

PATHLIGHT

A NEWSLETTER FOR PATIENTS WITH PULMONARY HYPERTENSION, THEIR FAMILIES AND PHYSICIANS.

VOLUME ONE NUMBER THREE NOVEMBER 1990

PATHLIGHT GROWS

by Teresa Knazik

Welcome, newcomers. We now have 35 members and anticipate many more in the coming months. Over 100 copies of issue #3 have been mailed, and many members are sharing copies with physicians and organizations in their communities, helping to spread the word about us. Several of our new members are patients of Dr. Rich at UIC Medical Center, and I want to thank Lisa Kaufmann, RN, for making copies of Pathlight available to those patients.

Would those who have so generously volunteered their time and talents please let me know what skills you can offer? Soon we will form committees to begin working toward our goals, and we will need leadership and committed individuals to do this. Please share any ideas that you may have to further our efforts. We are hoping to incorporate before the end of the year, and our goals are:

- ...to organize a national patients' association with regional and local support chapters.
- ...to make Pathlight accessible to those who need it but cannot afford it.
- ...to help family, friends, and each other understand the pain and fear we experience - coping.
- ...to educate the public about our disabilities.
- ...to promote awareness among family physicians who can aid in early detection.
- ...to encourage research and become informed of research in progress.
- ...to form a "collective voice" so that our needs may be heard among those who have more well-known disorders.

PRIMARY PULMONARY HYPERTENSION PATIENT REGISTRY

by Carol E. Vreim, Ph.D.

Division of Lung Diseases

National Heart, Lung, and Blood Institute

Bethesda, Maryland 20892

The first reported case of unexplained pulmonary hypertension appeared in medical literature in 1891. When a series of 39 patients with unexplained pulmonary hypertension was reported in 1951, the term primary pulmonary hypertension (PPH) was coined to describe this condition.

An outbreak of PPH in central Europe was traced to an appetite suppressant, and the World Health Organization met in 1973 to review the current state of knowledge on PPH. An international, multi-center study was recommended to collect data, but was never undertaken. In 1981 the National Heart, Lung, and Blood Institute at the National Institutes of Health in Bethesda, Maryland established a PPH patient registry.

The objective of the registry was to collect and analyze data on the natural history, cause, progression, and pathology of PPH. For purposes of the registry, PPH was defined as having a mean

pulmonary arterial pressure greater than 25 mm Hg at rest or 30 mm Hg during exercise and was unexplained by any secondary cause. Patients were carefully evaluated according to a standardized protocol to rule out any chronic pulmonary, cardiac or other abnormality which could have caused the pulmonary hypertension.

One hundred ninety four patients were enrolled in the registry from July 1, 1981 to September 30, 1985 and followed for at least 1 year and for as long as 7 1/2 years. Thirty two clinical centers throughout the U.S. participated. At the time the registry was begun, there was an impression that PPH affected almost exclusively women 20-40 years of age; however, the registry enrolled 121 females and 73 males. The female-to-male ratio was relatively constant for all ages. The patients enrolled ranged in age from 1 to 70 years, with a mean age of 36 years. The distribution of patients by race was similar to the general population with 12.3% being

black and 2.3% Hispanic.

The first symptom patients most commonly reported was difficulty in breathing (dyspnea). Other early symptoms included fatigue, chest pain, near fainting (syncope), fainting and palpitations. The mean time from onset of the first symptom until the diagnosis of PPH was 2 years. Although over 90% of the patients were diagnosed within 3 years of the first symptom, on occasion patients would state their symptoms had been present for as long as 20 years before the diagnosis was made.

At the time of enrollment, the patients were questioned extensively about their medical and family history in an effort to identify any factor(s) which might have caused PPH. No correlations could be found with cigarette smoking, use of appetite suppressants, occupation, place of residence, use of oral contraceptives, pregnancy, or prescription drugs. Six percent of the patients reported having

immediate family members with PPH.

Patients on average already had a three-fold increase above normal in mean pulmonary artery pressure, an elevated pressure in the right side of the heart, and a reduced cardiac output at the time they enrolled in the registry. The pulmonary artery pressure in patients who had symptoms for less than 1 year was similar to that in patients who had symptoms for more than 3 years. It was also found that patients whose only symptom was difficulty in breathing upon exercise already had high pulmonary artery pressures. This suggests that the pulmonary artery pressure rises to high levels early in the course of the disease. The duration of symptoms is not a reliable indicator of the severity of the disease, and the progression of the disease may vary considerably from one patient to another.

Tissue samples studied from the lungs of 58 patients revealed the following: 1) increased thickness of muscle in vessel walls; 2) appearance of muscle in the walls of small vessels that are normally nonmuscular; 3) increased number of cells and a deposit of fibrous tissue in the innermost layer of the vessel wall; 4) additional branching or dilations in the arterial walls; and 5) blood clots in the small vessels. All of these abnormalities result in a narrowing or closing of the cavity (lumen) in the blood vessel, which increases the resistance for blood flow through the vessels. No abnormality was found that is unique for PPH.

Because this study was designed as a registry, it was not possible to evaluate the effectiveness of therapy, which would have required a much more costly and complex study design. Of the registry patients, 19% were on chronic drug therapy at the time of enrollment into the registry and 83% received chronic drug therapy upon hospital discharge following cardiac catheterization for registry enrollment. Drug therapy consisted of one or more of the following: vasodilators, digitalis, diuretics, oxygen and anticoagulants.

Following enrollment into the registry 68% of the patients survived at least 1 year, 48% survived at least 3 years and 34% survived at least 5 years. The estimated median survival was 2.8 years (1/2 the patients lived less than 2.8 years, 1/2 lived longer than 2.8 years). It should be emphasized that over 60% of the patients already had moderate to severe disease at the time they enrolled in the registry. Twelve patients underwent heart-lung transplantation.

The registry provided data on the largest number of PPH patients studied to date. Because we still do not understand the cause nor have a cure for PPH, the National Heart, Lung, and Blood Institute remains committed to supporting basic and clinical studies of pulmonary hypertension.

SUPPORT CENTER

To all new members, welcome! The following volunteers offer their support and encouragement to anyone who would like to network.

Robin Weiss (PPH)
1505 Christina Lane
Northbrook, IL 60062 708/564-3022

Jerry McNair (PPH)
854 White Oaks Dr.,
Dixon, IL 61021 815/652-3021

LuAnne Washburn (PPH)
719 N 119th St.,
Wauwatosa, WI 53226 414/258-5355

Sue Kelly (PPH)
P.O. Box 1931
Tucson, AZ 85702-1931

Shawn Wiggins (PPH)
2033 Darrow
Evanston, IL 60201

Please welcome our new support volunteers: Jerry was diagnosed with PPH in June '88 while living in Colorado at an elevation of 8900 ft. He left immediately and his right atrial pressures dropped by 50%. Look for Jerry's article in the next issue. LuAnn was diagnosed with PPH in Aug '88 and is involved in the prostacyclin research program. She works part-time as a pediatric occupational therapist and participates in a cardiac rehab program. Sue is married, mother of 5, and has 11 grandchildren. She was diagnosed with PPH in July '86. Sue stresses the importance of corresponding with other patients who have PPH. She goes to college and will be sharing some of her poems with us. Shawn is 34 and has 2 children who are very helpful around the house. She is on digoxin and, like most of us, is still confused as to what she should and shouldn't do physically. Shawn, diagnosed in Oct '86, works 15-20 hours a week as a secretary and has a very optimistic attitude. Welcome, and thank you Jerry, LuAnn, Sue, and Shawn.

CHRONIC MAJOR- VESSEL THROMBOEMBOLIC PULMONARY HYPERTENSION (C T-E PH)

A Case History by Colonel William
W. Waters U.S. Army (Ret)

My problem started in Dec '77 while on a long road trip, because I failed to stop often to stretch my legs. Several days after reaching my destination I began to experience intense pain in my lower right leg. Several weeks and three misdiagnoses later, I was finally hospitalized with deep-vein thrombophlebitis. Unfortunately, the damage had been done. As I was to learn later, blood clots had formed in my leg and had migrated through my heart to build up a mass in the pulmonary arteries leading to both lungs.

Although I seemed to have recovered from the pain, I became increasingly short of breath. In July '81, I entered the hospital again to learn that the pulmonary arteries to both lungs were severely obstructed and that the internal pressures were four times normal (my introduction to the term "Pulmonary Hypertension" or "PH"). This led to a pulmonary embolectomy at Duke University in Dec '81. This surgery, while not totally corrective, gave me relief until the Spring of '88 when it seemed that suddenly I was becoming shorter of breath and more fatigued without any excess exertion.

Again, I was referred to a pulmonary specialist, who determined that the PH had returned. Unknown to me, he advised my wife that I was terminal. Various medications were prescribed without improving my situation. In Nov '89, I was hospitalized with fluids in my lungs. This turned out to be the turning point in my life. By a miracle of fate, Dr. Norbert Scharff, a cardiologist consultant entered my case. Although he too was not optimistic, he recalled reading of the successful work of Dr. Kenneth Moser's Lung

Center at the UCSD Medical Center in cases like mine. Through the personal efforts of Dr. Scharff, arrangements were made for me to be evaluated by Dr. Moser and his team at the Lung Center. On March 5th '90, I underwent a successful pulmonary **thromboendarterectomy** which not only saved my life, but has apparently extended it to normal life expectancy.

In conclusion, I wish to stress three points: first, embolic PH as differentiated from Primary Pulmonary Hypertension (PPH) may be subject to surgical "cure"; secondly, the capabilities of Dr. Moser's Lung Center are not as widely known among the professionals and potential patients as it should be; and thirdly, I am dedicated to correct the "second" point.

"C" HERE

by Dorothy Olson

Challenge is the backbone of our whole being. It is coping, acceptance and making the choice to make the best of things.

Choices. You are not responsible for your illness, however, you are responsible for how you live with it! Remember the old saying, "If life hands you lemons, make lemonade!!!" Don't allow yourself to sit around and mope.

Change: When faced with the feelings of your many restricted activities, don't think, "I can never do any of those things again." Think, "Maybe I can adjust and do things related to those activities that were so much a part of my life before."

Concentrate: When you have negative thoughts, think, "Cancel," then think about something pleasant or visualize a lovely place.

Courage: It sure takes a lot of this,

right?...facing up to all probabilities, the tests, the coping, changing your way of life.

Communication: Share your inner feelings. Your doctor can treat your physical problem, but your emotional feelings require communication and understanding with the people/family who share your many fears of the illness.

Coping: We need courage to cope, and I think happiness is a form of courage. We need to share that, too.

Consideration: The lives of our families and friends have been changed as well as ours. The problems we have are shared by them.

Cooperation: If your car was making funny noises, you wouldn't wait until it totally quit to fix it, would you? Tests are the only way that doctors can determine what is best for us. Little warnings can turn up in tests that may not be apparent to us.

Cultivate a sense of humor. Humor weathers all storms. Try not to worry or be fearful of the future. Worry is death of happiness. Try to bring some happiness to those around you. "You can't give a smile away. It always comes right back to you."

Dorothy Olson
3215 Monza Dr.
Sebring, FL 33872
813/471-0052

We encourage readers to discuss their healthcare with their doctors. Pathlight will not be responsible for readers' actions taken as a result of their interpretation of information contained in this newsletter.

DEAR DOCTOR

Dr. Rich,

Why are pulmonary hypertension and PPH more prevalent in women? Is PPH equally distributed among all races and ethnic backgrounds? Can a woman with pulmonary hypertension become pregnant and carry to term with no danger to herself or the unborn child? Robin Weiss, Chicago, IL

The reasons for the increased prevalence of pulmonary hypertension in women remains unknown. However, there are many diseases that have specific gender prevalence. Speculation hovers around the genetic and hormonal differences between men and women. Many of the collagen vascular diseases such as lupus and scleroderma also have increased prevalence in women and show many features common to pulmonary hypertension. Thus there might be some link in this regard.

The NIH PPH Registry that was conducted from 1981 to 1987 looked at ethnic and racial distribution of primary pulmonary hypertension. The prevalence of the disease in the United States did not seem to have any increased distribution with respect to either. There has been some suggestion in the medical literature that the prevalence is different from country to country, specifically that it might be higher in India. However, this has never been really tested.

All the medical literature today suggests that pulmonary hypertension carries an extreme risk to both the mother and fetus. Cardiovascular changes that occur in normal pregnancy put increased demands upon the heart and lungs which patient with pulmonary hypertension would not likely handle very well. There are many cases reported of death and cardiac arrest during labor and delivery in women with pulmonary hypertension. In addition, the reduced cardiac output would carry risk to the fetus and likely result in considerably increased fetal mortality. Finally,

there has been a widespread experience that women with underlying pulmonary hypertension can have marked exacerbation of the disease following pregnancy. For all of these reasons it has been a general recommendation that women with PPH do not become pregnant.

Stuart Rich, M.D.
Associate Professor of Medicine
Chief, Section of Cardiology
University of Illinois at Chicago

WANT ADS

Most cities have an agency which acts as a clearinghouse for support groups, helping people locate one they need in their community. I would like to request members to try and find out who is responsible for doing this in their communities, and let them know about us. Thank you.

~~I am interested in organizing regular meetings of patients with PH and PPH in the Chicago area for purposes of support, information, etc. We hope to find a meeting place with good parking and no stairs. If you would be interested in attending, write to: Edys Gordon, 4544 N. Cleveland Chicago, IL 60625. Please include your name, address, phone, and when you could or could not attend meetings, plus any other pertinent information.~~

Those interested in forming a Florida or SE regional support chapter of Pathlight Patients Association for Pulmonary Hypertension please contact Dorothy Olson or Teresa Knazik. Goals will include scheduling annual or semi-annual meetings and organizing intra-chapter and inter-chapter communications networks.

Wanted: Tips on coping with PH, recommended reading, articles of interest. When sending articles, please limit length to 300 words. There is no minimum. Please double space and indicate permission to edit if needed. Include an address where interested parties may obtain further information.

HEALTHY FOOD

by Edys Gordon

Most people know that fish is nutritious, but many people do not know how to cook it. I would like to share with you my favorite fish recipe. As with all of my favorite recipes, this one is both easy and delicious and calls for no salt.

Jimmy's Catfish--Serves 4

4 catfish fillets	dry mustard
1 small onion, thinly sliced	curry powder
1/2 green pepper, thinly sliced	3 Tbsp dry white wine(optional)
1 lemon, thinly sliced	

Wash and dry fish. Arrange the sliced vegetables in a single layer in a glass baking dish, and place the fish on top. Sprinkle fish with dry mustard powder and curry powder (& with wine, if desired). Arrange lemon slices evenly on fish. Cover tightly with plastic wrap; puncture with knife holes in several places. Microwave on HIGH for 2 minutes. Turn dish, and microwave for another 2 minutes. Let rest 1 minute. Check the bottom of the baking dish to test for doneness; when the middle of the dish is hot to the touch, the fish is done. If fish is not done, repeat 1 minute of microwaving, and checking, until done.

This recipe is also good with other white, flaky fish, such as cod, scrod, or whitefish. If you prefer, the fish may be baked or wrapped in foil and grilled. Since this recipe relies on the seasonings to impart zip without adding salt, be sure to sprinkle fish with plenty of mustard and curry powder.

LITERATURE

Books:

The Pulmonary Circulation: Normal and Abnormal by Dr. Alfred P. Fishman Contact: University of Pennsylvania Press Blockley Hall - 418 Service Dr. Philadelphia, Pennsylvania 19104, Phone 215/898-6261

Reprints:

"Primary Pulmonary Hypertension" by Dr. Stuart Rich reprint from Progress in Cardiovascular Disease, Vol XXXI, NO 3 (Nov/Dec), 1988: pp 205-238 Send self-addressed stamped envelope to: Stuart Rich, MD University of Illinois Cardiology Section P.O. Box 6998 Chicago, IL 60680
**Dr. Rich also has an article in last August's issue of Circulation "on fibrinopeptide A levels which has important implications on the management" of PPH.

"Treatment of Primary Pulmonary Hypertension with Continuous Intravenous Prostacyclin (Epoprostenol)" by Dr Lewis J. Rubin reprint from Annals of Internal Medicine Vol 112, NO 7, 1 April 1990 Send request to: Lewis J. Rubin, MD University of Maryland School of Medicine 10 South Pine Street Room 8-00 Baltimore, MD 21201

"Chronic Major-Vessel Thromboembolic Pulmonary Hypertension" by Dr. Kenneth M. Moser reprint from Circulation Vol 81, NO 6, June 1990 Address for correspondence: Kenneth M. Moser, MD Professor of Medicine UCSD Medical Center 225 Dickinson St. Pulmonary Division H-772 San Diego, CA 92103-1990

TIPS

by Dorothy Olson

To help occupy my time, I do couponing and refunding. It's amazing how much I can save, and it is something that can be done while sitting down! I save even more by shopping on "double coupon" days and watching for sales. It is not unusual for me to save 1/3 to 1/2 of my total grocery bill.

I truly enjoy playing Trivia contests with the radio stations. There is no cost involved. Prizes range from dinner at restaurants, amusement park tickets, and theater tickets to gift certificates, citrus fruits, and concert tickets.

Believe it or not, I ride a bicycle. It is geared so that I can ride it without using too much energy, and I don't think there is anyone in the world who can ride slower than I. I peddle a bit then coast. If I get tired I stop for awhile. One thing for sure, one couldn't lose any weight riding the way I do!

Speaking of weight, although diet helps, exercise is the true key to keeping it down. But for us, true work-outs are out of the question. I have found that I can do some exercising in the pool. Many of the things I cannot do out of the water, I can do in the pool. Check with your doctor, first, of course.

Something which is often overlooked and shouldn't be is our health in general. We are naturally concerned with the foremost problem, PH or PPH, but it is imperative that we remember other things can go wrong, also. Don't forget your regular physical.

THANK YOU

...Dr. Vreim for your informative article on the PPH Patient Registry; Dr. Rich for telling us more about PPH; members who volunteered their time, talent, and typewriters!

MEMBERSHIP NEWS

Pathlight is a quarterly newsletter for patients with pulmonary hypertension, their families, and physicians and offers support in coping with this rare disorder. Membership is free, but we ask for a ~~\$10~~ donation to help us meet production and postage expenses. Now, you may order back issues of Pathlight for \$1.50 per copy.

NAME _____

ADDRESS _____

CITY _____

STATE _____

ZIP _____

NEW MEMBERSHIP? _____

BACK ISSUES: #1 _____ #2 _____

AMOUNT ENCLOSED: _____

United Patients Association For
Pulmonary Hypertension, Inc. (UPA~~PH~~)
P.O. Box 061556
Palm Bay, FL 32906-1556

Mail to:

~~670 Teresa Knazik
1060 Pembroke AVE NE
Palm Bay, FL 32907~~

United Patients Association For
Pulmonary Hypertension, Inc. (UPA~~PH~~)
1060 Pembroke Ave. NE
Palm Bay, FL 32907

EDITOR

Teresa Knazik

ASSISTANT EDITOR & TREASURER

Robert Knazik

ADVISE COLUMN

Dorothy Olson

DIET & NUTRITION

Edys Gordon

CONTRIBUTORS

Robin Weiss

Col. W.W. Waters, U.S.A. (Ret.)

Sue Kelly

EDITORIAL ADVISOR

Maxwell Hull

COUNCIL

Scott Szpara

VOLUNTEERS

Carl Knazik

Jason Rife

"Hope is the
thing with
feathers that
perches in the
soul."

Emily Dickinson

"Dear God, be
good to me. The
sea is so wide,
and my boat is
so small."

(Breton Fisherman's
Prayer)

Contributed by Sue Kelly

HELP WANTED

In order for our support group to grow into a successful organization we need more volunteers to take over some of the duties. We need someone who would be willing to do some research and to correspond with professional advisors to help with the "Dear Doctor" column. We would like to include a nutrition and recipe column, so we are

Do stairs daunt you? Turn around and go up backwards. It may look a little strange, but it's easier to PUSH than to PULL. But be careful not to stumble. (A helpful hint from Dorothy Olson. If you know of any let's hear them!)

looking for someone who would be willing to try new recipes and then write about them. As we grow we will need more administrators and directors so anyone interested in helping out with the business duties involved in running a patients association,

please respond.

If anyone would like to contribute any information for publication - medical news, a recipe, a poem, a thought - anything at any time, it will be welcome. We will try to address everyone's needs be it a question that needs an answer, an article to publish, a shoulder on which to cry. Many of us are limited in what we can accomplish alone, but by contributing what we can we can succeed and see our efforts grow. This is your support group - we want to help each other.

EDITOR

Teresa Knazik

ASSISTANT EDITOR AND TREASURER

Bob Knazik

SUPPORT COORDINATOR

Dorothy Olson

VOLUNTEERS

Shirley Brown

Carl A. Knazik

PATHLIGHT SUBSCRIPTION NEWS

Pathlight is a quarterly newsletter for patients of pulmonary hypertension, their families and physicians and offers support in coping with this rare disorder. A one year subscription is \$5 in the United States. If this amount is not easily affordable please indicate so on the subscription form.

NAME

ADDRESS

CITY

STATE

ZIP

QUANTITY

@ \$5 each =

TOTAL

MAIL TO:

PATHLIGHT

C/O Teresa Knazik

1060 Pembroke Av. NE

Palm Bay, Florida 32907

WE REQUEST THAT DOCTORS DISTRIBUTE COPIES OF THE PATHLIGHT TO PATIENTS WITH PULMONARY HYPERTENSION.

ALSO, PATIENTS MAY WANT TO SHARE COPIES WITH OTHERS AND ARE ENCOURAGED TO DO SO.