



11. What machine was used for your procedure?	Treatment #	1	2	3	4	5	6
Gamma Knife (metal helmet)							
Linear Accelerator (LinAc) (metal frame on head)							
Peacock Conformal Therapy							
Proton Beam							
Cyberknife (no frame or helmet)							
Shaped-Beam							
Other (fill in) _____							

12. After the surgery, how did you feel? On a scale from 1 to 10, with 1 meaning really sick and 10 meaning feeling great, how did you feel...

Treatment #	1	2	3	4	5	6
a. Before the procedure						
b. The day of the procedure (or beginning of treatment)						
c. The day after the procedure (or end of treatment)						
d. One month following the total procedure						
e. Two months following the total procedure						
f. 3 months after						
g. 4 months after						
h. 5 months after						
i. 6 months after						
j. 7 months after						
k. 8 months after						
l. 9 months after						
m. 10 months after						
n. 11 months after						
o. 12 months after						

13. If you were not feeling as well one year following the procedure as you had felt before the procedure, please explain (add paper as needed) \_\_\_\_\_

14. Were you given medications to help with any symptoms?	Trt#	1	2	3	4	5	6
Steroids for swelling							
Anti-inflammatories							
Pain medications							
Anti-depressants							
Other _____							

15. Did the medications work as expected? (Yes/No)

16. If not, please explain (add paper as needed)

17. Did the treatment succeed?

The tumor stabilized (did not grow)

The tumor shrank or disappeared

The tumor grew

The tumor was treated again with SRS

The tumor was surgically removed

Other, please explain (add paper as needed) \_\_\_\_\_

18. If needed, would you have SRS again?  Yes  No

19. Would you recommend this procedure to someone else?  Yes  No

Comment: \_\_\_\_\_

20. Is there something you wish you had known before you undertook this procedure? (add paper as needed)

21. May we contact you to follow up on this questionnaire?  Yes  No

22. Please enter your contact information for follow-up:

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

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

City/State/Zip/Country \_\_\_\_\_

Telephone: \_\_\_\_\_

Email address: \_\_\_\_\_

**Thank you very much for participating!**

Mail completed questionnaires to: VHLFA, 171 Clinton Road, Brookline, MA 02445



## 20 Tips to Help Prevent Medical Errors

### Be an Active Member of your Child's -- or your own -- Health Care Team

Adapted from a Patient Fact Sheet. AHRQ Publication No. 02-P034, September 2002. Agency for Healthcare Research and Quality, Rockville, MD. [www.ahrq.gov](http://www.ahrq.gov)

Medical errors are one of the leading causes of death and injury. Rates of medication errors and adverse drug events for hospitalized children were comparable to rates for hospitalized adults in a 2001 study in the *Journal of the American Medical Association*. However, the rate for potential adverse drug events was three times higher in children, and substantially higher still for babies in neonatal intensive care units. Studies of medical errors outside the hospital are just getting underway.

This fact sheet is intended to help parents help their children avoid medical errors. It is good advice for any patient and their partner or advocate.

#### What are Medical Errors?

Medical errors happen when something that was planned as a part of medical care doesn't work out, or when the wrong plan was used in the first place. A 1999 report by the Institute of Medicine estimates that as many as 44,000 to 98,000 people die in U.S. hospitals each year as the result of medical errors.

Medical errors can occur anywhere in the health care system: Hospitals, Clinics, Outpatient Surgery Centers, Doctors' Offices, Pharmacies, and Patients' Homes.

Errors can involve Medicines, Surgery, Diagnosis, Equipment, and Lab Reports.

Most errors result from problems created by today's complex health care system. But errors also happen when doctors and their patients have problems communicating. For example, a study supported by the Agency for Healthcare Research and Quality (AHRQ) found that doctors often do not do enough to help their patients make informed decisions. Uninvolved and uninformed patients are less likely to accept the doctor's choice of treatment and less likely to do what they need to do to make the treatment work.

#### What Can You Do?

##### Be Involved in Your Child's Health Care

1. The single most important way you can help to prevent errors is to be an active member of your

child's health care team. That means taking part in every decision about your child's health care. Research shows that parents who are more involved with their child's care tend to get better results. Some specific tips, based on the latest scientific evidence about what works best, follow.

#### Medicines

2. **Make sure that all of your child's doctors know about everything your child is taking and his or her weight.** This includes prescription and over-the-counter medicines, and dietary supplements such as vitamins and herbs.

At least once a year, bring all of your child's medicines and supplements with you to the doctor. "Brown bagging" your child's medicines can help you and your doctor talk about them and find out if there are any problems. Knowing your child's medication history and weight can help your doctor keep your child's records up to date, which can help your child get better quality care.

3. **Make sure your child's doctor knows about any allergies and how your child reacts to medicines.** This can help you avoid getting a medicine that can harm your child.

4. **When your child's doctor writes you a prescription, make sure you can read it.** If you can't read the doctor's handwriting, your pharmacist might not be able to either. Ask the doctor to use block letters to print the name of the drug.

*cont'd on page 2*

#### Inside this issue!

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| Green Tea           | <b>Spanish, and Chinese!</b> |
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**5. When you pick up your child's medicine from the pharmacy, ask: Is this the medicine that my child's doctor prescribed?** A study by the Massachusetts College of Pharmacy and Allied Health Sciences found that 88 percent of medicine errors involved the wrong drug or the wrong dose.

**6. Ask for information about your child's medicines in terms you can understand--both when the medicines are prescribed and when you receive them at the hospital or pharmacy.**

- What is the name of the medicine?
- What is the medicine for?
- Is the dose of this medicine appropriate for my child based on his or her weight?
- How often is my child supposed to take it, and for how long?
- What side effects are likely? What do I do if they occur?
- Is this medicine safe for my child to take with other medicines or dietary supplements?
- What food, drink, or activities should my child avoid while taking this medicine?
- Is the dose of this medicine appropriate for my child based on his or her weight?
- When should I see an improvement?

**7. If you have any questions about the directions on your child's medicine labels, ask.** Medicine labels can be hard to understand. For example, ask if "four doses daily" means taking a dose every 6 hours around the clock or just during regular waking hours.

**8. Ask your pharmacist for the best device to measure your child's liquid medicine. Also, ask questions if you're not sure how to use the device.** Research shows that many people do not understand the right way to measure liquid medicines. For example, many use household teaspoons, which often do not hold a true teaspoon of liquid. Special devices, like marked oral syringes, help people to measure the right dose. Being told how to use the devices helps even more.

**9. Ask for written information about the side effects your child's medicine could cause.**

If you know what might happen, you will be better prepared if it does--or, if something unexpected happens instead. That way, you can report the problem right away and get help before it gets worse. A study found that written information about medicines can help people recognize problem side effects. If your child experiences side effects, alert the doctor and pharmacist right away.

#### *Hospital Stays*

**10. If you have a choice, choose a hospital at which many children have the procedure or surgery your child needs.** Research shows that patients tend to have better results when they are treated in hospitals that have a great deal of experi-

ence with their condition. Find out how many of the procedures have been performed at the hospital. While your child is in the hospital, make sure he or she is always wearing an identification bracelet.

**11. If your child is in the hospital, ask all health care workers who have direct contact with your child whether they have washed their hands.**

Handwashing is an important way to prevent the spread of infections in hospitals. Yet, it is not done regularly or thoroughly enough. A study found that when patients checked whether health care workers washed their hands, the workers washed their hands more often and used more soap.

**12. When your child is being discharged from the hospital, ask his or her doctor to explain the treatment plan you will use at home.**

This includes learning about your child's medicines and finding out when he or she can get back to regular activities. Research shows that at discharge time, doctors think people understand more than they really do about what they should or should not do when they return home.

#### *Surgery*

**13. If your child is having surgery, make sure that you, your child's doctor, and the surgeon all agree and are clear on exactly what will be done.**

Doing surgery at the wrong site (for example, operating on the left knee instead of the right) is rare--but even once is too often. The good news is that wrong-site surgery is 100 percent preventable. The American Academy of Orthopaedic Surgeons urges its members to sign their initials directly on the site to be operated on before the surgery.

#### *Other Steps You Can Take*

**14. Speak up if you have questions or concerns.** You have a right to question anyone who is involved with your child's care.

**15. Make sure that you know who (such as your child's pediatrician) is in charge of his or her care.**

This is especially important if your child has many health problems or is in a hospital.

**16. Make sure that all health professionals involved in your child's care have important health information about him or her.**

Do not assume that everyone knows everything they need to. Don't be afraid to speak up.

**17. Ask a family member or friend to be there with you and to be your advocate.** Choose someone who can help get things done and speak up for you if you can't.

**18. Ask why each test or procedure is being done.** It is a good idea to find out why a test or treatment is needed and how it can help. Your child could be better off without it.

**19. If your child has a test, ask when the results will be available.** If you don't hear from the doctor or the lab, call to ask about the test results.

**20. Learn about your child's condition and treatments by asking the doctor and nurse and by using other reliable sources.**

Ask your child's doctor if his or her treatment is based on the latest scientific evidence. For example,

treatment recommendations based on the latest scientific evidence are available from the National Guideline Clearinghouse™ or other Web sites such as healthfinder® at <http://www.healthfinder.gov>.

## Top 40 Under 40

The Atlantic City Weekly (New Jersey, USA) recently honored forty of the region's "best and brightest young leaders."

"If they make the rest of us look bad, we can't blame them. While many of us barely get by with our day-to-day lives, these 40 individuals have found a way to excel - often in the face of adversity. Our inaugural selection of the Atlantic City area's Top 40 Under 40 proves how rich our region is with talented, successful, charitable and community oriented young people."

Among them is **Scott D. Cannon -- Probation Officer, Superior Court of New Jersey**

The newspaper reports, "Though Scott is considered disabled, he repudiates his disease with a strong work ethic as an exceptional probation officer. Scott has a rare genetic disorder called Von Hippel-Lindau Disease. During his medical treatments, he completed his master's degree from Villanova University and volunteered at the Bacharach Rehabilitation Center. He received the Donald J. Sykes Award and a Certificate of Congressional Recognition from Rep. Frank LoBiondo. Scott is the only individual in the southern New Jersey region to get the Probation Association of New Jersey's Presidential Award."

Scott is the first in his family to have VHL. In 1993, at the age of 28, he was diagnosed with 15 brain tumors, four of which were successfully removed. In 1996 one of the remaining brain tumors had grown quite large. He was sent for embolization in preparation for brain surgery the following day. Embolization is a procedure where a sticky substance like putty is placed carefully in the blood vessels feeding the tumor, in an effort to block them and reduce the amount of blood in the tumor, making it easier for the surgeon to remove the tumor. Just as they completed the embolization, the

tumor ruptured, resulting in massive bleeding into his brain. The team whisked him into surgery for emergency removal of the tumor.

Miraculously, Scott survived, but with significant stroke-like damage. Through his own strong will, and with the help and loving support of his family, he learned to walk, talk, and read all over again.

His mother, Joan, says, "Scott is no ordinary human being. He claims to be accomplishing all he has out of spite. Whatever the doctors said he wouldn't or couldn't do, his goal was to prove that he would and he could."

Scott completed his Master's degree, and was accepted as a Probation Officer for the Superior Court in New Jersey. He had a third surgery in 2001, removing another four brain tumors and a large cyst, with no further deficits.

"Five years ago," Joan says, "a friend asked me, 'Don't you ever ask, why me, why my son?'" My answer was, no, because that would mean that I would want this to happen to you or to someone else and that would make me less than human. But it's more than that. I truly believe that this handsome, determined, strong willed human being is here to be an inspiration for all who see him. My purpose is to be a voice, not only for him, but for all of those who have VHL. Scott is my joy, my miracle."

*Note: The Top 40 Under 40 Review Committee included members of the staff of Atlantic City Weekly and the Greater Atlantic City Jaycees.*



### Recycle for VHL!

Attached to this newsletter is a mailing envelope in which you can mail empty ink cartridges and old cell phones instead of throwing them away. VHLFA will receive a commission for every ink cartridge and cell phone received.

There are over 200 million empty ink cartridges and old cell phones being thrown away annually. Nearly every cartridge except Epson has a value to a recycler. Thirteen brand names of telephones

have recycle value. Old laptop and handheld computers, including PDAs and Palmtops, can also be recycled.

Please send multiple items in each bag to help bring shipping costs down. If you have more to send, please call Brian Lesser at Cartridges for Kids (800-420-0235) to get additional prepaid bags or shipping labels.

Due to postal and customs regulations, this program operates only in the United States.

# Hello to the VHLFA family!

— Sharon H., Michigan

I wanted to write and share a couple of things with you. My family and I are fairly new to the wonderful group of people in the Von Hippel-Lindau Family Alliance. My daughter Kortney who is now 17, was diagnosed with VHL in January of 2004. It first started when she was 14. Although we didn't know at the time it was VHL, much less ever heard of VHL, she began to see what she called a "small black dot" out of her right eye. Once we took her for an eye exam with just our regular eye doctor, things went like wildfire from there!

But before I begin with our story, I'd like to inform you all of a couple of small, but meaningful things I have found to help inform family and friends about VHL and the VHL Family Alliance.

In most email programs you can add a signature to your emails. It's very easy to do, and if you use the email program at all, you'll find you can reach a lot of people by adding this signature. You simply type in what you want your signature to be. For example, mine is

**Sharon (last name and contact info)**

Support us in our fight with von Hippel-Lindau. You can help the VHL Family Alliance help us. Please visit: <http://www.vhl.org> to find out more about this rare disorder. MY FAMILY AND I THANK YOU.

Of course, you can add or delete anything else you want. I have had lots of positive comments on it, and it does encourage people to go and investigate. I also change the color to pink, to make it more catchy, and put it in *italics* and **bold** as well.

The other thing that I came across, was what they call a **CarePage**. I found it on the internet, when Kortney was getting ready to have surgery this past November. It can be found at [www.carepages.com](http://www.carepages.com). This is a wonderful site where you can invite people to go to, and keep updated on you or your loved one's condition. The creator of the page, or anyone you designate, can update the page. You can upload pictures of your own into the page, give them information on the patient's surgery, hospital information, contact information, and just about anything you want! It truly is a wonderful idea and we use it anytime my daughter has any new news, or is going in for any surgery. You have to check it out! It is a very user friendly website. [See note below.]

Since we were informed of Kortney's condition, which was about three years ago, many things have moved very quickly. The black spot I mentioned in the beginning of this article, of course turned out to be a retinal hemangioma. It took us two years to

actually find a good retinal specialist who could determine what was going on. We spent the first two years going to other doctors who simply did not know what it was or what was causing the complications with her vision, and of course, the condition was continuing to get worse.

The retinal specialist tried many things to help her vision, including laser surgery four times, and two steroid shots into her eye to dry up the fluids that the tumor was leaking. All that we were able to do was stop the tumor from growing for awhile.

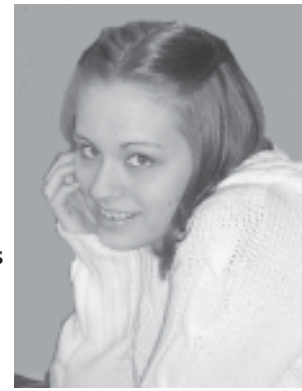
Her vision in her right eye was already down to, at the best, 20/400. That's what they call "finger count vision." The retinal specialist sent us for a second opinion to Will's Eye Hospital in Philadelphia, Pennsylvania. We saw two very excellent doctors out there, a husband and wife team, who basically said that what our specialist was doing was the best anyone could do, and also agreed with him that she should be DNA tested for VHL.

That was our next step. And indeed she tested positive. We then had myself, her sister, and her father tested, all of whose results came back negative. Hence, Kortney was a new mutation.

About two days before we got her DNA results back, she became ill with very bad pains in her stomach area. This turned out to be two cysts, one on her ovary, and one on her fallopian tube. We thought this was just coincidence, as we didn't have the DNA results back yet. Now that we know she has VHL, these were likely APMO's. But since we didn't know yet about the VHL results, the cysts they removed were not sent into the Brain and Tissue bank.

Not too long after all this, Kortney had a fainting spell, which caused us to take her to the emergency room by orders of her pediatrician. He is an excellent doctor, but also one who has never heard of VHL. We are finding this a lot! At the emergency room that day, they ran MRI's on her brain and cerebellum area, and found two very small tumors there, neither of which was large enough to worry about at this time. I could see then that we were on the way to a new lifestyle, many doctor appointments, and even hospital stays, and all the testing that would now be a part of the life of this very spunky 17-year-old young adult, who is never still!

From there, we underwent all the tests that come along with the routine testing in a VHL patient. Blood work and urine tests were coming back with



levels that were abnormal. By this time, it was summer 2004. After MRI's, blood work, three different 24-hour urine collections, and an MIBG, it was determined that she also now had a pheochromocytoma on her left adrenal gland. It was large enough that they wanted to remove it. This we did in November of 2004. She had a partial adrenalectomy. This phoe was the likely cause of the fainting episode.

She came through the surgery like a trooper. And it hasn't slowed her down, not one step!!!! She continues to be very active, and is involved in just about everything she can! She enjoys school now more than ever, tries harder than she ever did, and has a great attitude about having VHL. Her school has also been wonderful and has done many things to assist her with her vision problems. They have been more than accommodating when she misses school work due to the many doctor appointments, surgeries, etc. When I ask her about her VHL, and why she doesn't seem concerned, she just says, "I'm not worried, Mom. I know you are taking care of things right now for me!" I guess for now, this is OK with me. I don't want VHL to slow Kortney down, or burden her. I want her to continue to enjoy her young adulthood for as long as she can without worry. She is doing well enough at this point, that there is no need to have it any other way! In fact, she takes this much better than I do! Sometimes it can be very trying for me.

The eye specialist thinks that her retinal hemangioma may be trying to become active again. But we have all the faith in the world in this man, and know we can trust him to do his best. Finding a doctor you can trust, and being able to rely on them, is half the struggle sometimes. Especially with VHL. Because so many doctors have not heard of this, let alone know what to do, you need to find one that is informed of VHL. It takes work to find the right doctor, and to develop a good working partnership and level of trust.

***Melissa Thomas of Texas is competing in the Lake Placid Ironman Triathlon  
The Janus Fund offers a challenge grant to the top fundraisers for charity***

***Help Melissa do her Personal Best for YOU!***

***Help her meet the Janus Challenge and win more money for VHL research!  
Donate on page 15.***

I was very proud when Kortney took the initiative to help me make posters and distribute donation cans to her school and other local businesses to collect money to help fight VHL. She took pride in sending these monies to the VHLFA.

Three years ago, no one in this family had ever heard of von Hippel-Lindau. Now it's a major part of our life with Kortney, and will always be. But with the help of the Von Hippel-Lindau Family Alliance, the newsletters, the Handbook, and the very many wonderful people I have met through the VHLFA, I have faith someone will always be there for us. That's a very comforting thought. Thanks to all of you I have come in contact with to date, and all of you I will meet in the future. You are all very special people. I hope that this information I have shared with you will help you in some way.

— Sharon, Kortney, family, and friends.

**Editor's note:** [www.carepages.com](http://www.carepages.com) is an ethical commercial venture offering custom branded CarePages services to hospitals in addition to this public site. The web service is funded through advertising. As with any such offering, you should begin by reading its Privacy policy, and then read very carefully the full user agreement, and any of those pesky pop-up boxes that ask your permission. Take care to give your permission thoughtfully, not blindly. In order to earn money from advertisers, they will need to collect certain information from you. You have a right to know what information they are keeping, and what they intend to do with it, and just how public or private your pages will be. In some cases, they give you a web location with a cryptic name, and assume that you are giving that name only to the people you want to share it with. While the chance that someone would guess your URL is remote, people do "random dial" and could stumble on your page. Password protection offers greater security. You want to make sure that any such service you use is going to protect your name, address, and other information. You do not want to find your loved one's name and medical history searchable with a web search engine! There is an internet phenomenon known as a "blog" or "weblog", a kind of personal diary on the internet. Be very careful with these, as your information may become public property.



# How the Weather Affects Your Life

**Editor's Note:** None of the conditions listed in this article are linked to VHL. However, people with VHL do experience a number of symptoms similar to the conditions described here. Often they are caused by increased pressure in the brain or spinal cord, or scar tissue from prior surgeries, and can be made worse by weather conditions. Keep a log, and report these issues to your physician.

For as long as people have been aware of weather, they've pondered its impact on their health. The Greeks noted the effect of "hot and cold winds" on pain and illness 2,400 years ago. During the Civil War, physicians wrote about amputee soldiers sensing pain in their "phantom" limbs when the weather changed. And folk wisdom tells of people who "feel the weather in their bones."

In modern times, doctors continue to explore the connection between weather and pain, especially in relation to chronic conditions such as migraine headaches and arthritis.

While many people maintain that the weather affects how they feel, scientific studies linking weather to chronic pain don't always agree. Some doctors say it's a matter of perception; patients might feel worse on a rainy day, they say, just because it's gloomy. But others say the pain is very real. Not all people who suffer from chronic pain feel effects from the weather. Among those who do, reactions depend on the individual and can be hard to measure.

For those people who are sensitive to weather, *changes in weather* are generally more likely to affect them than specific weather conditions. Doctors who specialize in chronic pain sometimes suggest that patients keep a detailed journal of weather conditions to establish a possible relationship to their pain.

In particular, the following weather factors may contribute to aches and pains:

**Barometric pressure** is the weight exerted by the air around us. Rapidly falling barometric pressure generally signals the onset of stormy weather, and is believed to have a strong correlation with the potential for feeling aches and pains. Rising pressure may also affect some people.

**Humidity** is the amount of water vapor in the air. It can be expressed as "absolute humidity" (the amount of water vapor per unit of air) or the more familiar "relative humidity" (the amount of water vapor relative to the amount of moisture the air can hold at a given temperature). An increase in absolute humidity, especially in the summer, can lead to an increased potential for aches and pains. Some research also finds a correlation between dry, cold air and migraine headaches(1).

**Precipitation** includes not only rain, but also snow, sleet, hail or any other form of water that reaches the ground. It is considered a factor in aches

and pains because stormy weather accompanies changes in barometric pressure and humidity. For those sensitive to hot weather, such as some people with multiple sclerosis, rain can cool the atmosphere and may bring some relief.

**Temperature.** Rapidly rising or falling temperatures are a hallmark of big weather changes, indicating underlying shifts in barometric pressure. Extremes in temperature, not just changing temperatures, can also affect the potential for feeling aches and pains. Low temperatures may trigger migraine headaches, exacerbate circulatory conditions such as Raynaud's phenomenon (2) and contribute to arthritic joint stiffness. Cold weather has also been associated with an increase in asthma-related hospital admissions.

**Wind** often results from big shifts in weather, indicating that barometric pressure and other factors may be changing rapidly. Wind is also a health consideration because it can carry pollution and allergens far distances, therefore affecting people with respiratory ailments such as asthma.

Weather is considered a possible influence on a wide variety of health conditions. In some cases, such as migraine headaches, weather may bring on an episode. In other cases, weather factors may make existing pain worse.

(1) Drs. Matthews and Rappaport reported to the 10th Annual International Headache Congress (2001) that their patients could tell, based on the onset of their headaches, that there was a weather change coming the next day.

"People often implicate high temperature and humidity as the most common contributing factors," Birgeneau Prince said in a statement. "But our study suggests a combination of low humidity and cold weather is even more common than the opposite in triggering migraines."

(2) Raynaud's phenomenon is a condition in which blood vessel spasms disrupt blood circulation to the fingers or toes. The phenomenon, which turns one or several digits pale or bluish, can last minutes or hours, but generally does not cause tissue damage. Pain is uncommon, but there may be partial, temporary paralysis in the affected area. Emotional reactions and cold weather are two known triggers.

From a story by Lucas J. Mire, The Weather Channel, July 2001, Copyright 2005, The Weather Channel Interactive, Inc., [www.weather.com](http://www.weather.com)

***Has the VHL Family Alliance made a difference in your life? Please help to keep our services strong, and fund research. Thank you!***

# Ask the Family

*Excerpt from a discussion in the online discussion list, vhlfa@yahoogroups.com. Reprinted with permission of the authors.*

**Question:** If you have 3 small brain tumors, does it matter what size they are to start to react? I have always thought it doesn't really matter what size they are, they can react at any time or cause problems. It's a question my brother has been asking me, as he's having no luck asking the genetics people.  
— *Lesley B., Germany*

**Answer (1):** This is not a question for the genetics people. The best people to answer this question would be a neurologist or neurosurgeon who is looking at the scans in question. In the brain, it's not so much a matter of size, it's where it is and what it's sitting on. Size matters too, in that there is what they call a "mass effect" — the effect of excess bulk inside your skull. Depending how much extra stuff is in there (tumor, cyst, and/or swelling), displacing normal brain tissue, you can get symptoms just from the compression.

So tell your brother to go see a neurosurgeon or neurologist, get a scan, and have a heart-to-heart talk about what's there and what action (if any) may need to be taken.

I know it sounds scary to talk about brain surgery. But the best time to do it, if you need to, is while the symptoms are mild — present, but mild. You have headaches, you're starting to throw up, maybe you have a little difficulty walking a straight line, but you're not falling down or passing out, and have no slurred speech. Once the symptoms get more serious, then the surgery may be more difficult for the surgeon, and the recovery is going to be more difficult for you.

Scans are wonderful. People who had brain tumors in the 60's and 70's didn't have MRI's. We had to guess. He's throwing up and has headaches that won't quit, and we know he has VHL, so he must have a brain tumor, probably about there, so let's go in and see what we see. (Or worse, we don't know she has VHL so we continue to blame it on early menopause or hysteria or a bunch of other things until she goes into crisis.) Believe me, the scans alone are a blessing.

Today we can see precisely where the problems are, which way we can approach them most successfully, and plan a surgery that will do the best possible job. If we have a series of scans every year or two, we know that the tumors are growing at a particular rate, so we know we should plan to do surgery in the next few months. We can organize a team we feel confident about, and schedule it into our lives. With this carefully planned approach, people are having much better outcomes, with few

***United Way? write us in!***

***CFC? #0242***

***Please print out one or more posters from our website to hang in your office for workplace charity campaigns.***

or no deficits after the surgery.

So it doesn't pay to wait or play games or hope those headaches will quit, or wonder what might be causing you to throw up twice a day for a month — get thee to a neurologist and get the clear information and expert help you will need to get through this in fine shape!!! -- *Joyce Graff, Massachusetts*

**Answer (2):** I agree with Joyce. I agree that it doesn't help to wait and see if the problems will get better. Take action! Don't be so bold as to overreact to everything, but be aware of what's going on and stay on top of your own health. Don't live in fear of what might happen. Be open and honest with yourself and take action when it is needed; don't be ashamed to admit when something is not right.

I suffered from symptoms of a brain tumor for two years. The symptoms were passed off to me as many things, none of which were a brain tumor. I had headaches, dizziness, and routine vomiting. Many doctors told me that it couldn't be a brain tumor, it had to be something else. They were all wrong!

I had a 2cm brain tumor growing, sitting on my brain stem. If they had just thought logically that a girl with VHL might possibly have a brain tumor, and had actually given me an MRI, they would have found the tumor. Instead, they passed off all my problems to be related to some other reason. I am still sick, five years later, because we waited so long to take care of the tumor, not realizing what was going on. If they had just taken the right precautions and looked for one at the onset of the symptoms, I may have been spared all these problems.

The moral of the story is: don't beat around the bush. If you're having symptoms, of any kind, tell your doctor and get a scan. A simple scan may save you from a lifetime of problems. Even more, a scan may save your life. — *Christina D., California*

**Answer (3):** I have to agree with everyone here that has spoken on the benefits of not waiting for any type of test or treatment. I truly believe that this could have saved my brother-in-law's life. If he had known sooner or had surgery faster he may have been alive today. I believe in my heart that it was the fact that the treatments weren't given quickly and the fact that if he had known sooner that he had VHL he would have been here now. I hope that no one waits to have scans or any necessary treatments. It can definitely save your life. — *Mindy G., Mass.*

## Green Tea — Best as a Beverage

Many people around the world drink green tea in hopes of warding off cancer, heart disease and immune system ailments. Green tea is a popular beverage throughout much of the world and has been used medicinally for centuries in China and India. In areas where people drink a lot of green tea, there is a lower incidence of heart disease and some cancers.

In recent years, several manufacturers have produced dietary supplements containing highly concentrated extracts of green tea's active ingredients. The compounds are not toxic in large doses, but high concentrations may not necessarily be healthful.

A study completed by University of Mississippi researchers indicates that extremely high doses of green tea extract actually may activate -- rather than shut down -- genetic mechanisms that help certain tumors survive and grow.

"Drinking green tea still is good for you," said Yu-Dong Zhou, a molecular biologist at the university's National Center for Natural Products Research. "There are thousands of years of evidence on that, but the idea of taking the equivalent of hundreds of cups of tea a day is something that needs to be looked at carefully."

Zhou is principal investigator on a study that examined the effect of high doses of the active ingredients in green tea extract on hypoxia-inducible factor-1, or HIF-1, a key regulator of how tumor cells adapt to low-oxygen conditions. "At low concentrations, it doesn't seem to have this potentially negative effect as we saw in the lab," she said. "A lot more study needs to be done to see what the outcome will be in people who take high doses of these compounds."

"In theory, this effect on HIF-1 could suppress some early forms of tumors but may actually help other tumors -- especially some of the more aggressive ones -- survive and grow," Nagle said. "We can't really tell people for sure what will happen when they take these high doses," she said. "Our best advice is to be careful. This needs to be studied carefully, and it will take time to determine what the actual overall effects are."

Y-D Zhou et al, "Hypoxia-Inducible Factor-1 Activation by (-)-Epicatechin Gallate: Potential Adverse Effects of Cancer Chemoprevention with High-Dose Green Tea Extracts", *J. Nat. Prod.* 2004 67(12); 2063-2069.

Material from Newswise was used with permission in the preparation of this article.

## Smoking and kidney cancer risk

— Jay D. Hunt, Ph.D, *International Journal of Cancer*, 10 March 2005. *Reuters Health*, reprinted with permission.

The link between smoking and kidney cancer may be stronger than experts have appreciated.

Pulling together data from 24 studies conducted since the 1960s, an international research team found that the risk of kidney cancer was 38 percent higher among people who had ever smoked versus those who had never picked up the habit.

And the more smokers had puffed over a lifetime, the greater the risk to their kidneys — a so-called dose-response relationship that supports a direct link between smoking and kidney cancer.

In fact, Hunt told Reuters Health, before conducting this study, he had been skeptical that the relationship between smoking and kidney cancer was real — in part because past studies have yielded conflicting results. Even when an association has been found in individual studies, he noted, it has generally been "modest."

Hunt and his colleagues culled data from 24 previous studies conducted in North America, Europe and Australia, and used a mathematical model to estimate the overall risk of kidney cancer associated with various levels of smoking.

The researchers found that among men, those with a history of smoking were more than 50

percent more likely to develop kidney cancer than those who had never smoked. Among women, smokers had a 22 percent greater risk of developing the disease. Even light smokers — those who averaged fewer than 10 cigarettes a day — were 60 percent more likely than non-smokers to develop the disease. The odds were still higher with moderate smoking, defined as 10 to 20 cigarettes per day.

These findings diverge from previous estimates, according to Hunt, who said that modest levels of smoking have typically been thought to carry little to no risk of kidney cancer.

There was, however, a bright spot in the findings — namely, that the risk of kidney cancer appears to keep dropping in the years after a person kicks the smoking habit. The risk of the disease was substantially lower among former smokers who had abstained for 10 years or more, compared with those who had quit more recently.

This trend, Hunt said, can definitely be seen as "good news" for former smokers.

*Editor's note:* VHL itself is a risk factor for kidney cancer. People with VHL who smoke generally have more tumors and faster tumor growth. (Walther et al.)

**Book Review****Brain Tumors: Leaving the Garden of Eden**

*A Survival Guide to Diagnosis, Learning the Basics, Getting Organized and Finding your Medical Team, by Paul M. Zeltzer, M.D., Neurooncology. Shilysca Press, ISBN 0-9760171-0-5*

This book is the product not only of Dr. Zeltzer's medical training and experience with people with brain tumors since 1978, but also of his interactions with patients, answering questions on websites over a seven-year period. There is much of benefit in this book — as long as you remain keenly aware that there is nothing directly pertaining to hemangioblastomas.

“The perspective I used in writing this book came about after reading *Genesis: A Living Conversation*,” a work that started as a Public Television series in 1996. Acclaimed television journalist Bill Moyers gathered prestigious writers, film critics, clergy, scientists, physicians, and philosophers from many cultures and moderated a round table dialogue. This diverse group studied the stories of *Genesis* and discussed how our reactions to ordinary and familiar situations echoed many themes of these stories.” [p. 19]

“*Genesis* describes Eve questioning the highest authority figure. This parallels an individual's instinctual need to question a doctor's authority about diagnosis or treatment ... and risk the consequences. Other examples include the story of Noah's Ark: ordinary people today caught up in a torrential flood (in our case, following a diagnosis of a brain tumor) and reaching for the Ark of safety during the long journey toward health.” [p. 20]

“In spite of the diagnosis, many people plunge into a search for knowledge, questioning and sometimes challenging their physicians. Others choose to follow their doctors' recommendations without question and believe that Fate will determine the outcome. Which pathway is preferable for you?

“Resolving that conflict is the subtext of this book. How can you work with and understand those feelings, so that you receive the best care, are aware of the choices and alternatives, and do not feel punished? How do you learn to manage the chaos you feel and minimize the confusion so that you can make good decisions? I plan to teach you how.” [pp. 21-22]

The book offers good advice on organizing your information and keeping a notebook, which is a good methodology for all issues encountered with VHL. There is a very good section on using the internet to do research, and how to evaluate the truthfulness of different website. There is good information on how to seek a second opinion, and how to deal with the healthcare system. There are very helpful discussions of medications.

There is also a set of “job descriptions,” including a very good job description for a caregiver [pp. 90-92]

But for people with VHL, there is one major deficit in this book: it does not once mention hemangioblastoma. The thrust of the discussion is for people with malignant tumors who may benefit from radiation therapy or chemotherapy. At this time, neither of those approaches is helpful with VHL.

Hemangioblastomas represent only two percent (2%) of all brain tumors. In this book, there is no one mention of this kind of highly vascular tumor.

This should not keep you from deriving other benefits from the book, but do be careful in reading the recommendations for therapy.

For example, he recommends that “a biopsy should be performed to make sure it is cancer and not something else.” [p. 39] While pathology review of surgically removed tumors is always a good idea, biopsy is NOT recommended for a hemangioblastoma. In a biopsy, they remove a sample of the tissue from the tumor and look at it under the microscope to make sure they understand the nature of the tumor. In the case of most brain tumors, this is certainly good advice. But a hemangioblastoma is made up of capillaries. It is a tangle of blood vessels. If you take a snip of this kind of tumor, what do you think might happen? It will bleed. And bleeding in the brain is called a stroke. So, biopsy is a very bad idea for a hemangioblastoma.

In all other respects, however, there is a wealth of practical advice in this book. Just remember that he is not talking about your particular tumor type. Hemangioblastomas are not malignant, they do not metastasize to other tissues in the body, and they do not respond traditional kinds of chemotherapy and full-brain radiation. All the estimates about life expectancy in this book do not pertain to VHL.

It is important to remember this when reading in the press about new treatments for brain tumors. If hemangioblastomas are so rare that they do not even rate a mention in this book about “all” brain tumors, you can be sure that they are not included in advertising for new methods of stereotactic radiosurgery.

Zeltzer has some good groupings of families of brain tumors. However, hemangioblastomas do not fit into any of his categories. Stereotactic radiosurgery is mentioned only in passing, as a possible treatment for some kinds of brain tumors. But the

special issues involved in radiosurgery for hemangioblastomas are not discussed.

“There are a few important pointers about the politics of getting multiple opinion. First, you do not need to apologize for this; it’s your right! If questioned, you might say, “I’m getting several opinions in order to make the most informed decision possible about my treatment options.” If a doctor gives you a hard time or shows an attitude problem, don’t let it bother you. It’s immaterial if doctors approve or disapprove of your decisions. Remember, you are out of the Garden of Eden already. Now is the time to search for knowledge to make your life better. In many ways, you have to think selfishly and follow your own instincts — no matter what other people say or think.

- Tell your primary physician that you are seeking a second and third opinion.

- He may be helpful in referrals or in interpreting any conflicting information.”

And in the case of hemangioblastomas, don’t stop at second opinions from local doctors. Feel free to seek input from a skilled neurosurgeon with expertise in the vascular structures of the brain and experience specifically with hemangioblastoma.

### **Pancreas Study Open**

Anyone with VHL and a hard tumor in the pancreas may apply.

*Please contact:*

Steven K. Libutti, M.D., Senior Investigator  
Surgery Branch, Center for Cancer Research  
National Cancer Institute, Bethesda, MD  
Tel: 301-496-5049; Fax: 301-402-1788

## **Finding a Neurosurgeon**

The American Association of Neurological Surgeons, a professional organization representing this medical specialty, now offers an online program that helps users locate a board-certified neurosurgeon in their area. You can use the tool to see if the neurosurgeon suggested to you is board-certified, or you can create a list of board-certified neurosurgeons in your city, state or country, worldwide. This easy-to-use, free program can be found at [www.neurosurgerytoday.org/findaneuro](http://www.neurosurgerytoday.org/findaneuro)

If you are not sure of the importance of “board certification” the site offers an explanation, as well. Active members of this list may also be certified by the Royal College of Physicians and Surgeons of Canada (RCS) or the Mexican Council of Neurosurgical Surgery (MCNS). International members are certified by the certifying board in their country.

If you do not have access to the internet, their office may also be reached by telephone or paper mail:

The American Board of Neurological Surgery.  
6550 Fannin Street, Suite 2139  
Houston, TX 77030  
Phone: (713) 441-6015  
Fax: (713) 794-0207  
Email: [abns@tmh.tmc.edu](mailto:abns@tmh.tmc.edu)

If you have a physician you are happy with, who has done a good job for you, please share the name and contact information with the office. We will keep your doctor on our newsletter list, and will be able to supply the name to another patient in your geographic area. *Thank you!*

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### VHL Family Forum, Newsletter of the VHL Family Alliance

Volume 13, Number 1, April 2005, ISSN 1066-4130

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

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# Regional Support Meetings - Please Join Us!

## **San Antonio, Texas, May 21, 10-4**

A regional meeting will be held in San Antonio, Texas, May 21 at the Marriott Rivercenter.

The featured speaker will be Col. J. Michael Lamiell, M.D., of the U.S. Army Medical Corps, whose investigations of a large family in Hawaii from 1977 led us to the finding of the VHL gene.

Registration is \$25 (\$30 at the door), which includes lunch. See page 15.

## **Binghamton, NY, June 4, 10-4**

Cathy Clifford invites you to a regional meeting in the Community Conference Room at Lourdes Hospital in Binghamton. Anyone is welcome, especially people from Buffalo to Pennsylvania.

Featured speakers include Luba Djurdjinovic, M.S., Genetic counselor, and Joyce Graff, Executive Director of VHLFA. Registration is \$10 in advance (\$15 at the door), which includes lunch. See page 15.

## **Manuel de VHL en français - Folleto VHL en español - 中文使用手冊**

The VHL Handbook is now available in Spanish, French and Chinese!

Our thanks to Pierre Jacomet of Chile for the Spanish translation, and to Drs. Karina Villar and Myriam Gorospe for their close review.

Our thanks to Paul Bonneau of Canada for the French translation, and to Gilles Brunet and Dr. Stéphane Richard for their close review.

Many thanks to Dr. Kan Gong, Peking University Hospital, Beijing, for translating the Handbook into Chinese, and to Dr. Zhengping Zhuang of the U.S. National Institutes of Health for review.

## **Annual Meeting**

### **Chicago, IL, June 25, 10-4**

The Annual Meeting of the VHL Family Alliance will be held June 25 in the ballroom of the Union League Club of Chicago

Featured speakers include Dr. Daniel P. Dalton, Urology, Northwestern Memorial Hospital, and Dr. R. Loch Macdonald, Neurosurgery and Radiation Oncology, University of Chicago, and an in-depth discussion of genetics and pre-implantation testing.

Board members will be present, and two new board members will be presented to the membership for election.

Registration is \$25 (\$30 at the door), which includes lunch. See page 15.



An additional nine translations are under way throughout the world. These translations make this critical information available to patients and local doctors in their own language.

According to genetic estimates, there are as many as 30,000 people with VHL among China's 1.4 billion people. We are hopeful that the Handbook will help more of these people achieve a diagnosis, and get the help they need to manage their health.

All local language versions are downloadable from the internet, or available on paper from the Brookline office.

**Support Melissa's Triathlon for VHL, and meet the Janus Challenge! see page 15**



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