

# PATHLIGHT

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

VOLUME ONE NUMBER TWO AUGUST 1990

## ***PATHLIGHT WELCOMES NEW MEMBERS***

by Teresa Knazik

Welcome, new members and subscribers to "Pathlight." The repletion of Number 2 speaks for the enthusiastic response we received to the first issue. In this issue we are joined by two new support volunteers, Robin Weiss and Edys Gordon. Edys will also be our nutrition and recipe columnist. You will find details inside on the formation of two new regional support chapters! Chapter goals include organizing group meetings and forming a national communications network between chapters.

Many of those in need of support must live on strict budgets because of high medical costs that PH incurs and cannot afford the subscription fee. On the other hand, we have received donations from concerned individuals that will help in meeting our costs of producing "Pathlight." Because of their generosity we may extend complimentary membership to those in need. Please, don't hesitate to send for your subscription now. Payment is voluntary. In addition to patients, we would like to hear from their family members and friends, physicians and any others interested in our effort.

A message to our new members: feel free to contact any of the volunteers listed in "Support Center" for further encouragement, empathy and friendship.

## **NORD**

by Dorothy Olson with Teresa Knazik

The National Organization for Rare Disorders (NORD) is a non-profit organization dedicated to the prevention, control and cure of all rare "orphan diseases." It is a non-governmental group and works to achieve federal funding for research of rare diseases. It also acts as a referral service putting people in touch with others who may share the same rare disorder and supplying information to help us understand our illnesses. NORD also provides research grants. It is supported by the general public, foundations, corporations, and memorials to loved ones. If you want to join NORD, yearly dues are \$25.00, however, less may be paid if one is unable to afford this amount. NORD was established in the late 1970's to advocate the development and approval of "orphan drugs" to treat rare disorders.

Because these drugs were unavailable to those in need of them, a group of consumers and researchers testified before Congress requesting action. Jack Klugman's

brother noticed an article about this in the Los Angeles Times and brought it to the attention of Jack. In 1981 an episode concerning the plight of people with orphan diseases was featured on Klugman's show, "Quincy." This broadcast got things rolling, and NORD has been growing steadily since.

In January, 1986, McCalls Magazine printed an article on NORD and received thousands of inquiries. NORD was featured on the Phil Donahue Show in the summer of 1987 and the Sally Jesse Raphael Show in April, 1989. These and other programs have helped to educate the public about the need for increased support of research on Orphan Diseases.

If you would like to receive NORD's newsletter, the "Orphan Disease Update," or would like more information write to: NORD

P.O. Box 8923

New Fairfield, CT 06812  
(203)746-6518

## **THOUGHTS OF A PPH PATIENT**

by Shirley Brown

Before encountering PPH I gave very little thought to oxygen. It was a colorless gas that was essential to life. God provided air and I used it - plain and simple! After all air envelopes the earth, people breath it, airplanes fly in it, we have air beds, air rafts, air-conditioning, etc. But along came PPH and no longer is oxygen taken for granted. Now I am pleased when I can breath, walk and talk at the same time!

When I was diagnosed as having PPH my first thought was "things like this happen to other people but not to me." But it did happen to me and I have to live with it, so I decided to learn more about it. I went to the library but found only statistics because PPH is so rare. There was nothing written on "How to live" with the disease and so in total frustration I decided, "Okay, Shirley stop trying to find answers because there are no written answers." I decided to get out and prove that one can live a quality life

even with a rare and mysterious disease. My plans were to go on and live each day and make the very best of it.

There are so many things one can do if you just find another way to do them. I had always loved to read and so I began to read again. Cooking had been a hobby, so I began to watch cooking shows on TV. Then I would go into the kitchen and experiment with new gourmet recipes. I had done some painting in oils and watercolors, and although I was not an accomplished artist, I found I could teach my grandchildren to paint. So they come over one at a time and we paint in the patio. We have a swimming pool and though I can no longer swim, I can sit and watch others swim and put my feet in. I can no longer take walks, but if I am in need of fresh air I can ride my "scooter" (motorized wheelchair). I always loved clothes and shopping but it is impossible for me to walk the malls, so I began to order from catalogs that come to the house. I find lovely things, receive many compliments and take pleasure in telling friends that I shop by mail. Have you tried taking portable oxygen to the movie theaters to see one of the many new movies that are out this summer? It is a wonderful way to spend an afternoon.

What I am attempting to say is find a new way of doing things you enjoy and go for it! The world is full of wonderful people who will help if you will just ask. Go slow, but do everything you can. Do not let fear keep you from doing things you want to do, but remember not to over-do. One thing a day is enough. Until more is learned about PPH, let us not become statistics. We must learn "How to live with PPH" so we can help others with the same problem.

#### IN MEMORY

This issue of Pathlight is dedicated to the memory of Betty Walin.

### SUPPORT CENTER

If you want a helping hand in coping with PH, the following volunteers may be contacted for support:

[REDACTED]  
[REDACTED]  
[REDACTED]  
[REDACTED]  
[REDACTED]  
[REDACTED]  
[REDACTED]  
[REDACTED]

Teresa Knazik (PPH Patient)  
1060 Pembroke Av. NE  
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407/729-0256

Dorothy Olson (PH Patient)  
3215 Monza Dr.  
Sebring, FL 33872  
813/471-0052

[REDACTED]  
[REDACTED]  
[REDACTED]  
[REDACTED]

Our two new support volunteers are Robin Weiss and Edys Gordon, both from Chicago. Robin is 43 years old and an avid Judy Garland fan. She loves winter and thinks chocolate is divine. She likes to read, write, and collect Judy Garland memorabilia. Her therapy includes Capoten, Lazix, Lanoxicaps, and Theodur. She is a "doer" and can be very supportive. Welcome, Robin!

Edys is 42, married, and the "doting mother of a baby boy born in November." Before motherhood she was a researcher and lecturer of biochemistry and genetics at The University of Chicago. Now she works a few hours a week as a medical writer. Her PH was diagnosed in March, '89 "and is probably secondary to sarcoidosis, a disease which involves the formation of benign lumps in various organs, commonly lung tissue and very rarely in pulmonary arteries." Edys' therapy includes prednisone, Coumadin, Lasix, and supplementary oxygen 24-hours a day. Edys will be happy to research and explain medical terms. Welcome, Edys!

### NEW THOUGHTS

by Robin Weiss

Having been diagnosed with PPH last year has caused me to be drawn inward for some self-reflection and reevaluation. Each of us is born with a clean slate. Our capacity to love, hate and fear is not inherent but evolves through our life experiences and how we react and grow from them.

I believe that everyone has had a time when their ego, whether semideveloped or fully inflated, tells them they are special or blessed and that life will be picture perfect. Any serious illness can be devastating, but, to an individual who has experienced this "blessed syndrome," the devastation is more intense. The obvious question is, "Why me?" After months of working through the usual ego deflators and some extremely motivated research on PPH, I reached the conclusion, "Why not me?"

There is nothing notable or remarkable about me to distinguish me from the crowd, except perhaps the ability to create a sort of mental transfer into another person's shoes and view a situation from their vantage point. This has enabled me to touch many people and to offer them a superior look at themselves and circumstances. It has also been an enrichment to me.

My preliminary diagnosis of PPH was in October, 1989. The symptoms, I've since discovered, were consistent with the disorder. For the previous two months I'd displayed extreme cyanosis in my lips, fingertips and knees, water retention of 25 pounds, and experienced 6 or 7 fainting spells per day. Any physical exertion left my heart pounding out strange rhythms of tachycardia and left me breathless and fatigued. A cardiac cath in December confirmed the original diagnosis and I was placed on a regimen of diuretics, anticoagulants and medications to aid breathing while attempting to control the symptoms of the disease.

I am currently considering the benefits of Procardia but have not come to a conclusion regarding the 24 hour cardiac catheterization necessary to test the drug on my system. But, I have learned to take each day as an extension of the time allotted to me and that each of us is unique in our own multifaceted way. We are placed in the world to fulfill our special purpose and must go on with strength, and (utmost) with a desire to help others benefit from our experiences.

## "B" LEAF

by Dorothy Olson

Others' Belief In You. In the many conversations I have had with my PPH and PH friends over the years, I think one thing stands out. When we think of someone with a serious illness we seem to see someone with a "sickly" appearance. Most of us don't look sick, and it is almost as though others question the seriousness of our problem. It is depressing to us as it seems people "think" we are putting on. Yet, the more people are around us, the more they will definitely see the problem when we try to keep up. We must not let others try to force us to do more than we are really capable of doing. If you can, talk about the problem. It will make it easier for others to accept and begin to understand.

Belief In Yourself. Believing in your abilities to cope with and accept your medical problems and making the best of them is essential. You have to believe in yourself. Each of us comes to know our own limitations. We want to keep up, but it won't work and will only speed up the heart/lung deterioration. Some activity is good, but you know how much, and when you are tired, stop. We all want to keep up, but the smart thing to do is heed your body's warning. BE CAREFUL!

Belief In Supreme Being. We each have our own beliefs, and I will not

go into the religious aspects, but I will state that when I look outside and see the wonders of nature around me, I must believe that a supreme being was responsible. I am considered a long-time survivor so I have always felt that in God's plan I have been allowed to live longer in order to help others in need of support. God gives us a challenge to make the best of whatever comes our way. As Dr. Arnold Hutschnecher says in his book, *The Will To Live*, "I tried to walk without a dream, and I fell down."

## CLASSIFIED ADS

**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, please write to Edys Gordon (address in "Support Center"). 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 please contact Dorothy Olson or Teresa Knazik. Goals will include scheduling annual or biannual meetings and organizing intra-chapter and inter-chapter communications networks to aid in the dissemination of information.

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

## DEAR DOCTOR

Dr. Rubin,

What can you tell us about the drug, Prostacyclin? TK, Florida

*Prostacyclin* is a potent dilator of blood vessels, including those in the lung, and also inhibits the aggregation of platelets, which are sticky cellular components of the blood that may adhere to the inner wall of the blood vessels in the lung and, in some undefined way, contribute to the development of pulmonary hypertension. Prostacyclin has been used by several investigators for many years as a test of an individual patient's ability to respond to medications that dilate blood vessels. It is administered intravenously and has the advantages of being potent, short-acting and safe. Additionally, responses to prostacyclin acutely have been useful in determining whether an individual patient will respond to orally active medications.

The medication is administered using a portable syringe pump with the syringe attached to a Hickman catheter. ...to date we have found that long-term continuous infusion prostacyclin may improve the pulmonary hypertension, improve activity tolerance, and may possibly prolong life in this disease although this is less clear-cut at present. Importantly, we feel that prostacyclin may be particularly useful as a bridge to transplantation in severely effected patients.

Prostacyclin is presently an investigational drug with limited availability, however, an application by Burroughs Wellcome (the pharmaceutical company responsible for developing this agent) is under review by the Food and Drug Administration and we anticipate the ability to enter new patients onto prostacyclin by the fall of 1990.

**Lewis J. Rubin, M.D.**

Division of Pulmonary  
and Critical Care Medicine  
Department of Medicine  
University of Maryland Hospital

## HEALTHFUL FOODS

by Edys Gordon

Ever since my husband discovered that he had high blood pressure, our household has adopted a heart-healthy low-fat, low cholesterol, low-sodium diet. Food has always been very important to us and we feel that the food we now eat is delicious. It just took a bit of time and patience to get used to the changes.

One important change in our way of cooking has been to substitute yogurt for sour cream and mayonnaise. We made this change gradually, beginning with the substitution of yogurt for 1/3 of the normal amount of higher fat food. From the following table, you can see that the substitution of yogurt for nearly any amount of sour cream or mayo can help cut the fat by a lot. The table also shows that sour cream is much lower in fat and sodium, and therefore more healthful, than mayonnaise. All entries are for 1 cup.

	calories	fat	cholesterol	sodium
nonfat yogurt	130	0 g	less than 5 mg	170 mg
low-fat yogurt	140	4 g	15 mg	159 mg
sour cream	410	40 g	80 mg	123 mg
mayonnaise	1580	176 g	130 mg	1,280 mg

The following are some of my favorite recipes because they are very easy and because I find that healthful veggies go down much easier with a yummy dip. They can also be used as salad dressings or as sauces with other foods. If you are not yet ready for the all-yogurt approach, try using 1/3 yogurt with 2/3 sour cream. Once you get used to this, try 2/3 yogurt with 1/3 sour cream.

**Bombay Curry Dip** (I make this with all low-fat yogurt to serve with raw vegetables at parties)

- 1 cup low-fat yogurt, or yogurt and sour cream
- 1 tablespoon curry powder, or to taste
- 1 teaspoon honey

**Tivoli Dill Sauce** (good with salmon or green beans)

- 1 cup low-fat yogurt, or yogurt and sour cream
- 1/4 teaspoon mustard, preferably Dijon or other gourmet type (not hot mustard)
- 1 1/2 teaspoon dillweed

**Creamy Italian Dip** (I love this on raw zucchini fingers and green or sweet red pepper slices)

- 1 cup low-fat yogurt or yogurt and sour cream
- 2 tablespoons meatless spaghetti sauce, preferably salt-free (I use Enrico's, from my health food store)
- 1/4 teaspoon garlic powder
- 1/4 teaspoon oregano

**Guadalajara Mexican Dip** (good with cold chicken or in Mexican-flavored dishes)

- 1 cup low-fat yogurt or yogurt and sour cream
- 1 tablespoons salsa (I use LaVictoria medium hot salsa suprema)
- 1/4 teaspoons each ground cumin, chili powder, garlic powder
- 1 tablespoon chopped fresh cilantro (optional)

For all recipes, mix all ingredients well and chill for at least 1 hour before serving. For dessert, please try frozen yogurt as an ice cream substitute. It's delicious and creamy. The fat content usually ranges from 0 to 2%, compared to 11% and up for ice cream.

## OUR THANKS...

...to Dr. Rubin for telling us about his work with Prostacyclin, to our members for volunteering their time, talents and support, and to the generous individuals whose donations will help us to reach others.

## WORDS OF ENCOURAGEMENT

by Dorothy Olson

Make happiness a habit. We make our own happiness. We are not born with it.

The trials of life are far less important than how we deal with them.

Accept change and learn from it.

Survivors have a talent for life; they know we need to treasure the time that is, not was.

It is not easy to be a survivor, but well worth the effort.

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Edys Gordon

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Carl Knazik

## RECOMMENDED READING

***It's Always Something.*** Gilda Radner, 1989, Avon Books, paperback. Gilda's story of her struggle against "the most unfunny thing in the world" - cancer - includes her poignant message on coping with illness. T.K., Florida

***Jane Brody's Good Food Book: Living the High-Carbohydrate Way.*** Jane E. Brody, 1985, paperback or hardback. My favorite book of low-salt, low-fat recipes. E.G., Chicago

***Minding the Body, Mending the Mind.*** Joan Borysenko, Ph.D., 1987, Bantam Books, paperback. The former director of the Mind/Body Clinic of New England Deaconess Hospital, Harvard Medical School, explains how to take an active role in healing oneself by eliciting the relaxation response. E.G., Chicago

***Natural Health, Natural Medicine: A Comprehensive Manual for Wellness and Self-Care.*** Andrew Weil, M.D., 1990, Houghton Mifflin Co., hardcover. How to eat, drink, breathe, relax, and relate for maximum health, along with recommendations for natural treatments and home remedies for various conditions. E.G., Chicago

***Peace, Love and Healing--Bodymind Communication and the Path to Self-Healing.*** An Exploration, Bernie S. Siegel, M.D., 1989, Perennial Library, Harper & Row, paperback. A New York Times #1 best seller. E.G., Chicago

***When Bad Things Happen To Good People.*** Harold S. Kushner, 1981, Schocken Books. A healing message to help understand "Life's windstorms." T.K., Florida

## TIPS

*The tip* in the last issue about walking backwards up stairs also works when going up a hill or incline  
Dorothy Olson

*I often feel dizzy* when I first get out of bed. I have found that it helps if I do leg-lifts before I get up, then get up slowly and sit on the edge of the bed a few minutes. I wiggle my toes and swing my legs, then stand up while keeping my eyes straight ahead.  
..

Dorothy Olson

*Stay busy.* Activities are not always of a physical nature. Live for today, but look ahead and plan things that you can do or make.  
Dorothy Olson

*I keep* inexpensive, light-weight camp stools placed around the house, garage, and porch to sit upon instead of bending when I have to do work close to the floor or ground.

Teresa Knazik

*It is important* that we follow the doctors orders precisely when taking medications. He really can't help you and be in charge of your well-being if you are skipping doses or taking medications in an inappropriate manner. Some drug dosages need to decreased slowly, not abruptly. When a new medication is introduced, ask questions and be sure you understand what it is supposed to do and if there are any side-effects. In some cases, the doctor will have samples of new medications and can use them to see if they are helpful before you get a costly prescription filled.  
Dorothy Olson

*When you feel* really tired, it is imperative to slow down. When your heart beats too fast, your pulse races, or you can't seem to get enough air, stop right then and sit down. Pushing yourself is the worse thing you can do. Your heart is just not able to handle it  
Dorothy Olson

## PATHLIGHT 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 \$5 donation to help us meet production costs.

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CITY

STATE

ZIP

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*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.*