



VHL Family Forum



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Robot Does Brain Surgery

The first patient to receive frameless stereotactic radiosurgery with the Accuray Neurotron 1000 was treated June 8, 1994, at Stanford University with Dr. John Adler, neurosurgeon and Dr. Richard Hoppe, radiation oncologist. The Neurotron 1000 is a robot machine which brings some significant advancements to the field of stereotactic radiosurgery.

Stereotactic radiosurgery is a method for stunting the growth of a tumor without conventional surgery. Laser-beams of radiation are aimed at the "target site," the tumor, from hundreds of different angles. As each beam passes through healthy tissue, that tissue gets the low one-beam dose. Where the hundreds of beams come together at the tumor, the tumor receives the sum of the dosages of all those beams, effectively performing laser surgery on the tumor.

Stereotactic radiation is still somewhat controversial in the neurosurgical community. Conventional microsurgery is still considered the first choice for tumors of the cerebellum, where surgical risks are relatively low. Times when you might consider stereotactic radiosurgery¹ are

- When the tumor is deep in the brain
- When there are recurrent tumors
- Where there are multiple tumors which cannot all be handled in one surgery
- Where the patient is frail and would not tolerate conventional surgery very well
- For preventive treatment of small asymptomatic tumors, before they develop a cyst

The best candidates for stereotactic radiosurgery are tumors which are 2-3 cm. in size. If there is an associated cyst, it will not get better quickly, but will shrink over some months. Sometimes a shunt or other method of shrinking the cyst can be used in combination with radiosurgery to manage the cyst.

A number of VHL patients have undergone stereotactic radiosurgery for treatment of hemangioblastomas of the brain, and are generally delighted

with the procedure. There are many treatment centers around the country using either gamma knife or linear accelerator machines.

David I. of California recently underwent gamma knife stereotactic radiosurgery treatment at Stanford for his second VHL hemangioblastoma. The first was successfully treated with conventional surgery. "I am tempted to say it was fun," he says, "but that is not the right word. It certainly was interesting and relatively painless.

"My appointment was for eight o'clock and by 10:30 my wife and I were having brunch at a nearby restaurant! I looked at her across the table and said, 'Do you realize I just had brain surgery?' We found the thought almost amusing. I had two small band-aids on my forehead where the frame had been attached and other than that, the casual observer would have no way of telling there was anything unusual about me. I had no aftereffects from the surgery, no nausea, no headache. I felt as well as I had going in that morning, I even stopped in at my office and visited and will be returning to work this morning. And I had brain surgery yesterday morning!"

Using today's treatment machines, the patient must wear a "helmet" throughout the treatment which is screwed to the skull to keep it fixed in place. CT and MRI testing are used with the helmet in place to determine the precise relationship between the tumor and the frame. These measurements are then used

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Join us in Boston, April 1995!***

Recipes Needed!

The Florida Chapter invites you to contribute your favorite recipes to the Very Happy Life Cooking Book. Great Chefs from throughout the world-wide VHL community -- including you, your friends and significant others -- are sending us their best secret recipes.

The Cook Book will be available for purchase at the Annual Meeting in Boston in April.

Send recipes to:

Gale L., Florida Chapter, VHLFA
1990 Harmon Avenue
Winter Park, FL 32789-5306

during the treatment to aim the beams at the tumor.

Using "target tracking" software from the space and missile tracking programs, the Neurotron 1000 follows the target site and adjusts for patient movements up to 1 centimeter (one-third inch) and stops automatically if the patient moves more than that. This keeps the beam focused precisely on the "target" and *eliminates the need for the frame*.

It is possible to fix the frame to the bony skull and be confident that the tumor will stay in that fixed relationship to the skull. But there is no such bony structure which can be used to cause a rigidly fixed relationship around, for example, a kidney tumor. Up to this time, this has limited the application of this methodology mostly to brain tumors.

Dr. Adler has treated a number of VHL patients with hemangiomas of the brain, and is hopeful that the frameless treatment can be applied to spinal and kidney tumors as well. "While designed initially to treat cancerous tumors of the brain," Dr. Adler says, "the Neurotron 1000 system is also being designed for treatment of other body sites. This achievement has the potential to dramatically change the treatment of many solid malignancies which represent the vast majority of cancers. Compared to alternatives, *Accuray's* precision radiation treatment promises to be a more effective, less painful and less expensive therapy for many cancer patients."

Today's machines move in only one or two planes, so that they deliver a spherical dose. If the tumor is not spherical, but is irregular in shape, overlapping spherical doses to cover the tumor result in some areas of undertreatment, and some of overtreatment. As a result, not all irregular tumors can be treated today with stereotactic radiosurgery.

The Neurotron 1000 moves on six planes or axes, which allow it to plan a dose distribution the shape of the tumor and treat the entire tumor area evenly.

This project has been funded in part by the Murray Foundation, with the express intent of creating

advancements which will facilitate treatment without invasive surgery of VHL hemangioblastomas of the brain, spine and other organs. "People with VHL," says Mike Murray, "often have to deal with a series of invasive surgeries which themselves take a physical and mental toll on the human being. The Neurotron 1000 will provide a non-invasive approach to treating many of these tumors until we have a genetic treatment or cure for VHL."

This machine is also expected to lead to reductions in the cost of treatment by making it possible to treat more patients in the same length of time. Today, the treatment team is intensely involved with a single patient during an elaborate setup procedure which has to happen after the fixing of the frame. With the Neurotron 1000, the CT's and MRI's can be done in advance so that the time in the treatment room is minimized.

Dr. Joseph G. Depp, president and CEO of *Accuray*,² stated, "The first patient treatment officially launches clinical trials which are required by the FDA before commercial marketing of the system can commence in the United States. During the next few months, *Accuray* will be installing additional prototypes at Newport Diagnostic Center in Orange County, California; Simmons Cancer Center at the University of Texas in Dallas; Shadyside Hospital in Pittsburgh, Pennsylvania; Georgetown University Medical Center in Washington, D.C.; and the Cleveland Clinic in Cleveland, Ohio. A prototype will also be installed by Marubeni in Japan to provide clinical data for Japan's Ministry of Health."

1. For an excellent laymen's explanation of central nervous system hemangioblastomas and surgical options, see *Advances in Treatment of CNS Lesions*, a presentation by Dr. Haring J.W. Nauta, University of Texas Medical Units, Galveston, and Dr. John Adler, Stanford University Medical Center, on the audio tapes of the VHL Family Alliance meeting in Kansas City, April 1994. See page 15 for ordering information. 2. *Accuray*, 1715 Wyatt Drive, Santa Clara CA 95054 (408) 982-9900. □

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To order, see Page 15

The audio tapes of the Kansas City conference were great! I picked up all kinds of information I'd missed when I was there. The audio quality is really excellent -- they're a pleasure to listen to! Many thanks to Brian the sound man for a job very well done. -- *Fran M., Michigan*.

Science Isn't Enough

Charles Wilson, M.D.

When you or someone close to you has a diagnosis of a brain tumor there are three immediate effects. One is practical -- how will this affect your life? The second is physical -- you may have some after-effect from the tumor or its treatment. And the third, on which I will dwell, is psychological.

I'd like to give you ten very specific things that you can do . . . not instead of conventional medicine, but in addition to it.

And the theme of this is that there are certain things you can't control -- you can't control that you have a tumor, you can't control where it is. But there are things you can control. So, rather than be dismayed and depressed over those things that you can't control, focus on things over which you do have some control -- and I can assure you that in these things I will recommend there is real power.

1. Seek psychological help

On the front page of today's San Francisco *Examiner* is a headline: "It's Mind and Matter," and it's a story about sports psychology. These are world-class elite athletes who often scoffed at the idea of needing any kind of psychological help, but you'd have to be pretty out of it not to recognize that sports psychology is a big thing. The Oakland A's, for instance, have a full-time sports psychologist.

So there has to be a power in psychologically examining ourselves...who we are...our fears...our strengths...that helps us in dealing with our lives and whatever life may have brought us. But, I think more important from your standpoint, is that this type of examination can empower you in a way that has often been shown to increase your chance of survival and have a profound effect on longevity.

By this (seeking psychological help) I mean learning coping skills and how to manage stress, whether by meditation or whether by some breathing exercise -- but something you can be taught to do that gives you some sense of having control over what's happening.

2. Socialize, socialize, socialize

Socialize with your friends, with your family. If you are socially isolated with cancer of the breast your mortality is twice as high as those who are not socially isolated -- a fairly impressive statistic.

If a patient has at least one confidant, a person to whom they can tell everything...the bad and the ugly...the fears...someone with whom they can absolutely spill everything, their chances of surviving over any given period of time is approximately doubled. And there are those who say a pet can have the same effect.

3. Join a support group

Find some place where you can go to express your worries, your concerns -- someplace where you can

laugh, where you can bond with people who happened to end up in the same boat that you find yourself in.

4. Learn about your disease

Initially it may be very stressful. You read about brain tumors, you see a picture of a scan, you can picture your own scan, you may get tight in the throat or feel sick in the stomach. But once you have learned something about your disease you become a partner in the team of people who will determine your future -- that is your treatment and your management. Over time, learning about your disease improves your ability to cope.

5. Grieve

Elizabeth Kubler-Ross says you've got to grieve. It's a terrible thing that's happened. Your family grieves, you grieve, and it's okay, it's natural; but you've got to do that and then get on with life.

6. Face your own mortality and cope with dying

As I whizzed by my 60th birthday -- and I'm looking forward in a few months to my 65th -- in a way I came in touch, finally, with my own mortality. I realized that life is finite. Life can be beautiful, but it certainly is not forever. A part of the end of life is dying and it's okay to think about it; it's okay to wonder about it -- but it's not okay *not* to think about it.

7. Try to learn something about imaging

I used to think this was not important, but I'm now persuaded that for some people (maybe not for you and certainly not for all), it can be very powerful. There are many ways of doing it. For instance, in San Francisco, Dr. Martin Rossman¹ has introduced interactive guided imagery, which is just one form.

8. Decide to develop some new interest or some new activity

Despite all that's happened, decide that you're going to develop some new interest or some new activity. Maybe you've always wanted to go down to Glide Memorial Church in San Francisco and become a volunteer. Maybe you've always said, "I'd like to take a class in Greek mythology," or maybe you'd like to learn jazz saxophone, write a book, or learn to paint. Do something that's different, that's compelling, and that will give you a sense of new accomplishment.

9. Exercise and eat properly

You may say, "I have a weak arm or a weak leg," or "I'm on Decadron and don't have much energy." Well, if you want to be inspired, just look in some day on the special Olympics or watch a wheelchair marathon. Exercise gives you a sense of discipline; it gives you a sense of control, and it makes you feel better. Psychiatrists learned long ago, that probably exercise for depression is more effective than the

couch. And eat properly. You should be healthy. Just because you have a brain tumor does not mean that the body that happens to be harboring this tumor should be allowed to go along without maintenance and improvement. Vitamins will help.

10. Get in touch with some higher being.

It can take many forms, it can be many spirits. It will give you a strength and it will give you a peace. Physicians are now learning that when religion matters to a patient, they should take that seriously.²

The mind can be a powerful determinant in healing and recovery. Believe that you can beat this disease against all odds. Focus on those things in your life that you can control. But above all, put your trust in a higher spirit, because the strength that you can

find in a spiritual experience has power beyond your imagination.

It's a beautiful day, it's the first day in the rest of your life, and I hope that my God and your God will bless you all.

This article is excerpted from the closing address of the Third Biennial Brain Tumor Conference delivered by Dr. Charles Wilson, Professor and Chairman of the Department of Neurosurgery at the University of California, San Francisco, and a leading pioneer in the field of brain tumor research. Dr. Wilson's talk was entitled *Science Isn't Enough*. Reprinted with permission of the National Brain Tumor Foundation, 1-800-934-CURE. 1. Martin Rossman, M.D., *Healing Yourself*, New York, Pocket Books, 1994. 2. See also *Beyond the Relaxation Response: How to Harness the Healing Power of your Personal Beliefs* by Herbert Benson, M.D., with William Proctor (New York, Times Books, 1984). □

Greetings from East Berlin

-- Peter and Sylvine Z, Berlin, Germany

When Joyce Graff and Peggy Graham attended the VHL Symposium in Freiburg in April, they had the privilege of meeting a number of people from Germany who had VHL. Whenever VHL families find one another, they can't wait to tell their stories. Even with language difficulties, we shared our stories — a few key words, some sign language, lots of clasped hands and hugs. Dr. Otmar Wiestler and Dr. Luitgard Neumann helped with translation, everyone worked with whatever English or German they could muster, and we shared.

Peter and Sylvine Z. drove ten hours to Freiburg from East Berlin. After the Second World War, when Germany was divided into occupational sectors. In 1961 the Berlin Wall was built. For 28 years it stood not only as a barrier within the city, but as a symbol of the political wall between the Soviet East and the West.

Peter was born at the end of the War, in January 1945, in a bomb shelter. He grew up in a small town west of Berlin, just east of the border between East and West Germany. His parents were both physicians.

The inhabitants of East Berlin and East Germany were not able to go to the West once the Berlin Wall and the Wall between East and West Germany were built in 1961. Starting in 1980 people could apply for a permit to visit the West in case there were important family affairs, like the burial of a relative. One of Sylvine's brothers living in West Germany was able to visit the Z. family a few times, but they were not able to return the visit.

While Peter was studying at the University in the 1960's he began to have headaches which came more and more frequently. The university clinic told him he was simply studying too hard, that he should relax and enjoy college life more. But he had been doing that all

along. When the headaches became more severe, he went back to the clinic. This time they found a brain tumor. His parents had him transferred from the university clinic to the well-known University hospital Charité in East Berlin. He almost died in the ambulance. In the Charité he was diagnosed with VHL. He was operated on for the brain tumor, received very good care, and was followed there for a number of years.

He went back to college, finished his program, and became an engineer, working in radiation protection for a nuclear power plant.

In the middle 1970's he began experiencing weakness in his arms. His fingers cramped up and he had unbearable pains in the left side of his back. After the removal of a spinal tumor the problems disappeared.

Peter met Sylvine in 1970. She grew up east of Dresden. They were married in 1976. At that time one didn't know that VHL was hereditary. She studied to be a physical therapist in Berlin. When she studied about von Hippel-Lindau disease, she never imagined that her own husband would be affected.

In 1992 Peter had another brain tumor. The recovery from this one was slower — two months in the hospital, and then a month in a rehabilitation center. Sylvine came every day after work to visit and cheer him along.

They have a 16-year-old son, a bright and vigorous boy who was off in Switzerland with his class on a field trip. "But not for skiing," Sylvine said. This was a tourist experience, not a skiing trip. For people who were unable to travel outside their country for forty years, it is very exciting for their teenager to go to another country with his schoolmates. When we went together to visit nearby Colmar in France, it was the second time Sylvine had been out of Germany. Each

Resources

There has recently been a dramatic reduction in the cost of **Voice Activation/Recognition software**, making it much more accessible to individuals. Complete software and hardware outfitting for your computer is now about \$2,000.

DragonDictate 30K is voice recognition with a 30,000 word vocabulary. It is available for the IBM PC for \$995. The user speaks into a headset, and DragonDictate transcribes from the verbal dictation. When you first begin to use it, you spend some time "training" the software to understand your particular pronunciation. When it transcribes something wrong, you correct it, and it learns. DragonDictate is excellent for people who have difficulty writing or typing. It can be used for editing or to control interactive software. Call Dragon Systems at 617-965-5200 or CompuCare Business Services, Inc., Box 578, Peterborough, NH 03458.

Similarly, Pre-Dict-ate for the Mac offers Voice Navigator, voice recognition, and word-prediction software which permit the user to control the Mac with voice commands. Call Apple Computer Aisle 17, 1-800-600-7808.

Options for Independence. For people with impaired mobility, and/or speech problems, a reference guide "The Best 25 Catalog Resources for Making Life Easier," by Shelley Peterman Schwarz, highlights 25 mail order companies offering hard-to-find products for people who would benefit from making their lives easier. These products are often not readily available in regular stores.

Ms. Schwarz writes a syndicated column, "Making Life Easier," which appears in more than 25 newspapers and magazines across the United States. Through her experiences Ms. Schwarz found that people with physical limitations were often frustrated because they didn't realize adaptive devices and products were available to help them, let alone where to find them. Categories of products available include cooking/preparing food; Making your home more accessible; Communicating; Enjoying leisure/recreational activities; Completing lawn/garden work. Most of the catalogs are available free of charge.

US \$10.45 postpaid, Canada \$11.95 from Shelley Peterman Schwarz, 933 Chapel Hill Road, Madison, Wisconsin 53711 (608) 274-4380. Please allow 4-6 weeks for delivery.

Dialing the phone. The telephone company has a number of clever adaptive devices to assist people in dialing the telephone. For people who need more assistance, the telephone company offers "Dial Zero" service. Once you have registered with the telephone company as eligible for this service, you simply press zero, and the operator will connect you with any number for the direct-dial rate. □

Peter and Sylvine Z. from Berlin

new step — beyond East Berlin, beyond East Germany, beyond Germany itself — is a new adventure.

Last fall one of Peter's sisters, a pediatrician, read in the *Deutsche Arzteblatt*, a journal for physicians, an article about VHL by Dr. Hartmut Neumann of Freiburg. She advised Peter to see Dr. Neumann. Peter arranged an appointment at the Albert-Ludwigs University Clinic for a full check-up. For the first time, Peter had a thorough examination for VHL involvement throughout his body. They found some cysts in his pancreas and kidney, neither of which required treatment. He has one spinal tumor which causes problems. He is very happy to know the full story at last, and to have a full idea of what is happening, not just a series of surprises.

Peter and Sylvine came to Freiburg for the Symposium to learn more about VHL. In particular, they are interested in DNA testing. They have sent Peter's blood sample off to Dr. Hiltrud Brauch in Munich for analysis. They want to find out whether their son inherited the faulty VHL gene or the healthy one, so that they will know how much screening he needs to stay healthy. Peter's parents' blood is also being tested, since neither of them has shown any signs of VHL. It may be that Peter is the first in his family to have a flaw in the VHL gene.

As a physical therapist, Sylvine is very health conscious and works hard to keep them on a healthy diet — lots of vegetables and vitamins, and little meat. Peter says he lost all fear of death after an experience once with a coma. He has a strong will to live, and a great optimism about medical progress and improvements in medical techniques. He and his family live a healthy, happy life.

Our thanks to Gerlind Banskberg, Stuttgart, for translation assistance in the preparation of this article. □

Von Hippel-Lindau: Ocular Complications

by Lloyd M. Aiello, M.D., and Jerry Cavallerano, O.D., Ph.D., Beetham Eye Institute, Joslin Diabetes Center, Boston, Massachusetts

In 1904 Eugen von Hippel, M.D., described retinal *angiomatosis retinae*, the retinal angiomas we now recognize as occurring in von Hippel-Lindau disease. About 30 years later Arvid Lindau, M.D., described angiomas of the spine and cerebellum. We now recognize von Hippel-Lindau disease (VHL) as a genetic condition which involves the growth of angiomas in vascular-rich areas of the body, including the retina, the light-sensitive tissue of the eye which is important for our vision. In addition to the eye, the lesions frequently affect the central nervous system, the pancreas and the kidney, as well as numerous other organs in humans.

Von Hippel-Lindau disease is transmitted genetically in an autosomal dominant fashion. Consequently, a child of a person with von Hippel-Lindau disease has a 50% chance of having the condition. The expressivity of the disease is often delayed, and a person who has the disease may not be diagnosed for many years. While an angioma from VHL can grow in the brain, spinal cord, adrenal glands, kidney, pancreas, and other organs, an angioma in the eye is the only VHL hemangioma that can be directly observed.

Review of Retinal Anatomy

The human eye can be considered as a forward extension of the brain. Embryologically, the eye develops from the same tissue as the brain. The human eye functions by focusing images on a light sensitive tissue within the eyeball. This tissue, the retina, is highly vascularized, receiving nutrients from blood vessels within the retina itself and from circulation in the underlying choroid. (Figure 1) The retina is able to take the light signal that is focused on its surface and convert the signal to a neural message which is conducted to a cortical area of our

Fig. 1: Illustration of the human eye. brain that is responsible for sight.

The sclera, which is the "white" of the eye, provides the eye with a degree of rigidity. The eye is usually divided into two main sections, the anterior segment and the posterior segment. The anterior segment includes the cornea, the iris (the colored portion of the eye), and the lens of the eye. The internal structures of the anterior segment of the eye are bathed with a fluid called aqueous.

Behind the lens of the eye is the posterior segment of the eye. The posterior segment is filled with a highly viscous substance called vitreous. The vitreous pushes against the retina of the eye. The light sensitive retina of the eye contains blood vessels which deliver oxygenated blood to the eye (arteries) and blood vessels which drain deoxygenated blood from the eye (veins). Capillaries are small vessels where arteries and veins meet. In a healthy eye, fluids are well contained in the vessels of the retina. In some disease conditions, fluids, lipids, and other molecules leak from the vessels in the retina. These leaking areas can be identified by a special procedure called fluorescein angiography, where a fluorescein dye is injected into a vein in a person's arm, and the flow of the dye can be traced in the retina of the eye. In a healthy eye,

the fluorescein does not leak out of the vessels. In disease cases, such as retinal angiomas, dye can leak into the retina.

When angiomas are examined using fluorescein angiography, the classical lesion will show leakage of dye from the tumor. The feeder vessels are easily visualized, and shunting of blood through the tumor is likely to be present. Blood flow is generally rapid through the tumor, and there is poor perfusion or blood flow through capillaries surrounding the lesions. Fluorescein dye which has leaked from the tumor will collect in the retinal tissue, causing hyperfluorescent areas of fluorescein pooling -- an area which "lights up" on the photograph.

Retinal Disease in VHL

A retinal angioma observed during a routine eye exam may be the first clinical sign that a person has VHL, and may be a reason for conducting additional specialized testing. Other patients may experience other VHL issues long before retinal angiomas occur. Even if no eye lesions occur early, it is important to continue screening throughout the patient's life.

In the earlier stages (see below) of development, the angiomas are usually asymptomatic, causing no change in visual acuity. The lesions are painless in the early stages. Angiomas in the eye may be solitary or multiple, and the angiomas occur in both eyes approximately 50% of the time. These retinal capillary angiomas develop in approximately 57% of persons who have systemic lesions of VHL (lesions in other organs of the body).

Retinal angiomas arise in the capillary bed of the retina, in the vascular tissue between the arterioles and venules in the circulatory system. (Figure 2) The initial appearance of the lesion may be as a small micro-aneurysm, which then

may assume the appearance of a red nodule and eventually a larger, orange-red tumor. Arterio-venous shunting in the tumor leads to dilatation and tortuosity [twisting] of the artery supplying blood to the tumor and the vein draining the tumor. When the angiomas are untreated, the tumor will leak plasma and other blood constituents into the retinal tissue, which leads to retinal hard exudates, edema, serous retinal detachment, bleeding, and, in some cases, eventually blindness.

Natural History of VHL Eye Disease

Different researchers have identified various stages of retinal angiomas in VHL. It is important to remember as we describe these stages that progression is not inevitable. The ophthalmologist will be working with you to halt or slow the progress of the disease.

The lesions can be single or multiple and there is considerable variation in presentations and progression through the various stages. While the different categories vary depending on the categorization method used, five clinical stages can definitely be identified as follows:

Stage 1: Preclassical. (Figure 3) In the preclassical stage small capillary clusters can be seen in the retina. In this preclassical stage the lesions appear as simple red spots or micro-aneurysms, similar to those

associated with diabetic retinopathy or hypertension, among other diseases. These lesions are easy to overlook, especially if the retinal examination is conducted without pupillary dilation. These lesions are most likely to be asymptomatic (i.e., no change in sight or vision is noted).

Stage 2: Classical. (Figure 4) Although the presentation is variable, the classical retinal hemangioma is a knot of vessels with a large artery as a feeder vessel leading to the hemangioma and a large vein as a draining vessel. In the early stages of the classical lesion, the hemangioma may appear as a nodule with normal-sized vessels on either side of the nodule. The nodule can be either light or dark reddish or pale grey in color. As the lesion develops, the feeder vessels can become 2 to 3 times the size of a normal retinal artery or vein. These vessels usually assume a tortuous configuration. Eventually the lesion becomes elevated, usually growing toward the vitreous of the eye, and subsequently leading to the next stage of the disease. Depending on location in the retina, these lesions may be symptomatic, although most peripheral retinal lesions at this stage remain asymptomatic.

Stage 3: Exudative. In the exudative stage of the hemangioma, leakage of fluid across the walls of the lesion results in retinal edema, retinal exudates, and leaked proteins and plasma from the blood. The accumulation of fluid which has leaked from the fenestrated walls of the angioma cause retinal swelling and edema, which leads to the next stage of the disease. Depending on location, lesions which have reached the exudative stage may also be asymptomatic. Lesions which affect the macula, the area of the retina responsible for fine visual tasks and sharpest acuity, are likely to cause symptoms. These symptoms may include blurring of vision, loss of visual acuity, or distortion of vision. In some cases, straight lines

may appear warped or distorted.

Stage 4: Retinal detachment. In the retinal detachment stage of the disease, the retinal exudates and edema result in a lifting of the light sensitive retinal layer from the underlying tissue. Localized retinal detachments are likely to occur in all but the smallest angiomas. This retinal detachment has a tendency to spread, particularly as the lesions permit additional exudations. Symptoms similar to those described for stage 3 above are usually present at this stage. A retinal detachment may cause a curtain-like effect across the field of vision, or sudden, marked loss of all or some vision.

may appear warped or distorted.

Stage 5: End stage. In the end stage of the disease, the retinal detachment has progressed. A long-standing retinal detachment can result in changes in the retinal tumor itself. Glial or scar tissue is likely to form. A painful glaucoma is likely to develop, vision is permanently and completely lost, and the eye becomes phthisical or shrunken.

Treatment of Retinal Angiomas in VHL

Various strategies have been attempted to prevent visual loss or blindness from VHL retinal angiomas. Some of these treatments have resulted in limited success. Early, unsuccessful treatments included X-ray irradiation, which did

Figure 2: An illustration of a retinal angioma, showing interwoven capillaries arising between the arterial and venous system of the retina.

more harm to healthy tissue than to the tumor itself. Other types of radiation treatment using radioactive seeds resulted in some limited success, but results were variable and unpredictable. Diathermy has proven to be somewhat successful, but a major breakthrough was achieved in the mid-1950s by Professor Meyer-Schwickerath of Germany with photocoagulation of the tumor itself.

The advent of the argon laser improved the ability of ophthalmologists to treat the tumors, and in some cases the feeder vessels, with a significant level of success, especially when the tumors were treated in the earlier stages and when the tumors were comparatively small. Direct treatment of the tumors and the feeder vessels with laser photocoagulation is now the treatment of choice for tumors that are less than 2 millimeters in diameter. Various lasers have been used for this photocoagulation with a high rate of success, including the dye yellow laser. Multiple laser treatment sessions are usually necessary to obliterate a lesion fully. Regrowth of a lesion is not uncommon. In general, early diagnosis and treatment of the tumor result in favorable prognosis for destruction of the tumor and preservation of sight. Cryotherapy, either alone or as an adjunct to laser photocoagulation, is also an important part of the armamentarium [set of options] for the treatment of angiomas.

Side effects and complications of laser treatment include hemorrhage, retinal detachment, increased leakage into the retina, retinal holes, loss of visual field, or wrinkling of the retinal surface. In general, the benefits of laser treatment for angiomas outweigh the risks of treatment, since many untreated lesions usually result in painful blindness.

When lesions are too large to be treated with lasers, other methods have been successfully undertaken. Eye wall resection, which involves the surgical removal of the tumor

Figure 4A: This angioma shows a large feeder vessel leading to the angioma with a large vein as a draining vessel. Figure 4B: A fluorescein photograph showing the large feeder vessel and drainage vessel of a VHL angioma. The angioma appears bright white in this photograph since it has filled with fluorescein.

through a cut in the sclera of the eye, is a drastic procedure with significant risk, but has been successful for treating a number of large tumors, some as large as 4.5 millimeters.

Examination Guidelines

During routine eye examination, any patient with a retinal lesion suggestive of VHL, and any person with a family history of VHL who has a suspicious retinal lesion, should be referred for a systemic neurological and abdominal evaluation. Relatives of a person diagnosed with VHL should also be screened for any manifestations of the disease. Ocular lesions are frequently the first diagnosed lesions, but VHL lesions can be present in the cerebellum, medulla, or pons of the brain, or in the spinal cord. Visceral tumors may be present as cysts of the kidneys, pancreas, adrenal glands, or epididymis. Treatment of retinal tumors is with photocoagulation (usually with a laser), cryosurgery, or penetrating diathermy. Other treatments may be attempted if the tumor is too large. Possible complications of the procedures include serous detachment of the retina, macula or optic nerve damage secondary to treatment, or failure of the lesion to respond to treatment.

All persons with diagnosed VHL should have a thorough eye exami-

nation at least once a year. It is important to note that early diagnosis and treatment for retinal lesions of VHL can result in significant savings of vision. Family history and retinal examination for other family members of a person with VHL are advisable. Since VHL is a multisystem condition, a person who has a retinal hemangioma should be referred for additional medical evaluation, and people with VHL in other systems should be referred for eye examination. Silent lesions in other organs of the body may be identified, and early treatment may prove life saving. Genetic counseling, family screening, and construction of a family tree are advisable.

Additional Reading

Bedell, A.J. "Angiomatosis Retinae." *American J of Ophthal* (1931); 14:389-411.
 Welch, R.B. "Von Hippel-Lindau Disease: The Recognition and Treatment of Early Angiomatosis Retinae and the Use of Cryosurgery as an Adjunct to Therapy." *Tran Am Ophthal Soc* (1970); 68:367-423.
 Hardwig, P., Robertson, D.M. "Von Hippel-Lindau Disease: A Familial, often lethal, multi-system Phakomatosis." *Ophthalmology* 1984; 91:263-272.

Acknowledgements: Figures 1 and 2 courtesy of Vincent Giovanucci, O.D. Figures 3 and 4 courtesy of Denis Fleming.

Driving Tips for Monocular Individuals

by Singular Vision Outreach, submitted by Cindy D., Missouri

Many newly monocular visioned individuals are concerned about their ability to drive. Singular Vision Outreach (SVOR) volunteers contributed these helpful suggestions to ease the uncertainty. Because of our individuality, a suggestion may work well for one person and not be helpful to another. Singular Vision Outreach hopes that you will find several helpful tips from this list that will get you back in the driver's seat.

Devices for the car

Devices can be added to your vehicle that help compensate for the loss. Most of the devices listed can be obtained from local auto part stores or auto part supply catalogs.

1. Have sideview mirrors on both sides of your car.
2. Convex bubble mirrors can be a valuable addition to a sideview mirror, especially on trucks and vans. (Be aware that objects viewed are closer than they appear.)
3. Obtain rearview mirrors that offer multiple or panoramic views.
4. Install curb feelers.
5. Backup warning beepers can be installed to alert others to your actions.
6. Have a rear window wiper and rear window defogger.
7. Use window wipers with double or triple blades.
8. Install Quartz halogen headlamps for increased night visibility.
9. Items such as hood ornaments, hoods with a center ridge, and antennas can be used as reference points on your vehicle to aid in lane position, location of curbs, etc.
10. A sunglasses case that mounts to the dash provides a place for, and easy access to, a good pair of sunglasses.
11. Clear plastic lenses, mounted in the rear windows of vans, allow the driver to view nearly straight down behind the vehicle.
12. Equip motorcycle helmets with a bubble face mask.
13. Liquid window treatments that repel water can be applied to windshields.
14. Do not put decals on windows.
15. Do not place stuffed animals held on by suction cups on your windows.

Aids for the driver

1. Purchase a good pair of sunglasses. Sunglasses with ultra-violet blocking lenses as well as neutral gray or green, polarized lenses can be purchased. Glasses should be the correct refraction. Ask your health care professional for advice on your specific needs.
2. Check with your doctor about glasses designed for night driving.
3. Avoid using photogrey lenses as they are slow to react to changes in light.
4. Never wear sunglasses at night.

Hints for driving and parking

1. To enhance depth perception, as in viewing a photograph, consider larger objects as closer than smaller objects.
2. Objects that rapidly increase in size are getting closer. *Slow Down!*
3. To maintain safe distances, be able to see pavement between the hood of your vehicle and the rear bumper of the vehicle in front of you.
4. Never switch lanes without turning your head. If unsure what is in your blind spot, avoid lane changes.
5. Stay in the middle lane of the highway to avoid merging and exiting traffic on your right. For those who have lost vision in the left eye, travelling in the far left lane can eliminate concern for cars on your limited sighted side.
6. Enlist the help of your passengers as spotters when navigating difficult driving situations.
7. Leaving windows slightly open can enable the driver to hear cars that may not be seen.
8. Hang a tennis ball from the roof of the garage so that it hits the windshield when the car is in the proper location.
9. Use a point of reference when pulling into a garage. Paint a mark on the side of the wall or use a window for a guide.
10. When parallel parking, sight the parking meter in your rearview mirror and use it as a reference point.
11. When parking a truck with a camper, use the handle on the camper shell as a reference point.
12. If use of mirrors is difficult when parallel parking, turn and raise your body to view the rear of the car when backing.

Safety tips

1. Know your vehicle's capabilities and limitations. Avoid dangerous situations.
2. Keep windows and windshields clean and clear.
3. Keep wipers in good condition and washer tank full.
4. Avoid drinking from a glass or cup while driving. If you do drink, use a straw.
5. Slow down early for stop signs.
6. Travel on well lighted streets.
7. Avoid having tinted or smoked glass on your vehicle.

An informal survey concluded that monocular drivers have fewer accidents and their accidents are usually minor ones. As one Singular Vision Outreach volunteer commented, "I'm just like any other driver on the road. I've just taught myself to concentrate on my driving. I'm cautious and I leave margin for error." Sound advice for every driver behind the wheel!

For information or extra copies of *Driving Tips* write to Singular Vision Outreach, P.O. Box 1451, Maryland Heights, MO 63043. See also *A Singular View, The Art of Seeing with One Eye* (3rd edition, 1985), by Frank B. Brady, P.O. Box 4653, Annapolis, MD 21403 □

DNA Testing

by Dr. Eamonn R. Maher, University of Cambridge, England

The mapping of the VHL gene to the tip of the short arm of chromosome 3 in 1988 was the first step towards developing a DNA test for VHL disease, and the isolation of the gene last year seemed to promise that DNA testing would become available for many families.¹ So how far have we progressed toward this aim?

By 1992 the position of the VHL gene had been pinpointed so accurately that it became possible to track a faulty gene through a family using DNA markers that were close to the gene. Although the gene tracking (or genetic linkage) approach could be as much as 99% accurate, there were a number of limitations. Firstly this technique can only be used when there are two family members known to be affected by VHL disease. It requires blood to be available from several key relatives. Even in families with a suitable structure for gene tracking, DNA markers may be uninformative for predictive testing. However our experience of using gene tracking in Cambridge to determine the risk of 166 people with a family history of VHL disease was that in 86% of cases we were able to produce a low risk (1% carrier risk) or high risk result (99% carrier risk). So, in families with a suitable structure, DNA testing by gene tracking will usually be informative.

What about those families in which gene tracking is uninformative or not possible? The isolation of the VHL gene held out the hope of DNA testing by testing directly for the faulty gene (mutation) rather than gene tracking. Furthermore direct mutation analysis is more accurate than gene tracking. During the past year we and other research teams have been trying to determine in what proportion of families it would be possible to detect a mutation. Recent results suggest that the faulty gene can be identified in 70-75% of patients. A wide variety of mutations have been identified, but once a mutation is identified all family members can be screened.

It is well known that whereas phäochromocytoma² is rare in some VHL families, in others it is the most common complication of VHL. By comparing the types of mutations in families with and without phäochromocytoma we have found that some types of mutations are associated with a higher risk of phäochromocytoma than others. Thus when a large piece of the gene is missing (deletion) or the mutation is predicted to cause a shortened protein the incidence of phäochromocytoma is low. Missense mutations (when a single amino acid is changed) have a higher risk of phäochromocytoma and some missense mutations are particularly prone to cause phäochromocytoma. This work needs to be con-

firmed, but it suggests that in the future it may be possible to identify patients who are particularly likely to develop phäochromocytoma and so allow careful screening for this complication.

The work I have described is still at a research stage, but over the next two years DNA testing by direct mutation analysis will be introduced on a service basis for the 70-75% of patients in whom the faulty gene can be identified. The main advantage of DNA testing is that relatives shown not to be gene carriers can be reassured and spared repeated screening. However there are disadvantages and anyone wanting to undergo DNA testing should carefully consider the possible effects of a high risk result on insurance and employment prospects, etc., and emotional repercussions within the family of finding a child or relative is at high risk. Genetic testing of children is controversial, but in the case of VHL our policy has been to offer testing from age 5 years as after this the DNA result can be used to modify screening and so that test can benefit the child. Clearly even when DNA testing is possible it requires careful thought and should not be viewed as a simple blood test!

1. See the March and June 1993 issues of *VHL Family Forum* for background on genetic research, linkage analysis, and the finding of the VHL gene. Back issues available, see page 15.

2. A tumor of the sympathetic nervous system, usually in the adrenal gland. See the *VHL Handbook*, and the December 1993 issue of the *VHL Family Forum*.

Sources of DNA Testing

If you are not already enrolled in a research project, your genetic counseling center will probably need to contact one of the following laboratories to obtain DNA testing for you:

U.S.: Ms. Corinne Boehm, DNA Diagnostics Laboratory, Center for Medical Genetics, Johns Hopkins Hospital, Baltimore, MD 21205, Tel: 1-410-955-0483; Fax: 1-410-955-0484.

England: Dr. Eamonn R. Maher, Clinical Genetics, Addenbrooke's Hospital, Hills Road, Cambridge CB2 2QQ, U.K., Tel: 44-223-216446, Fax: 44-223-217054.

Germany: Dr. Hiltrud Brauch, Labor für Molekularpathologie, Klinikum rechts der Isar, Trogerstraße 32, D-81675 München, Germany, Tel: 49-89-4140-4592; Fax: 49-89-4180-5215. □



Great News from Down Under!
The VHL Handbook is now available
translated into Australian! Contact
Jennifer Kingston, address on page 14.

New Board Members

**Join us at the Annual Meeting
Boston, April 1995!**

At the meeting in Kansas City in April the following four people were elected to the Board of Directors. Please support them in their efforts to lead the activities of the Alliance.

Patti Kohlen, California

"I live in the central coast area of California, having moved to the United States from England in 1972. My husband, Ken, teaches architecture at the local university and my daughter, Corinne, attends junior high. I share a second-grade teaching position with another teacher. I also have my own art consulting business, selling the work of many of our local artists and the major art publishers. I am actively involved in our small town, especially in the arts and with our Chamber of

Commerce.

"In October 1992, I was diagnosed with VHL shortly after my 44th birthday and 20th wedding anniversary. Within seven months of diagnosis I had three surgeries: spinal, brain, and kidney. I am tremendously pleased to be working with the VHL Family Alliance in the capacity of Public Relations Chair. Through teamwork and networking we can further education, research and support and move towards our goal of making VHL stand for Very Happy Life."

Patti has been a very active member, and since February 1994 has been Chair of the Public Relations Committee. She has undertaken a number of programs to gain name-recognition for Von Hippel-Lindau disease in dignified ways. She has taken courses in Public Relations to enhance her own skills in this area, and prepared the press kit and press releases for the Kansas City meeting.

Peggy Marshall, Mississippi

"I am a Day Care Provider and have been in business for the past fifteen years in our home in Corinth, Mississippi. I really love taking care of children and have found them to provide the best therapy possible when going through the difficult times that VHL sends our way. I am part of four generations of family with VHL, including my mother, two sisters and a brother, daughter, granddaughter and two nephews. We first became aware of VHL in 1962.

"My husband, who has been my life support for 33 years, is right by my side in support of the VHL Family Alliance. We have lived all over these United States from coast to coast and know first hand the difficulties of dealing with minimal medical information, and the need for knowledgeable professional medical attention.

"We are proud to be involved and look forward to being an active part of the VHL Family Alliance, adding our support in any way we can."

Peggy and her husband Don initiated the Mississippi/ West Tennessee Chapter last spring, making it the oldest and one of the most active chapters. Since the geography is very wide-

spread, the chapter operates primarily by telephone and mail. Don handles production of materials for the Alliance. Peggy's knowledge, courage and steady hope are inspiring, and her gentle wisdom is always welcome.

Altheada LaVerne Johnson, Brooklyn, New York

"I am a Registered Dietitian with a master's degree in Clinical Nutrition. In my last job I worked as a Nutritionist in the out-patient department of Newark Beth Israel Medical Center in Newark, New Jersey. There, I did nutrition counseling with pregnant, breast-feeding and post-partum women. I am a member of Delta Sigma Theta Sorority, which focuses on public service and community activities. I am working on forming the New York Chapter of the VHL Family Alliance.

"I have been married to Fred for ten years and my favorite hobbies are crocheting and other needle work. I was diagnosed with VHL in 1988 and have had spinal cord, eye, and brain involvement.

"I was honored to be elected to the Board of Directors of the VHL Family Alliance. The rarity of this disease can make one feel very alone. Having the knowledge and support of the Alliance is very comforting."

As her first major effort to build the New York Chapter, Altheada undertook a mailing to 1000 physicians in the State of New York in the specialties which would treat VHL. As her Board project, she is working on outreach to U.S. minorities affected with VHL.

Thomas D. Rodenberg, Esq., Missouri

"I am an attorney practicing law in the state of Missouri. My mother and several of my brothers and sisters are affected with VHL. I too have been affected. Therefore, my involvement with the Alliance is very personal and heart-felt.

"As an attorney, my interest is in legal issues and insurance issues related to VHL. More specifically, my goals for my time on the Board relate to legislative activity as to health care concerns for persons with VHL and insurance coverage issues for persons who currently have health care coverage. I believe it is very important that persons with VHL demand the coverage which their policies afford and work to see that health care laws protect persons with genetic conditions. As an additional goal while on the Board, I intend to be involved with fund-raising from charitable organizations so that the work of the Alliance can move forward.

"My wife, Kathy, and I live in Independence, Missouri. We have four boys, James (age 5), Benjamin (age 4), Daniel (age 2) and Mark (age 6 months). I look forward to the opportunity to assist the Alliance as a member of the Board." □

Introducing our Medical Advisory Board

There are twelve medical professionals listed on page 15 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. In the next few issues we will introduce them to you. 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.

Lloyd M. Aiello, M.D., Massachusetts

Dr. Aiello is an Associate Clinical Professor of Ophthalmology at Harvard Medical School and the Founder and Chief of the Beetham Eye Unit of Joslin Diabetes Center in Boston. He holds a concurrent appointment as Chief of Ophthalmology in Surgery at New England Deaconess Hospital. He serves as President of the New England Ophthalmological Society and as a member of the National

Diabetes Scientific Advisory Committee, Diabetes 2000, of the American Academy of Ophthalmology.

A graduate of Harvard College and the Boston University School of Medicine, Dr. Aiello began his career as a physician in the Navy. He has spent the majority of his medical career doing research in diabetic eye disease and the use of lasers in medicine and biology. He has done extensive work on the use of lasers for the treatment of diabetic retinopathy and has made considerable contributions to this field.

He has been following a number of families with VHL in the Boston area for some thirty years. As a medical adviser to the Alliance he shares with us his expertise in the application of laser to retinal diseases, and his richness of experience with von Hippel-Lindau in these patients. His son, Lloyd Paul Aiello, has recently joined the Beetham Eye Unit as an ophthalmologist.

Debra L. Collins, M.S., Kansas

Debra Collins is a genetic counselor in the Division of Metabolism, Endocrinology, and Genetics at the University of Kansas Medical Center. She is also an Assistant Professor and Director of the Genetics Education Center, designing and implementing genetics education programs for science teachers in the secondary schools.

She has served on the Board of Directors of the

American Board of Medical Genetics (1989-1994) and of the National Society of Genetic Counselors (1982-1991), and as President of the NSGC 1988-89.

She graduated from the University of Texas in Austin, and earned her master's degree in Genetics at Sarah Lawrence College in Bronxville, New York. She served as an Advisory Board Member of the Human Genetics Graduate Program, Sarah Lawrence College, 1985-1992.

She has participated in numerous scientific articles, including several on VHL, such as "Computed tomography screening of the abdomen in the von Hippel-Lindau syndrome" (*Amer J Roentgenology* 139:505-510, 1982); "Diagnosis and management of asymptomatic renal cell carcinomas in von Hippel-Lindau syndrome" (*Urology* 21:2:146-150, 1983); "Von Hippel-Lindau disease maps to the region of chromosome 3 associate with renal cell carcinoma" (*Nature* 332:268-269, 1988); "Genetic flanking markers refine diagnostic criteria and provide insights into the genetics of von Hippel-Lindau disease" (*Proc Natl Acad Sci, USA*, 88:2864-2868, 1991); and "Von Hippel-Lindau syndrome: Screening at-risk family members and genetic counseling" (*Birth Defects Original Article Services*, Vol. 19(3):208, 1983).

Ms. Collins has been a valued Advisor to the Board of Directors of the VHL Family Alliance since its inception.

Haring J. W. Nauta, M.D., Ph.D.

Dr. Nauta was born in the Netherlands in 1946. The following year he moved with his family to Zurich where his father had accepted a post in Neuroanatomy. After four years in Zurich, the family again moved, this time to the Washington, D.C., area. Dr. Nauta's father imparted an enthusiasm for the neurosciences which was infectious to many people.

By the time he obtained his bachelor's degree from Duke University, Dr. Nauta was already well aware that his future lay in some aspect of the neurosciences. He then went to medical school at Case-Western Reserve University and while there was enrolled in the M.D./Ph.D. program, doing his dissertation under Dr. Raymond Lasek. At Case-Western Reserve Dr. Nauta was introduced to the neurosurgical operating room by Dr. John Jane, who imparted an immediate enthusiasm for neurosurgery and neuroscience research.

His research work involved stereotaxy, the accurate location of a definite circumscribed area within the brain, with minimal damage to the remainder of the organ, by moving a probe or electrode along coordinates for measured distances from certain external points or landmarks of the skull. This led, in turn, to a natural progression towards human stereotaxy.

Dr. Nauta then did an internship in surgery at the Toronto General Hospital and a residency in Neurosurgery at the University of Toronto. There he was introduced to functional stereotactic neurosurgery by Dr. Ronald Tasker. After completing residency, Dr. Nauta joined the faculty at

the University of Toronto and practiced for two years at the Toronto Western Hospital. He then went to the University of Texas Medical Branch in Galveston, Texas. In 1985 he joined the faculty in neurosurgery at the Johns Hopkins University where his energies were primarily focused on vascular neurosurgery and stereotactic neurosurgery, including stereotactic needle procedures in the CT scanner and LINAC-based radiosurgery.

In April 1993 he returned to Galveston to become the Chairman for Neurosurgery at the University of Texas Medical Branch. His plans are to help neurosurgery make more use of intraoperative imaging and treatment planning.

R. Neil Schimke, M.D., Kansas

Dr. Schimke has been with the University of Kansas Medical Center since 1967. He is Professor of Medicine and Pediatrics, and has been Director of the Division of Metabolism, Endocrinology, and Genetics since 1977, where he has built up a highly respected practice and regional resource center for inherited cancers, including von Hippel-Lindau disease. He served for five years as Chief of Medicine

at Kansas City Veterans Administration Hospital.

Dr. Schimke has held a number of leadership roles, including service on the Board of Directors of the American Board of Medical Genetics, 1983-1986. He currently serves on the Editorial Board of the *American Journal of Medical Genetics*.

He has participated in more than 150 scientific papers and books, including "Genetics and the Practicing Physician" (*J Ks Med Soc* 69:1, 1968); "CT screening of the abdomen in von Hippel-Lindau disease" (*A J R* 139:505, 1982); "Proximal 3p deletion in renal cell carcinoma cells from a patient with von Hippel-Lindau disease" (*Cancer Genet Cytogenet* 27:345-348, 1987); "Von Hippel-Lindau disease maps to the region of chromosome 3 associated with renal cell carcinoma" (*Nature*, 332:2265:269, 1988) and a book, *Genetics and Cancer in Man* (Churchill-Livingstone, Edinburgh, 1978).

Dr. Schimke is a native of Kansas, graduating from Leavenworth High School and the University of Kansas. He won a Fulbright Scholarship in Theoretical Organic Chemistry, under which he earned a graduate degree at the Rheinische Friedrich-Wilhelms Universität, Bonn, Germany. He also studied Mammalian Genetics under Dr. Elizabeth Russell at Jackson Memorial Laboratory in Bar Harbor, Maine.

He was recently designated "Medical Alumnus of the Year" (1993) at the Kansas University Medical School. He lives in Leavenworth with his wife and three children.

Robert B. Welch, M.D., Maryland

Dr. Robert B. Welch is Chairman Emeritus of the Department of Ophthalmology of The Greater Baltimore Medical Center, and Associate Professor of Ophthalmology and Former Director of the Retina Service at the Wilmer Ophthalmological Institute, Johns Hopkins University School of Medicine, Baltimore.

A native of Maryland, he graduated from Princeton

University and the Johns Hopkins University School of Medicine. He is a charter member of the Retina Society, and served as its President 1981-83. He served in the U.S. Navy and was Commended by the Army in 1986 for Outstanding Contributions to Army Ophthalmology.

His paper, "Von Hippel-Lindau Disease: The Recognition and Treatment of Early Angiomatosis Retinae and the Use of Cryosurgery as an Adjunct to Therapy," published in 1970 by the American Ophthalmological Society, is still a significant reference in the field. He too has cared for a number of families with VHL for thirty years, and has been involved in a number of papers, courses, and presentations on VHL, including a chapter on fluorescein angiography and VHL in *Interpretation of the Fundus Fluorescein Angiogram*, ed. Patz and Fine, Boston, 1977.

Dr. Welch was recently honored as President by the American Ophthalmological Society. □

In Memoriam

Frau Lena Chemin-Petit, daughter of Dr. Eugen von Hippel, passed away June 3 in Bad Soden, Germany. Frau Chemin-Petit kindly allowed us to publish a photograph of a portrait of her father in our June 1993 issue. In return, we

added her to the mailing list. This past April, she sent greetings which we read at the conference in Kansas City:

"Last year we wrote to you how glad we were to hear that someone from our family tree had worked in such an important subject and discovered the basis for your enormous medical movement, which we admire so very much.

Many things have developed — the research work was most effective, the Conference in the Kansas University and the Symposium in Freiburg — what a wonderful development!

Thank you very, very much for keeping us posted on your work. Best wishes for the future."

Mrs. Chemin-Petit was a very fine cellist, married to a famous German conductor and composer. The very talented von Hippel family includes a number of notable physicians, scientists and academics, several of whom are now in the United States. We are grateful to Dr. Eugen von Hippel for his astute observations and description of VHL, and to Frau Chemin-Petit and the von Hippel family for their kind interest in the progress of this organization toward better management of VHL.

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Toll-free in the United States and Canada: 1-800-767-4VHL

Email: MCI: Joyce Graff 3996438; internet: joyce.graff@zko.mts.dec.com

Editor: Joyce Wilcox Graff, 1-617-232-5946 (eve)

Adviser: Debra L. Collins, M.S., U. Kansas Med. Center, 1-913-588-6043

171 Clinton Road, Brookline, Massachusetts 02146 U.S.A.

Telephone: 1-617-232-5946; Fax: 1-617-734-8233

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Postmaster: Please send address changes to VHL Family Forum, 171 Clinton Road, Brookline, MA 02146.

Membership, Feedback and Order Form

Name: _____

Address: _____

City: _____ State: _____ Zip: _____

Country: _____ Fax: _____

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

My check includes: \$ _____ for dues (\$25 Family members & Friends, \$35 Professionals)
 \$ _____ tax-deductible contribution
☐ \$100 Special Friend ☐ \$250 Sustaining Members ☐ \$1000 Lindau Society
 \$ _____ tax-deductible contribution in honor of... (see below)
 \$ _____ for audio tape sets: Teamwork for Health (see below)
 \$ _____ for VHL Handbooks (@\$2 for members, \$4 non-members)
 \$ _____ for back issues @ \$1 for mem, \$2 for non-mem - index on request
 \$ _____ for T-shirts @ \$12 each (see below) - \$15 outside US
 \$ _____ Total enclosed

Please make checks payable to VHL Family Alliance. Our tax-exempt number is 04-3180414. -- Thank you!

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

All Members receive a membership certificate, 3-4 issues of the Forum this year, and copies of all Alliance publications. Free subscriptions are available where the dues are a hardship.

Donations greater than \$250 receive a special sponsor certificate.

☐ Audio version available if needed for a handicapped member

I 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

Friends and Family Contributions

☐ Please send a supply of _____ (number) puzzle brochures so I can share them with friends and family members who might like to contribute to the Alliance.

Teamwork for Health, Audio tape sets from the 1994 Annual Meeting

Boxed set of seven 90-minute cassettes, boxed, with program and handouts

Includes presentations on kidney, pancreas, adrenal glands, family stories, coping skills, stereotactic radiosurgery for brain lesions, coordinating medical care, DNA testing, current research

☐ Please send _____ copies of the boxed set (\$ 40 members, \$80 non-members)

VHLFA T-Shirt -- Beautiful ivory T-shirt with VHLFA logo, S, M, L, or XL \$12 each

☐ Please send _____ shirts total -- sizes: #__ Small; #__ Medium; #__ Large; #__ XL

VHLFF Back Issues - \$1 for members, \$2 for non-members. Index available free on request

☐ Please send issues for (dates) _____

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

I am enclosing a donation ☐ In Memory Of ☐ In Honor Of

Honoree's Name _____

Occasion: _____

Please send card to (name and address) _____

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

In Britain, send to VHL Patient and Relative Contact Group, 114 Longfield Rd, Littleport, Ely, Cambs CB6 1LB

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

FUNd Raising in Mississippi and VHLFA T-Shirts

The Mississippi Chapter recently sponsored a raffle to raise money for VHL research and education. Not only was it a lot of fun, they succeeded in raising awareness of VHL throughout their community. Peggy Marshall and the Alliance were featured on the front page of their local newspaper, and friends and local businesses pitched in to help.

Peggy is shown here with Randy Ferrell drawing the winning ticket at Ferrell's Appliance store in Corinth. The prize, a video tape recorder, was won by Melinda Hamlin of Rienzi, Mississippi. Melinda, who was in bed for a few months, expecting twins, was delighted with her prize. Her twin boys, Matthew and Mark, arrived safely in August.

The new VHLFA T-shirt Peggy is wearing was designed by Altheada Johnson of the New York Chapter and WC Enterprises of Delaware. It makes a perfect sport shirt or bathing suit cover-up, and helps to spread the word about the Alliance. Your T-shirt purchase helps to fund the educational programs of the VHL Family Alliance. To order, see page 15.

Peggy Marshall and Randy Ferrell drawing the winning ticket at the VHLFA Mississippi Chapter raffle. Special thanks to Peggy and Don, Hazel, Tammy, Terry, Sheryl, and all the special friends in Corinth and Alcorn City, Mississippi. VHLFA T-shirts are available on page 15. Call for bulk orders for your fund-raising event.



Plan to join us for the Annual Meeting, Boston, 1995!

VHL Family Forum

Newsletter of the VHL Family Alliance
171 Clinton Road
Brookline, MA 02146

Address Correction Requested

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