



# VHL Family Forum



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## Kidney Study Shows Effectiveness of Conservative Treatment

*Review of "Treatment of Renal Cell Carcinoma in von Hippel-Lindau Disease: A Multi-Center Study." by Frank Steinbach, Andrew C. Novick, et al.*

Physicians at eight prominent U.S. medical centers pooled their findings on the treatment of 65 patients with VHL kidney tumors over the last fifteen years to determine the effectiveness of different approaches to treatment. The key question was whether it was better to deal aggressively with tumors, removing the entire kidney, or to remove only the tumors and leave the remainder of the kidney, preserving kidney function. Opinions have differed widely on this point.

VHL tumors of the kidney are closely related to the kinds of "sporadic" renal cell carcinoma (RCC) tumors which occur in about 23,000 people each year in the general population. But they are different in many significant ways. In VHL, there tend to be more tumors, usually on both kidneys, and occurring at a younger age. They tend not to grow as rapidly as in sporadic RCC. In the general population, the average age of onset of kidney cancer is 61; in VHL, it was report by Maher et al. as 44 years. In this study, the mean age of diagnosis was 36 years. With better diagnostic methods and more careful screening programs for people at risk, VHL kidney tumors are being found at younger and younger ages.

While it is important to avoid the risks of cancer spreading, or metastasizing, to other places in the body, it is also important to design a life treatment plan which will keep those with VHL healthy throughout their lives, and preferably on their own kidney power. This study looked back at the results 5-15 years after surgery, to see how patients fared with the different kinds of treatment.

They confirmed that the keys to managing VHL kidney tumors are to find the tumors early, deal with them appropriately, and keep up a conscientious program of follow-up.

They found that the 49 people who had "nephron-

sparing" or kidney-saving surgeries did very well.

Every one of these patients was alive five years after surgery, and 81% ten years later. While 51% of these patients had more kidney tumors within the next 5-15 years, only 2 had metastastatic cancer. This is very different from sporadic RCC, where recurrence usually means the cancer has spread. Thirteen of these people had an additional nephron-sparing surgery, 6

“ . . . when nephron sparing surgery is technically feasible, this approach can preserve renal function for an extended interval without compromising cancer-free survival in most patients. ”

have not required additional treatment yet, and 6 have had a complete removal of the remaining kidney. Recurrence is likely due to microscopic RCC that was not removed in the initial operation.

Most of the patients in the study (54 out of 65) had tumors on both sides. Most of the tumors were found during routine screening; 37% were found because of symptoms, which usually means they already had more extensive involvement. Only one patient had metastatic disease when the kidney tumors were diagnosed.

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A total of 68 kidneys in 49 patients were treated with nephron-sparing surgery, removing the tumors and leaving as much functioning kidney as possible. Eight patients were treated with removal of one entire kidney, and another 8 with removal of both kidneys. After surgery, all patients returned for check-ups on a regular basis for up to 11 years.

In the course of the study, 15 out of 65 patients lost kidney function and required dialysis. Six of these had kidney transplants within two years of beginning dialysis. All the grafts were successful. Nine of the patients have been managed with dialysis alone.

This study suggests that "when nephron sparing surgery is technically feasible, this approach can preserve renal function for an extended interval without compromising cancer-free survival in most patients. However, patients treated in this manner should be advised of the importance of close postoperative surveillance and probable need for repeat renal surgery in the future." Recurrence typically does not happen for several years, and the risk of having an associated metastasis appears to be small.

When patients were diagnosed due to symptoms, they usually already had more extensive involvement. This underscores the need for pre-symptomatic screening in people with VHL at risk for kidney tumors. Where the condition was diagnosed early, more conservative treatment had a much better chance of success.

Where removing all remaining kidney tissue is required, kidney transplantation can be used successfully as effective replacement therapy.

This multi-center study, headed by Dr. Andrew C. Novick, Chairman of the Department of Urology at the Cleveland Clinic Foundation, included sixty-five patients from the Cleveland Clinic (19), the Mayo Clinic (18), the University of Iowa (11), the University of Southern California (6), Brigham & Women's Hospital, Boston (4), U.C.L.A. (3), Johns Hopkins (2), and the University of Washington (2).

Frank Steinbach, Andrew C. Novick, Dave P. Miller, Horst Zinke, Richard D. Williams, Greg Lund, Donald G. Skinner, David Esrig, Jerome P. Richie, Jean B. deKernion, Fray Marshall, and Christopher L. Marsh, "Treatment of Renal Cell Carcinoma in von Hippel-Lindau Disease: A Multi-Center Study." *Journal of Urology*. □

"I want to let you know just how much Marston Linehan and I enjoyed the recent VHL Family Alliance meeting. Dr. Linehan and I returned to NIH invigorated by the meeting -- with renewed enthusiasm for research on von Hippel-Lindau disease. You might put an announcement in the newsletter that family members who want to visit the research laboratory in Frederick, Maryland, are always welcome."

— Berton Zbar, M.D., U. S. National Cancer Inst.

## NIH Renews VHL Research

Dr. Berton Zbar and his team at the U.S. National Institutes of Health (NIH) are looking for VHL families not previously seen at NIH, to assist in their renewed effort on VHL research. They will analyze the VHL gene of a family and provide a "road map" so that additional family members can be screened easily.

"We are interested in families that have not previously been seen at NIH. We are particularly interested in families with asymptomatic children who are at risk of developing VHL. I would suggest that you have someone from the families contact me in Frederick, Maryland, on Fridays at 301-846-1288. We will be examining both affected and unaffected family members." □

## VHL Tissue Bank Opens

by Sally Wisniewski, Project Coordinator,  
University of Maryland

It is an honor to us to be the official Tissue Bank for the VHL Family Alliance. We can assure you that without exception all tissue received through referral from the VHL Family Alliance will go exclusively for the study of VHL unless other arrangements have been made through communication with you.

We are always encouraged by working with groups such as yours which evidence such a deep commitment to further research on the cause and treatment of the various developmental disorders.

I believe that The Brain and Tissue Banks for Developmental Disorders will be of great service to those members of the VHL Family Alliance who have concluded that the greatest gift that they can make to further medical research is the donation of tissue either at the time of surgery or at the time of their death. The availability of such tissue will encourage new research studies that are currently not feasible and will bring us closer to the day that disorders such as VHL are no longer a mystery.

A tissue donation program works through the combined efforts of support groups, their membership and the research community. We have found that this sort of cooperation fosters a sense of alliance, comfort and hope in all the diverse individuals who are working towards the same goals. We hope that you too find this to be true.

Please feel free to call either Dr. Zielke or myself at any time if questions arise. Our phone, 1-800-847-1539, is answered on a 24-hour basis. Thank you once again for your confidence.

Phone: 1-800-847-1539; Fax: 410-706-0020

E-mail: btbumab@umabnet.ab.umd.edu

H. Ronald Zielke, Ph.D., Director

Sally Wisniewski, B.A., Project Coordinator □

# Tissue Bank: A New Aid for VHL Research

by William C. Dickson, Chair, Research Management Committee

We are constantly striving to increase the level of VHL research. Once considered only "an obscure medical curiosity", VHL is becoming one of the most important diseases in the study of cancer. The discovery that the VHL gene is mutated within the cells of clear cell renal cell carcinoma in the general population should tend to raise VHL research to a new level. While it is estimated that there are only about 7,000 people with VHL in the United States, it is estimated that more than 25,000 people will develop clear cell renal cell carcinoma in the United States each year.

As the level of VHL research increases, the need for VHL tissue for research also increases. It is here that we can help. The VHL Family Alliance, in conjunction with the Brain and Tissue Bank for Developmental Disorders of the University of Maryland, is proud to announce the establishment of a VHL Tissue Bank. Tissue donated by VHL patients will be held in the Tissue Bank until an approved research project has need for it.

If you have been diagnosed with VHL, and are contemplating surgery, you can help the research community by donating any surgically removed tissue to the VHL Tissue Bank. All cost and arrangements for recovery and transfer of tissue will be taken care of by the Tissue Bank. If you would like to help the VHL research effort, please fill out the Donor Registration Form and mail it today. All information will be treated in the strictest confidence. Pre-registration makes the process simple in the event of surgery. Simply contact the tissue bank, give them the name and contact information for the surgeon and the date of surgery, and the Tissue Bank will make all the necessary arrangements. Even if you are not already pre-registered, arrangements can be made by contacting the Tissue Bank.

Give a gift that only you can give, and help promote research on VHL.

Researchers interested in access to tissue on file should submit research proposals to William C. Dickson, VHL Family Alliance Research Management Committee. 10613 Creamcup Lane, Great Falls, VA 22066, (703) 759-3665, Fax: (703) 759-7992, or e-mail: [vhires@pipeline.com](mailto:vhires@pipeline.com) ☐

## Donor Registration Form -- Tissue Bank for VHL Research

I, \_\_\_\_\_, wish to register myself (or a dependent minor) as a VHL tissue donor with the Brain and Tissue Banks for Developmental Disorders at the University of Maryland at Baltimore. This donation grants permission for the Brain and Tissue Bank to make every attempt within its means to coordinate recovery of surgically removed tissue of the above named donor. Further, if death should occur, I (do or do not ) hereby grant permission for recovery of brain and other tissues. All tissue is donated for the expressed purpose of furthering the research of von Hippel-Lindau disease.

Donor name _____	Next of Kin _____
Address _____	Address _____
City _____	City _____
State _____ Zip _____	State _____ Zip _____
Phone Evening _____	Phone Evening _____
Phone Day _____	Phone Day _____
Donor's Date of Birth _____	Sex _____ Ethnic Group _____
If the Donor was diagnosed with VHL: When? _____	By whom? _____
Brief Medical/Family History (add paper as necessary):	

If you (the Donor) are not diagnosed with VHL, are you the parent/relative of someone who is?

Yes, No Describe relationship \_\_\_\_\_

Signature of Donor or Legal Guardian: \_\_\_\_\_ Date \_\_\_\_\_

Feel free to include any further relevant information.

**Please mail to:** Brain and Tissue Banks for Developmental Disorders  
University of Maryland at Baltimore, Department of Pediatrics  
655 West Baltimore Street, Room 10-009 BRB  
Baltimore, Maryland 21298-2964

# Plain Talk about Stress

-- Louis E. Kopolow, M.D.

You *need* stress in your life! Does that surprise you? Perhaps so, but it is quite true. Without stress, life would be dull and unexciting. Stress adds flavor, challenge, and opportunity to life. Too much stress, however, can seriously affect your physical and mental well-being. A major challenge in this stress-filled world of today is to make the stress in your life work *for* you instead of against you.

Stress is with us all the time. It comes from mental or emotional activity and physical activity. It is unique and personal to each of us. So personal, in fact, that what may be relaxing to one person may be stressful to another. For example, if you are an executive who likes to keep busy all the time, "taking it easy" at the beach on a beautiful day may feel extremely frustrating, nonproductive, and upsetting. You may be emotionally distressed from "doing nothing." Too much emotional stress can cause physical illness such as high blood pressure, ulcers, or even heart disease; physical stress from work or exercise is not likely to cause such ailments. The truth is that physical exercise can help you to relax and to handle your mental or emotional stress.

Hans Selye, M.D., a recognized expert in the field, has defined stress as a "non-specific response of the body to a demand." The important issue is learning how our bodies respond to these demands. When stress becomes prolonged or particularly frustrating, it can become harmful--causing *distress*, or "bad stress." Recognizing the early signs of distress and then doing something about them can make an important difference in the quality of your life, and may actually influence your survival.

**Reacting to Stress** To use stress in a positive way and prevent it from becoming distress, you should become aware of your own reactions to stressful events. The body responds to stress by going through three stages: (1) alarm, (2) resistance, and (3) exhaustion.

Let's take the example of a typical commuter in rush-hour traffic. If a car suddenly pulls out in front of him, his initial alarm reaction may include fear of an accident, anger at the driver who committed the action, and general frustration. His body may respond in the alarm stage by releasing hormones into the bloodstream which cause his face to flush, perspiration to form, his stomach to have a sinking feeling, and his arms and legs to tighten. The next stage is resistance, in which the body repairs damage caused by the stress. If the tress of driving continues with repeated close calls or traffic jams, however, his body will not have time to make repairs. He may become so conditioned to expect potential problems when he drives that he tightens up at the beginning of each commuting day. Eventually, he may even develop a physical problem that is related to stress, such as migraine headaches, high blood pressure, backaches, or insomnia. While it is impossible to live completely free of stress and distress, it is possible to prevent some distress as well as to minimize its impact when it can't be avoided.

**Helping Yourself** When stress does occur, it is important to recognize and deal with it. Here are some suggestions for ways to handle stress. As you begin to understand more about how stress affects you as an individual, you will come up with your own ideas of helping to ease the tensions.

**Try physical activity.** When you are nervous, angry or upset, release the pressure through exercise or physical activity. Running, walking, playing tennis, or working in your garden are just some of the activities you might try. Physical exercise will relieve that "up tight" feeling, relax you, and turn the frowns into smiles. Remember, your body and your mind work together.

**Share your stress.** It helps to talk to someone about

your concerns and worries. Perhaps a friend, family member, teacher, or counselor can help you see your problem in a different light. If you feel your problem is serious, you might seek professional help from a psychologist, psychiatrist, social worker, or mental health counselor. Knowing when to ask for help may avoid more serious problems later.

**Know your limits.** If a problem is beyond your control and cannot be changed at the moment, don't fight the situation. Learn to accept what is--for now--until such time when you can change it.

**Take care of yourself.** You are special. Get enough rest and eat well. If you are irritable and tense from lack of sleep or if you are not eating correctly, you will have less ability to deal with stressful situations. If stress repeatedly keeps you from sleeping, you should ask your doctor for help.

**Make time for fun.** Schedule time for both work and recreation. Play can be just as important to your well-being as work. You need a break from your daily routine to just relax and have fun.

**Be a participant.** One way to keep from getting bored, sad, and lonely is to go where it's all happening. Sitting alone can make you feel frustrated. Instead of feeling sorry for yourself, get involved and become a participant. Offer your services in neighborhood or volunteer organizations. Help yourself by helping other people. Get involved in the world and the people around you, and you'll find they will be attracted to you. You will be on your way to making new friends and enjoying new activities.

**Check off your tasks.** Trying to take care of everything at once can seem overwhelming, and, as a result, you may not accomplish anything. Instead, make a list of what tasks you have to do, then do one at a time, checking them off as they're completed. Give priority to the most important ones and do those first.

**Must you always be right?** Do other people upset you--particularly when they don't do things your way? Try cooperation instead of confrontation; it's better than fighting and always being "right." A little give and take on both sides will reduce the strain and make you both feel more comfortable.

**It's OK to cry.** A good cry can be a healthy way to bring relief to your anxiety, and it might even prevent a headache or other physical consequence. Take some deep breaths; they also release tension.

**Create a quiet scene.** You can't always run away, but you can "dream the impossible dream." A quiet country scene painted mentally, or on canvas, can take you out of the turmoil of a stressful situation. Change the scene by reading a good book or playing beautiful music to create a sense of peace and tranquility.

**Avoid self-medication.** Although you can use prescription or over-the-counter medications to relieve stress temporarily, they do not remove the conditions that caused the stress in the first place. Medications, in fact, may be habit-forming and also may reduce your efficiency, thus creating more stress than they take away. They should be taken only on the advice of your doctor.

**The Art of Relaxation** The best strategy for avoiding stress is to learn how to relax. Unfortunately, many people try to relax at the same pace that they lead the rest of their lives. For a while, tune out your worries about time, productivity, and "doing right." You will find satisfaction in just *being*, without striving. Find activities that give you pleasure and that are good for your mental and physical well-being. Forget about always winning. Focus on relaxation, enjoyment, and health. If the stress in your life seems insurmountable, you may find it beneficial to see a mental health counselor. *Be good to yourself.*

-- U.S. National Institute of Mental Health.

□

# Tumor Growth and Radiology

by Hartmut P.H. Neumann, M.D., and Jörg Laubenberger, M.D., Departments of Medicine and Radiology, University of Freiburg, Germany

Many patients with VHL now follow the advice of their experienced physicians and have regular examinations for early detection and treatment of newly developing or small known lesions. The main targets of these re-investigations are hemangioblastomas of the cerebellum or spine, tumors of the kidneys and pheochromocytomas, usually but not always in the adrenal glands. The techniques used are classical CT or MRI, and for both i.v. contrast (gadolinium) should be used.

In the Freiburg VHL clinic and at patient-provider conferences such as the one recently held in Burlington, Massachusetts, we are repeatedly confronted with situations like this: "I just had my check-up and was told that my kidney tumor increased in size by 25%! Since there is such dramatic growth, I was advised to undergo surgery."

Growth of 25% sounds terrible. Having money in the bank at 25% is extremely high interest. But for a kidney tumor of 2 cm. in diameter, 25% growth means a diameter of 2.5 cm., which is still not very much.

I always want to know the growth exactly. Not being a radiologist myself, I have had many discussions with our radiologists on this subject. There are two major aspects we recognized.

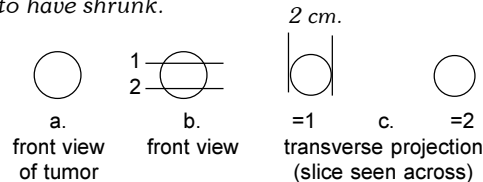
1. It is very important, and often not easy, to use exactly the same methodology for the test, including having exactly the same Computed Tomography (CT) slices for comparison. Patients have to stop breathing for every CT slice, but they do not stop breathing in exactly the same place each time. The air volume in the lungs is not always exactly the same, and the position of the kidney, which moves during breathing, is slightly different. Even if you use the same slice depth, you may be taking a very different picture of the tumor (see Figure 1). Consequently the tumor may appear larger or smaller, simply because of the position of the image.

2. Techniques are improving rapidly. Most medical centers now have CT scanners of the third or fourth generation. With each improvement in design, the resolution of the imaging has improved a great deal, and the pictures are clearer. We also have larger films showing larger images. If the scale changes, the larger picture may give the impression that the tumor is larger, when that is not the case. All CT images have a built-in absolute scale, to provide a ruler to measure the structures in the image, which must be referenced to calculate the relative size of the tumor in each picture.

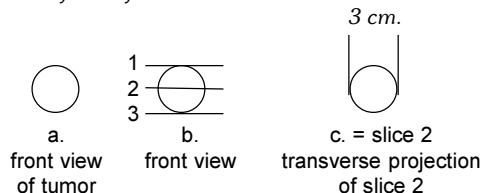
The improved resolution of the images enables us also to see very small tumors which could not be seen on first or second generation scanners. When we see these smaller lesions, we must inform the patient. "You have not three but six tumors in your kidney." The patient thinks: "There is new growth, I have three new tumors." But the radiologist cannot definitely say that those tumors are new unless he used a similar machine and has ruled out the breathing effect.

Furthermore, the improved image quality also adds to the impression that known tumors may have grown, since

**Figure 1.** Illustrations A and B show the possible technical error in measurement of CT scans caused by breathing. If A were taken before B, the tumor would seem to have grown; if B were taken before A, it would seem to have shrunk.



**A.** A 3 centimeter tumor (approximately one inch in diameter), as it might be shown on a scan. If images were taken at positions 1 and 2, the tumor would measure 2 centimeters. Figure 1c shows the view of the face of the slice, as it would appear in the scan. Picture slicing an orange at position 1 as shown, then turn the orange up so that you are looking at the round cut face of the slice.



**B.** If images were taken at positions 1, 2 and 3, the tumor would measure 3 centimeters.

they can now be seen to have sharper margins. Patients should be aware that imaging is not precise.

In Freiburg, we prefer to have imaging controls avoiding radiation. Kidney lesions are now followed with MRI imaging. Because of the breathing problem, we developed a special program. We regularly perform imaging while the patient holds a breath for 12 seconds, which is comfortable even for disabled patients. In one cycle half of the kidney is imaged with 8 slices of 8 mm. thickness without any interslice distance. Two of these sequences cover the whole kidney. To measure the craniocaudal extensions a third coronal sequence is performed routinely. By application of two even faster sequences it will hopefully be possible to avoid the administration of i.v. contrast medium.

Helical or spiral CT is a new development covering an entire volume during one breathhold cycle: for patients with electrical implants, such as pacemakers, and other situations not permitting the use of MRI, helical CT is an alternative. Contrast application is required, and a disadvantage, particularly where multiple follow-up screenings are required, is the use of radiation.

Of course, all lesions which VHL patients have done some growing, and some are still growing. But growth may be less than a first glance impression might tell us. Different decisions may have to be made for each affected organ whether surgery has to be recommended or can be postponed. I prefer to have very well informed patients. I feel it facilitates making decisions about what to do. □

# Teamwork in Action in Burlington!

by Altheada Johnson, Brooklyn, New York

On Friday night VHL patients, family members, friends and medical care providers were introduced and reacquainted with one another. On Saturday and Sunday we got down to the business of learning how best to live with VHL, with screening recommendations and the most recent treatments.

All the medical care providers were very open to questions and discussion, and did their best to provide answers the conference attendees could understand. This was beautifully demonstrated on Saturday when Drs. Novick and Libertino were joined by Drs. Neumann and Linehan in a panel discussion on the treatment of kidney cysts and tumors. Often, explaining VHL can be very technical and controversial. This panel discussion demonstrated the wide range of opinions and medical treatments available to VHL patients.

The Sunday Meditation was very inspiring and thought-provoking. VHL is a serious disease, but we must remember to count our blessings and keep the humor in our lives. A large part of Sunday's session was devoted to DNA, whether to test or not. Here again, we found that there is no one right answer. The answer each individual chooses must be right for them.

Parting really is a sweet sorrow. I know I didn't want to leave. This meeting was the first time many VHL patients had laid their eyes on another person with this disorder. The camaraderie, sharing of stories and friendships, will never be forgotten. □

## Members line up to ask questions.

"We really enjoyed the weekend and are so pleased to know the Alliance exists. It was so neat to be with other people like me that have VHL."

-- Pam. M., Ohio

"First of all, let me congratulate you on the conference! I regret that I was only able to attend the Saturday morning part but I certainly expanded my knowledge about VHL exponentially just during that time. It was also both moving and inspirational to hear the kind of acute and directed questions that came from the audience. It was certainly an illustration of the kind of partnership that can move a field forward very quickly." -- Hilary Worthen, M.D., Cambridge Family Health, Cambridge, Mass.

"Thank you! A million thanks! Had we not been present at the conference we would not have been aware of the connection to VHL of some of my son's problems. I feel the very real presence of God in our lives as he places us in the hands of angels -- the caring doctors and the countless VHL people of courage we met. . . . It was Craig who moved us to tears. Quiet courage. I think that's the one most significant impression I took away with me -- the understanding of what real courage is. I saw it everywhere, expressed as smiles in the face of chronic pain, and grit in the face of overwhelming diagnoses." -- Evelyn S., Florida

"It was a pleasure to see you and to be able to participate in the VHL Second International Conference. My experiences were most rewarding, and I enjoyed the opportunity to meet other members of the VHL Family Alliance. You and your co-workers are to be congratulated on orchestrating a superb conference, which provided opportunities for education and learning, socializing and growth. It was indeed a pleasure to have a small part in your program." -- Jerry Cavallerano, O.D., Ph.D., Joslin Diabetes Center, Boston.

# Highlights from the Boston meeting

by Joyce Wilcox Graff and Laurel Newson

The Town of Burlington declared it VHL Awareness Week. The Lahey Hitchcock Clinic sent out press releases. One hundred twenty-three people gathered from near and far to talk about von Hippel-Lindau disease. Patients and family members came from as far away as Ireland and Hawaii; five genetic counselors from the U.S. and Canada; twenty-four physicians from the U.S., England, Wales, and Germany. Attendees ranged in age from infants to two people with VHL in their sixties. Jim D. from Burlington, a patient of Dr. Aiello for the last thirty-five years, introduced the panel on eye involvement. Betty B., a teacher from Tennessee and a recent transplant recipient, came with her three adult daughters.

On Friday night when we gathered for a reception, it was the first time many of those present had ever met another person with VHL. This was a time to get acquainted, share stories, and make new friends.

On Saturday we heard lectures on clinical aspects of VHL. Dr. Peter Dempsey and Dr. Andrew Mancall from the Lahey Hitchcock Clinic spoke about the anatomy of VHL lesions of the brain and spine, and the latest imaging techniques. Dr. Peter Black of Brigham & Women's Hospital showed an amazing video demonstrating techniques in computer modeling for treatment planning, and the use of shunts to control cysts.

Ms. Hetty DeVroom of the National Institutes of Health shared the current status of their research on endolymphatic sac tumors (ELST), which can cause hearing changes in people with VHL. They estimate that some 10-15% of people with VHL have ELST in the inner ear. They are still working to determine the best treatment for these; meanwhile, no treatment is recommended without consultation with Dr. Edward

Oldfield at NIH, as the wrong treatment could damage more hearing than it might save.

Dr. Black and Dr. Haring Nauta of the University of Texas spoke about stereotactic radiosurgery, and what we are learning about the strengths and limitations of this interesting procedure.

Not every patient and not every tumor is a good candidate for stereotactic radiosurgery. While there is no visible cut, nonetheless it is a surgical procedure which essentially cauterizes the hemangioblastoma deep in the brain. The treated tissue swells after surgery, sometimes causing new or increased symptoms for some months which needs to be managed with steroids. The treatment needs to be carefully planned to ensure that the anticipated swelling is sure to be manageable, and to make sure that the patient is not sensitive to the medication, especially since large doses may be needed to manage the swelling.

Dr. Lloyd Aiello and Dr. Jerry Cavallerano from the Joslin Diabetes Center and Dr. Mami Aiello Iwamoto of Brigham & Women's Hospital provided

an excellent tutorial on VHL in the retina, and Dr. Aiello shared a preview of the Joslin Vision Network, a computer-based imaging network which will allow Joslin to consult with millions of diabetics -- and potentially VHL patients too -- throughout the United States and Europe.

Dr. John Libertino from the Lahey Hitchcock Clinic spoke about VHL in the adrenal glands, and Dr. Andrew Novick of the Cleveland Clinic shared his research, which is reported in greater detail on page 1 of this issue. There are many possible treatments for VHL in the kidney. The trick is to work out a lifelong treatment plan to keep the patient healthy and on his own kidney power for as long as possible.

After Dr. Novick's talk, Dr. Hartmut Neumann of Freiburg and Dr. Marston Linehan of the U. S. National Cancer Institute began dialoguing with him about his findings, almost as if the rest of us were not in the room. The dialogue developed wonderfully, letting us listen in while four of the world's leading experts on VHL in the kidney discussed the pros and cons, and then took questions from the audience. We continued until Dr. Novick had to run to catch his plane back to Cleveland.

Kathy H. from Massachusetts, Dr. Diana Wheeler from Wales, Dr. Neumann from Germany.

L to R: Dr. Hartmut Neumann, Germany; Hetty DeVroom, Maryland; Dr. Lloyd Aiello, Boston; Dr. Eamonn Maher, England; Dr. Andrew Novick, Cleveland; Dr. Berton Zbar, Maryland; Dr. Marston Linehan, Maryland; Dr. James M. Lamiell, Texas; Dr. John Libertino, Boston.

On Sunday, David Torres, Fred Johnson, and Craig Warnick shared with us their thoughts on the importance of their faith in sustaining them through the rough spots in their lives. As one member said, "spiritual support is just as important if not more important than scientific support."

The rest of Sunday's presentation was devoted to genetic research and testing. Dr. Eamonn Maher of Cambridge, England, and Dr. Linehan showed us how they found the gene and the kinds of findings emerging from the mapping of the gene. Dr. Maher and Ms. Jane Green from Memorial University in Newfoundland brought charts showing the improvements in life expectancy brought about by clinical screening, and improvements in quality of life through DNA testing to rule out VHL in relatives at risk. Once a relative has had DNA testing and shown not to have the mutated VHL gene, then one can with confidence discontinue clinical screening in that person, assure them that their children could not be affected, and free them of this complication in their childbearing decisions. Nonetheless not everyone chooses DNA testing for themselves or for a child under 18. "Sometimes, at least for a while, people feel it is better not to know," Dr. Maher said. They prefer to continue clinical screening.

For people in the United States, who are not in a system of universal health care coverage, the decision to obtain DNA testing is further complicated by concerns about insurability. Dr. Philip Reilly of the Shriver Institute and Mr. Richard Saltus of the Marfan Syndrome Foundation related examples from people with other genetic conditions, and spoke about the legal and ethical concerns. There were many questions from the audience.

Ms. Jane Green shared stories from the Newfoundland VHL families which showed the improvements in length and quality of their lives through better clinical and DNA screening. While fifteen years ago the

majority of people with VHL died in their thirties and early forties, today most people are living at least 15-20 years longer. The Canadian health care system, like U.S. Health Maintenance Organizations, are eager to spend their money wisely, and do not want to do expensive imaging tests unless they are truly warranted. But clearly, waiting for symptoms is unwise in VHL, particularly in the case of kidney and adrenal tumors. Her research has shown that through DNA testing they are able to rule out VHL in a large number of family members who would previously have been followed with clinical tests annually for thirty years, as a precaution. Not only does this save money for the health care system, it also improves the quality of life for these people, relieving them of the concern about VHL.

With that saving, they are able to afford to follow people who do have the mutated gene more closely. In finding the tumors at an early stage and dealing with them appropriately, they have been able to keep people much healthier for many more years. In fact, it had been thought that two of these families had pheos but no kidney tumors. As their life spans have increased, however, it seems that the kidney tumors develop later on in life, though there are significantly fewer kidney tumors than pheos in these families. In sum, her research shows that the investment in more expensive imaging studies for preventive screening far outweighs the cost of dealing with tumors which are farther progressed and more likely to cause disability.

The families were all thrilled to be together, and were very moved by the whole-hearted involvement of the professionals. One family wrote, "We had the best experts in the world, the most caring physicians in the world. The doctors were wonderful, and we give them credit for their dedication to VHL." The physicians in turn were amazed at the quality of the questions from the families, and at the amount of learning they gained through interacting with the families.

As Gloria P. from Ireland said, "I arrived in Boston feeling very alone and nervous, and I left feeling confident and strong. I have come home feeling able to handle anything. It's the first time since my daugh-



ter was diagnosed 6½ years ago that I feel like this. It was good to talk to other people that understand and to hear the doctors being so knowledgeable and interested in VHL."

When Joyce opened the meeting on Saturday, she said that she was confident that the cure for VHL was in the room, and would come as a product of Teamwork among the VHL families and health care professionals. By the end of the meeting, we could all feel the energy that would bring that into being. As Dr. Zbar said, "Dr. Linehan and I returned to NIH invigorated by the meeting -- with renewed enthusiasm for research on von Hippel-Lindau disease." □

**Audio tapes available  
Ordering info on page 15.**

"How proud you must be to look back, not just at the obvious success of a great conference, but at all the accomplishments made possible because of the VHL Family Alliance. . . . But the real reward lies ahead -- the reward of seeing, if not a cure, significant breakthroughs in the treatment of VHL."

-- Bill Dickson, Virginia

"I wish to congratulate you and the other hardworking board members and volunteers on the superb conference in Burlington! I thoroughly enjoyed myself and learned a thing or two on kidney and eye involvement in VHL. The degree of involvement and the quality of information from the health care professionals was truly exceptional. . . . I returned home with increased realization of the magnificent work that can be done by volunteers and the enormous challenge patients with VHL must face each day." -- Hetty L. DeVroom, R.N., B.S.N., CNRN, Surgical Neurology Branch, National Institute of Neurological Disorders and Stroke, Bethesda, Maryland

We'd like to thank Joyce and Laurel and all the doctors who participated in the Patient/Provider Conference. Without caring individuals like yourselves, we would be doomed. You've given us hope, guidance, understanding, and most of all love. Thanks again for all you've done for us. -- Cindy & Greg M. and children, Texas

## Boston Family Reunion

by Peggy and Don Marshall, Corinth, Mississippi

The VHL Family Alliance Second International Conference in Boston was the highlight of another year in our Family. We went with great expectations and left with an even greater sense of caring and sharing.

The opportunity to interface with the VHL family of members and medical professionals was heartwarming and informative. As the conference progressed we were able to meet new family members and to establish that sense of warmth and comfort of knowing we are on target, meeting both our personal and the VHL Family Alliance goals.

Several aspects of the conference stand out as we reflect on this past conference. The first conference in Kansas City was the beginning of a learning process for us to be able to take our story public, share our concerns, build our future, and to learn to interface with the medical professionals outside of the examining room. We met many new families for the first time and found that we all share the same feelings. We were impressed with the grace of each family in the face of crisis. Boston begins a new chapter in our Family with an even greater understanding of VHL. We met new families that are now experiencing the joy of knowing they are not alone, but share a common link with many others. We also had the opportunity to meet friends from a year past and share in their joys and tears. We feel privileged to be a part of this family reunion.

Another aspect of the Boston Conference was the chance to communicate with the medical professionals we depend on. They are a large part of our Family and share in our concerns. The presentations were superb in form, content, and presentation. We came away with a renewed vision of the activities taking place to identify and treat VHL. For so many years we floundered in a sea of darkness wondering if someone out there was really concerned about our future and the future of our children. We saw quite emphatically that progress is being made and there is a real concerted effort to control the effects of VHL.

The one aspect that stands out in our minds is the communication that took place during the breaks and intermissions. Families were talking to each other, sharing names, phone numbers, and stories. Every one had the opportunity to discuss their concerns with others and establish that bond that will last a lifetime. We learned we are part of a large caring and sharing family that can support each other through the most trying of times.

We are looking forward to the coming year, and to our next Family Reunion in Hawaii. □

# Take Charge of your Medical Records

Your medical records — housed in your doctor's office and in any hospitals where you've been a patient — are confidential. However, medical information given when you apply for health, life, or disability insurance is often stored in a central data base called the Medical Information Bureau and is shared among health insurance companies. This material may also be disclosed (with your permission, though sometimes without) to government agencies, schools, and employers. Yet, ironically, these same records may not be readily accessible, or not available at all, to you, the patient. You should have a complete, up-to-date set of your records. That way, if your doctor retires or dies and has not provided for the transfer of records, you'll be able to bring your records to a new doctor. Thus the continuity of your care will be ensured and costly tests will not have to be needlessly repeated. And by obtaining a copy of your records, you may also become a more informed medical consumer in general, gain more control of your health care, and perhaps establish a more open and equal relationship with your doctor.

In the past, many doctors and health-care providers argued against patients' right of access to medical records, since these files are often highly technical — and possibly confusing and alarming. However, in recent years, thanks in large part to lobbying by consumers' groups, about half of all states now guarantee access to medical (and often mental health) records. To obtain a copy of your records, simply contact your doctor's office or the hospital where tests or procedures were done. Even in states without full guaranteed access, most doctors and hospitals will honor requests for copies. There may be a charge for photocopying and mailing. A good way to keep records is to regularly request copies of test results and notes from your doctor after each visit.

Specific laws and procedures concerning medical records vary from state to state. Public Citizen's Health Research Group, the nonprofit consumer advocacy group, has published a 66-page booklet called *Medical Records: Getting Yours*, which includes a summary of your legal rights in each state, and what to do if a request is denied or if the records are incorrect. It is available for \$10 from Public Citizen, 2000 P Street, NW, Suite 600, Washington, DC 20036.

The Medical Information Bureau, the central

clearinghouse that insurance companies rely on to pool the data you supply whenever you apply for insurance, contains files on millions of Americans. Your records there can play a key role in whether you're able to get medical or life insurance and how much you have to pay for it, so it's important to make sure they are accurate. You can get a free copy of your file by calling 617-426-3660 or by writing to P.O. Box 105, Essex Station, Boston, MA 02112. — *University of California at Berkeley Wellness Letter, November 1993.*

## Cautions Regarding Stereotactic Radiosurgery

While stereotactic radiosurgery seems a very promising alternative, there are real concerns about its limitations. Not all tumors are good candidates for this treatment, and sensitivities to certain medications may rule out this therapy for some people. It is *critically important* that you discuss possible post-treatment effects with your medical team before you undergo treatment. Please call the hotline, 1-800-767-4VHL (U.S. and Canada) or write to us for the latest list of questions to ask your doctor, to make sure that stereotactic radiosurgery is right for you. In many cases, it is in fact safer to choose conventional surgery. □

## Online Services

by Fred Johnson, Computer Committee  
**VHLBBS/UMC available via Telnet**

The VHLBBS free bulletin board sponsored by the Global Ministries of the United Methodist Church may be reached at 1-212-222-4724 or

telnet hwbbs.gbmg-umc.org

ftp hwbbs.gbmg-umc.org

Anonymous ftp access is possible by responding to the *name* prompt with "Anonymous" and as the *password* enter your email address.

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For technical assistance, or to ask about access charges, call 1-800-ROCK-WEB.

Georgia on the Net

Write to Eva and Chris and the Georgia chapter at GA VHL from American Online, or gavhl@aol.com

Have fun! □

Find information about VHL on the World Wide Web! Connect to the VHLFA Home Page, at URL  
<http://kumchttp.mc.ukans.edu:80/instruction/medicine/genetics/vhl/vhlhomep.html>  
<http://neurosurgery.mgh.harvard.edu:80/vhl-fa.html>

## Introducing our Medical Advisory Board

There are thirteen medical professionals listed on page 14 who may not always be very visible to you as readers, but who are very visible to the members of the Board of Directors. They very generously lend us their expertise as consultants on difficult questions from members, in the writing or reviewing of material for this newsletter, in presenting or helping design presentations for our annual meeting, and in advising us on various aspects of our programming. We continue here the introductions begun in the September issue. We have purposely sought out people with depth and breadth of experience with VHL — people whose formal training has been enriched by working with a number of patients with VHL over a number of years.

### John A. Libertino, M.D.

Dr. Libertino is Chairman of the Institute of Urology at the Lahey Hitchcock Clinic, Burlington, Massachusetts. He has written or co-authored some 85 articles, has contributed to nearly forty textbooks, and has edited or co-edited another five textbooks. He is the series editor of the textbook series *International Perspectives in Urology*.

Educated at New York University and Georgetown University School of Medicine, he did his internship at Strong Memorial Hospital in Rochester, New York, and his residency in urology at Yale-New Haven Medical Center in New Haven, Connecticut. He was awarded a Cardiovascular Research Fellowship at the National Institutes of Health and Georgetown University School of Medicine in Washington, and the Harvey Cushing Fellowship in Surgery at Yale University School of Medicine.

Dr. Libertino serves on the Medical Advisory Board of the VHL Family Alliance.

### W. Marston Linehan, M.D.

Dr. Linehan is Head, Urologic Surgery, Surgery Branch, National Cancer Institute, Bethesda, Maryland. Dr. Linehan is author or co-author on over 170 scientific publications published or In Press, most dealing with kidney cancer. His primary work is the study of the genetic basis of familial and sporadic kidney cancer. This

work, which began eleven years ago with Dr. Berton

Zbar, led to the discovery of the VHL gene. Dr. Linehan is currently studying the VHL gene and performing studies to determine why inactivation of this gene leads to von Hippel-Lindau disease and how this knowledge will lead to better forms of diagnosis, prevention and therapy of this disease. Dr. Linehan is initiating clinical trials to evaluate new forms of preventative therapies in patients with von Hippel-Lindau disease.

### Andrew C. Novick, B.S., MDCM

A native of Canada, Dr. Andrew Novick has been with the Cleveland Clinic Foundation since 1974. He graduated in 1972 from McGill University in Montreal with a combined degree in Science and Medicine. After completing residencies in Montreal, he went to Cleveland for his Urology residency, and has continued on their staff. He is Chairman of the Department of Urology and Professor of Surgery at Ohio State University School of Medicine. He has served as head of the Section of Renal Transplantation (1977-1991), and as Chairman of the Organ Transplantation Center (1985-1992).

He is an active member of the American Urological Association and has led studies in various management techniques for renal cell carcinoma, including a movie on "Partial Nephrectomy for Renal Carcinoma" (1987), and evaluations of the effectiveness of Interleukin-6 and ALT versus Interferon Alpha. Drs. Novick and Libertino collaborated on a volume on *Adrenal Surgery (Urologic Clinics of North America, vol. 16, no. 3, August 1989)*. His ten-year study of von Hippel-Lindau has been published in the *Journal of Urology*. □

### Teamwork Pin

We are proud to present the VHL Teamwork Pin. Melissa Minster of J. J. Minster's Jewelers in Newark, Delaware, created this beautiful design and etched it onto silver or gold-filled disks. Minster's is donating the first 100 pins to raise money for VHL education and research. Order yours on page 15.

We are very grateful to Melissa and to Minster's for this very generous and meaningful donation. Families and health professionals: working together, we can all have a Very Happy Life. □

# New Board Members

## **William C. Dickson, Great Falls, Virginia** *Chair, Research Management Committee*

Bill is retired from the field of research and development. He developed a model to improve the production and scheduling procedures of a large military industrial complex, which was widely implemented in the military and private sectors. He was a member of a think tank in the Office of the Secretary of the Navy and received a commendation from the Assistant Secretary of the Navy for Research and Development.

He subsequently worked on projects for the Department of Health and Human Services, for which he received a commendation from the President. He worked as a consultant for the Logistics Management Institute in Washington, D.C., and consulted with Johns Hopkins Hospital.

Bill's son-in-law and grandson have VHL. Bill will be heading our Research Management Committee, working to foster increased research on VHL.

## **Lois Erickson, Bloomington, Minnesota** *Chair, Development Committee*

Lois Erickson, Bloomington, Minnesota, candidate for re-election. Lois has served as Secretary to the Board for the past two years, and was the National Chairperson for the Kansas City meeting.

"I am married and have two children: Carmen and Chad. I have VHL and so does my son Chad. I work at Knutson Mortgage Corporation in the Master Servicing Department. I have many hobbies and think of myself as a healthy person.

"I am delighted to be a member of the VHL Family Alliance, and think the organization has given me a purpose in life. I don't want anyone to be an uninformed as our family was for 40 years. I will do my best to talk about VHL to everyone who will listen. I am thankful for each day that I feel well and can continue going 100 miles per hour!"

## **Terrence Redding, W. Palm Beach, FL** *Development & Research Database Committees*

Terry is a past W. K. Kellogg Fellow in adult and higher education. He is nearing the completion of the Ph.D. and should finish this summer at the University of Oklahoma with a degree in Higher Education and a minor in Marketing. He is currently the Director of Corporate Computer Training, Palm Beach Atlantic College, West Palm Beach, Florida. Terry also operates his own consulting firm,

R & R Advantage, Educational Research and Marketing, which specializes in marketing not for profit organizations.

Terry and his wife Barbara have six children, one grandson, and have been foster parents to ten newborn infants. Barbara is a past La Leche League Leader, Religious Education Teacher, and she and Terry were active in Catholic Folk Choir prior to their recent move to Florida.

Both express strong appreciation for the work of the VHL Family Alliance. "For the first fourteen years of Barbara's illness, we felt isolated. Now, through the efforts of the Alliance, we know we are not alone."

## **Audrey J. Tobin, Faribault, Minnesota** *Treasurer*

Audrey is a personal banking officer at Norwest Bank in Faribault, Minnesota, where she has worked for the past seven years. She has been recognized as one of the top ten loan officers in her region every year, and has twice earned recognition at the National Level.

Audrey and her husband John owned and operated two Montgomery Ward Catalog Stores and a Mobile Home Park twelve years prior to that. Her husband is a Small Business Manager Instructor at Riverland Technical College in Faribault. They have two children, ages 25 and 20.

Audrey has also been recognized as an Outstanding Jaycee Woman for Minnesota. She has held several treasurer positions and at one time paid bills and reconciled seven checkbooks in one year. Audrey enjoys bridge, golf, bicycling, and the sun!!! VHL has been in Audrey's family for over sixty years and a worry for the past thirty years. Audrey is a sister to Lois Erickson, who also serves on the Board of Directors.

## **David Torres, Kapaa, Hawaii** *Chair, Kidney Dialysis & Transplant Support Group; Co-Chair, VHLFA 1996 International Meeting in Honolulu*

David Torres is 39 years old, born and raised in Hawaii where he resides with his wife and two children.

He graduated from high school in 1974 and enlisted in the U.S. Navy shortly thereafter. He was diagnosed with VHL in 1978 as a result of screening by Drs. Lamiell, Hsia, Salazar, and others.

In February 1980 he became Kapaa's first resident to receive a successful renal transplant.

He worked with the Department of Education from 1985 through 1987. He was ordained by the Garden Island Missionary Baptist Church on December 18, 1986, and has been the pastor of the Hanapepe Missionary Baptist Church since it was organized in April 1989.

He joined the VHL Family Alliance almost immediately after it was organized and has been active in Hawaii in informing and sharing the information the VHL Family Alliance provides with other members of his large affected family. He has been a speaker at both the 1994 and 1995 conferences of the VHL Family Alliance. □

## Hawaii Conference 96

The Second International Symposium on VHL will be held in conjunction with the Third International Patient/Provider Conference of the VHL Family Alliance in Honolulu, June 17-21, 1996, hosted by the John A. Burns School of Medicine and the "Hawaii family." The genetic mapping of this large family, begun in 1977, started the genetic research effort which resulted in the finding of the VHL gene.

A medical symposium is a forum where scientists gather to discuss the many ways of approaching a problem, in hopes of coming up with better medical management and new paths toward a cure. The first Symposium, held in Freiburg, Germany, in 1994, resulted in several new scientific articles and new energy in the research community around VHL.

By combining the Symposium with the VHLFA meeting, people who would like to participate in both kinds of meetings will be able to do so in one trip.

On Wednesday, June 19, 1996, a special day at the conference is being devoted to Family Issues in VHL. The Rev. David Torres and Ms. Patti Kohlen will be co-chairing the Family Issues Day. The other three topical days are "Consumer Issues about Familial Cancers," chaired by Debra Collins; "Medical Issues in VHL," chaired by Drs. Hartmut Neumann and James M. Lamiell; and "Scientific Issues in VHL," chaired by Dr. Berton Zbar.

In order to provide topics of interest to you, please let either David or Patti know what issues you would like covered. Rev. David Torres, P.O. Box 749, Kapaa, HI 96746; Ms. Patti Kohlen, 359 Los Cerros Drive, San Luis Obispo, CA 93405, E-mail: pkohlen@violin.calpoly.edu □

## Volunteers Honored

Don Marshall was honored at the Burlington meeting for his unstinting service as chairman of the Publications Committee. "In fact, Don is the publications committee." Most people know his name since he sends out the new member packets, and information packets for physicians. Don arranges printing services for all Alliance publications for the best possible price, and makes sure they are in the right place at the right time. In his exhibit at the Burlington meeting, he displayed samples of our publications in four languages, and some amazing statistics: in only two years we have distributed more than 5,000 Handbooks and 30,000 brochures to people in more than 20

## California Chapter

— Dawn Cerf, Chair, California Chapter

Members drove between 2 and 5 hours, with an average drive of four hours, to attend the first meeting of the California Chapter held in San Luis Obispo on April 8, 1995. With 31 people attending, we wasted no time in getting acquainted, sharing our experiences, bonding as friends, and learning as much as we possibly could.

This was the first time most of the attendees were able to meet others with VHL outside their own families. Unfortunately, our speaker and facilitator both canceled just days before the meeting, but thanks to Patti Kohlen's networking we then got two other speakers on the subjects of kidney dialysis and physical therapy relating to spinal surgery, who proved to be excellent speakers.

Dawn Cerf,  
California Chapter Chair

Our meeting lasted over four hours, and because we were not ready to say goodbye, we continued as a group at a local restaurant for dinner for an additional three hours. The overwhelming feeling of the attendees was that of surprise that they enjoyed themselves so much. We did have a great group with a terrific sense of humor. We laughed almost as much as we talked. Everyone is already looking forward to our next meeting in San Francisco in 1996, and several members are also planning to attend the Hawaii conference next year. □

countries. "We couldn't have done it without Don's caring and efficient partnership."

The First Annual Minster Volunteer Service Award was presented by Susan Warnick, incoming Chairman of the Board, to Joyce Graff. "We are all grateful for Joyce's vision and leadership. While I had envisioned a nice little local organization, Joyce was thinking world-wide -- and making it happen." "Well," said Joyce, "for thirty years the doctors told me and my husband that VHL was so rare, he was one of only about nine affected people in the world. So I thought we should look world-wide and find all nine. So far we have found nearly 4,000 people with VHL." Dr. Aiello read his version of "You've Got a Friend," a song by Carole King, modified to "All you have to do is call, and Joyce Graff and the VHL Family Alliance will be there. You've got a friend." □

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 W. C. Enterprises, New Castle, Delaware  
 Mr. & Mrs. John L. Wood, Tennessee  
 Marsha Zabarsky, Lahey Hitchcock Clinic  
 Dr. Robert Zamenhof, Boston  
**... and from all the Members of the VHL Family Alliance**

# Membership, Feedback and Order Form

Name: \_\_\_\_\_

Address: \_\_\_\_\_

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Country (if not USA) \_\_\_\_\_ Fax: \_\_\_\_\_

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☐ My employer will match my contribution. I have enclosed the appropriate forms.

*All Members receive 3-4 issues of the Forum during the year, and one copy of each Alliance publications. Free subscriptions are available where the dues are a hardship.*

☐ Audio version available if needed for a handicapped memberI am a ☐ VHL patient ☐ VHL family member ☐ Supporting Friend☐ Professional (physician, nurse, dietitian, social worker, etc.)☐ My occupation is \_\_\_\_\_☐ I am interested in participating in a local support group☐ **Membership** (\$25 per mailing address, \$35 for medical professionals)☐ **Tax-deductible donation** to support the work of the Alliance☐ 1994 Conference audio tapes (7 tapes & album) #\_\_ @ \$40 \$ \_\_\_\_\_☐ **1995 Conference audio tapes** (12 tapes & album) #\_\_ @ \$60/set \$ \_\_\_\_\_

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\_\_\_\_(1) Brain &amp; spinal; \_\_\_\_ (2) Stereotactic Radiosurgery; \_\_\_\_ (3) Eye;

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Total sessions: #\_\_ @ \$7.50 \$ \_\_\_\_\_

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## In Honor Of . . . donations (minimum \$5 each):

I am enclosing a donation ☐ In Memory Of ☐ In Honor Of ☐ OK to list in VHLFF

Honoree's Name \_\_\_\_\_

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Please send card to (name and address) \_\_\_\_\_

## Return order form to: VHL Family Alliance, 171 Clinton Road, Brookline, MA 02146

Sales of materials are used first to fund local activities. If you are in one of the countries with a local affiliate group, please send money to one of the following addresses:

**In Britain**, send to VHL Patient and Relative Contact Group, 114 Longfield Rd, Littleport, Ely, Cambs CB6 1LB**In France**, send to VHL France, 87, rue de la République, 95240 Corneilles-en-Parisis, France**In Italy**, send to Alleanza VHL, Loc Malvicina, 19, 15066 Gavi (AL), Italy**In Australia**, send to VHL Family Alliance, 2/51 Musgrave St, Yarralumla 2600, Canberra, ACT, Australia

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Every  
contribution  
helps!  
Thank you

# VHL Volunteers Win Hope Award

Fred and Altheada Johnson of Brooklyn, New York, were awarded the 1995 Hope Award for Family Volunteers at the 5th Biennial Symposium on Minorities, the Medically Underserved, and Cancer, in Washington in April, honoring their outstanding work as volunteers for the VHL Family Alliance.

Altheada, who has VHL, lost the use of her legs following an unsuccessful spinal surgery. "Despite Altheada's physical limitation," says Joyce Graff, "she works diligently with physicians to raise awareness of this multi-system disorder to increase the probability that a doctor will consider a diagnosis of VHL. She is an excellent speaker and advocate for people with VHL. On the hotline, she is a great listener, a great source of comfort and encouragement, and an excellent resource for families." A registered dietitian, Altheada also works with the International Center for the Disabled in New York on nutritional programs.

Altheada has been chairperson of the New York chapter of the Alliance for two years, and serves on the Board of Directors. She and Fred established the VHL On-line Bulletin Board under the auspices of the United Methodist Church in New York, one of their many efforts to raise consciousness about VHL and identify persons who many have the disease.

Fred and Altheada were presented with a beautiful trophy, and a cash award which they donated to the VHL Family Alliance, to promote education and research on von Hippel-Lindau disease. At this conference Fred and Altheada were able to share their experience with VHL with people who had never heard of this disorder. "We were able to put VHL in the ears of many Very Important Doctors and others in cancer care around the nation."

"Attending this conference taught me a lot," says Altheada. "On top of all VHL may force us to deal with, we cannot forget to do breast self-examination, get routine Pap smears, and be screened for prostate cancer. Every VHL patient is a cancer survivor. Having cancer of any kind is not a death sentence. With early diagnosis and appropriate treatment we can all have Very Happy Lives." □

*Altheada Johnson, with Robert Guillaume and Roscoe Lee Browne, co-hosts of the Congressional reception.*

*1995 Family volunteers  
Fred and Altheada Johnson.*

## VHL Family Forum

Newsletter of the VHL Family Alliance  
171 Clinton Road  
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