# VHL Family Forum

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# Caution Urged for Stereotactic Radiosurgery

*by Gail S., Tennessee, with Joyce Graff*

When people hear that my sister and mother and I have all had Stereotactic Radiosurgery, or Gamma Knife surgery, they want to know – what do you think of it? Would you recommend it? Well, it depends.

We have learned a lot in the last two years about when stereotactic radiosurgery is wonderful, and when it is not a good choice. It sounds so seductive – you go in for day treatment, like having dental work, and then go have lunch in a restaurant – wow! Comparing that to going through an open surgical procedure, it sounds great. But there's a lot they don't tell you up front.

It was Dr. Oldfield from the National Institutes of Health who suggested to my sister Carolyn that she consider stereotactic radiosurgery (SR) for her three brain tumors. He felt they were of a size that is usually a good target size, and in a good position so that the post-operative swelling would not be a problem, and none of them had a cyst. She checked with the clinic in Memphis, and had the procedure in the fall of 1996. She had some headaches, but little negative reaction to the treatment. At her 90-day checkup, they were already seeing some improvement, some shrinkage of the tumors! We were thrilled for her – and envious.

"Do me, too," our mother said. Seeing Carolyn's success, our mother, Pat, asked the treatment center if they could treat her two brain tumors with SR too. They did the treatment in December 1996, and the treatment went well. One of the tumors had a cyst, but our SR team felt that once the tumor shrank the cyst would shrink also. Mother was thrilled to avoid having another open brain surgery.

At Mother's checkup in May 1997, the report from the MRI was that the tumor was gone, though the cyst was still there. Mother's results seemed to be good, and Carolyn was still doing fine.

"Do me, too," I said. I had one hemangioblastoma with a cyst which was not causing symptoms, but this seemed such an easy way to get rid of it, I thought I might as well get it done too.

Being in the research protocol at NIH, I was in touch with some of the nurses and researchers there, and the word got back to Dr. Oldfield. The day before the procedure, Dr. Oldfield called me, and said he thought we should talk more about stereotactic radiosurgery before I went ahead with the treatment. While Carolyn's tumors were just the right size and placement for successful treatment, mine, he said, with its cyst, was not a good candidate for stereotactic radiosurgery. While the treatment might shrink the tumor, it would not do so immediately, and there was danger that the cyst might grow more before the tumor was controlled. He warned me that there was danger for me in proceeding with this treatment, that for this particular kind of tumor he would recommend open surgery.

Like so many other people with serious illness, I was in a quandary. I had one physician whom I greatly respect telling me there was danger. I had another physician, at the gamma knife center, telling me he could successfully treat the lesion. My mother and sister had had good outcomes, and my mother's lesion had a cyst. How was I to make the decision? I read articles in Medline. There was one sample of ten patients with cystic hemangioblastoma. Six had problems following SR, two had urgent problems, one needed emergency surgery. But, I rationalized, ten is not a very big sample, and the bad outcomes wouldn't happen to me.

I called the physician who would perform my SR treatment to tell him about Dr. Oldfield's call. He conducted research of how own, consulting the national SR database. He read the article I had found in Medline.

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*Choices, cont'd from page 1.*

He found no other data indicating that cystic hemangioblastomas were not good candidates for SR. He urged me to continue with my plans.

I felt committed to go forward the following day with the scheduled procedure. And most of all, I wanted it to be successful. I wanted to believe that I could have the same success my mother and sister were enjoying. I didn't want to have surgery, didn't want a "zipper" in the back of my head. So I went ahead with the procedure, went out to lunch after the procedure, and felt I had made the right decision.

Then the nightmare began.

In June 1997 my mother began to have headaches – big headaches. They prescribed steroids, and kept increasing the dosages, but it wouldn't stop. There was more and more pressure in her head. Her brain was swelling. Over the summer on several occasions she had serious reactions to the large amounts of decadron she was taking. They changed the medication, but her brain continued to swell. Headaches, double vision, difficulty walking – things we now know were caused by the long-term swelling reaction from the radiosurgery. The steroids caused problems of their own – heavy dosages, reactions to medication, rapid weaning off the medication and reactions from that withdrawal, diverticulitis – altogether a bad scene. Mother was now completely bedridden and in the hospital.

With all the crises around my mother's health, I didn't go for my three-month checkup. After all, I was feeling fine, and it was Mother we were worrying about. By September I too started to have headaches – bad ones – and on October 23 they did surgery to remove a 31 mm cyst from my brain. The cyst had continued to grow and had to be surgically removed after all.

Mother was having incredible pain. At first it was blamed on a "bad pillow" or other normal hospital complaints, but finally my sister and I convinced the hospital that Mother was not a complainer, and they should do an MRI. Once they took a look at the MRI they scheduled emergency surgery to remove the tumor and the enlarged cyst, which had continued to grow. She continues to have dizziness and weakness in her legs, and her brain still shows signs of swelling, 17 months after the SR procedure. She is still being weaned from the steroids.

What we learned was that stereotactic radiosurgery can be wonderful, or can be terrifying. Thanks to learning accumulated over the past five years of experience at a number of SR treatment centers, by NIH, and by the VHL Family Alliance members sharing their experiences, there are now some pretty good guidelines for when SR will likely be successful, and when it is inadvisable.

The guidelines agreed at the Bethesda Focus Meeting on Stereotactic Radiosurgery are that the hemangioblastomas that respond best to SR are 10-12 mm. or smaller and where there is no cyst creating pressure inside the skull.

It is important to understand how it works. You have to know too that hemangioblastomas are very rare tumors, and the amount of experience most centers have in treating hemangioblastomas is very limited. Hemangioblastomas account for only two percent (2%) of all brain tumors. And hemangioblastomas react very differently to SR than hard tumors. That is why it is critically important that we share our experiences with the Alliance, and make sure that all these outcomes – both good and bad – contribute to our learning and create better outcomes for people in the future. For example, adverse reactions to the drugs, like decadron, had not been reported in papers about stereotactic radiosurgery, because the procedure had gone well: it was the drug that caused the problem. The Alliance, however, has collected information about four such cases. Remembering that there are not yet 100 patients who have had stereotactic radiosurgery for hemangioblastoma, four is a significant number. Through the Alliance, we can share the real human experience of the procedure, and whether we as humans feel the procedure was a success. How much of an interruption was this procedure in our lives? Did we get the outcome we wanted? Was it worth it?

> We think that patients with VHL disease who have had their diagnosis established and who present with small (<3 cm) solid hemanbioblastomas without significant mass effects are reasonable candidates for radiosurgery... Microsurgical resection remains the treatment of choice for the vast majority of symptomatic cystic hemangioblastomas .. because of the need to eliminate mass effects ... For rare patients with very numerous lesions, treatment is limited to symptomatic and radiographically enlarging tumors, to minimize excessive radiation. -- *Chang, Adler, et al., "Treatment of Hemangioblastomas in von Hippel-Lindau Disease with Linear Accelerator-based Radiosurgery,"* Neurosurgery *43:1, July 1998.*

Essentially, SR zaps the tumor with beams of radiation from hundreds of different angles, so that as each beam passes through normal tissue that tissue gets only a very small dose of radiation. Where the beams meet at the target site, the tumor gets the sum of the dosages of all the beams, the "surgical dose", which is essentially a radiation burn that intends to kill the tumor. But it doesn't happen immediately. As with any burn, it swells for a while, then heals, and hopefully leaves the tumor dried up and dead. With burns you've had in the kitchen, the swelling goes on for a few hours, or a few days, and then begins to heal. But with a radiation burn, the cycle of swelling and healing can go on for months, or even years, depending on the dosage delivered. Hard tumors simply dry up and crumble; but hemangioblastomas swell.

There are ways to reduce the amount of swelling. Sometimes they will "fractionate" the dosage, or divide it across a number of sessions. Dr. Haring J. W. Nauta compares the effect to the difference between spending two hours on the beach in Miami on the first day of your winter vacation, versus spending 10 minutes in the sun twice a day for the week. You will have spent that same two hours at the beach, and had the same exposure to

the sun's radiation, but you will probably not burn. This is "fractionating" your dosage of the sun's radiation.

I encourage you to talk with your doctors – a serious, open-minded talk – with both a neurosurgeon and a radiation oncologist, and ask them all the hard questions that are in the *VHL Handbook*. Those questions are there for a reason – they represent the hard learning of families like mine whose outcomes were not so great because they didn't have all the information up front. Don't be pig-headed, as I was. I wanted to avoid surgery – I had it anyway. I got my zipper after all, but it's well hidden by my hair, and it's not so bad.

Get a second opinion, or even three. Ask one of the doctors with experience in treating hemangioblastoma to review the treatment plan before, not after it happens. Once the radiation dose has been delivered, they can't take it back. You're on the roller-coaster, and you have to do the whole ride.

And if you do have the procedure, be very sure to go back for follow-up as scheduled, don't minimize the importance of those follow-ups, even if you're feeling good. Make sure they are monitoring the pressure inside the skull, especially if you experience headaches, vision disturbance, or other neurological symptoms. With careful monitoring, there is a better chance of controlling negative consequences.

Carolyn continues to do well. For her, sterotactic radiosurgery was indeed the right choice. Her checkups show that the tumors are drying up as planned.

Pat H. and her daughters Carolyn H., and Gail S, Tennessee

I'm back to work and doing fine after my brain surgery, six months after SR. In retrospect, my cystic lesion should not have been treated with SR. While the radiation may not have caused the cyst to grow, it also did nothing to slow down its natural tendency to grow. The best alternative for me from the beginning was open surgery.

My mother continues fragile and in poor health, though she is coming back slowly from a nearly fatal experience, and four months in the hospital, beginning six months after SR. Of course there is no way to know what might have happened if she had had open brain surgery in the first place, but it could not have been any worse than this. ❑

***Questions to ask the Doctor***

*(1) Get both opinions.* We strongly urge you to consult with a physician who is good at *both* conventional micro-neurosurgery *and* stereotactic radiosurgery. It is *not* enough to speak only with someone who practices only SR. Be sure to talk with someone who is expert in the other method and get that view. In many cases, it is safer to approach a tumor with conventional surgery. You need a team of medical professionals who can help you to evaluate fairly the pros and cons of both procedures and decide which is better for you in this particular situation at this particular time.

*(2) How big is the tumor?* Recommendations are *not* to treat a hemangioblastoma larger than 2-3 centimeters at the very most (optimal size is 1.0-1.2 cm). Size is not the only issue, but it is a very important issue. The larger the tumor, the more energy it will absorb and the more it will swell.

*(3) Is there cyst or other source of mass effect?* "Mass effect" is the effect of having some additional mass in your skull. This could be from a cyst, swelling, or from the tumor itself. If there is already extra pressure inside your skull, SR is probably not a good idea, since the additional swelling caused by the procedure would compound the mass effect.

*(4) Where is it?* Once treated, there will be swelling (edema) of the tumor and surrounding tissues. What this means to you is that the treated tumor will get bigger before is gets smaller, and depending how much room there is for it to expand, your symptoms may increase before they get better. What position is the tumor in? When it swells, what symptoms may occur? How will the doctor propose to control the swelling? How can you work in partnership with the medical team to minimize the swelling and get through the swelling period? Note that this period of swelling is not measurable in days but in months. Ask your doctor how long you should expect this swelling period to last.

*(5) What are the dangers to surrounding tissues?* There is usually some margin of healthy tissue that will be irradiated with a therapeutic dosage. What tissue is within that margin? What would such damage do? If the tumor is in a position where there is fluid beside it, then there is some "margin for error," but if it is in a critical spot, then effects to the healthy tissue can be significant.

*(6) How many tumors do they propose to treat?* What is the sum of the radiation to which you would be subjected? If more than one tumor is to be treated, is it wise to treat them all at this same time? Will the combined swelling of the various tumors cause a dangerous situation? Is it better to treat them one at a time? Pacing the treatment can be critical to managing the post-treatment swelling.

*(7) What medication(s) would the doctor propose to use* to manage the post-treatment period? Have you taken this medication before? Can they test you for sensitivity to the medication before the treatment, to make sure that you are not likely to have an adverse reaction? The worst problems we have seen from stereotactic radiation involve sensitivities to the medication.

*(8) What experience does this team have with treating hemangioblastoma*, as opposed to other solid tumors? Hemangioblastomas react differently to radiation treatment. It is important to get someone with experience in treating hemangioblastoma to participate in reviewing the treatment plan prior to the beginning of treatment. If you cannot find someone in your area, we can suggest some sources of second opinions. This should be welcomed by your team, as it is for their protection as much as for your own. ❑

# At Second Glance

### *When the look fits, experts in ocular prosthetics shine*

*By John Beifuss,* The Commercial Appeal*, Memphis, Tennessee, reprinted with permission.*

*[Editorial note from Peggy Marshall:] This article is very important to read if you have a blind or visually impaired eye that your ophthalmologist feels is untreatable and will most likely have to be removed. Eventually the increase of pressure , deterioration, color change, cataracts, redness and pain will lead to the decision to have the eye eneucleated (removed). This is not the end of the world and will not leave you scarred for life.*

*A caller to the 800 Line recently related her concerns about, "What happens after my eye is removed?" Her doctor said, "We will worry about that after the eye is removed." The fact is, the time to address the issue of an artificial eye (prosthesis) is* ***before*** *surgery -- with a Certified Occularist! There are decisions to be made about the choice of available implants.*

*As you will read in the following article, the type of implant determines the type of prosthesis. This needs to be a well informed decision between you, the Surgeon and the Ocularist. This is a very difficult time, but the better informed you are the better you will feel about your surgery. Once your eye is removed and you are fitted with a prosthesis, the positive change in how you feel about yourself is enormous. A good Ocularist will make you wonder why you waited so long. I know -- I have been there!*

Dave Parrott lost his right eye when he was 2.

"We were living over on Vance, and I was across the street playing with some kids, and a 10-year-old kid had a bow-and-arrow set he'd gotten for his birthday," said Parrott, 42. "He was kind of playing William Tell, and he shot my eye out.

"I remember him in front of me, and I remember the arrow, I sure do. I do not remember the pain."

Readers, however, no doubt can feel Parrott's pain: Ouch. But don't cry for him, no matter how many good eyes you have left to produce the tears. Because one man's injury is another man's fashion accessory.

Parrott, in fact, flaunts his Cyclopean status with a series of "novelty eyes" that he wears most days instead of a more traditional artificial eye of matching brown.

"I've got a cat's eye, with a vertical pupil," Parrott said. "That's my favorite. And I have a black one, with an Egyptian god in the middle. I've got that one because I used to be in the Carnival krewe of Ptah.[1] And I've got my red one that I wear at Halloween that has a horizontal, gold-colored pupil.

"I do have a brown one that matches my remaining eye almost perfectly, but I'll usually put that one in only when I'm out of town on business. Normally, the one I really wear out and about is my cat's eye.

"I'm one of those people, if you've got a handicap, have fun with the darn thing. I have a blast."

The makers of these so-called glass eyes are Bob and Rob Thomas, a father-and-son team with offices at Kirby Parkway and Poplar. The father, 60-year-old Bob Thomas, is Tennessee's only certified fitter and fabricator of "ocular prosthetics."

"It's a fun place to work," said the son, 27-year-old Rob Thomas, brandishing an artificial eye that stared with a lifelike if glassy gaze through a dark pupil and a lovely hazel iris. The iris was set like a jewel inside a realistically veiny and discolored sclera (the white of the eye).

Although most people still refer to false eyes as "glass eyes," most artificial eyes since the late 1950s have been made from hard acrylic, not glass. Also, these prosthetics are not marble-like orbs that fit into the socket like a round plug. Rather, they are convex shells that fit over an implant in the socket like a fat contact lens.

In fact, the fitting and fabrication of artificial eyes is an art-cum-science similar to denture-making or the manufacture of other false body parts.

"Years ago, there used to be stock eyes," said Bob Thomas, a former president of the American Society of Ocularists[2] who has been making eyes for 39 years. "The patient would go into the eye fitter's office, they would look in a drawer and pick out a pre-made eye to fit the socket."

Now, however, ocular prosthetics are painstakingly detailed to match the patient's remaining eye as closely as possible – except, of course, when a patient like Parrott asks for a "gag eye" (gag as in joke, not choke, although Parrott admits that some who see his novelty orbs experience the latter reaction).

The American Society of Ocularists keeps no statistics on how many people in the United States wear artificial eyes, but the number is greater than most might think. The Thomases' current patient roster includes more than 1200 people from the region and scores more from around the world, including many children from St. Jude's Children's Research Hospital who have lost an eye to cancer.

To meet the demand, Thomas Ocular Prosthetic Laboratories, Inc., operates satellite offices in Little Rock, Nashville, Jackson, Mississippi, and Johnson City, Tennessee. The main office, which opened in 1988 after Bob Thomas moved from Birmingham, is at 1900 Kirby Parkway.

The laboratory looks like a cross between an eye doctor's office and a special effects studio. The Thomases wear doctor-style white coats and maintain a neat and magazine-laden waiting room, but the back offices reveal trays filled with sample eyes and plenty of tools for sanding, painting and mold-making.

Acrylic eyes were developed after World War II, when the world no longer was able to import German-made glass eyes. Nowadays, eyes are made through something ocularists call the "modified impression technique," which is similar to denture manufacture.

Bob Thomas, in fact, was a dental technician in the Navy before he made the career shift to eyes, first working as an apprentice with a New York firm. Patients usually are referred to the Thomases about six to eight weeks after their "enucleation", the surgical removal of the damaged eye.

The surgeon inserts a spherical implant in the socket that "helps make up for lost volume," Bob Thomas said. The latest implants are porous, which allows blood vessels to grow into it, like roots growing into one of those styrofoam planters. This "vascularized" implant is able to move somewhat, in coordination with the remaining natural eye.

To measure a patient's socket, the ocularist uses a small, hollowed device, shaped somewhat like a contact lens with a handle. While inside the socket, this device is injected with an alginate similar to that used by a dentist to make an impression of teeth. The resulting mold, which conforms to the shape needed for the artificial eye, is the first step in a series of processes that eventually result in a painted and cured "eye" that is converted from wax to acrylic.

During the process, a "pupil" – a tiny black plastic circle – is added to the mold. The iris and sclera are painted, which requires the Thomases to really shine as artists (little of the "white" of a person's eye is actually white).

"Veins" are created through the use of minuscule red cotton threads. Said Bob Thomas: "We had one guy who said, 'Put a few extra veins in there so I can have a few extra beers and my wife won't know it."

Creating an artificial eye is a day-long process. The cost of an eye is usually about $1500, although novelty eyes aren't as expensive because "they don't have to be as exact," Rob Thomas said.

The Thomases also create other prosthetics, as necessary. Their scrapbooks are filled with before-and-after photographs of patients who have suffered through all sorts of injury. For instance, some customers have lost not just an eye but an entire facial region. This requires the manufacture of fake cheeks, brow ridges, noses and other parts. These often are attached to a pair of glasses, so they can be removed easily at home.

Artificial eyes basically are held in place through the pressure of the fit, and by the eyelids. They should be cleaned at home every few months with a mild facial soap. In addition, patients are asked to make a yearly visit to their ocularist, to have their artificial eye polished, sterilized and thoroughly cleaned of "protein buildup". A good eye should last five to seven years.

How are natural eyes lost? The causes are many, the Thomases said, including disease, infection, birth defects, gunshot wounds, yard darts, paperclips shot across classrooms and errant golf balls. "When your mother tells you not to run with scissors, listen," Bob Thomas said.

William B. Walls, 74, of Parkway Village, who visited the Thomases for his annual checkup last week, lost his left eye in 1985 after it developed malignant melanoma. "You have to learn to drive all over again, I'll tell you that," he said of the change in depth perception, as Bob Thomas extracted the artificial eye from its socket with a tiny suction cup attached to a handle.

Now, however, Walls is perfectly comfortable with his artificial matching eye. "Nobody even notices it."

Walls, however, doesn't go in for novelty eyes, unlike many of the Thomases' patients.

Many people want eyes associated with their favorite sports team, Bob Thomas said. He said he has manufactured orbs decorated with the University of Tennessee logo, the fleur-de-lis of the New Orleans Saints, and a Texas longhorn.

Thomas said one person ordered a "formal eye" with a diamond for a pupil. Another asked for a matching eye to be installed inside a clear sort of window in his bowling ball, "so when he lined up to bowl, the eye was actually looking right at the center bowling pin."

He said an ocularist is something of a counselor as well as a craftsman. "You get people coming in here, they're contemplating suicide, they refuse to look at themselves, they can't imagine life with one eye," he said, "But after they get used to it, sometimes they have the opposite reaction."

He said new patients often learn to accept their loss by talking with satisfied veteran patients, like Parrott.

"Many times when someone loses an eye, especially as an adult, they're kind of down about it," Parrott said. "It takes a while before you even feel like making a joke about it. I think it's healthier to have fun with it, quite honestly.

"You get all kinds of neat reactions," he said of his novelty eyes. "You have people that will look at you and the first thing they want to do is grab you by both shoulders and say, "Don't move – I want to look at that eye." And then you have other people that are frightened of you, they just shun away from you.

Human nature doesn't change, but Parrott is glad technology does. He said one of his old matching artificial eyes, which he had owned for years, was a victim of good intentions. "The way you cleaned those things then was you boiled 'em with a little saltwater. My wife, Monika, was doing it for me and forgot about it and left it on the stove. It melted."

1. The Annual Memphis Cotton Carnival features a parade with fanciful floats created by groups of friends, often with Egyptian themes echoing the name Memphis.

2. An ocularist earns certification by serving a five-year apprenticeship under a Board Certified ocularist. With modern methods of training and fabrication, the artificial eye is now a very natural looking device, hand-painted to match the remaining eye. A list of Board Certified Ocularists in the United States can be obtained from the American Society of Ocularists, 493 - 8 th Avenue, San Francisco, CA 94118, +1 (415) 221-5765; fax: +1 (415) 221-0755; e-mail: aso@zapsmith.com. For a listing of societies of ocularists in other countries, please see http://www.generation.net/~ocuplast/wooc.htm. Special thanks to Bob Thomas of Thomas Ocular Prosthetic Laboratories, Memphis, for his help in the preparation of this article.

# Advocacy and You

***Part 3, Keeping Posted***

*by Don Marshall*

*If not me...Who? If not now ....When?* Keeping up to date with your health issues is paramount in maintaining the best health care possible for you and your family. Recently President Clinton is quoted as saying " *We've made a lot of progress in health care reform, but we've got a long way to go. It hurts us all if we are shelling out money for health care coverage and then we can't get quality health care."* How can we follow the changes that are occurring every day that effect the health care we receive?

There are several sites available that keep us abreast of proposed changes to health care legislation. The National Organization for Rare Disorders (NORD) provides a periodic report titled *NORD ON-Line.* The staff at NORD maintains constant contact with the Federal Legislature. They work to keep us informed about any and all medical issues that will affect our health care. The task is monumental considering the many different committees that have an impact on health care: the U.S. House and Senate Appropriations Subcommittees on Labor, Health, and Human Services and Education, Federal Drug Administration, Senate and House Subcommittee on Agriculture, Rural Development, Food and Drug Administration and Related Agencies, Social Security Administration, and several others. There are seventeen Senate and nineteen House of Representative committees that have the power to generate legislation. The best time to affect an issue is as early in the process as possible. *NORD ON-Line* is an excellent way to keep in touch with impending health issues. NORD may be contacted at +1 (203) 746-6518 or on the web at www.nord.rdb.com.

The *Alliance Alert*, published by the Alliance of Genetic Support Groups, is a monthly publication dedicated to providing timely and useful material to its constituency. Included in the Alliance Alert are several website resources. The Alliance may be contacted at 1-800-336-GENE or on the web at www.geneticalliance.org.

Other web sites of interest are: www.towson.edu/~bhalle/disable.html. This site covers Disability Issues, the Media, and Communication. The National Parent Network on Disabilities (NPND) maintains a site at www.npnd.org. Issues affecting Disabilities, SSI, Consumer Health and the President's Advisory Commission on Consumer Protection and Quality in the Health Care Industry are the focus of recent material. You can contact them at npnd@cs.com to get on either their e-mail or fax list. You can receive a copy of their Friday Fax each week.

Be sure to check our web site www.vhl.org and the *VHL Family Forum* to keep abreast of issues that concern all of us.

Remember we not only have the right but the responsibility to keep ourselves informed and to let our elected officials know how we feel about our health issues. Please contact the VHL Family Alliance whenever you learn of issues affecting our VHL family so we can keep everyone else informed. Call the 800 Line, e-mail us at info@vhl.org or send us informational material. The who is *me* and the when is *now!*

# Happy 5th Anniversary, VHL Family Alliance

*by Peggy Marshall, Mississippi*

Don and I have been reflecting back on the past five years on our involvement in the VHL Family Alliance. This is a very special event for all of us and a perfect opportunity to share what it has meant to us personally.

I was invited to a VHL Family Alliance support meeting in June of 1993. The invitation came from Joyce Graff and the meeting was to be held at her parents' home in Memphis, Tennessee. Our daughter and my sister from Arkansas also received invitations, but would not attend unless I agreed to go. My husband, Don, expressed strongly his concerns that this meeting might not be a good idea. He felt (like others who are reluctant to attend a support meeting) that this might not be a positive experience. He was and is concerned that I maintain as positive an attitude as possible and live every day to its fullest. He felt being among other VHL patients would only lead to negative thoughts about living with VHL. As I searched for the reasons to meet Joyce, I realized that I would not be satisfied until I knew what the goals of the VHL Family Alliance would be. My family felt isolated having a disease that most physicians had never seen. Meeting with Joyce, her family and friends that day in June changed my life, strengthened my faith, and gave me hope for the future.

Immediately on my return from Memphis, Don saw a new commitment in my life and he joined in the enthusiasm I felt, knowing that we had finally found someone with the leadership, knowledge and initiative to help VHL patients and families to deal with VHL. Words can not express how much Don and I appreciate the energy that Joyce has given to keep the organization alive. Thank you, Joyce, from all of us!

As Chair of our 800 Line Committee, I, along with Altheada J., Barbara R., and Eva L., have had the pleasure of talking with hundreds of individuals about VHL. We feel it is one of the most rewarding aspects of our involvement with the VHL Family Alliance. For


*Peggy Marshall and Joyce Graff at the Memphis meeting.*

many callers, it is their first contact with another person with VHL. Our hearts have been touched over and over again. It is overwhelming to know that you have been part of saving an individual from a total nephrectomy and dialysis by recommending a second opinion. The VHL Family Alliance has developed a long reaching world-wide support mechanism through our Medical Advisory Board, major medical facilities as well as the National Institutes of Health to bring VHL from a paragraph in a medical textbook to a manageable and treatable condition.

The VHL Family Alliance has published a *VHL Handbook* that is so thorough it is used as a resource for medical teams and health maintenance organizations worldwide.

We have formed 28 State Chapters and have seen caring and dedicated volunteers give tremendous energy and time to the goal of improving diagnosis, treatment and quality of life for all of us with VHL. We also have 22 Clinical Care Centers, domestic and international, to better serve patients in the coordination of their medical professionals to provide the necessary screening needed for treatment of VHL. Sixteen international Affiliate Chapters have been formed to make patient knowledge and care a world-wide effort.

The VHL Family Alliance has been able to fund three impressive research teams in the past two years to work toward our goal of finding a way to control VHL. We will continue to focus our efforts to fund research until that breakthrough becomes a reality. All of these accomplishments could not have happened without each volunteer on the team doing their part. It has truly been a team effort. We have grown from three families to over seven thousand people in five years. We are as excited today as we were five years ago when we said "Yes, we want to help."

The VHLFA support system is growing stronger every day and we are making a difference! **Happy Anniversary, VHL Family Alliance!!**

# Understanding Adopted Children and Families

*reviewing the* Family of Adoption[1] *by Joyce Maguire Pavao*

Dr. Joyce Maguire Pavao is a pioneering and nationally recognized family adoption therapist who, through her first book *The Family of Adoption*, demonstrates that there are predictable and understandable developmental stages and challenges for all adopted people.

Pavao lays them out age level by age level, showing, for instance, how and why day-dreaming is a normal strategy for adoptees and how particular academic subjects may create pain for adopted children.

Pavao argues that all adoptive parents, as well as teachers and therapists and all who work with children, must come to understand these developmental stages as normal -- though challenging -- for adopted children. She writes with equal insight of the "birth rites" of both biological and adoptive parents; of how adoption does not cure infertility, but childlessness; and of healing rites for birth parents who must give up the parenting of their child.

Pavao is an "important voice in the spirited public debate over what 'the best interest of the child' is in the imperfect, bureaucratically untidy world of adoption," says Joseph P. Kahn of *The Boston Globe*.

Joyce Maguire Pavao is executive director of the Center for Family Connections and founder of PACT (Pre/Post Adoption Counsulting Team) in Cambridge, Massachusetts. The Center holds training sessions for parents, professionals, and friends of Adoption and Complex Families, as well as intensive clinical training sessions for Clinicians. A summer session will be held July 13-17 in Provincetown, Massachusetts. Workshops on birthparent loss, single parent adoptions, medical issues, search and reunion, and other adoption topics will be held one Friday each month during the year. For information, contact the Center for Family Connections, P.O. Box 383246, Cambridge, MA 02238, +1 (617) 547-0909 or 1-800-KINNECT; Fax: +1 (617) 497-5952; E-mail: kinnect@aol.com.

1. To be released August 1998, by Beacon Press, Boston, MA, http://www.beacon.org. Other useful contacts: Adoptive Parents for Open Records , 9 Marjorie Drive, Hackensack, NJ 07840. American Adoption Congress (ACC), 1000 Conneticut Ave, NW, Suite#9, Washington, DC 20036. Council for Equal Rights in Adoption (CERA), 401 E 74th Street, Suite 17-D, NY, NY 10021

***VHLFA Internet Web site:*** **http://www.vhl.org**

*VHLFA Patient support line,* 9-9 Eastern U.S. time, +1-800-767-4VHL or +1-617-232-5946 or info@vhl.org

*VHL Tissue Bank, 24 hours a day,* 1-800-847-1539.

# Seattle! Paris!

As this issue goes to press, the Seattle meeting is almost here! We hope to see you there.

The Paris meeting is looking very exciting. Dr. Stéphane Richard in Paris has organized an outstanding meeting, with more than 100 physicians already registered to attend. The special registration fee for families is 200FF for one day, or 500FF for the three day meeting, including a special gala dinner one night. The full registration packet is on the internet at www.vhl.org/meetings or may be requested from our phone or e-mail hotlines. Several families are staying at the Comfort Inn Mouffetard. Please join us September 16-18!

# Welcome MGH!

We are delighted to welcome the Massachusetts General Hospital in Boston to our Clinical Care Program. Dr. Katherine B. Sims, Neurogenetics, is the sponsoring physician for the VHL Clinical Care Center there. You can reach her at the Developmental Neurogenetics Clinic, +1 (617) 726-5732, Fax: +1 (617) 724-9620, E-mail: sims@helix.mgh.harvard.edu.

A VHL CCC is not a building, not even a day-of-the-week, rather it is a "state of mind." These Centers are familiar with VHL, offer the full range of diagnostic and clinical services people with VHL need, and have agreed to ensure that care is coordinated across specialties. They are active members of our worldwide "chain of information," helping to support local physicians in obtaining expert advice on this rare disorder. The complete list of clinical care centers is in the back of the *Handbook* and is also maintained on the internet at www.vhl.org. The MGH Neurogenetics service was the original sponsor of our website.

For questions or feedback on the CCCs please contact Ellen Lydon, R.N., Chair, vhlccc@vhl.org.

# Angiogenesis in the News

Discussion of Angiogenesis Inhibitors in not new to this newsletter. We have been tracking Dr. Judah Folkman's work on angiogenesis inhibitors for several years. The hemangiomas of VHL are essentially the "food supply" of a cancer tumor, without the hard tumor itself. A hemangioma is that nest of abnormal blood vessels that you see in the diagrams, without the tumor part. In shrinking the nest of blood vessels, you shrink the hemangioma.

In 1994 a number of VHL Family Alliance members gave urine samples to Dr. Folkman to test for chemical tracers that would tell him whether angiostatin or endostatin might be effective in the treatment of VHL. The answer at that time was that those two drugs were not quite the right ones, but nonetheless the theory is exactly what we need -- we just need a more specific drug.

Angiogenic factors (those which lead to the development of new blood vessels, or hemangiomas) are hypoxia (when there is too little oxygen, the body builds more blood vessels to bring more blood to bring more oxygen), tissue injury (where increased blood flow is needed for repairs), and vascular endothelial growth factor (VEGF). Thus the VHL protein is up-regulating an angiogenic factor and down-regulating an inhibitor of angiogenesis.

Through biomedical research at many institutions we are beginning to understand the function of the VHL protein and what drugs might be able to intervene in the chemical processes in the cell, bolster or replace the function of the missing VHL protein, and prevent the formation of tumors.

Dr. Edward Oldfield and others reported several years ago that VEGF, a powerful angiogenetic factor, was found in high quantities around brain and spinal tumors. Dr. Maartje Los reported to the Dutch VHL/MEN meeting in 1997 that VEGF is found in excessive quantities in the ocular fluid and renal cyst fluid of VHL patients, much higher than in the blood. These and other studies indicate that the VHL gene might be regulating VEGF expression in living tissues.

There are some new drugs which have been shown to manipulate the levels of VEGF in mice, and there is reason to believe that they might be helpful for people with VHL. "But it's tricky," says Dr. Richard Klausner, Director of the National Cancer Institute. "The testing has to be very carefully conducted because we don't want to cause bleeding. Testing these drugs on people with VHL is a delicate business, and will take at least a year. It won't be as dramatic as what you have seen recently in the news." The National Cancer Institute is now writing a protocol for use of one of these new drugs with renal cell carcinomas, including VHL. A similar study is expected to start in Boston for people with brain and spinal involvement in the fall.

# A Few Words to the Wise

*by J. E. Brody*

*Focus on* eating more vegetables, fruits and whole grains. Plan your meals around these most health-protective, naturally low-fat foods and use high-protein animal foods more as condiments than as the centerpiece of a meal.

*Sacrifice* a little of your free time to prepare nutritious meals at home. The only way to be sure of the nutritional quality of your meals is to make them yourself. Cook in batches and freeze well-labeled individual or family portions for wholesome instant meals later on.

*Avoid rigidity.* Any food, however caloric or fatty, is O.K. now and then. The secret to a successful, healthful diet is variety, not limitation; moderation, not elimination, and gradual, evolutionary change, not revolution. Consider changing one meal every week to something more nutritious. By year's end, you will have a new way of eating and be much less likely to miss your old, less wholesome habits.

*Use exercise*, meditation, yoga and other reliable relaxation techniques to help reduce the stress in your life. Stress undermines the immune system and can increase susceptibility to infections, heart disease and cancer. There is nothing to gain and much to lose by countering stress with health-damaging habits like cigarette smoking, drug abuse or excessive alcohol drinking.

Devote at least a half-hour a day to an activity you enjoy that makes you breathe harder and perhaps sweat a little. The benefits to your body--your heart, bones, blood-sugar level, digestion and more--are almost too numerous to mention. And recent studies indicate that regular exercise also protects against cancer. An equal dividend is the feeling of well-being you get after exercising. Even when mentally or physically drained from the day's demands, I head for a pool each evening to swim for a half-hour, then return home a renewed and more relaxed person.

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Yesterday we learned that two of my daughters do not have the gene for VHL. An answer to my prayer. With this knowledge, we now do not have to fear that the gene has been passed on to their four children.

Since I learned of VHL in 1979, I have tried to learn everything I could for the sake of my children but it was my association with the Alliance that finally gave me the information and resources to proceed to genetic testing. I know that this information is not for every family but for ours it was a godsend. We can now focus on keeping my youngest daughter who does have VHL as healthy as possible and supporting her while she battles VHL.

Thanks for all your help and I know it will continue. -- *Donna N., Tennessee*

# Teen Talk!

A series of questions and answers has been posted on the new Teen Page on the internet at http://www.vhl.org/teens. The site includes special "beginner materials" prepared for people who have been diagnosed through DNA testing and who have no symptoms. Our goal is to provide information so that you can manage your health, and stay at least as healthy as you are today!

***We need you*** to help grow the Teen Page. Please contribute your ideas, let us know what you need.

# Diet and Health

A new booklet *Diet and Health: Recommendations for Cancer Prevention*, is available free from the American Institute for Cancer Research, based on the World Cancer Research Fund report, *Food Nutrition and the Prevention of Cancer: a Global Perspective*, prepared by a worldwide panel of experts on diet and cancer.

This 34-page booklet has practical suggestions with explanations for their advice, and includes sensible discussions of vegetables, fat, salt, food storage and handling, cooking methods, dietary supplements, alcohol and tobacco.

Single copies of *Diet and Health* are available free by contacting the American Institute for Cancer Research, 1759 R Street, NW, P.O. Box 97167, Washington, DC, 20090-7167. 1-800-843-8114 or 202-328-7744.

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I am recovering quietly at home since my successful operation. My wife and I can't thank you enough for what you and the Alliance have done to promote VHL since you started five years ago. Without your action, I would be on hemodialysis today with a lesser quality of life. Thanks for everything. – *Paul B., Canada*

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It's impossible for me to express how grateful I am for the VHLFA and the entire international VHL team. Whether or not any cure or other advancements personally benefit me in my lifetime, you have helped me, and many others, to lighten up a very dark room and get on with *living! – Nelson B., Utah*

***Publication from the Dutch Symposium***

The special issue of the *Journal of Internal Medicine* on the MEN and VHL meeting in 1997 in Holland will appear in June 1998. Altogether 26 articles will be published as well as a report on the opinion poll.

# Artist and Entrepreneur

*by Pierre Jacomet, Chile*

*Editor's Note: In the order section of this issue you will see an original recording by Pierre Jacomet, an award-winning Chilean concert pianist and philosopher who has VHL. After several surgeries for multiple pheochromocytomas, he is no longer able to play the piano. Nothing daunted, however, he has learned to express his classical piano music with the aid of a synthesizer. While the notes are made by the synthesizer, the programming, expression, and musicology are entirely those of this talented man. He is expected to speak to the VHL Symposium in Paris this fall. These recordings were donated by the artist to the VHL Family Alliance, to raise money for a cure. -JWG*

I was born in Valparaiso, Chile, on March 24 1933, of French parents, and hold double citizenship. At the age of nine months I was kidnapped but immediately recovered. Kidnapping was a fashionable crime in Chile in those days, after the kidnapping of the Lindbergh baby. My late father told me once that my character changed completely after this episode, of which I still keep a memory, a sort of photo without any sensation attached.

I started feeling bad — that is, different — beginning about the age of seven. I had *pavor nocturnus* [fear of the dark] and other unexplainable fears. My mother was an excellent pianist, painter and a superb sculptor with gold medals and all that. My piano lessons started with her. Then we moved to Argentina where I pursued my studies at the Beethoven Conservatory, and then with Maestro Scaramuzza.

When I was 15 my parents divorced. Father remained in Buenos Aires and I came back to Chile with my mother. I finished high school and went to College were I started studying Law and Economy. These studies were interrupted because I got married at 21. As children started coming I had to earn a living. The piano was not sufficient to support the family, although I already had finished my studies at the Conservatory and had also studied composition with Maestro Gustavo Becerra-Schmidt who is now in Germany.

I went into business on my own. At the same time I started what was going to prove a long and futile search to determine that was wrong with me. After several wrong diagnoses I was labeled a neurotic and spent 3000 useless hours of psychonalysis. I of course studied psychoanalysis myself during that time, studies that brought me to philosophy and theology.

When I was 33 I opened an office in Buenos Aires where I settled with my wife and 7 children at the age of 38. I became a successful businessman but my marriage went to pieces and in 1972 I divorced my wife who had never gotten used to Argentina.

In 1975 one of my children was abducted. It was horrible. We got him back after six sinister days, paying the ransom. He has never completely recovered from this experience. Then came the military coup d'Etat in Argentina. My business was flourishing at that time and I had offices in New York, Paris and Rotterdam. In November 1978 there was a quasi war between Argentina and Chile. As you may know Argentica was, during that time, under the grip of a brutal military dictatorship. Those current authorities called me to their headquarters and forced me to surrender all my businesses at gunpoint. They subjected me to a long and insufferable psychological torture (following me with unidentified cars, threatening my family, abducting my brother in law, raping one of my nieces, and so forth).

In 1980 the French ambassador got me out of Argentina but all my family stayed there. I went to New York where I lived with Susan (who is now my wife) until 1982. I studied languages and literatures (Spanish, Italian, German, French and Portuguese, which were the languages I spoke). I abandoned business and dedicated myself to studying (ancient Greek, Latin, etc.). We moved to France where we stayed until 1984. I decided to start a new family.

We came back to Chile in February 1984. By then we had one daughter. I started again with the piano and recovered most of my technique. I gave a couple of concerts and was preparing Ravel's *Gaspard de La Nuit* and Stravinsky's *Petrushka* plus Beethoven's *Opus 101*. But since the military episode I had continuous crises. My blood pressure went down to 70/35, with abundant sweating and tachycardia several times a day. This had started in 1979 and was getting worse and worse. Again everything was attributed to neurosis, which made me feel quite an idiot. Besides, all psychotherapies were ineffectual.

Finally, in 1989 I was diagnosed as having Multiple Endocrine Neoplasia (MEN) and two ugly pheochromocytomas* were extracted from my neck. The first surgery was badly performed in Chile and some neck nerves were severed, which left me with a clumsy right hand (I can shake hands, but not play the piano

*Pierre with two of his daughters in Chile.*

anymore). The second surgery was done in New York City with a partial removal of my larynx and no secondary effects.

Only this year Dr. Richard Haber, from Mount Sinai Hospital in New York, diagnosed VHL. Studying my family group he found kidney cancers, blindness, brain tumors and pheochromocytomas, which are possible results of VHL. He did not find any medullary thyroid cancer, which is characteristic of MEN. In retrospect I have lived with symptoms of VHL from age 7 to age 65, without a diagnosis. My family has had VHL for at least four generations without a diagnosis. The childhood fears, the "neurosis", the cardiac symptoms — all had a physical cause: those pheochromocytomas. Once they were removed the symptoms disappeared.

Right now I feel fine. I do have an angioma in my cerebellum but it's not causing any symptoms. With help from Altheada Johnson on the VHLFA hotline I sent my scans to Dr. Oldfield at the U.S. National Institutes of Health in Bethesda. Apparently the MRI was done without contrast and he was not able to see the angioma clearly. He advised me to repeat the MRI, which I intend to do in New York at the end of May.

I continue to record my music with the aid of a synthesizer, and dedicate myself to study, translating philosophic and scientific books. My recording of Bach's *Goldberg Variations* was very well received in Germany. My newest field of interest is Molecular Biology. I live very quietly in a small town on the Chilean coast, near Viña del Mar. In all I have nine children, seven grand children. Life has gone by very fast and has been an incredible adventure. It still is. And wonderful too.

***Footnote:*** Pheochromocytomas (called pheos [FEE-ohs] for short, most commonly occur in the adrenal glands, but may occur outside them, along the Zuckerkandl bodies of the endocrine system, roughly along a line from the groin to the collar bone on both sides. Pheos can cause excessive sweating, cardiac symptoms, and untreated pheos can cause damage to the heart and vascular system. Pheos in the neck in particular can cause neurotic or even psychotic episodes. ❑

# Lifetime Achievement Award to Mike Murray

At its meeting in Seattle, the VHL Family Alliance presented a Lifetime Achievement Award to J. Michael Murray. Mike, 43, died May 7 at his home in Princeton, New Jersey, after a long battle with von Hippel-Lindau disease. A private investor and a director of the Murray Foundation, Mike devoted many years of his life to philanthropic causes related to his illness. Recording for the Blind & Dyslexic, Memorial Sloan-Ketering and Bascom Palmer Eye Institute of Miami and Palm Beach, Florida, were among the many institutions which benefited from his tireless energy and the support of the foundation. Mike was honored at the Seattle meeting for his lifelong contribution to improving diagnosis, treatment, and quality of life for people with VHL.

Under his leadership, the Murray Foundation provided funding for research into enhanced imaging techniques for change detection in breast and kidney tumors. He provided funding for development of the Cyberknife stereotactic radiosurgery technology at Stanford University. He provided co-funding with VHLFA to Dr. Diana Griffith, Dr. William Kaelin, and Dr. James Gnarra for their research on VHL. The Murray Foundation also provided funding for research on Retinal Transplantation under Dr. Robert Aramant, a program with the potential to prevent people suffering of retinal diseases from becoming blind, or to help restore eyesight.

"The support of Michael Murray and the Murray Foundation made possible several breakthroughs in VHL research including the demonstration that the normal VHL protein prevents blood vessel growth by inhibiting vascular endothelial growth factor (VEGF)," said Dr. Kaelin. "We must never lose sight of Michael's vision." Those of us who knew him and worked with him will cherish his warm voice, his wry sense of humor, and the creative force he contributed to this work.

I sent you an email about a patient newly diagnosed with VHL. Thanks to you and to the leads that came from the leads you gave me, I was able to finally convince my patient and his physicians to consider an alternative to bilateral total nephrectomy. He had surgery in January, and his urologist removed only one and one-third kidneys--my patient is cheerful, feeling well and back to work, and I appreciate your help very much. -- *Linda Randolph, M.D., Medical Director, Prenatal Genetics, Alfigen/The Genetics Institute*

# VHL Eye Care Survey

*by Joyce Graff*

The American Optometric Association asked us to revisit our recommendation in the VHL Handbook that people at risk for VHL should be seen regularly by an ophthalmologist. They argue that more people in the United States see an optometrist than any other eye care professional. In many places there are not enough ophthalmologists to go around and it would be more convenient and less expensive to go to an optometrist. Is it really necessary to recommend an ophthalmologist? We decided to collect some feedback from the membership.

While we were at it, we asked the members whether in their opinion an ophthalmologist is enough? Is there a case to be made for going straight to a retinal specialist?

A questionnaire was put up on the VHL website in March, and kept there for six weeks. A total of 46 people replied to the questionnaire: 37 people with VHL, 3 people at risk for VHL, 4 other VHL family members, one ophthalmologist, and one other concerned citizen. 43 people (93%) see professionals regularly for eye care.

Of the people with VHL who responded, 25 (68%) have diagnosed lesions. At least once a year, 35% of them see an optometrist, 87% see an ophthalmologist, and 65% regularly see a retinal specialist. All three of the people at risk for VHL see an ophthalmologist annually.

Diagnoses were made 11% (5) by optometrists, 26% (12) by retinal specialists, and 63% (29) by ophthalmologists. At the time of the diagnosis, each had seen another eye care professional an average of 3.5 months prior to the diagnosis.

63% of all respondents recommended that people at risk for VHL be followed by an ophthalmologist. 12 respondents (26%) recommended going directly to a retinal specialist. Only 11% (5) felt that an optometrist would be sufficient to diagnose VHL. Among people with VHL, 65% (24) recommended an ophthalmologist, 30% (11) recommended a retinal specialist, and only two people (5%) felt that an optometrist would be sufficient to diagnose early VHL lesions in people at risk. Both of these respondents noted that their diagnosis was made by an optometrist, but neither of them has an eye lesion, so diagnosis was not made based on finding lesions.

### *Comments*

This data is consistent with the experience of the hotline team in speaking with families over the past five years. The comments left with the questionnaires explain the issue very well.

"If there is a hereditary risk of VHL, I would recommend going directly to an ophthalmologist for all check ups and if there are eye lesions then a retinal specialist. The retinal specialist I see recommends an optometrist for visual acuity exams and necessary corrective eyewear."

"Because of good medical care and numerous laser procedures, I have 20/20 vision."

"Seeing an optometrist is when a new pair of glasses is needed. Angiomas in my eye are being looked after by an ophthalmologist."

"I have had a history of angiomas in both eyes. Presently I have 3 stable ones in right eye. I have had cryo in right eye and laser in left. I have found that sometimes these lesions are difficult to spot unless by an retina specialist or ophthalmologist. I would not trust my boys to be tested by a optometrist since these are not the usual areas an optometrist would study and because they are so hard to spot."

"An Optometrist referred me to an Ophthalmologist who specializes in the retina. He did the diagnosing. My Optometrist described her capabilities as 'looking into a room through the keyhole' — she could only see so much. To see all the room you need to go to an Ophthalmologist. I feel anyone at risk for VHL should see an Ophthalmologist."

### *Delay = Visual Impairment*

All too often there were delays in spotting that first angioma, and delayed treatment can mean loss of vision.

"The Optometrist only referred me to a retinal specialist when I complained of seeing "cobwebs" and floaters in my vision. I only see the Optometrist for a new prescription for my glasses."

"We had an evaluation for our daughter with an optometrist who said her retinas were clear when she actually had a large hemangioblastoma in the right eye that was diagnosed at the NIH approximately 12 months later."

"I was seen by an optometrist one month before I went to NIH. He said he did a complete check and there were absolutely *no* retinal angiomas in either eye. I saw the retina specialist at NIH and I had one in each eye. After that experience, I have gone to a retinal specialist only."

"My Optometrist missed my lesions for five years and I paid because of it. I would avoid them and go right to Ophthalmologists every time"

"My tumors were almost missed by an ophthalmologist. It was only after I asked him to look carefully for these tumors that he found one. (There were two.) They were treated by a retina specialist and I see him for yearly checkups."

### *Challenges of Managed Care*

"Managed health care coverages generally will not allow a person to see a specialist without a referral,

which means a problem has to be found first by the Primary care physician or optometrist before a ophthalmologist or retinal specialist can be seen. This will certainly delay effective treatment."

"Without being seen by an ophthalmologist it would have been worse. However it does put a strain on the pocket book."

We realize that there are cheaper options. We discussed this issue at length prior to publication of the first Handbook, and again prior to the publication of the second edition. However because of the number of stories like these that exist in our community, we felt that we could not stop with the cheap solution, we needed to recommend that an ophthalmologist be used. Degrees do not make the difference. There are very talented and conscientious optometrists who will do at least as well as most ophthalmologists, and optometrists do often make diagnoses of VHL (27% of the time in this group alone). But as a general rule, it is statistically more likely that an ophthalmologist will find lesions at an earlier, more treatable stage than an optometrist.

***Conclusions***

The people who said they did not see an optometrist, and saw an ophthalmologist or retinal specialist instead, would surely correct themselves to say that when they go to the ophthalmologist or retinal specialist, that check-up almost invariably includes a session with an optometrist who takes care of the refraction, pressure testing, and other first-line eye screening. While many diagnoses of VHL are made by optometrists, ophthalmologists are more likely to find lesions early enough to successfully conserve vision.

The experience of our members has led us to the belief that most optometrists cannot be relied upon to find retinal angiomas early enough to conserve vision. Even many ophthalmologists may not find very small angiomas.

Retinal specialists are even more likely to find early tumors, but because of the additional expense and the availability of services, referral to a retinal specialist is usually not warranted until at least one lesion has been identified, or at least until a clinical diagnosis of VHL has been made.

We therefore confirm our recommendation that all people at risk for VHL visit at least once a year an ophthalmologist familiar with VHL who will do a thorough check of the retina and its equator.

This is not to diminish the essential role of the optometrist in eye care. Every person (with or without VHL) should see an optometrist regularly for basic eye care (refraction, pressure testing, first-line screening). People under the care of an ophthalmologist or retinal specialist will generally see an optometrist as part of that care. ❑

# VHL: Quest for a Cure

An idea born at the Boston meeting bears fruit at the Seattle meeting. VHL: Quest for a Cure will be shown for the first time at the opening of the Seattle meeting. This 15-minute video provides an overview of what VHL is, the importance of preventive care, and our search for a cure. Thanks to producer Fred Simon, and Cary Schwanitz and KOAT-TV, Albuquerque, New Mexico, for the special narration and editing that give it its polish, and to all the VHL families who sent in photos for a family album at the end. *VHL: Quest for a Cure* is suitable for viewing by medical audiences as well as the general public. 15 minutes, $20. See page 15 for ordering details. One copy will be sent to each Chapter.

# Thanks to Lois Erickson

Lois Erickson will be retiring from the Board of Directors this year. Lois has been one of the Directors of the VHL Family Alliance since its birth in 1993. She has served in a number of roles: membership, fundraising, professional education, and on the 800 line, working to build the Alliance. We are very grateful to Lois for her five years of faithful service. We look forward to continuing to work with her in her on-going role as Chairman of the Minnesota chapter. Lois lives with her husband Donald in Bloomington, Minnesota, and has two children: Lois and Chad, and one grandchild.

# Thanks to Madge Hall

Madge Hall has resigned from the Board, but continues as Chairman of the Oklahoma chapter. She is moving into retirement in Oklahoma, and has stepped down from the Public Relations Committee. We thank Madge for her service during the past year.

## Show Us Your Stuff!

***Photos - Drawings - Paintings***

We are seeking art work for the 1999 VHL Calendar! They can be any photograph, drawing, or painting done by a member of our community. Please send *only copies*, as the submission cannot be returned.

Please send by 15 August to: Pierre & Lisa Bonneau, 1761 W. Waterbuck Drive, Phoenix, Arizona 85742

Twelve winners will be chosen, and will be notified by September 1. Winners will be featured in the 1999 VHL Calendar.

***This issue is dedicated to the memory of Sharon Regal Pastway*** (mother of Tania Durand) who will be greatly missed by all who knew and loved her. This issue has been dedicated to her by the donations of loving family members and friends in a final tribute to her life long battle with VHL.

***Our thanks for Contributions . . .***

Carolyn Bishop, Teresa Blazich, Kathy & Andy Braden, Lorraine Burns,
Dr. John Cabell, Janice Capinegro, Chase Manhattan Bank, Cathy Clifford, Audrey & Donald Clifton, Richard B. Cohen, Conrail Corporation,
Hazel Dalton, Mrs. Dalton's Sunday School class, Alexander Dimont,
Constance Fiala, Joan & Bruce Freed, Karen Freed, Cynthia Griffin,
Betsy Hansen, Kelly Heselton, Carole Hohmeier, Melvin Holm, H. L. Huff,
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Terry Trim, Jeffery Vance, M.D.,
Walmart, Corinth, Mississippi, Tim & Heidi Walter, Lisa Whitten, Andrea Witte,
Beverly Yoder

***In Honor of . . .***

Terry Blazich, by Jo Anne McClamroch, Bernard L. Shipp
Charles Cannon Jr on his 60th birthday, by Ethel Kligerman, Pauline Swartz
Alice Coday, by Stephanie Armacost
Mary Christine Donohue on her 2nd birthday, by Evelyn & Bob Werner
Lois Erickson, by Temple Inland Foundation
Joyce Graff, by Nadine Chase, Morton Morgenstern
Kelly Heselton, by Paul Downes
Camron King, by John King
Becky Lima, by Jean Lima
Becky & Jean Lima, by Julie Jordan
Jean Lima, by Becky & Peter Lima
Dr. W. Marston Linehan, by Patti & Ken Kohlen
Gale Lugo, by Evelyn & Bob Werner
Bob Lydon on his birthday, by Mary Swanston
Cia Manolatos, by Patti & Ken Kohlen
Fran Mott, by Pauline & Donald Kwiatkowski
Tammy Nash, by the Rienzi Baptist Church
Cathy, by Sandra & Kenneth Noel
Korrina Sanchez, by Commonwealth Land Title Co., Houston, Jed Manocherian
Pat Stepper, by her daughter, and by Mary Gaydos
Cathy Stolle, by Patti & Ken Kohlen
Rick Werner, by Evelyn & Bob Werner
Harry H. Wilcox on his 80th birthday, by Joyce Graff & Rachael Morgenstern
Dr. Berton Zbar, by Patti & Ken Kohlen

***In Loving Memory of . . .***

Rondo Anderson, by Mary Dufrene
Nelson Boschen, Sr., by Nelson Boschen
Bernard Christensen, by Carol Ausloos, Karla Benton, Jean Boelter, Tondra Brunk, Jean Christensen, Mardene Cosgray, Carol Erdmann, Jacqueline Fenton, Richard Harley, Katherine Hynek, Betty Jacob, Nancy Klatt, Gerald Linzmeyer, Peggy & Don Marshall, John Melchior, Beverly Menghe, Patricia Millot, Hugh Murphy, Roberta Peterson, Rebecca Stewart
John Christoff, by Christine MacDonald
Dr. Charles Clarke, by June & Harry Wilcox
Barbara Eiseman, by Sandy Sharpshair
Dr. Alexander Fedinec, by Joyce Graff
Carolyn Flanagan, by Carl W. Hedberg
Great-Aunt Ruth Fowler, by Peggy & Don Marshall
Eldon Frerking, by Vicky & Stanley Palmer
Allen Fry, by Charles & Dorothy Mayo
Gustave Gertz, by Claire Duquette, Ruth Fortune, Richard Gertz, Martha Guzzardi
Charles Hall, by Beavercreek Church, Wilma Back, Irene Bechtold, Richard Blaylock, Gene Carus, Leonard & Betty Daws, Gerard Insurance Agency, Diana Hall, Diane Holcomb, Helen Johnson, Mary Keifer, Betty Lewis, Dorothy McIlvaine, Martha Miley, Beverly Moon, Patricia Rowland, Virginia Rudduck, Clarine Swayne, Geraldine Townsend, Mary Wuest, and the Andrea Circle neighbors
Rob Harding, Australia, by Joyce Graff, Jennifer Kingston
Joe Taylor Jobe, by Peggy & Don Marshall
Beatrice Johnson, Fred's mother, by Altheada & Fred Johnson
C. Michael Kruse, by Dennis Bentson, Barbara Cabezas, Cornelius Coughlin, Anne Denton, LeBoeuf, Lamb, Greene & MacRae, Marc Maloney, Thomas McCaskill, Lynn McConnell, Frances Murphy, James Nicholls, Nancy Norman, Douglas Reid, Sheila Simpson, Richard Soble, Sally Sposato, Stratfield School PTA, Leslie Okin, Margaret Vandenberg, Christopher Woolley
Tim Mason, by Jan Mason and the Orchestra of the Age of Enlightenment
Alice Masse, by Micheline Masse-Lysell, Louise Bhattachattaya, Paul Bonneau, Gertrude Briere, Fondation Martineau Drapeau, Claude Dufresne, Hubert Palardy
Leslie Meese, by Kathleen Mylecraine
Meg Moody, by Abbott Laboratories, and by her father-in-law Thomas Moody on her birthday
J. Michael Murray, by Joyce Graff
Anna Mae Noel, by Anton Noel
Sharon Regal Pastway, by Ann Babson, Tania Durand, Gail George, Sharon Luttazi, Leslie Meau, Kelly Regal, Stacey Regal, Anthony & Jill Valentino
Raymond Pulley, Sr. and Jr. by Marguerite Golding
Matthew Rider, by Rita Miller
Robert Schumacker, by Helen Dankenbring
Mrs. Dale Wendler, by Joyce Graff
Robert Jr. & Thomas Werner, by Evelyn & Bob Werner
Diane Wilcox, by Sidney & Dorothy Cohn

***For their gifts and services . . .***

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***.... and other members of the VHL Family Alliance***

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### *God's Living Trees*

*-- poem and illustration by Sherri D., Tennessee*

I thank God for his Word I see!
We are to stand, in this world like a tree!
Strong and rooted in his Word!
Telling others what we've heard.
Summer or winter, sunshine or rain,
God is with us through every pain.

Our leaves may wither and fade away,
Our branches may get broken, and fall astray,
Our trunk may lean, or be bowed down!
God promised to always be around!
Have faith in God and His love.
He loves us all the same,
Thank Him and praise his name!

---

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# Chapter News

### *Michigan, by Fran Mott*

The Michigan chapter has been having success with frequent small get-togethers of 2-14 members, usually for a Saturday brunch in a public restaurant, in different locations around the state. This offers people an opportunity nearby to meet others with VHL.

A state-wide social was given by the Michigan Chapter on May 2, 1998, in Grand Rapids at the Breton Health Plaza. There was an abundance of coffee, juice, donuts, and informational handouts from many sister organizations for all who were able to attend. Attendees heard a very beneficial presentation on stress management, given by Gerri Navarre, MSW, ACSW, a supervisor for the Patient and Family Counseling Department of Metropolitan Hospital in Grand Rapids. Everyone, including Ms. Navarre, had an opportunity to learn about VHL and the VHL Family Alliance as they looked over displays and tables lining one entire side of the room and selected information to take home. Along with an enlightening slide presentation and open discussion on approaches for managing stress, Ms. Navarre brought along numerous topic-related books for us to look over and provided everyone with a Stress Audit from the Biobehavioral Institute of Boston to determine their individual level of susceptibility to stress and factors that contribute to it. New faces were seen, much was learned, and an enjoyable morning was had by all.

### *California, by Dawn Cerf*

Thirty-eight members of the California Chapter gathered on April 18, 1998, on the beautiful campus of Stanford University, for their fourth annual state-wide meeting. About half of the attendees were returning members, and it was the first VHLFA meeting for the other half.

Our speaker was Dr. John Adler of Stanford's Department of Neurosurgery. Dr. Adler has previously spoken at VHL nationwide conferences and is on the cutting edge of VHL stereotactic treatment of the brain and cervical spine. He inspired us with information on current and future possibilities of treatment, including the possibility of using stereotactic radiosurgery on all parts of the spine and eventually internal organs. His talk ended with a tour of Stanford's new Cyberknife machine.

Members also gathered for a roundtable discussion of experiences, concerns, and coping methods. Many issues surfaced, including the emotional issue of childbearing. Everyone agreed that the recent VHL Family Forum newsletter handled the topic extremely well.

It was a wonderful day of knowledge, networking, and shared feelings. Dinner at a nearby restaurant afterwards allowed some of use to have more time to socialize. One quote from our meeting's evaluation form sums up the general feeling of everyone: "It is nice to know the support and knowledge are out there. Thank you!"

**VHL Family Forum**

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171 Clinton Road
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