Robert A. Naseef, *Special Children, Challenged Parents* ”

In the words of James May, Project Director, National Fathers' Network: "With enormous insight and eloquence Bob Naseef blends his understanding as a psychologist with his real life experience of raising a son with autism. The result is a book which moves and teaches us at the same time--an intelligent, authentic, and impressive achievement."

Joan F. Goodman, Ed.D., University of Pennsylvania Graduate School of Education and author of *When Slow is Fast Enough*, had this to say: "Never have I read a book that combines so profoundly the experiences of a father—agonizing, embarrassing, frustrating, exhilarating, sometimes even blissful—with the insights of an experienced therapist and the wisdom of a scholar."

The book is receiving praise from many disability organizations. The National Psoriasis Foundation wrote that "the honesty and compassion with which it is written translates easily to any condition."

For more information about *Special Children, Challenged Parents* you are invited to arrange an interview or to request a press pack, respond to this message or contact the author. The book is available through your local bookseller or through the VHL Online Bookstore <http://www.vhl.org/bookstore>.



Chapter News

Regional meetings in Germany and France were very successful.

The Massachusetts Chapter had dinner with Dr. Mika Niemelä from Finland. The Indiana Chapter heard from speaker Margie Bryan, R.D., speaking on general nutrition. The Louisiana Chapter took time to socialize this year, sharing ideas among members. The Minnesota Chapter

met in Bloomington. The New York chapter welcomed Theresa Iodice, Physical Therapist from the International Center for the Disabled, who worked with members on relaxation techniques. The Tennessee Chapter heard from Jay Platt at their meeting in Maryville, near Knoxville.

Dan Riconda, Genetic Counselor, (see at right) from Arnold Palmer Hospital for Children & Women, spoke to the Florida Chapter at their meeting in Orlando.



In addition to the printed booklets, the Handbook is now available in Adobe Portable Document Format (pdf) in English and Spanish so that it can be copied over the internet and printed on any printer.

"Your site has been valuable for us...great job! I especially love the handbook. The PDF format does work well for us, thank you!" -- Heidi Sue Adams, MS, Kalispell Regional Medical Center - Medical Library, Kalispell, Montana

Finding Hope Within

by Amy G., Arkansas

"That is why we never give up. Even if our bodies are dying, our spirits are being renewed every day." -- *2 Corinthians 4:16*.

I've lived with von Hippel-Lindau (VHL), a rare (or rarely diagnosed) familial cancer, for 15 years. With VHL, abnormal blood vessels grow in little knots of capillaries called angiomas, or hemangioblastomas, which can cause problems. The disease is different for everybody. Mine has resulted in two brain surgeries, spinal surgery, two partial nephrectomies, and an eye laser treatment.

Everyone leads a complicated life, even without the variables of chronic illness. Those living with a multi-symptom disease, however, have times where we envy those with a cancer that is well researched. VHL has no middle, just a beginning and an end. Symptoms come in spurts. Sometimes they crop up after years of inactivity. Sometimes they appear several times within a year. People die when their bodies become overwhelmed by disparate tumors and problems associated with the disease.

Because VHL has so many varied symptoms, it is difficult to condense into a narrowly worded definition, and even harder to diagnose. Over the years, doctors in good faith have given me wrong information. The depth of medical knowledge on VHL is depressingly slim; even today, many doctors still have not heard of it. While I was suffering from my first brain tumor, my doctor insisted nothing was wrong. I told him I thought I had a brain tumor. I'll never forget retching into a specimen cup while he told me "70 percent of his female patients had psychosomatic illnesses." I checked myself into the hospital days later and told a resident what was happening. He ordered a CT scan, which confirmed my suspicions. I didn't feel vindicated. Just grateful. Sometimes half the battle is finding a doctor willing to listen.

I've survived episodes of illness, surgery, and recovery while remaining stubbornly fixed on the ideal

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Sometimes half the battle is finding a doctor willing to listen.

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of getting back some semblance of a normal life. Throughout the years, it has become up to me to redefine "normal." I've learned what I can about the disease, joined an information and support group, and interviewed dozens of doctors to form the right medical team. In the process, I've learned fundamental truths. I can stand up to doctors, technicians, and surgeons. When I take myself seriously, I gain more

self-assurance. And, I am incapable of using a bedpan, even if it means waiting ten hours before going to the bathroom.

I've lost friends and a first marriage to the cracking-up-sorting-out-coming-to-terms-baggage that accompanies anyone who lives with chronic illness. Gone are the certainty of bearing a healthy child ... the ability to continue the athletic pursuits I love ... a level

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To have hope you must find hope within -- often at the precise moment in your life that hope appears most fleeting.

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of independence and privacy ... at times, personal dignity and modesty ... and the privilege of what I call false positives -- such as taking for granted the fact that you will wake each day and your body will work effortlessly.

At times, I mourn the loss of self-assuredness that preceded this illness. It's hard weathering things that most people normally do not confront until old age -- or, if they are lucky, never at all. But self-pity can be such a bore. After awhile, I force myself to enjoy life, even if I have to pretend a little. Then one day, I begin to re-appreciate all I have and what I can do. I find activities and hobbies that keep me active and engaged. I resolve feelings of meaninglessness and loss and move on. I become balanced and happy again.

Despite knowing this, last summer I lost my sanity for awhile. I had spinal surgery and it left me with unrelenting pain. After months with no end in sight, I went a little crazy. It surprised me. Being the veteran of five previous surgeries, I thought I had gone through just about everything. As time dragged by, I realized with a sinking heart I had not. I was unable to sleep more than an hour at a time before pain forced me out of bed. Unable to tolerate much of anything, I limped around the house all hours of the day and night until I grew so exhausted sleep overcame me.

When my disease had flared up in the Fall of 1997, I became distraught and frightened. The spinal surgery had been hanging over my head for years. The doctors had wanted to hold off as long as possible, for fear the surgery might leave me paralyzed. I was bewildered. With my husband sorting through the myriad tasks of making airline reservations, scheduling doctor visits, and working through the red tape of the insurance company, I went about my daily obligations and arranged my work schedule. In between, I tried to nurture my embattled body. Now I look back and see it was wrong to squeeze concern for myself in between work demands and the realities of life. Today, I know I should have stepped back, breathed deeply, and

carved out more quiet time to absorb and confront the illness, and appreciate and nurture my body. Eventually, time, faith, physical therapy, and medicine sorted it out. But I had to re-realize past lessons learned. The luxury of perspective helped me remember.

To have hope you must find hope within -- often at the precise moment in your life that hope appears most fleeting. When I first learned of my chronic illness, the hope that sprang within was born of courage and determination; not just to survive, but to prevail. I have gained so much the scales have tipped in counter-balance. My marriage and relationships with family and friends are more authentic. I've walked through fire and am purified and humbled by the experience.

I know my will power and inner strength is ironclad when I must call upon it, and my spirituality is growing stronger. I am constantly learning how to appreciate, develop, and nurture this gift. Because this lesson was not learned easily, it is all the more precious. Finally, I vow that this disease will never define my life. But it does *refine* it.

Ending the Madness

Chunks of me are taken out
with Pac Man precision
several holes in my brain, my spine
nibbles to both kidneys
a black laser hole in my eye
Sometimes I feel my soul
leaching out of the openings
a precious resource leaching
from my very bones
rendering me tired and helpless
like an old clock winding down
When I am feeling stronger, however,
I think my spirit plugs up these holes
and the armor it grows
in my skin and bones supports me
I will combat this evil that grows
unwelcome and unwarranted tumors
and cysts, it won't defeat me
The trick is to end the madness
while continuing to generate love for the body
leaving bones, tissues, and organs intact.
-- AGL

This essay was prepared for a contest among cancer patients offered by the Thsepong project, a fundraising project for the Cancer Association of South Africa (CANSA), Pretoria, South Africa. Thsepong means "place of hope" in South Sotho.

Where did you hear about VHLFA?

The most favorite answer heard on the hotline is "I got your number from Excy's chest!" She copied it off his T-shirt at the beach in North Carolina.



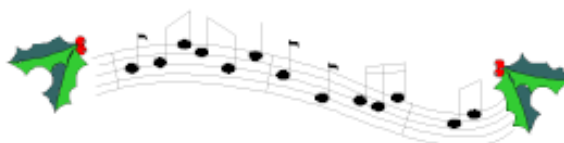
Amy and her husband Excy in the dappled light of afternoon in their back garden.

Resources

There is an excellent multi-media glossary of **genetic terms** available at <http://www.nhgri.nih.gov/DIR/VIP/Glossary/> In addition to written explanations and pictures, there are also audio explanations.

United Resource Networks offers a continuum of products and services designed to help people who have received a **transplant** manage their transplant benefit. From the nation's leading transplant network, to medial appropriateness review services, United Resource Networks offers clients the opportunity for improved outcomes and significant cost savings associated with transplantation. See <http://www.urnweb.com> or call 1-800-847-2050 (U.S.)

Our **VHLFA German affiliate**, Vereins für von der von-Hippel-Lindau (VHL) Erkrankung betroffener Familien e.V., has a beautiful new homepage in German, thanks to the efforts of member Andreas Beisel. There is an online discussion group vhl-de@egroups.com, and chats are held in German about once a month. See <http://www.hippel-lindau.de>



Stress -- More or Less?

How Stress Can Work For, Not Against You

You've heard it all -- stress is bad for you, stress can be good for you, too much is distressing, a little a blessing -- what does it all mean? The right amount of stress is good for you -- but what's the right amount?

Understanding Stress

First, it helps to know that stress is just our bodies' reaction to a new situation. Our bodies react in the same way whether the situation is welcome or unwelcome. Our hearts beat faster, our bodies produce a natural stimulant called adrenaline, we breathe more rapidly, we are more alert, and we may perspire more. All of these responses can be good for you when followed by periods of relaxation. They can give you the "competitive edge" in sports, keep you alert and concentrated for a test, or can literally save your life in an emergency. But, when your body is constantly wound up and can't relax, these same responses can lead to increased blood pressure, anxiety attacks, and a wide range of physical disorders.

How Much is Too Much?

There's no set formula for figuring out how much stress is too much -- one person's stress may be another's "motivation." However, if you experience stress-related symptoms such as insomnia, appetite changes, excessive fatigue, depression, headaches, stomach ailments, or muscle tension, you may be feeling the effects of an overly stressed lifestyle -- all stress and no relaxation. The key to bringing the stress in your life to a manageable level is to try to avoid or eliminate unnecessary stressful situations. And, when you can't avoid them, to be sure to balance stressful experiences with periods of relaxation.

Eliminating Excess Stress

Some stressful situations can be eliminated by creative problem-solving. For instance, if long lines at the shopping mall drive you up the wall, consider shopping from catalogues, over the phone, or during "off" hours. You might even consider turning "wasted" time standing on line (or sitting in traffic) into extra leisure time -- bring along a book, a magazine, a crossword, or even your knitting. Listen to the radio or bring along a cassette player and a favorite tape. Or, take along a pad and pencil and try your hand at sketching. You may not be able to avoid all stressful situations, but you can turn many into positive experiences.

Making Stress Work for You

There will always be minor hassles (and major ones, too) that can't be avoided, eliminated, or

changed. But they can be balanced by periods of relaxation. You can break the cycle of continuous stress by planning time for relaxation. Five minutes of stretching, deep breathing, a warm bath, uninterrupted silence, may be all you need to break the pattern of tension and fatigue. When you set aside time to relax, you'll feel refreshed, revived, and better able to handle the situations and challenges you face each day.

Adapted from an article from Parlay International.

Eight Gifts

That Do Not Cost A Cent

1) *The gift of Listening:* But you must REALLY listen. No interrupting, no daydreaming, no planning your response. Just listening.

2) *The gift of Affection:* Be generous with appropriate hugs, kisses, pats on the back and handholds. Let these small actions demonstrate the love you have for family and friends.

3) *The gift of Laughter:* Clip cartoons. Share articles and funny stories. Your gift will say, "I love to laugh with you."

4) *The gift of a Written Note:* It can be a simple "Thanks for the help" note or a full sonnet. A brief, handwritten note may be remembered for a lifetime, and may even change a life.

5) *The gift of a Compliment:* A simple and sincere, "You look great in red," "You did a super job" or "That was a wonderful meal" can make someone's day.

6) *The gift of a Favor:* Every day, go out of your way to do something kind.

7) *The gift of Solitude:* There are times when we want nothing better than to be left alone. Be sensitive to those times and give the gift of solitude to others.

8) *The gift of a cheerful disposition:* The easiest way to feel good is to extend a kind word to someone, really it's not that hard to say, Hello or Thank You.

Friends are a very rare jewel, indeed. They make you smile and encourage you to succeed. They lend an ear, they share a word of praise, and they always want to open their hearts to us. Show your friends how much you care.

Anniversary

by Charlene W., Massachusetts

Well -- today is a special day. As Mark so kindly pointed out to me this morning, this is the one year anniversary of my cancer diagnosis! A whole year ...

By the way -- I mentioned this in my support group this morning, and it was the unanimous decision that, since this is an anniversary, I should **celebrate!** Now, Mark, I know you are really busy and probably won't be home until late tonight, so I won't insist on dinner and dancing. Flowers might be nice, though!

Anyway, since it is a significant milestone for me, I thought I would set down a few of my thoughts about this incredible year. Now I don't often say it, but I will admit that there has been a lot of hell in my life since this diagnosis, but that isn't what I want to think about today. Despite the pain and fear and horror, I feel like I have gained a lot too, and that is what I want to share.

I feel rather like, in the car trip of life, that this last year has been a wierd detour. I was suddenly sent off the main highway onto an unknown road, without any warning at all. (Well, okay, there was warning, but I couldn't read the signs.) It is a twisty road and there was no way to turn around. I don't know where it comes out, or if it ever even joins the main highway again. It is not a road I would choose and I am still not thrilled about being there. However, even though I would never have chosen this road for myself, I can't help but notice that along the way there are some glimpses of unbelievably beautiful scenery. And I find myself driving a little slower, and admiring these views a little more, and I think when I get to wherever this road is leading me, I will like that view a whole lot better for what I have seen here.

I have also learned a lot in this last year, about myself, about others, and about life in general. I know I can't put it all into words, but I would like to share some of the most important lessons with you:

- You never know how much you can stand until you are put to the test.
- It is okay to cry in front of other people and to let others cry in front of you.
- Being "just human" is okay -- even when others seem to expect you to be superhuman.
- Even in the darkest moments, you can always find something to laugh at.
- Sometimes the only way to take control of your life is to let go of all control and put yourself into someone else's hands.
- When you have mouth sores, grape jello is truly the food of the gods.
- At 3 am, a kind-hearted nurse truly is an angel.
- Doctors are human beings -- just people like the rest of us.

- It is okay to ask for help.
- Don't ever leave anything white on a hospital bed, it *will* get carted away with the laundry.
- There is no way to be sneaky or tip-toe quietly when you are wearing floppy slippers!

Finally, the most important thing that I have learned this last year is that there is a lot of love out there in this world. I always knew that, but until my life changed with this cancer I never really experienced it. For a very long time, I have tried very hard to love people, but a part of me was always afraid to open up and let people love me back. I was always afraid of being hurt or rejected, or worst of all, letting the love in only to have it go away. Of course, there were always some exceptions -- thank you, Mark, my angel! -- but those were hard-fought battles.

After I was diagnosed with cancer, my whole world changed. Suddenly I found that I couldn't do it all by myself and that I had to depend on people -- both physically and emotionally. So, for the first time since I was a child, I just let go of the fear and let people in. What a revelation! Not only didn't the love go away, it got stronger. In letting people love me, I was able to learn how to truly love them. In opening up myself to the world I was able to let the world open up to me. And that truly is the greatest give this disease has given me. I like myself better, I like the world better, and I think I have learned how to really love.

This is not to say that I still don't have bad days, that there aren't times when I just want to throw a pot at anyone who walks into the room. It just means that now I can feel like that and still know that the love will be there when everyone comes out from cover -- and that I will still be able to love when the pots come flying at my head.

And now, I know I have said it before, but today it means even more -- thank you to each and every one of you for being there with me this last year! Thank you for all your prayers and all your love -- and every single kick in the pants you have given me when I deserved it. And most of all, thank you for opening yourselves and letting me love you the way you have loved me.

Here's to many more years of love and lessons!

for a long time
it had seemed to me that life was about to begin
real life --
but there was always some obstacle in the way
something to be got through first
some unfinished business
time still to be served a debt to be paid
then life would begin
AT LAST
it dawned on me that the obstacles
were my life

- b. howland

Ask the Family

Stereotactic Radiosurgery

Question: What's it like to have stereotactic radiosurgery?

Answer: Went in for stereotactic radiosurgery on Tuesday, 7 Sept, 1999. This was my third stereo since my 1994 diagnosis. Outside of some scheduling glitches with the MRI machine, the day proceeded very smoothly. I was escorted into my prep room at about 9:30, and was released from the hospital at 4:30. Two tumors were treated during this visit.

I once thought that having the frame attached was the worst part of the whole day, but that was pretty ho-hum. I've recently been having some back soreness, though, so it turned out that lying on that uncomfortable metal table during the procedure was a real challenge.

Outside of some fatigue after returning home that evening, which dissipated with a good night's rest, I was 100% back to my regular schedule the next morning, including a full day's work.

These were two tumors: one 12.5 mm and the other 6 mm. Both were comparatively larger than the previous scan. Neither one had a significant cystic component. I have no symptoms from these tumors. The strategy here is to stop their growth before they become symptomatic.

I have heard stories of people having less than glowing reports of their stereo experiences. I write just to add my voice to the mix. As long as this procedure keeps my tumors from becoming a problem, it is a welcome inconvenience. My experiences would prompt me to recommend the approach to others whose diagnoses fit the stereo profile.

-- David I., California

Heart Problems?

Question: Has anyone ever had problems with their heart after being diagnosed with VHL, such as with heart palpitations, chest pains and such? If so, did these symptoms start after surgery or before? There are two of us diagnosed with VHL, who are not related, who have these symptoms. We wanted to know if anyone else is experiencing these problems. -- Maria S., Pennsylvania

Answer: I have VHL and have had things occur after a brain stem surgery which seemed like symptoms of improper heart function. Heart palpitations were actually heard while I was in the hospital (felt by me too!) and were rather frightening. Several tests were done to be sure there was nothing serious going on. I was told that they would not hurt me; that I should let the doctor know right away if I had any changes or other symptoms of heart trouble and that the symptoms might just go away. Well, they lasted a

few more weeks, gradually lessened and eventually went away.

Since a brain stem surgery eight years ago, I also had (and still get) fairly severe chest pains -- actually an extreme sensation of pressure (like a truck on top of your chest) is a more accurate description of what it feels like for me. Early on, I had been rushed into emergency several times, hooked up to EKGs and kept overnight for fear of heart attack because my blood pressure (BP) was very also high at the time. I'm still here -- no heart attack to this day. It seems the BP went up because of the fear and the pain, along with the fact I had a problem (that took years to identify) of widely fluctuating hypertension from a tiny bit of damage to the brain stem during that surgery. Pheos were ruled out with testing. My GI system was greatly affected by the surgery to the brain stem, and certain spots along the GI trail simply do not work right. The esophagus is part of the GI system, and what happened was esophageal spasms. They apparently can feel just like a heart attack can, according to the doctors. Now when it happens I think: "Darn it", try to relax a bit so I don't make it worse, and go about my business. Of course a quick prayer that it won't really be a heart attack this time still does cross my lips on the longer episodes. But tests today show no heart damage, so I probably need not worry after all this time.

Everyone is different! If you feel you may be experiencing heart problems, then you *must* get thoroughly checked out immediately by well qualified doctors to be *sure* it is not something serious. Good news will help put your mind at rest before you give yourself a heart attack worrying about it! Talk to your doctor about your concerns. Heck -- talk to several of them. It never hurts to get other opinions when you are very concerned about such an important issue. -- Melanie M., Massachusetts

Pain Control

Question: One of our members suffers from a great deal of pain. She is already using morphine adhesive plasters, and these don't always help. Does anyone out there have any experience with pain treatment? -- Richard H., Denmark

Answer: In regard to your friend that has so much pain, it reminds me of my recent experience after spinal surgery. For almost two weeks after surgery, I had some nasty back spasms that I was told were unusual in their duration and intensity. In the end, I am not sure that there was one magic answer, but I did learn one very valuable lesson about pain management....

While it is important to listen carefully to all the medical doctors have to say about your own particular situation, there are also a number of other approaches

you can take on your own. Relaxation therapy, acupuncture, meditation, music, etc. can all have very real results in pain management. There are many sources of information about this to find guidance. -- Larry S., Pennsylvania

Ask the Experts

Organ Donor Registration

Question: When I went to renew my driver's license this year I was handed an organ donor registration card. I would like to think that when I die someone else might benefit from a few of my "spare parts" -- but I'm not sure whether VHL would rule me out as a donor. I know that my kidneys would not be right for anyone, but what about corneas, skin, heart? What should I do with this card? -- Tania D., Canada

Answer: The best advice is to sign your donor card and ensure that your family members are aware of your desire to donate, and are willing to facilitate the donation when you die.

Each patient is evaluated on a case-by-case basis at the time of their death for all donation options (research and transplantation). The Organ Procurement Organization (OPO) will be called and will evaluate the decedent's medical condition at the time of death. They will discuss all available donation options with the family (or decision maker) at the time of death.

What can be donated will vary depending on the cause and manner of death. Medical/Social history will also play a role and we would consult with the next-of-kin or designated decision maker to determine medical suitability. Tissue bank and Eye bank criteria vary depending on the bank with which the hospital is affiliated.

Remember tissue criteria is much stricter than organ donation. Tissue is life enhancing ... organ donation is life saving...

A surgeon or patient may choose to take an organ that is less than perfect if the patient's only other option is to die. With tissue, on the other hand, we want to limit the risks, since the decision to receive a tissue transplant (cornea, bone, skin) is usually not life threatening.

-- Lori E. Brigham, Executive Director, Washington Regional Transplant Consortium, +1 (703) 641-0100 Lori@WRTC.org

Editor's note: Remember too to register with the VHL Tissue Bank, to donate tissue needed for research. Pre-registration shortens the process of donating surgically removed tissue, or of communicating with the Tissue Bank when someone dies. Even if your tissue does not qualify for transplantation, it will be very helpful for research. Call 1-800-847-1539 U.S. or see <http://www.vhl.org/aboutvhlfa/tissue.htm> to register or to make arrangements.

Mutation Numbering Scheme

Question: In newer papers there is a new numbering system, where the first amino acid of VHL (codon) is number 1. I understand that the original numbering scheme (with 505 mutations and 712 mutations) was based on the initial cloning of the VHL gene which did not precisely describe the start and stop markers of the gene. Since many families who participated in the early studies know their mutation best by the old numbers, how do we convert from the older to the newer notation? -- Joyce Graff, Mass.

Answer: It is quite simple. If you talk about amino acids (AA), you need to use the formula: old AA - 71 = new AA. If you talk about nucleotides, the formula is: old nucleotide - 213 = new nucleotide. It takes three nucleotides to make one amino acid, so there are three times as many nucleotides as amino acids.

For example, if you talk about 505 (old nomenclature) it is now nucleotide 292 corresponding to AA 98 (292/3). 712 (old nomenclature) is now nucleotide 499 or AA 167 (499/3).

I hope that my answer will help you to understand our new data.

-- Christophe Bérout, Ph.D., Genetics, Necker Hospital, Paris, France, and manager of the Universal VHL-Mutation Database on the internet at www.umd.necker.fr

Chat Schedule

Online discussion groups are up and running in English (107 members), German (38 members), Spanish (8 members), and French (7 members).

VHL Chats are being held in English and German. Please see the calendar at <http://www.egroups.com/lists/vhl-de> for the schedule of chats in German.

English-language chats will be held at the following times in the next three months at <http://www.egroups.com/lists/vhlfa>

You must be a member of the eGroup in order to participate. Please note that while we guard people's confidentiality as much as possible, there is only moderate security at eGroups.

If you are very concerned about your privacy, you may wish to create a free e-mail account as an alias.

12 Dec, 2 pm: Adoptees with VHL, Ann Hughes

19 Dec, 8 pm: Taking care of yourself, Dr. Gary Wood

26 Dec, 8 pm: Family, with Dr. Gary Wood

9 Jan, 2 pm: How VHL influences sexual feelings in a relationship

23 Jan, 8 pm: Getting over Brain Surgery

6 Feb, 2 pm: Support for the "unaffected" spouse

20 Feb, 8 pm: Pain management

5 March, 2 pm: Pros and Cons of DNA Testing

19 March, 8 pm: Dealing with Denial

26 March, 2 pm: Open Forum

This roster is subject to change. Please confirm the schedule in the online calendar at eGroups "vhlfa"

Update on Clinical Trials

Editorial, by Joyce Graff

Clinical trials are always bumpy roads. While the announcement from Dr. Kaelin (see box) is disappointing, we should point out that Novartis is being *very responsible* with this drug. This is not the only drug awaiting trial. There are alternative drugs "in the pipeline" that we expect to pick up where this one left off, but it will slow down the process a bit.

The patients taking PTK are doing fine. The snag came in Novartis' long-term testing on dogs. Not every drug is put through long-term testing this early, so it is very much to our benefit that this long-term testing be done. A few of the animals developed complications after long-term use. They are now evaluating which of the related drugs also in preparation might have the same positive effects without the negative side-effects.

Please remember that this is **not** the only drug, that there is still plenty of hope on the horizon, it's just always a bumpy road. This is what drug testing is all about -- better we should find out the downside now among other animals rather than find out 10 or 20 years from now when someone we thought was doing well winds up with some unwelcome side effect.

The data from Phase 1 trials of this and similar drugs are relatively reassuring, with only a few mild side-effects from the current treatments, and no hemorrhages (always the biggest fear). Because of this, there is hope that we can get permission to involve people with VHL at earlier stages in the testing of subsequent drugs. This may mean that there will not be very long delays for the people in the "compassionate exemption" category who were anxiously awaiting the start of this trial because their condition is imminently life-threatening.

If you need additional information about the Novartis PTK78 trial, please contact Dr. William Kaelin, Dana Farber Cancer Research Institute, 44 Binney Street, Boston, MA 02115 USA; +1 (617) 632-4747 (Sarah); Fax: +1 (617) 632-4381; E-mail: sarah_alwardt@dfci.harvard.edu

Meanwhile, the Sugen trial continues, and Sugen is open to enrolling additional local physicians to allow local administration of SU5416. Long-term studies have not yet been completed for SU5416 (scheduled for 2000), but SU5416 is now in Phase 3 for VHL and other conditions. See article in VHLFF, September 1999 (<http://www.vhl.org/newsletter/vhl1999/99cbclin.htm>). Early feedback from two people with VHL is that there is no perceptible tumor shrinkage, but it's early yet. Linda W., whose primary problem was pressure on the optic nerve that was steadily reducing her vision, reports that her vision is much improved even though the tumor does not show much if any shrinkage; Geoff M. reports that his symptoms have not been relieved. So there are no clear results at this time.

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October 18, 1999

Dear Sir/Madam:

This letter is to update you on the Novartis PTK787 trial. Unfortunately, there have recently been some unexpected findings in ongoing animal toxicology studies. It is not known whether similar side effects would be observed in humans.

To date, the people treated with PTK787 have tolerated the drug well. No new patients will be treated with PTK787 and those who were being treated with PTK787 have been told to stop, until the results of the animal studies can be interpreted in greater detail.

Where do we go from here? If studies are resumed, we will probably allow VHL patients in Phase I if they have progressive symptoms for which there are no standard therapies. Otherwise, they will be advised to wait until phase II. For patients who can not, or should not delay treatment, there is the option of treatment with other anti-angiogenesis agents. It is not known whether these other agents will show similar results to PTK787 in animal studies. I am sorry to have to disappoint you with this news. Pitfalls emerge in bringing any new drug to the clinic, especially drugs with novel or new mechanisms of action. I am still confident that some form of VEGF inhibitor can be successfully developed and that such a compound might help patients with VHL disease. I will write you again as I learn more.

Sincerely,

William G. Kaelin, Jr., M.D., Associate Professor of Medicine, Dana-Farber Cancer Institute and Harvard Medical School, Assistant Investigator, Howard Hughes Medical Institute

SU5416 has to be administered intravenously twice a week in 4-week cycles. We do not yet know the consequences of going off any of these drugs. Also, we have noticed that the restrictions in entering trials (e.g. SU5416) may involve excluding people who have been on another anti-angiogenic therapy trial, so people should not rush to judgment in enrolling in a trial. Unless their situation is pretty scary, we would suggest they wait.

Two additional Sugen sites have been added, in France and Poland:

- Dr. Adrian Harris, Med Oncology, Churchill Hospital, Headington, Oxford OX3 7LJ England, U.K. Tel: +44 (0)1865 226184; Fax: +44 (0)1865 226179; E-mail: v.berry@icrf.icnet.uk

- Dr. Karol Krzystolik, Dr. Karol Krzystolik, Dept Genetics, Pomeranian Academy of Medicine, ul. Powstancow Wlkp. 72, Szczecin 70-111 Poland. Tel/Fax: +48 (91) 4828 450; E-mail: karol@polbox.com

- Dr. David Palchak, 1184 Grand Avenue, Arroyo Grande, CA, USA, 93420, Tel: +1 (805) 473-8983; Fax: +1 (805) 473-8735; E-mail: dpalchak.pol.net

- Dr. Stéphane Richard, Oncogenetics, Kremlin-Bicêtre Hospital, 61 rue Gabriel Péri, Le Kremlin Bicetre Cedex F-94276 FRANCE, Tel/Fax: +33 (1) 49 59 67 28; E-mail: stephane.richard@kb.u-psud.fr

- Peter Hirth, Executive VP, Drug Research & Development, 230 East Grand Ave., South San Francisco, CA 94080 USA; Tel: +1 (650) 553-8707; Fax +1 (650) 553-8303; E-mail: peter-hirth@sugen.com

Considering a Clinical Trial?

You should feel free to ask any questions or discuss any issues about the study at any time. When talking to your doctor you may wish to ask the following questions:

The Study

1. What is the purpose of the study?
2. Why do researchers think the approach will benefit patients?
3. Who will sponsor the study?
4. Who has reviewed and approved the trial?
5. How will scientists check the study results and safety of participants?
6. How long will the study last?
7. What will I be responsible for if I participate?

Risks and Benefits

1. What are my possible short-term benefits?
2. What are the possible long-term benefits?
3. What side effects are a short-term risk?
4. What are my possible long-term risks?
5. With my type of tumor (or tumor risk) what other options do I have?
6. How do the potential risks and benefits of this trial compare with those options?

Participation and Care

1. What kinds of treatments, procedures and/or tests will I have during the trial?
2. Will they hurt, and if so, for how long?
3. How do the tests in the study compare with those I would have outside the trial?
4. While in the trial will I be able to take my regular medications?
5. Where will I receive my medical care?
6. Who will be in charge of my care?

Personal Issues

1. Could participation in this trial affect my daily life?
2. Can I talk to other people already enrolled?

Cost Issues

1. Will I have to pay for any part of the trial such as tests or the study drug?
2. If so, what will the charges likely total?
3. What will my health insurance plan now cover?
4. Who in the study can answer questions from my insurance company?

Tips for Talking with your Doctor

1. Let your doctor know you need to set aside time for a discussion.
2. Take along a family member or friend for support and for help in asking questions or remembering answers.
3. Plan ahead: bring a list of questions, but don't hesitate to ask any new questions that may occur to you.

4. Write down your doctor's answers so you can review them later.

5. Consider bringing a tape recorder
Editor's note: For additional tips on doctor/patient communications, order a free copy (plus \$2 postage) of the publication "TEAMWORK: A Cancer Patient's Guide to Talking with your Doctor" from the National Coalition for Cancer Survivorship by calling 1-877-NCCS-YES (1-877-622-7937) a free call in the U.S. See also the book How Are You? Manage Your Own Medical Journey by Patricia Foote available from the VHL Bookstore at <http://www.vhl.org/bookstore> or see page 15.

Reprinted with permission from *In Touch Cancer Prevention and Treatment: The Good Health Guide*, June/July 1999.



This issue is dedicated to:

Elnora Weaver Voth, a compassionate and gracious woman who departed this life unexpectedly in September at the age of 67. Elnora was a nurse and midwife. She served as a volunteer in Vietnam with the Mennonite Central Committee 1959-62. She will be truly missed by her friends and Church family. One of her major concerns in life was VHL which plagued her family, especially her son. It is our prayer that these gifts will contribute to your research. -- her friends

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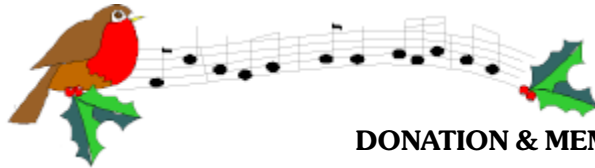
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What's Ahead for the 21st Century?

In 1903, Orville Wright telegraphed home the triumphant news of the first powered flight from the small town of Kitty Hawk, North Carolina. Orville and his brother, expert bicycle mechanics, had invented a flying machine that did more than glide, it actually drove through the air, as a boat propels itself through water. In the course of this century we have seen powered flight progress from Kitty Hawk to space flights to the moon and beyond.

What advances await us in the 21st century? As we approach the new Millennium, it is an exciting time to be alive. We stand on the threshold of astounding advances in the fields of genetics and medicine. In medical terms, our Kitty Hawk occurred in 1993 with the discovery of the VHL gene.

In the movie *Star Trek IV: The Voyage Home*, the Star Trek crew goes back from the 24th Century to the 20th to pick up a whale and save the Earth from destruction. While there, one of their men is injured, picked up in a 20th century ambulance and taken to the hospital. Terrified at what could happen to their shipmate in such a "primitive" hospital, the crew goes off to rescue him. Dr. McCoy passes a woman moaning on a gurney in the hall, bemoaning her kidney failure. He presses his hand-held transceiver to her arm, hands her two pills, and goes along on his mission. Later in the movie we see the same woman, joyfully declaring she is cured. Is this science fiction, or a Jules Verne prediction of our future?

Our wish for the New Millennium is that we will achieve a cure for VHL, a simple way to balance the VHL protein levels in the body and avoid the consequences that occur in this condition. We wish you joy and health and happiness for the new year, and for the 21st Century. -- *Your VHLFA Family*



Chat on the Internet! See page 11.

Changing Addresses?

Please let us know when you move -- we don't want to lose touch with you!

The postal service is getting increasingly fussy about the accuracy of addresses sent via bulk mail. Your newsletter will be returned to us if your apartment number is wrong, or if the street name is misspelled. Whenever possible, please send us your full 9-digit zip code extension -- and be sure it's right!

Last year we spent over \$400 on postage due returns because people forgot to notify us when they moved. Please help us to reduce this expense in the coming year. Thank you!

VHL Family Forum

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