

A photograph of a lush green field. In the foreground, there are many white daisies with yellow centers growing among tall green grass. The background is filled with more tall green grass and some dark, shadowed areas, possibly trees or dense foliage.

PHA's STORY

The Early Years: 1987-1999

PHA'S STORY

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Why a History of PHA?

New volunteers, PHA Board members, potential funders, and PHA staff members have asked how the Pulmonary Hypertension Association got its start. How did a handful of very sick patients and a few highly specialized doctors start paving the path that now leads around the world? How did they set a model of patient/doctor cooperation that is being copied by other disease-related associations? What follows is an inspirational story of synergy, serendipity, altruism, self-interest, love, and determination. Our mistakes are recorded here as well as our triumphs, in the hope that others can learn from them. You will find both dry recitations of facts and journal-like entries that capture the experience of being a PH patient or physician.

Those interested in just one facet of PHA's history, or who want to know what role some individual has played, may use their computer to do an Edit/Find search. Those interested in more recent events can find them on PHA's website: phassociation.org.

This history was written late in 2005, and is the joint effort of many persons, including Judy and Ed Simpson, Dorothy and Harry Olson, and Pat and Jerry Paton, Gail Boyer Hayes, Dr. Carol Vreim, Dr. Bruce Brundage, and Dr. Greg Elliott. They and others volunteered many hours of work, trying to fix memories to paper—or at least into printable patterns of pixels.

Cover photo: During the great Alaskan earthquake of 1964, the second largest earthquake ever recorded, the spot shown in this photo literally collapsed under the feet of the people once living above it. It was a scene of total destruction. But today it is again a place of life and hope. So, too, has PHA brought hope back into the devastated lives of many PH families. Photo by Gail Boyer Hayes.

1987-1989

PATIENTS AND RESEARCHERS BEGIN TO FOCUS ON A DEADLY DISEASE

The Patient Front: Tentative Blueprints for a Patient Organization

Dorothy Olson was changing planes in Cleveland—running for a gate and gasping for breath—when two flight attendants caught her by the arms and told her that she looked too sick to fly. It took some convincing, but the normally healthy Dorothy finally agreed to stay overnight in a hotel before flying home to Indiana. Several days later, while looking out a window and admiring a bunting bird, she fell across her bed and was unable to move a muscle. That was in



**The Olsons
Back Then**

1978. The bunting bird sticks in her mind because it was at that moment her life skidded around a sharp corner. Dorothy was 53 years old, married, and the mother of three grown daughters and a son who had died young. She earned a living in office management, as a troubleshooter.



Dorothy Olson today

Dorothy was hospitalized, and for five weeks her problem baffled doctors. In the sixth week, a young resident timidly suggested the possibility of something called pulmonary hypertension (PH), a disease that he vaguely remembered reading a paragraph or two about in a medical text. PH was thought to be so rare that a doctor would seldom, if ever, encounter such a patient. Her doctors were

unable to tell her anything about pulmonary hypertension, however, or why she had it. They did tell her that--if she was very lucky--she might live another two years. After a while, she recovered enough strength to leave the hospital in a wheelchair, and she found her new motto: "If it's gonna be, it's up to me."

Dorothy, the troubleshooter, wasn't satisfied with what the doctors were able to tell her. She and her husband, Harry, moved to Florida, and when she was physically able to, she contacted libraries, university physicians, and the National Organization of Rare Diseases (NORD) and begged for information on PH. No one knew very much. Dorothy did learn that there were then said to be two types of PH, primary PH (PPH) and secondary PH (PH caused by another disease). It made no difference which type she had, she was told, because the grim prognosis was the same. She was in and out of the hospital and so sick that she had out-of-body experiences and was put on a ventilator for a while.

Doctors finally figured out that blood clots in her lungs were at least the major cause of her PH, and a Greenfield filter was put in her inferior vena cava vein and she was told to take warfarin to reduce the likelihood of new clots forming.

Meanwhile, she wrote letter after letter, and made phone call after phone call. This was long before the Internet existed, and the letters and calls took time and energy, things she had little of. She begged doctors and NORD to give her name to patients who might want to be in contact with other PH patients. The pickings were slim. For nine lonely years, Dorothy Olson kept sending her message out, hoping, like the SETI search for extra-terrestrial life, that there was someone friendly out there who would answer. She hoped to find other PH patients interested not just in friendship, but in putting out a newsletter, printing a directory of PH patients and physicians

with experience in treating the disease, organizing support groups, and attracting physicians and researchers interested in looking for a cure.

Finally, a few responses began to trickle in. All were from women. Too often, she would get a name only to learn the patient had died. The American Lung Association put her in touch with another patient, Karen Cavanaugh. Like Dorothy, Karen had plans for forming a patient association with similar goals. But when Karen moved to Lima, Peru, Dorothy's letters to her went unanswered.

In the fall of 1987, through NORD, Teresa Knazik, a patient with what was then called PPH contacted Dorothy. She was eager to help. Teresa ("Terr") and her husband, Robert ("Bob"), were both in the Air Force and were based in Florida, not too far from where Dorothy lived. Teresa had a background in journalism, and Bob in data processing. Shirley Brown, also a PPH patient, became the third person in the original "pen pal" group. Things began to click.

The Professional Front: Creation of the Patient Registry and Publication of a Paper on Calcium Channel Blockers

Professional awareness of PH was also growing. An Austrian physician, Victor Eisenmenger, had described the symptoms of PH associated with congenital heart problems in the late 1800's. In 1973, WHO sponsored the first medical conference on the disease. In 1976, two British researchers, Sir John Vane and his colleague Dr. Salvatore Moncada, discovered prostacyclin, a vasodilator produced in the lungs. Dr. Vane received a Nobel Prize in medicine for this discovery (and for also showing that aspirin decreases the production of prostaglandins, thereby reducing pain).

It was the discovery of prostacyclin that led to the development of epoprostenol and other laboratory-made versions of prostacyclin. The first use of such drugs was to treat peripheral vascular disease. Now, of course, they are an important treatment for pulmonary arterial hypertension.



Sir John Vane

A handful of other researchers had also done brilliant work related to the disease, and communicated intermittently with one another. There was really no broad coordinated effort to tackle PH head on, however, until the National Institute of Health's National Heart Lung and Blood Institute (NHLBI) formed the Primary Pulmonary Hypertension Patient Registry in 1981 (its official birth name was the Patient Registry for the Characterization of Primary Pulmonary Hypertension).

Dr. Carol Vreim, who had a PhD in physiology and experience as a researcher on the pulmonary circulation, participated in the formation of the Registry from the beginning. Her involvement with PH patients began in 1981, about the same time as recruitment for the Registry began. She is presently the Deputy Director of the Division of Lung Diseases, NHLBI, and served on PHA's Scientific Advisory Board (which has since morphed into the Scientific Leadership Council) from 1992 to 2002. Today she is the NHLBI liaison to the Scientific Leadership Council and to PHA, and she provided the following history of the Registry.



**Dr. Carol Vreim, Deputy Director
of Division of Lung Diseases, NHLBI**

The concept for a patient registry for PPH came from a Working Group on Pulmonary Hypertension* convened by NHLBI's Division of Lung Diseases on September 14, 1978, the same year that Dorothy Olson collapsed across her bed. The purpose of the Registry was to collect data on the natural history, etiology, pathogenesis, and treatment of PPH. (Today the correct name for this type of PH is pulmonary arterial hypertension [PAH], and PAH has been divided into two sub-types: idiopathic PAH [IPAH] and familial PAH [FPAH]. The outdated, somewhat broader, term PPH will be used in this history, however, because applying the new label retrospectively would be confusing and sometimes misleading.)

The registry concept was developed into an initiative and taken to the Division of Lung Disease's Pulmonary Diseases Advisory Committee (PDAC) in 1979. The following year, the idea was enthusiastically endorsed, and then approved, by the National Heart, Lung, and Blood Advisory Council. A request for proposals for a data and coordinating center to run the Registry, and to provide support for a pathology core, was issued in May 1981.

Dr. Alfred P. Fishman, a member of the PDAC, played a key role in establishing the Registry. As a young man, Dr. Fishman had brought the artificial kidney to the U.S., opening the way to the widespread use of dialysis. He worked in laboratories headed by Nobel Prize winners and was himself acclaimed for his work on the interplay of the heart, kidneys, and lungs. He was intrigued by PH, both as a basic and clinical researcher and as a doctor treating patients and recognized the need for developing treatments. The African lungfish was one of several animal models that interested him, because of that animal's ability to survive for years in a state of suspended animation without food or



Alfred P. Fishman, MD

water. Might it be possible to slow the metabolisms of PH patients until therapies to help them could be found? This was just one of his many ideas. PH, he believed, would not be explained by a single cause or mechanism. Now an elder in the field, he is still bristling with research ideas.

Back in the 1980s it was very hard to locate PPH patients, which made it almost impossible to collect information on them. So Dr. Fishman was instrumental in convincing Dr. Claude Lenfant, Director of the NHLBI, to take the Registry initiative to the Council. Without Dr. Fishman's help, the Registry would probably never have been implemented.

In September 1981, a contract for the Data and Coordinating Center was awarded to the School of Public Health at the University of Illinois, Chicago. The project would be under the direction of Dr. Paul Levy, an epidemiologist, and Dr. Stuart Rich, a cardiologist. A subcontract for a Pathology Core went to the University of Pennsylvania under the leadership of Dr. Giuseppe Pietra. Through ads in medical journals and mass mailings to grantees, the Division of Lung Diseases solicited volunteer clinical centers to enroll patients into the Registry. To become a Registry center a clinic needed to: enroll a minimum of three patients a year, be willing to share data, have quality-controlled hemodynamic and laboratory testing, and have endorsements from its sponsoring department and institution.

The Registry components were:

- The data and coordinating center at the University of Illinois.
- A pathology core center at the University of Pennsylvania that did all the pathologic examinations and qualitative and quantitative studies of lung tissue samples. The samples were obtained by clinical centers from biopsies, lungs replaced during a transplant, and autopsies, and were forwarded to the pathology core center. Tissue samples from 58 PPH patients were eventually examined. These studies revealed several distinct pathological patterns of pulmonary vascular disease in these well-characterized PPH patients.
- The clinical centers.

- A steering committee** responsible for developing the patient reporting forms and manual of operations, and for overseeing the activities of the Registry throughout its duration. This group also authored the four articles derived from the Registry.
- A project office at the NHLBI, which was the sponsor of the study and responsible for the overall conduct and funding. The NHLBI project officer (Dr. Vreim) made sure the study was functioning properly—that its recruitment goals were met, regular steering committee meetings held, and data was monitored. She made site visits to the data and coordinating center, monitoring their activities, and ensured sound fiscal management.

To be eligible for enrollment into the Registry, a patient needed a mean pulmonary artery pressure greater than 25 mm Hg at rest or 30 mm Hg during exercise, and normal pulmonary capillary wedge pressure less than or equal to 15 mm Hg, as measured by catheterization of the heart. All causes of secondary PH had to be excluded. For enrollment, the following supporting data had to be submitted: demographic data (age, sex, race, height, weight); results from a chest radiograph, pulmonary function tests, and a lung perfusion scan or pulmonary angiogram; arterial blood gases; intracardiac left-to-right shunts as determined by angiogram, hydrogen curve, indicator dilution text, or oximetry. It was also necessary to know each patient's baseline hemodynamics (right atrial pressure; systolic, diastolic, and mean pulmonary artery pressure; pulmonary capillary wedge pressure; systemic systolic and diastolic pressures; and cardiac output). The investigators were asked to submit follow-up data every six months. They were reimbursed \$150 for each enrollment form and \$25 for each follow-up form submitted to the Data and Coordinating Center.

All PH patients and researchers owe a huge debt of gratitude to the 187 patients, many of whom were very sick indeed, who enrolled in the study from July 1, 1981, to September 30, 1985, and underwent this battery of tests. Follow-up continued through August 8, 1988.

This was the largest number of PPH patients that had ever been studied, and for the first time, all the data were prospective rather than

retrospective (taken from patients' old medical records). The findings were more confirmatory than startling, although some found it remarkable that it took so long for patients to get diagnosed after their symptoms appeared (mean = 2.03 years, median = 1.27 years). A few patients had symptoms for up to 20 years before being diagnosed. This might be a reason so many patients have advanced PPH before the disease is diagnosed.

Before the results came in, it was often said that the disease occurred almost exclusively in young women. But about 30 percent of the patients in the Registry were male, and 9 percent were 60 years of age or older. Only 6.4 percent (12 patients) had first-order blood relatives with PPH.

It was disappointing that the Registry data provided almost no clues about the cause of the disease, which had been the hope when the Registry was begun. Nevertheless, four major publications resulted from the study. The articles dealt with baseline characteristics of the patients, histopathology (microscopic changes in diseased tissues), acute administration of vasodilator therapy, and the survival rates of patients with PPH. To mark the end of the Registry, the investigators held an international meeting on PPH in Philadelphia in 1987.

It was the Registry, and the survival statistics it generated, that made it possible to evaluate the effectiveness of treatments for PH that later became available. The Registry also drew together a core of physicians and researchers interested in PH and helped to stimulate awareness of, and interest in, this devastating disease. Many young doctors who became future leaders in the PH field were involved in the Registry, including Robyn J. Barst, Jim Loyd, John H. Newman, Harold Palevski, David Ralph, Stuart Rich, Sharon Rounds, and Lewis Rubin. Dr. Vreim believes the greatest contributions of the Registry were to publicize the disease and heighten awareness, and to provide patient data that have since been used as

historical controls for many other studies. Prior to the Registry, almost no one (including physicians) had heard of PPH.

The Registry also played a subtle but important role in patient education. Letters and phone calls from PPH patients seeking information arrived at the Registry as soon as it was established. In April 1988 Karen Cavanaugh contacted Dr. Vreim and expressed an interest in forming a patient association. Karen had been diagnosed with PPH the previous year, when she was living in Tampa, Florida. She sent Dr. Vreim a letter she had drafted, addressed to other patients and urging the formation of a patient association. Because Karen was then living in Peru and postage would be very expensive for her, she asked Dr. Vreim to distribute copies of the letter to patients. Dr. Vreim had to tell her that, because of confidentiality rules, she could not distribute it to patients. She did, however, send the letter to all the physicians involved in the Registry.

In the letter Karen outlined her ideas for what the patient organization would do:

- Provide a directory of patients with addresses and phone numbers
- Produce a newsletter with contributions from patients and physicians
- Serve as a point of contact for PPH researchers who needed to collect patient information in their work to find a cure for PPH
- Act as a collective voice to promote greater public attention and research funding
- Organize meetings or get-togethers for patients

Karen's ideas were very similar to those percolating in the mind of Dorothy Olson. Like Dorothy, Dr. Vreim lost touch with Karen Cavanaugh at this point.

In November 1988, Dr. Vreim heard from Teresa Knazik, who had somehow gotten her name from Karen Cavanaugh. At the time, Teresa was mainly interested in starting a database to collect data from PPH patients

(her husband was a computer programmer and they had a friend who was an expert in database design). Teresa also expressed hope that Karen would be successful in establishing a patient organization. Dr. Vreim wrote to Teresa and her friend, the database expert, telling them about the recently completed NIH patient registry, and suggested that it was unlikely anything new would be uncovered by doing another database at that time.

The next time Teresa Knazik wrote to Dr. Vreim, she sent her a copy of the first *Pathlight*.

Publication of Paper on Use of Calcium Channel Blockers to Treat PPH

Another significant development on the professional front was the publication in July 1987 of an article in *Circulation*, "High-Dose Calcium Channel-Blocking Therapy for Primary Pulmonary Hypertension: Evidence for Long-Term Reduction in Pulmonary Arterial Pressure and Regression of Right Ventricular Hypertrophy." The authors were Dr. Stuart Rich and Dr. Bruce Brundage. This was the first study ever to show that a treatment could substantially lower pulmonary artery pressures. It would take many years, however, for this finding to seep into the awareness of the medical community and begin improving the lives of those PPH patients who respond well to these drugs.

**Working Group Membership: Stephen Ayres (St Louis), Edward Bergofsky (Stony Brook), Kenneth Brigham (Nashville), Charles Mullins (Dallas), and Lynne Reid (Boston).*

***Steering Committee Membership: Edward Bergofsky, Chairman (Stony Brook), Stephen Ayres (St. Louis), Bruce Brundage (San Francisco), Katherine Detre (Pittsburgh) Alfred Fishman (Philadelphia), Roberta Goldring (New York), Bertron Groves (Denver), Paul Levy (Chicago), Giuseppe Pietra (Philadelphia), Lynne Reid (Boston), Stuart Rich (Chicago), Carol Vreim (Bethesda), and George Williams (Cleveland).*

1990

THE FOUNDATION IS LAID & *PATHLIGHT* STARTS TO SHINE

The original name for the organization, the United Patient's Association for Pulmonary Hypertension (UPAPH), was chosen by Teresa Knazik. (UPAPH was pronounced U-path). The name Pulmonary Hypertension Association was considered, but discarded; to Teresa the acronym PHA meant Public Housing Authority.

Teresa and Dorothy started a newsletter for PH patients, which Teresa edited and Dorothy christened *Pathlight*. The first issue was published in May 1990. As Teresa said in the lead article, "With *Pathlight* we hope to encourage each other, offer emotional support, exchange hints and tips in coping with daily tasks, and keep informed about research and therapies."



Teresa Knazik

Teresa and Robert Knazik had been full-time college students when Teresa was diagnosed with PPH. Robert also worked full-time in the Air Force and Teresa raised their toddler son. When Teresa got sick, they had to give up hikes in the mountains and long walks on the beach, and Robert dropped out of college for a while and the Air Force moved the family to Florida. It was a low point for the young family. As Robert wrote in the first issue of *Pathlight*: "I was totally absorbed in Terr's illness and the very bad prognosis we had received for her. I didn't know what to do or how to act. I was just going through the motions at work...." Robert and Teresa worked their way out of their funk. But his article helped spread the word that UPAPH/PHA was going to deal with the concerns of not just patients, but also their spouses and other loved ones. It has been said that PH does not strike

just one member of a family. (Later in this history Bob writes more fully about the impact of PPH on their lives.)

Fifty copies of that first *Pathlight* were made at Kinkos and mailed to every PH patient UPAPH knew of. (Robert's mother copied and mailed *Pathlight* for the first few years.) It wasn't a slick publication—there were cross-outs and the three pages were stapled together-- but it found an eager readership. The unsinkable Dorothy Olson wrote the first of her many columns on ways PH patients could keep their spirits up. She began with "A" ideas, such as Accept, Adjust, and Attitude in the first issue, and worked her way through the alphabet to "S" (Stand Up and Be Heard, Sharing, Self-Esteem, etc.) in the summer 1995 issue. Douglas Moodie (a doctor at the Cleveland Clinic), answered a question in the first issue about children and PH. Shirley Brown contributed a from-the-heart piece about the frustrations of coping with PH. "It would be wonderful to talk with someone who actually has PPH," she wrote. "Having such a rare disease leaves one to discover everything for oneself...." Still, she added, "I have everything but good health and yet I have so much more fun than some healthy people." Her comments touch on both the purpose of PHA and the upbeat attitude of so many of our members.

Another article was about the Foundation for Pulmonary Hypertension, which was organized by PH patient and mother of three, Mimi Hansel and her husband Stephen Hansel in 1984. Their Foundation helped fund work under the direction of Dr. Alfred P. Fishman at the University of Pennsylvania to develop a standardized model of PH in sheep. (Mimi died from PPH in 1985, and repeated calls to the telephone number listed for the Foundation [504-533-5888] got a busy signal, perhaps because of Hurricane Katrina.)



Front page of the very first *Pathlight*.

Also in that first issue was a short list of PH patients who had recently died. Such lists would grow longer in the future and are often the first thing read when each issue of *Pathlight* arrives. Finally, on the back page, readers were invited to join UPAPH. The suggested contribution was \$5 a year, but if a patient was on a tight budget the subscription/membership was free.

The first patient to join UPAPH and subscribe to *Pathlight* was Robin Weiss from Northbrook, IL. A PPH patient diagnosed in 1989, she contributed to the second *Pathlight* (published in August 1990). Robin wrote about her anguish after her diagnosis with PPH: "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?'" Another patient to respond was Edys Gordon (a PPH patient from Chicago), who became *Pathlight's* nutrition and recipe editor. Lewis Rubin (a PH doctor and researcher then at the University of Maryland) wrote about early interest in a new drug, prostacyclin, which might help PH patients.

The medical world was beginning to take notice of this active group of patients. UPAPH had 35 members by November, and the suggested amount for a membership donation was raised to \$10.

Contributors to the third issue (November 1990) included Dr. Carol Vreim from the NIH, who offered a history of PH, the PPH Registry, and findings of the Registry; and Stuart Rich, a doctor/researcher then treating PH patients at the University of Illinois in Chicago, who answered a question about PH and pregnancy (not surprisingly, he thought it unwise for a PH patient to get pregnant).

1991

THE FORMATION OF CRITICAL MASS & A FIRST STAB AT INCORPORATION

A Tale of Two Sisters

Meanwhile, in another part of the country, two sisters who lived 300 miles apart and who had not seen each other in a long while, got together for a visit. It was the older sister, Judy Simpson, who traveled from Illinois to Indiana to make the visit possible. When Judy asked her sister, Pat Paton, where she should sleep, Pat said, offhandedly, "Take my bed. I haven't been able to sleep lying down for years."

This news stunned Judy, who was an RN. She learned Pat was also having some other health problems, and urged her to go to the Mayo Clinic in Rochester, Minnesota, to learn what was wrong with her. It was at the Mayo, in December 1987, that Pat was diagnosed with PPH. She was given what was then the standard advice: to get her affairs in order, because she would live only another 6 weeks to 6 months. This critically ill woman had been running two Dairy Queens, handling their finances, and training workers to make Dilly bars, bulb-shaped cones with a perfect twist on top, and correct change!

A new kind of bond formed between the sisters. Together, they learned all they could about this virtually unknown disease and sought ways for Pat to adapt her lifestyle to her diminished reserves of energy. Judy knew a thing or two about the heart and circulatory system from when she taught pediatric nursing, so she went to the Mayo library and read all she could find about PH. She learned that some doctors were trying calcium channel blockers on PPH patients, and Pat decided to volunteer as a guinea

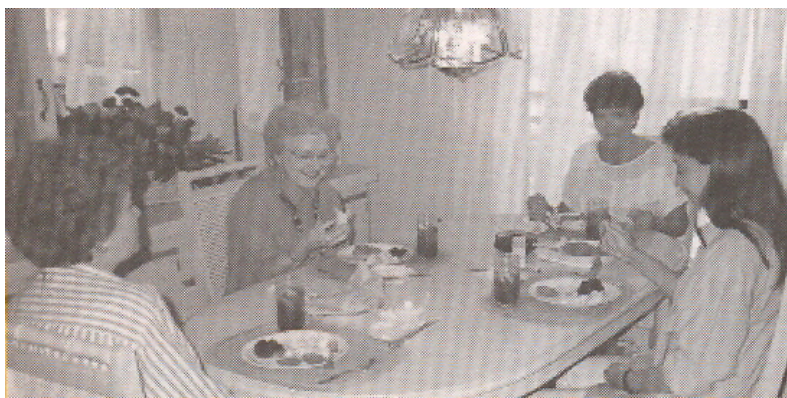
pig. The experiment was done in the Cardiac Critical Care Unit at Mayo, and the pills seemed to work for Pat. (It is now known that calcium channel blockers help only a small percentage of PPH patients, and that giving them to an inappropriate patient can cause death. Pat was just plain lucky.)

Judy knew her sister needed support from others who had PH. She wrote to NORD, but got no names. In 1989, by happenstance, one of her pupils knew someone with PH and they all met. Pat and Judy became more determined than ever to find a way for more patients and their families to meet.

When it looked as if Pat would live for some time, she and her husband, Jerry Paton, began wintering in Florida in 1988, and in 1990 they decided to move there permanently. They sold the Dairy Queen franchises and Jerry gave up his job selling ads for radio and television stations.

The “Kitchen Table” Organizational Meeting

Pat wrote another letter to NORD, and this time NORD responded and the sisters got the names of Dorothy Olson and Teresa Knazik. It was a great stroke of luck that Pat, Dorothy, and Teresa were all living in Florida. Pat organized a meeting of the three patients and their husbands at Pat and Jerry’s new home. Judy and her husband, Ed Simpson, traveled from Illinois to attend. This was the famous “kitchen table” meeting of January 12, 1991; those present are now recognized as the founders of PHA.



From left: Judy Simpson, Dorothy Olson, Pat Paton, Teresa Knazik

It was an unusually synergistic gathering of talent. Dorothy Olson brought her determination, optimistic outlook, skill as an art major, and problem-solving ability. Harry Olson fully supported Dorothy and kept her running at full speed. He also cheerfully filled in wherever he was needed, often serving as a member of the Board. Pat Paton contributed her business skills and acumen, and her husband Jerry was equally adept at reading a balance sheet. Teresa Knazik, as mentioned, could write, edit, and prepare a newsletter, and her husband Bob had business skills and was an early computer enthusiast. Judy brought her medical knowledge to the table--her training as an RN and 20 years as a teacher of nurses. And Judy Simpson and Ed Simpson together had started and run the Presbyterian Marriage Encounter in Illinois, where they worked with volunteers on relationship problems. Before that, Ed had been a professor at Northern Illinois University for 30 years. His specialty was adult education, and he trained business and community leaders in skills such as building relationships. Dorothy has said, "If not for Ed and Judy and their expertise, our organization would never have been successful."

More definite plans for UPAPH were drawn up and tasks assigned to those present. It was agreed that establishing credibility for UPAPH was paramount. Ideas for what UPAPH might do were bandied around. The group wondered if they could possibly form an organization that might:

- Create a network of support
- Keep patients/families informed on current treatments, medications, and research
- Encourage the formation of patient support groups
- Maintain and build contacts with physicians caring for PH patients
- Encourage and support research on PH
- Educate patients and families about the disease

No officers were then chosen, nor tasks assigned; the participants went home and mulled things over. In March, Terry called Judy and said that she had been "appointed" president, because she was healthy and had the most energy.

For the next eight years, the organization would be run entirely by volunteers, often working on kitchen tables. All the surviving founders, with the exception of Bob Knazik, are active in PHA to this day. Dorothy, the oldest patient among the group, still attends Board meetings and conferences. Although her eyesight is failing and her PH has returned to some extent since the thromboendarterectomy that saved her life, her courage is unwavering.

Shawn Wiggins – Organizer of First PH Support Group



Shawn Wiggins, center, with her "reasons for being"—her grandkids Toni and Christopher

In 1991, Shawn Wiggins, a patient of Dr. Stuary Rich's from Chicago, organized the first PH support group meeting. The single mother of two children, Shawn worked part time as a nurse. After she was diagnosed, but before meeting any other PH patients, Shawn spoke with Dorothy Olson on the phone. "She sent me voice recorded tapes for several years," Shawn wrote, "as I believe she did with several others early on." This act of kindness meant a lot to her.

When Shawn visited Dr. Stuart Rich, she watched for other PH patients in his waiting room. Her low-key, optimistic personality drew people to her. Unlike most of the early long-term survivors, Shawn did not respond well to calcium channel blockers, and at first had to treat her symptoms with only digoxin and foroseimide. She was fortunate, however, in having very supportive parents.

In 1989 two important events in her life occurred: she and Jerry McNair organized the first PH support group anywhere in the world, and in November she began using infused epoprostenol. The meeting was held in Shawn's home in Evanston, Illinois. She is probably the longest surviving patient using infused epoprostenol. Today she is doing great, her pressures remain stable, and her activity level is almost normal. She works part-time as a health aide (a job that requires her to travel to sunny Florida with some clients), walks a mile or two daily, and does light-weight training. She does miss being able to swim.



Jerry McNair, co-host of the first PH support group meeting.



**Ed and Judy
Simpson**

The Florida Founders probably learned of the Chicago support group because someone in the group read about UPAPH in a *Pathlight* given to them by Dr. Rich, and contacted Teresa, the editor, and then received a personal call or letter from Pat.

The Chicagoland PH Support Group met for the second time at the Simpson home in DeKalb, Illinois, in October 1991. It had about eight patient/family members drawn from all socio-economic levels. Later it grew to over 65 members. A young patient from Wisconsin who came to the second meeting, LuAnne Washburn (more on her later), was involved in a very early clinical trial at the

University of Maryland of a man-made version of prostacyclin, epoprostenol, which was given by continuous infusion into the right side of her heart. An awkward 50 cc syringe attached to a spring that she wore at her waist controlled the infusion. Her health had improved dramatically with this medication and she was a great inspiration to everyone.

***Pathlight* Continues**

Four more issues of *Pathlight* were published in 1991, and its circulation and list of contributors grew until it was reaching readers in about a dozen states. By early 1991, UPAPH had 53 members. In an editorial, Teresa told about how her South Florida support group believed that patients and family members should be the leaders of a national PH association, but that they should nurture a medical advisory board to include prominent professionals in the field. Dr. Alfred P. Fishman contributed an article on lung and heart-lung transplants, and Dr. Kenneth Moser (University of California, San Diego) fielded a question about the different types of PH.

By the May issue, UPAPH had 73 members. Dauna Leigh Bauer wrote about how contaminated L-tryptophan had given her and others PH, Gisel

Reyes (who had died in February) wrote on having PH secondary to lupus: "Remember that lying in bed won't make anything go away, and it just fuels despair. If I find I cannot get up out of bed I sit up on it." *Pathlight* was becoming a memorial to those who passed on as well as being a tool for the living. Shirley Brown, one of the three original pen pals, died in April following a lung transplant, and her passing was noted. The first memorial contributions made to *Pathlight* were in her memory; other early contributions were made in memory of Gisel Reyes and Shirley Brown. These gifts established an important precedent.

The August issue was mailed to 106 members from many different states. Ohioan Marilyn Dirr wrote about her lung transplant in Pittsburgh, and Sue Kelly wrote about going back to college two years after she was diagnosed with PPH. She hauled her oxygen around with her to classes and learned shortcuts and the locations of elevators. "I AM HAVING FUN!" she wrote. Sue was married, the mother of five, and the grandmother of eleven. Sue's writing skills would prove to be very useful to UPAPH. Dr. John H. Newman (St. Thomas Hospital, Nashville, Tennessee) answered a question about the impact of diet, drinking, and smoking on PH. (He said that a balanced diet is good, smoking is a "horrible habit," and it's probably okay to drink small amounts cautiously.) UPAPH also launched its first "lobbying" effort—asking members to collect signatures to support another patient group, the Organ Transplants of Southwestern Michigan, in asking for a U.S. organ donor awareness stamp. It took a long time, but such a stamp was released in 1998.

In November, *Pathlight* reported on the incorporation of UPAPH and its new officers (see below), and included an article (twice, on two different pages!) on the second meeting of the pioneering Chicago area support group. There was much discussion of the experimental prostacyclin continuous infusion pump that could be fit into a fanny pack, and the annoyance of having strangers say, "But you look too well to have a

disability," when PH patients used handicapped parking. These would continue to be topics of interest to PH patients for years to come. A "Tiny Tip from Pat" told how she always pushed a shopping cart around a store even if she didn't have a thing to put in it, because it gave her something to rest against. That's a little trick many PH patients use. It is also indicative of the nature of the early *Pathlights*, with their heavy emphasis on tips for living with PH and features to lift patients' spirits. This issue again urged readers to support organ donor awareness efforts.

The Struggle to Incorporate UPAPH

Bob and Teresa Knazik and Dorothy Olson worked on incorporating UPAPH. Teresa felt strongly that UPAPH should be incorporated in Florida. With Bob taking the lead, they did the paperwork and met with an attorney who had offered to help in exchange for a reduced fee of a few hundred dollars. They were dismayed when they discovered that he had incorporated UPAPH as a for-profit business, and not as a tax-exempt charity under section 501 (c) (3) of the Revenue Code. Action against the attorney was considered, but organization funds were scarce, and UPAPH's new Board of Directors decided that a lawsuit would cause too much strain on the patients involved. Later (see 1992) a special situation created an urgent need to obtain a nonprofit status.

Officers and Board of Directors

The officers of the new corporation were:

President	Judy Simpson
Vice-President	Teresa Knazik
Secretary	Dorothy Olson
Treasurer	Bob Knazik

Directors-at-Large were:

Pat and Jerry Paton	Harry Olson
Ed Simpson	Edys Gordon
Carla Gordon (Edys' sister)	

The UPAPH Board members financially supported the Association. All travel (yearly Board meetings were required under the Florida incorporation), phone bills, postage, copying, etc., were paid for by individuals. Memorials were the only donations the Association received besides the suggested donation for *Pathlight* (which did not even cover the production and mailing costs of the newsletter).

UPAPH was now handling enough correspondence that it began to flood the Knazik home, so the organization obtained its first post office address: UPAPH, P.O. Box 061556, Palm Bay, FL 32906-1556.

That Personal Touch

In 1991, Pat Paton either talked or wrote to all patients with pulmonary hypertension that UPAPH learned about. She kept a large United States map with pins identifying the location of each patient, and when patients wanted to network she put them in touch with one another. Patients had to first sign a release allowing UPAPH to give out their names. Judy Simpson sent a hand-written thank-you note to every single person who made a donation. By the end of 1991, 500 letterhead note cards had been ordered on which to write thank-you notes to donors, and acknowledgement cards to families whose loved ones had been memorialized.

Forming Connections with Other Organizations

Judy Simpson was asked to attend FLOLAN/PPH Investigators and Coordinators meetings at the Burroughs Wellcome Company as a patient advocate. She attended meetings of the primary investigators during the Phase III clinical trials of epoprostenol (the trade name Flolan had been selected).

At these meetings she learned much of interest to PH patients. Her work with these investigators would lead to appointments on other advisory groups.

1992

TIES ARE STRENGTHENED TO OTHER PATIENT GROUPS AND TO THE MEDICAL/SCIENTIFIC COMMUNITIES

Forming More Connections with Other Organizations

In 1992, Judy and Ed Simpson met with Paula Brazeal, president and founder of the Leukodystrophy Foundation. (Paula's stepdaughter was a student in Judy's pediatric nursing class.) Paula also served on the Board of the National Organization for Rare Disorders (NORD). She helped the Simpson's work through the "dos" and "don'ts" of a newly organized not-for-profit. Paula's wise counsel saved UPAPH several years in time and probably prevented some costly mistakes of the sort that any new organization might make. For example, Paula urged the Simpsons to select a medical board quickly and to ask physicians at the very top of the field to serve. The Simpsons thought this was gutsy, because nobody knew about UPAPH, but it worked. Paula further urged them to be vigorous in any patient services that UPAPH offered and to be ready for opportunities as they appeared, even if UPAPH wasn't sure they could manage them at the time. Ed must have been listening closely, because he proposed an international conference long before any Board member thought such a thing possible. It's easy for an organization to slide back into being reactive rather than proactive, and UPAPH/PHA sometimes does this. We have to keep reminding ourselves to be bold.

Annual Board Meeting

The annual meeting was held in January, in Sebring, Florida, at the Olson's home. Those present included:

Officers:

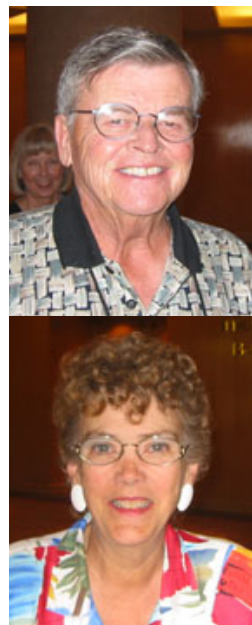
President.....Judy Simpson
Vice President.....Ed Simpson
Secretary.....Dorothy Olson
Treasurer.....Bob Knazik
Assistant Treasurer.... Teresa Knazik

Directors-at-Large:

Edys Gordon	Carla Gordon
Pat and Jerry Paton	Harry Olson

Tasks were assigned as follows:

- *Pathlight* editor, Teresa Knazik
- Patient networking, Pat Paton
- UPAPH member directory, Eleanor Bails
- Scientific Advisory Board liaison, LuAnne Washburn
- Creative writing, Sue Kelly
- Publications and public affairs, Teresa Knazik
- Donations, Judy Simpson
- Mail, Mary Knazik
- Organ donor awareness, Marilyn Dirr
- Volunteer coordinator, Bonnie Dukart
- Chicago region support group, Shawn Wiggins
- Florida support group, Dorothy Olson



**Jerry and Pat
Paton**

It was at this meeting that Ed Simpson made the bold suggestion that UPAPH hold an international conference that would bring together PH patients who may have never met another PH patient, their families, and physicians with knowledge of PH. This suggestion stunned the rest of the Board, including his wife, who was president. The UPAPH treasury was small, holding only \$858.31. And most known volunteers were present at the Board meeting. Nevertheless, Ed said he would organize a UPAPH conference for 1994 and was given approval to do so. Several months later, Teresa

Knazik's sister, Peggy Wessinger, came on board as conference co-chairperson.

Lists of PH Doctors and Patients Prepared

A list of physicians who treated PH was compiled by Judy and made available to patients. Ed drew up a questionnaire, which was mailed to every PH patient UPAPH had an address for, to gain information about their medical condition and to get written permission from those who wished to network with other patients. (At this point, every patient UPAPH heard of was automatically a "member.")

Scientific Advisory Board Created

Physicians who had written about PH in medical journals, PH patients' physicians, and other professionals working to care for PH patients were asked to become members of a new Scientific Advisory Board (SAB); the early SAB was very inclusive. (Years later, its name was changed to the Scientific Leadership Council [SLC]). PHA has never had a refusal from someone asked to serve on the SAB/SLC. In 1992 this distinguished group included:

Linda Clayton, Pharm D (Burroughs Wellcome Co.)
Alfred P. Fishman, MD (University of Pennsylvania Medical Center)
Valentine Fuster, MD, PhD (Harvard Medical School)
Frank Gold, MD (Peoria, IL)
Lisa Kaufmann, RN (University of Illinois, Chicago)
Walker A. Long, MD (Wellcome Research Laboratories)
Michael D. McGoon, MD (Mayo Clinic, Rochester, MN)
Douglas S. Moodie, MD, MS (Cleveland Clinic Children's Hospital)
Kenneth M. Moser, MD (University of California, San Diego)
Harold Palevsky, MD (University of Pennsylvania Medical Center)
Stuart Rich, MD (University of Illinois, Chicago)
Lewis Rubin, MD (University of Maryland)
Warren Summer, MD (Louisiana State University)
Carol E. Vreim, PhD (NIH's NHLBI)
Angela Wurser, RN, MSN (University of Pennsylvania Medical Center)
Diane Zwicke, M.D., University of Wisconsin Medical School

Publication of Follow-Up Study on Calcium Channel Blockers

In July of 1992, Stuart Rich, MD, Elizabeth Kaufmann, RN, and Paul S. Levy, ScD, published a paper in the prestigious *New England Journal of Medicine* showing that calcium channel blockers helped PPH patients who responded to them to live longer. (More recently, they reported that these patients continue to respond 20 years later.) The paper also described the survival advantage of using warfarin anticoagulation along with calcium channel blockers, even if calcium channel blockers were not effective.

UPAPH Becomes a Not-For-Profit Organization in the Nick of Time

A paid attorney was hired and UPAPH refilled, by addendum, for charitable status. On February 11, 1992, it was incorporated as the United Patient's Association for Pulmonary Hypertension (UPAPH), a not-for-profit organization, with officers/directors as follows:

Officers:

President.....Judy Simpson
Vice-President.....Ed Simpson
Secretary.....Dorothy Olson
Treasurer.....Bob Knazik
Assistant Treasurer.....Jerry Paton

Directors-at large:

Pat Paton Harry Olson Scott and Gay Szpara*

*(*Gay Szpara had PPH. She and her husband Scott were very involved for a few months; as Gay became sicker, they dropped off the Board.)*

There was a special urgency now behind obtaining a 501 (c)(3) status, because a foundation wanted to make a grant to UPAPH, but would do so only if UPAPH was a legal nonprofit. The background story is interesting; it is also the story of Dauna Bauer.

The Dauna Bauer Legacy

Dauna was born in Washington State, but spent much of her life in the Washington, DC, area. She developed PH as a result of eosinophilic myalgia syndrome (EMS). She had gotten the EMS from contaminated L-tryptophan. Dauna was a good friend of Heather Foley, who served as the unpaid chief of staff for her husband, Tom Foley, who was Speaker of the U.S. House of Representatives. Heather Foley was a woman whose phone calls were always returned. She called NIH on Dauna's behalf, and that led to Dauna being put in touch with Dr. Carol Vreim. It was Dr. Vreim who told Dauna about UPAPH.

Carol and Dauna also became friends. "Dauna was a very intelligent, talented person," Dr. Vreim says. "She came from a family of very modest means, but had accumulated significant assets on her own and had also received a personal injury settlement from Showa Denko, the Japanese company that manufactured the tainted L-tryptophan." Dauna was only 47 when she died in December 1991, of complications following a lung transplant. She had had a distinguished career in government service as a management analyst, was active in EMS support groups, and helped spur congressional hearings into the sale of unregulated supplements.

When Dauna died, she left \$5,000 to UPAPH, but expressed her desire that UPAPH obtain an IRS not-for-profit status so her estate could obtain the tax advantages. UPAPH had 60 days to do so.

The Knaziks scrambled to gather the necessary paperwork, and on July 10, 1992, the IRS granted an advanced ruling giving UPAPH a not-for-profit status retroactive to March 9, 1992. The Bauer estate issued the check to UPAPH in late 1992.

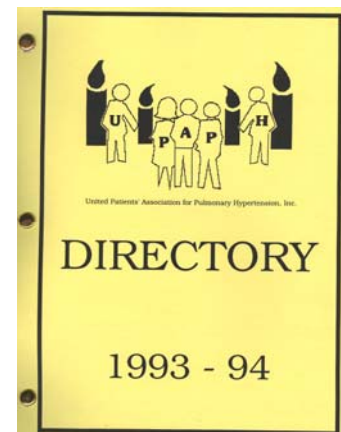
Assets not otherwise disposed of by Dauna's will became the Dauna Leigh Bauer Foundation. UPAPH/PHA is deeply grateful to Dauna Bauer. Her gift not only goaded UPAPH into quickly attaining a not-for-profit status, but the Foundation made another donation to UPAPH/PHA in December 1994, and has made an unrestricted contribution each year thereafter. Because she was thoughtful enough to do some estate planning, Dauna Bauer's legacy continues to enrich the lives of many people.

All donations to UPAPH were tax deductible once it became a legally recognized charity. It is easier to raise money if you have such a status, and UPAPH was growing fast, as was its budget. It's total revenue for 1992 (aside from the Bauer gift) had been just under \$3,000. Of that, \$22 went for a corporate seal, which has probably never been used.

Growth to 141 Members; First Press Releases; PH Patient Directory Prepared

UPAPH now had 141 members and had to get a new, larger post office box and a bulk-rate mailing permit. The permit was useful when the group mailed out its first press releases in December 1992, and sent complimentary copies of *Pathlight* to physicians at most of the teaching hospitals in the U.S.

A directory of patients' names and addresses was compiled by Eleanor Bails. Eleanor, an elementary school science teacher in Ohio who was diagnosed with PPH in 1989 at the age of 36, was one of the first patients to try calcium channel blockers. Fortunately, she was also one of the PH patients who respond well to these drugs, and she experienced a wonderful improvement. Before listing a patient in the directory,



Eleanor made sure they had given their written permission to be included. Because of patient networking enabled by the directory and *Pathlight*, some of the patients involved in the epoprostenol (Flolan) clinical trial that was underway were able to contact each other during the trial and compare notes. Investigators and physicians were not particularly happy about this. The Board decided to discontinue the patient directory in 1994 because of concerns that violations of privacy and unauthorized use of the directory might occur.

A Third Support Group Started

Bonnie Dukart, a New Jersey resident, was diagnosed with PH in 1982 when she was 25 years old. For some time thereafter, calcium channel blockers allowed her to continue a successful career in banking, and she was promoted to vice president of a large New York bank. In 1992, she learned about UPAPH over the Internet and volunteered to help. After visiting the Chicagoland Support Group (which included people from Illinois, Michigan, Wisconsin, and Indiana), she started a group in New Jersey. During this busy year, UPAPH assigned volunteers like Bonnie to tasks in areas that interested them. (Bonnie herself was appointed volunteer coordinator.) UPAPH responded to volunteer opportunities as they arose even though, at times, the group was not certain that the task at hand could be accomplished. Over the years UPAPH/PHA has continued to seize opportunities, ready or not. Volunteers with skills that UPAPH/PHA badly needs often materialize as if by magic. Some volunteers work their way up in the organization and become Board members and officers.

NHLBI Booklet of Pulmonary Hypertension Published



After the creation of the PPH Registry, the NHLBI's Division of Lung Diseases and the Office of Prevention, Education and Control of the NIH had a huge number of inquiries about the disease. In response, they published a booklet, "Primary Pulmonary Hypertension." It was created by Dr. Carol Vreim and a science writer hired by the NHLBI. Through Dr. Vreim, the booklets were made available to UPAPH to distribute to PPH patients; many doctors also requested them. Tens of thousands of the booklets were printed. For a long time this was the most comprehensive information on PPH that was designed for patients. The booklet was revised in 1996 by staff from the NHLBI's Office of Prevention, Education and Control, with input

from Dr. Vreim. Several PPH experts reviewed all versions prior to publication. (Today, everything goes on the NIH website; printed versions are no longer available.)

***Pathlight* Highlights**

For the first time, *Pathlight* contained a story about an upcoming PPH clinical trial (epoprostenol/Flolan, Phase III) so that patients interested in participating could contact their physicians.

Edys Gordon, *Pathlight's* food editor, wrote for the summer issue on how her lung transplant changed her life. She told how she had to wear a hat and suntan lotion to avoid sun exposure, and avoid crowds, most salad bars, and cut flowers in stale water. Her article offered many helpful tips for other transplantees or those considering a transplant.

The back page of the summer issue had information on the Orphan Drug Act and urged readers to ask their senators to support a 1992 amendment to it. The original Act, which became law in 1983, created incentives to spur private companies to develop treatments for rare diseases in markets with so few patients (fewer than 200,000 in the U.S.) that they would otherwise remain “orphan” drugs with no one to push for their development. The law gave a longer marketing exclusivity period, a tax credit, etc.

NORD believed that drugs that cost up to \$30,000 to \$350,000 per year per patient were so profitable that they would be developed without incentives, and wanted to see the law returned to its original intent—to encourage the development of drugs of little commercial value.

The Travels of Tess

In the February 1992 issue, a new contributor was Tess de Guzman, who wrote about the agony she went through to get a correct diagnosis. A petite, sunny-tempered woman, Tess was working as an R.N. in an ophthalmology and urology surgical unit, when in 1989 she suddenly had difficulty breathing and fainted. In 1990 she had gallbladder surgery and a cardiologist was consulted because of her abnormal echocardiogram. That cardiologist told her she needed a new tricuspid valve. She said, "No thanks." After 20 months of shortness of breath and fainting, and hints from other doctors that her problem was all in her head, Tess finally had a cardiac catheterization and pulmonary arteriogram and other tests that led to the correct diagnosis of PAH. She put off having the only remedy then available—a heart/lung transplant. A calcium channel blocker (Cardizem), anticoagulants, and a diuretic, kept her alive for years. She now uses sildenafil in addition, and takes medicines for the diabetes she developed along with several other medical problems.

As of this writing, Tess has been to every one of PHA's conferences, and she and her husband, Deven Anthony, have traveled all over the world. Since her diagnosis Tess has visited Puerto Rico, the Virgin Islands, St. Marteen, St. Martin, Canada, Mexico, mainland China, Hong Kong, Taiwan, Thailand, Japan, Singapore, Malaysia, Korea, the Phillipines, Nassau, Freeport, Spain, Portugal, Morocco, Gibraltar, Germany, Switzerland, Austria, Hungary, Poland, the Czech Republic, the Slovak Republic, France, Greece, Turkey, and Costa Rica. She has gone to several of these places twice. So what if Deven had to carry her piggyback up the Great Wall of China?



Tess de Guzman (far left) with travel companions at Mt. Giewont, Poland

1993

CONFERENCE PLANNING & A CLINICAL TRIAL OF THE FIRST REAL PH DRUG

Board Activities in 1993

Relying on old *Pathlights*, here is a list of officers and at-large directors in 1993:

Officers:

President.....	Judy Simpson
Vice President.....	Ed Simpson
Recording Secretary	Dorothy Olson
Treasurer.....	Robert Knazik
Assistant Treasurer.....	Teresa Knazik/Jerry Paton

Directors-at-Large:

Eleanor Bails	Marilyn Dirr
Carla Gordon	Edys Gordon
Harry Olson	Jerry Paton
Pat Paton	LuAnne Washburn
Shawn Wiggins	

The January meeting was held at the Knazik home in Palm Bay, Florida.

At the meeting it was reported that:

- An application had been made to join NORD.
- UPAPH would be listed in the next issue of Self-Help Clearing House (a source of information on support groups for people with certain health problems and other issues).
- The PH Patient Directory prepared by Eleanor Bails had been mailed to PH patients.
- *Pathlight* had a current mailing list of 188.

Plans for the first UPAPH conference, to be held in the greater Atlanta area in June 1994, were discussed.

After the Board meeting, Judy Simpson traveled to a Burroughs Wellcome meeting in Research Triangle Park, North Carolina, to learn about developments in the clinical trials of epoprostenol.

In April, the Board met in the morning at Lyn Simpson's home (Lyn is Judy and Ed's daughter) in Aurora, Illinois, a suburb of Chicago. LuAnne Washurn (more about her below) was elected to the Board of Directors. In addition to the usual sort of Board business and reports, agenda items included a plan to create "new member packets" for PPH patients, which would include a cover letter, the NIH booklet on PPH, a *Pathlight*, and a form for ordering back issues of *Pathlight*. Patients with PH secondary to another disease would get all but the PPH booklet. An advertising policy for *Pathlight* was discussed, but tabled. It was decided to search for a volunteer to manage a library.

There was discussion of the feasibility of forming UPAPH chapters. An amendment to the bylaws that would regulate chapters was proposed, and run in the summer, 1993, issue of *Pathlight*. It was never adopted. (The precise relationship of support groups to the national UPAPH/PHA organization continues to be a topic of discussion at PHA Board meetings.)

Afterwards, the Board attended a presentation on PPH by Dr. Stuart Rich, in Chicago.

UPAPH now had its first logo. LuAnn Washburn designed the concept and Kathy Knazik donated her graphic arts experience:



The interlocking figures represented the unity of patients with PH and the candles symbolized a "pathlight" of knowledge passed on to one another.

Finances

At the start of 1993, the treasury balance was \$1,164.12, most of which was from donations. By December 1, 1993, UPAPH's nest egg had

grown to \$21,083.67. The increase was due mainly to general donations. Board members still paid their own expenses to attend Board meetings, as well as the costs of telephone calls, postage, copying, etc. The UPAPH treasury covered the newsletter, mailings, pre-conference expenses, and office supplies.

The Epoprostenol (Flolan) Clinical Trial, Phase III

Many of UPAPH's members with PPH were involved in the clinical trial of epoprostenol (Flolan), a drug made by Burroughs Welcome. Sixteen medical centers in the United States, and one in Canada, participated. The bulky official title of the study was "A Multicenter, Open Label, Randomized, Parallel Comparison of Safety and Efficacy of Chronic FLOLAN Infusions Plus Conventional Therapy Alone in Patients with Severe Primary Pulmonary Hypertension: A Twelve Week Study." It was "open" because doctors and patients knew who was getting the Flolan. It was decided not use a placebo, because the drug was administered through a catheter that had to be inserted into the heart, and this invasive procedure could not be justified in the control group. Therefore, the control group merely continued on their conventional therapy. (A similar trial of infused treprostinil [Remodulin] now underway in India is a true "blind" trial with catheters inserted into PAH patients getting a placebo.) Patients who had gotten PPH as a result of having another disease were excluded from this trial, which was difficult for some desperate patients to understand. Before starting the trial, patients were evaluated and given a New York Heart Association Classification:

Class I: Patient has no symptoms during ordinary physical activity.

Class II: Patient is comfortable at rest but physical activity is somewhat limited by undue breathlessness, chest pain, fatigue, or near fainting.

Class III: Patient has no symptoms while resting, but physical activity is greatly limited by breathlessness, chest pain, fatigue, or near fainting while doing routine activities.

Class IV: Patient is breathless and tired even while resting and can do no physical activities without symptoms. Signs of right-heart failure.

Patients with symptoms that placed them in Class III or IV received a heart catheterization and a challenge test for responsiveness to epoprostenol. The names of the 81 patients who were able to tolerate the drug went into a computer at Burroughs Wellcome and were randomly designated to either (1) have a Hickman central line placed and be started on epoprostenol, or (2) continue with their regime of current medications (control group).

Judy Simpson was with her sister Pat Paton at the Mayo Clinic when the doctors slid a catheter into Pat and did an epoprostenol challenge. While they were still in the cath lab, the docs dialed up Burroughs and the sisters learned that the computer had randomized Pat into the control group. There was no way to appeal the computer's decision. To know if the medicine really worked, it was essential to have a randomized trial. This necessity was hard on doctors as well as patients. *Twenty percent of the patients in the control group died during the 12 weeks of the trial.* Dr. Robyn J. Barst had four patients die at her center who were in the trial, but not getting the active drug. "It was one of the worst things I've had to deal with as a physician," she says.

Those getting the epoprostenol received it, the clutter of related supplies, and supervision in using everything, from the medical centers involved in the study.

Several measurements were used to judge the usefulness of epoprostenol. The imperfect 6-minute walk test is the one most patients

remember best. All the patients who received Flolan survived, and most had some improvement. Flolan, originally thought to be a bridge to transplantation, eventually proved to be much more--it enabled many patients to avoid a lung transplant altogether.

The results of this early trial were published in the February 1, 1996, issue of *The New England Journal of Medicine*. Burroughs Wellcome was very responsive to the PH community during this time. Since Flolan has to be given by a central line as a continuous infusion, it is an exceedingly difficult drug to administer and requires intensive patient teaching. It is possibly the most complicated medicine that patients had ever had to learn to self-administer. If not handled with sterile procedures, there is a great risk of infection.

Nurses played a vital role in educating and supporting the patients; they were referred to as the patients' lifelines. UPAPH's members had many questions. Walker Long, MD, of Burroughs Wellcome, and nurse Jessica Mendoza the PH nurse coordinator working with Dr. Lewis J. Rubin at the University of Maryland, flew to a support meeting in Chicago to personally answer questions. Dr. Long entered the room, walked to the blackboard, and wrote down his home phone number. He said he would be available around the clock.

UPAPH's members networked, and soon support groups played an important role in affording PPH patients using Flolan the opportunity to talk to other PH patients and to nurses about the new drug.

Planning the First Conference

Ed Simpson and Peggy Wessinger, co-chairs of the first International UPAPH Conference, searched for a suitable hotel to host it. (Peggy was Teresa Knazik's sister. She had recently retired from Bell South, where, serendipitously, she had worked as a conference planner.) Facility requirements were:

- Proximity to a major airport
- An elevation at or near sea level
- Hotel rooms close to meeting rooms
- Refrigerators in rooms for Flolan
- Ability to accommodate oxygen and oxygen concentrators
- Moderate cost
- A staff able to meet the unique needs of our patients

Amazingly, they found all this and more at a beautiful facility called the Evergreen Conference Center. It was in Stone Mountain Park, just outside Atlanta, Georgia.

Letters were sent to 208 cardiologists and pulmonologists inviting them to attend a breakfast on June 8, 1993, sponsored by Bell South (Peggy Wessinger's former employer), to learn about the conference. In spite of the offer of a free breakfast, only two persons showed up: Dr. Clint Lawrence, of Emory University, and one home-health-care worker. This was discouraging. Dr. Lawrence agreed to be the admitting physician if any patient needed hospitalization during the upcoming conference.

Ed and Judy Simpson drove to Stone Mountain Park, and they and Peggy Wessinger talked with the hotel staff about the unique conference UPAPH was planning. The site was stunningly beautiful and tranquil. Not far away was a humongous granite dome that gave Stone Mountain Park its name. The mountain has a carving on its side of three Confederate heroes of the Civil War. Conference attendees who wished to would be able to attend a laser light show at the mountain after dark. (It was a dazzling show, but did make a few Yankees sort of nervous.)

Planning began in earnest. Ed's secretary at Northern Illinois University, Mary Risseuw, took on the responsibility of writing the several hundred letters required for the conference. Ed called on Dr. Walker Long (Wellcome Research Laboratories) for guidance; Dr. Long's advice was to contact Dr. Alfred Fishman (University of Pennsylvania) to give the conference keynote address. Dr. Long believed that if the distinguished Dr.

Fishman supported the conference, other physicians would follow his lead “like dominos.” This was sage advice. Every single doctor who was invited to the conference accepted the invitation! (Dr. Fishman later arrived at the conference expecting the usual sort of scientific gathering. He had brought his standard technical slides. A good-natured man, when he saw all the patients and their families he redid his slides and talk, and although his presentation was still pretty deep for some patients he was the glue that pulled the conference together.)

“In Memory Of” Cards

UPAPH ordered 500 printed “In Memory Of” cards. These were needed because of the growing UPAPH membership and the high mortality rate among our members. Handwriting an entire card had become unmanageable. The cards thanked people for their donations in honor of, or in memory of, UPAPH members.

ACCP Consensus Statement

In July 1993 an American College of Chest Physicians (ACCP) Consensus Statement on Primary Pulmonary Hypertension was published in the organization’s journal, *CHEST*. * Dr. Lewis Rubin (then at the University of Maryland) was chairman of the group developing the statement. This was a major step forward in physician education about IPAH. *CHEST* made an unusual concession by giving UPAPH permission to make copies of the article, instead of buying them from *CHEST*. (Dr. Rubin also chaired the subsequent ACCP Panel, which convened in 2003 and published its guidelines in July 2004 as a supplement to *Chest*.)

*Rubin LJ, Barst RJ, Kaiser LR et al. “Primary Pulmonary Hypertension—Report of the ACCP Consensus Committee.” *Chest* 1003; 104:236-250.

SAB Members in 1993 and 1994

(The names of new members are in boldface type.)

Robyn J. Barst, MD (Columbia/Presbyterian Babies Hospital)
Linda Clayton, Pharm D (Burroughs Wellcome Co.)
Alfred P. Fishman, MD (University of Pennsylvania Medical Center)
Valentine Fuster, MD, PhD (Harvard Medical School)
Frank Gold, MD (Peoria, IL)
Lisa Kaufmann, RN (University of Illinois, Chicago)
Jillian Kirkpatrick, RN (Columbia/Presbyterian Babies Hospital)
Walker A. Long, MD (Wellcome Research Laboratories)
Michael D. McGoon, MD (Mayo Clinic, Rochester, MN)
Douglas S. Moodie, MD, MS (Cleveland Clinic Children's Hospital)
Kenneth M. Moser, MD (University of California, San Diego)
Harold Palevsky, MD (University of Pennsylvania Medical Center)
Stuart Rich, MD (University of Illinois, Chicago)
Lewis Rubin, MD (University of Maryland)
Warren Summer, MD (Louisiana State University)
Carol E. Vreim, PhD (National Institutes of Health)
Angela Wurser, RN, MSN (University of Pennsylvania Medical Center)
Dianne L. Zwicke, MD (University of Wisconsin)

***Pathlight* in 1993**

In a new feature, "Sounding Board," LuAnne Washburn fielded medical questions and got responses from PH doctors. A resident of Wisconsin, Luanne had PPH yet worked part time as a pediatric occupational therapist and participated in a cardiac rehab program. When her insurance company refused to pay for the vasodilators that were temporarily extending her life, or for the transplant she might need, her plight was reported in Newsweek and elsewhere. Her brother wrote movingly about her in the summer issue of *Pathlight*. Not surprisingly, LuAnne volunteered for the early epoprostenol research efforts, and was a great inspiration to other patients. The "Sounding Board" continued until 1997 when it transformed into the "Research Corner."

Pathlight continued to run detailed articles on epoprostenol (Flolan). Reports on research development and patient experiences with the drug kept readers up-to-date.

The spring issue was largely devoted to the hows and whys of support groups. Chicago, Florida, and Northwest (New Jersey-New York) support groups were meeting, and Families of Children with PH was formed by Pam Hummel, from Minnesota, whose son had PPH. She made contact with 18 PH families and sent information on PH to major institutions.

In summer, *Pathlight* focused on PH families. Sue Kelly began a series on how to keep a writer's journal, and the benefits of journal-keeping for PH patients.

Fall and winter issues featured a "Kids Korner," edited by Pam Hummel and written by her son Ben, 12, with items for younger PH patients. Ben wrote about how the Make-a-Wish Foundation had sent him and his family to Walt Disney World. (Ben got a double-lung transplant in 1995.) UPAPH had begun getting general donations; gift acknowledgments now filled three-quarters of a page.

A discussion of NORD, its mission, services, etc. was featured in the winter issue, which also listed the names of the 14 PH physicians and 8 medical professionals planning to attend the first conference. There was also a list of the 18 centers then conducting epoprostenol (Flolan) clinical trials.

On November 30, 1993, the name *Pathlight* was placed on the Trademark Principal Register by the U.S. Patent and Trademark Office. This meant that no one other than UPAPH could use this name. (Several years later a business offered to buy the name *Pathlight*. The Board declined.)

1994

A BIG LEAP FORWARD WITH THE FIRST INTERNATIONAL CONFERENCE

200 PH Patients and Doctors Come Face-to-Face

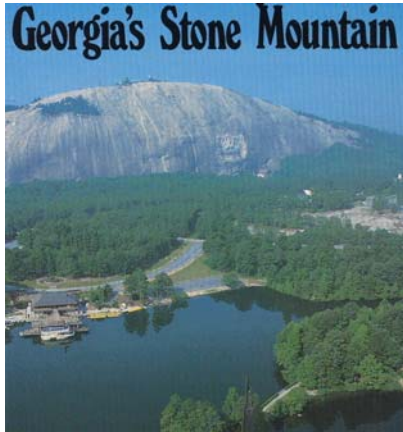
It is impossible to overstate the importance to the organization of the first International Conference. It was at the conference that UPAPH firmly made its mark on the consciousness of patients and physicians, and set the cooperative, optimistic tone that has since characterized its operations.



“Hearts and Hands” was the theme, and the conference was held June 17-19, 1994, in Stone Mountain, Georgia. Prior to the conference, Walker Long, M.D. (Wellcome Research Laboratories) advised Ed Simpson on the medical components. The Board had decided it was very important to promote doctor/patient interactions at the conference. Letters were sent to presenting physicians requesting permission to have their sessions videotaped. All agreed to this, but did not want written transcripts—doctors often speak more frankly, and speculate more willingly, at the conferences than they would in a medical journal. This openness is valuable to both patients and physicians.

About 200 persons attended the conference, which lived up to its ambitious name--it really was “international.” Sixty-eight patients came, including one from Australia. Most had never before another PH patient, and some cried with emotion and embraced near strangers. As patient Patricia Murphy wrote for *Pathlight*: “The ‘ice breakers’ when you met someone were...questions like, ‘What medicines are you on?’ and ‘Who is your doctor?’

When I told others that my doctor was Vic Tapson, most would say, 'That hunk!!'" Thirty-two health-care professionals, in addition to handsome Dr. Tapson, attended. Some came from England and Canada.



Judy Simpson met ahead of time with hotel staff supervisors to explain about PH and the special concerns of PH patients. She told them, for example, that many attendees would be in wheelchairs and/or using oxygen, and that many used an unusual medication called Flolan that had to be kept cold and that discontinuation of this medicine could result in a user's quick demise.

Such pre-conference briefing has proved to be important, and has been done as well at all of the following conferences. Some medical problems cannot be planned away; two patients using Flolan were hospitalized with central line infections. (A small number of medical emergencies have occurred with regularity at PHA conferences.)

These were UPAPH's goals at that first conference:

- For patients to gain first-hand information from experts
- For patients to interact with physicians and health care professionals to help them understand problems that PH patients encounter
- To assist researchers who are gathering data
- To network--to connect doctors and patients with others in similar circumstances or with similar interests

Research Done at the Conference

Two weeks prior to the conference, Ed Simpson received a call from Dr. Greg Elliott (University of Utah) asking permission to draw blood and obtain family histories from patients at the conference. Ed contacted the Scientific Advisory Board and checked out Dr. Elliott's credentials (he passed with flying colors) and permission was granted--with the stipulation that the blood samples and information gathered be shared with other physicians

doing PH research. Soon Dr. John Newman and Dr. Jim Loyd from Vanderbilt University called and became a part of the research group. Next, Dr. Jane Morse (Columbia Presbyterian Medical Center in New York City) called. All these physicians were looking for genetic ties among PPH patients and families in their effort to find a causative gene/genes. The unusual part of this story is that these physician/researchers heard about our small association and sought us out. The UPAPH conference would be the largest gathering of PH patients ever held; it offered a rich opportunity to further ongoing research.

Many patients and family members attending the conference agreed to participate in the blood draws and give information on their family histories. A research fellow working with Dr. Elliott, Dr. Gary Alexander, was particularly skilled at dealing with patients gently and with empathy, and helped to make the effort a success. As a result, five new families with inherited PH were identified and researchers moved a step closer to locating the "PH gene" (more on this below). Patients have had an opportunity to participate in PH research at all the succeeding conferences. Their generosity with their blood and time was instrumental in enabling the identification of a PPH gene in 2000. As patient Tammy Chapman wrote after the conference, "I thank the research people for taking my blood. It's not everyday that you want to have someone prick you with a needle, but believe me, it was a satisfying experience to know that there are people sitting behind microscopes working on PPH."

Volunteers and Generous Donations Helped Slash the Cost of the Conference

Burroughs Wellcome underwrote much of the cost for patients/family members to come. For a PH patient, registration, plus two nights' lodging, plus six meals were only \$90! For a PH patient and one significant other, the cost for all this was \$250. (The regular registration fee was \$300 per

person.) Ed Simpson was able to get oxygen services/supplies for all patients needing it at no cost to the patient. At the close of conference, the Evergreen Conference Center bill was \$13,000 more than UPAPH had in its treasury. Ed Simpson and Jerry Paton had their credit cards ready; however, the Conference Center wrote off the debt.

Volunteers did most of the work. Ed brought video equipment from Northern Illinois University and arranged for volunteers to videotape all conference sessions (no support group sessions were videotaped). To do all this, Ed signed up family members: daughter and son-in-law Lyn and Tom Becker, his son Greg, and his sister and brother-in-law Sherry and Steve Skelton. The Patons' daughter and son-in-law, Julie and Dave Hendry, also helped. Charles Lybrook, a cousin of Pat Paton and Judy Simpson, printed and donated the conference programs. Forty-five patients and/or family members served as moderators, support group facilitators, and room hosts. Peggy Wessinger gathered many door prizes and goodies for bags given to attendees. Seven patients were on panels. The medical presenters were, of course, volunteers as well. After the conference a young man in Illinois—at no charge to UPAPH--put leaders on the videotapes and did minor editing.

Conference Highlights

As at subsequent conferences, awards were presented. Awards given in 1994, 1996, and 1998 were determined by the UPAPH Board on the basis of service to the mission of the Association and, more importantly, of improving the lives of PH patients and families. (Beginning in 2000, a Board committee was established to select winners from a pool of nominees, using written criteria. Any PHA member may nominate someone for an award.)

Outstanding physician.....	Walker Long, M.D. (Wellcome Research Laboratories; Dept. of Pediatrics, University of North Carolina, Chapel Hill)
Outstanding patient.....	Teresa Knazik
Corporate award.....	Burroughs Wellcome

A huge card was prepared for Burroughs Wellcome on which patients and family members wrote messages of gratitude such as: "Thanks to BW, I've seen my daughter walk, talk and now she starts 1st grade! Thank you for the second chance for both of us;" and "Thank you very much for all you have done...actually thank you for my life." Burroughs Wellcome made replicas of the card for their employees working on Flolan, most of who had never seen a PH patient.

The "Journeys" presentation was outstanding. Six patients and their physicians each described their journeys with PH. The six patients sat on risers and the physicians sat on the floor. This arrangement unexpectedly seemed to set the tone of the conference for patient/doctor interactions. Gabriel Miyara, a patient and founder of the PPH Research Foundation, spoke briefly about her Foundation. (At this time UPAPH was unable to support research as an Association.) Patients provided the entertainment in the form of skits, music, and acrobatic dance. Connie Schweitzer, a pretty teen with PPH, did a dance that included flips. Her Flolan pump was so skillfully hidden under her leotard that some in her audience doubted she really wore one. (Connie got a terrible line infection at the conference, but it cleared up after a brief hospitalization, and early in July she left for a trip to Germany, flying by herself, with her prostacyclin and I.V. antibiotics in her carry-on luggage.) Connie was a great inspiration to other patients. More inspiration came from Dr. Jerry Patterson, a motivational speaker, who spoke on "Cabbages and Kings: Hope Springs Eternal." Dr. Patterson refused to take his usual fee for speaking. Larry Wessinger took photos during the conference and presented a beautiful videotape set to music, "Moments in Time," at the conclusion of the meeting.

A New Paradigm Is Created

Ed Simpson summed up the first conference as the "start of putting UPAPH on the map as an effective self-help association that can exert social and political action." Tim Higenbottam, M.D. (then at Papworth Hospital, Cambridge, England) said, "This has been a meeting of enormous cooperation, which I think has been a hallmark, not simply between patients and doctors...but it has also involved the pharmaceutical industry." Stuart Rich, M.D. (then at the University of Illinois, Chicago) said, "I have never been to a meeting that was organized by patients before, and it truly appears to have been a rather unique experience. All of us felt enriched and came away with a great sense of optimism. The meeting was expertly coordinated and run, and was as useful and positive an academic experience that I have had in my career." And a patient said, "This conference was the first time that a doctor told me there was hope that I might live more than a year or two. The doctors who have been treating me are totally out of date."

Board Activities in 1994

The officers in 1994 were:

President.....	Judy Simpson
Vice President.....	Ed Simpson
Recording Secretary.....	Dorothy Olson
Treasurer.....	Robert Knazik
Assistant Treasurer.....	Jerry Paton
<i>Pathlight</i> Editor.....	TerestaKnazik, then Mark Taylor Murphy
Corresponding Secretary.....	Teresa Knazik

Directors-at-Large were:

Eleanor Bails	Bonnie Dukart
Gary Dukart	Harry Olson
Pat Paton	LuAnne Washburn
Peggie Wessinger	

Marilyn Dirr, Shawn Wiggins, Edys Gordon, and Carla Gordon had resigned from the Board. (Edys' passing was noted in the spring 1995 *Pathlight*.)

Board members were still paying their own expenses to attend meetings, but started receiving some reimbursements for copying and postage. Before the conference, in January, the UPAPH Board met in Indiantown, Florida, at the home of Pat and Jerry Paton. When the Board met at the Patons', friends from Indiantown provided luncheon for the group, which included any UPAPH members who wished to attend.

The Board met again at Stone Mountain, Georgia, immediately after the conference. It was decided that videotapes of conference sessions would be edited, copied, and made ready for sale at \$39.50 per 27-hour set. They would also be available at no charge, except for postage, through UPAPH's new lending library. (Deborah Cane would be the new librarian. She was the mother of twins, both of whom had PH. One had died of the disease and the other was under treatment.) Appreciation was expressed to Mary Risseuw for the hundreds of letters she prepared for conference. Due to increased membership, Pat Paton would no longer be able to talk to/write to each new patient member; the Board decided to divide the U.S. into eight areas with an Area Coordinator in each area to respond to initial inquiries. Pat offered to coordinate these areas. Bonnie Dukart assumed responsibility for organizing these same areas for political activity. Area Coordinators would maintain patient lists and coordinate patients for networking purposes.

It was further decided that UPAPH would look for office space in the Palm Bay area, and (because of the great success of the first conference) that the UPAPH International Conference would be held biannually.

On September 24, a Board conference call meeting was supported by a grant from Support Works. Mark Murphy was elected to serve on Board. It was decided that the membership list would be maintained by Eleanor Bails on the computer; that Bob Knazik would continue as treasurer and would gather mail; and that Mary Knazik would send renewal notices to

members not responding to the renewal notice in *Pathlight*. "Journey" articles from patients would be compiled and sent out with *Pathlight*. A donation was received to purchase secretarial equipment. Gloria Blodgett agreed to be volunteer hours coordinator. The U.S. now had ten district areas. In 1994, 420 thank-you and acknowledgement cards were sent.

Finances and membership. UPAPH's total revenue for 1994 was \$60,819.15. It's total expenses were a few thousand dollars more than this. Membership at the end of 1994 was 321 patients and families, 77 professionals, and 13 organizations.

A Snapshot of Committee Chairs in 1994

Advisory Board Liaison.....	LuAnne Washburn
Creative Writing.....	Sue Kelly
Directory.....	Eleanor Bails
Donations.....	Judy Simpson
Librarian.....	Deborah Cane
Mail.....	Mary Knazik
Networking.....	Pat Paton
Organ Donor Awareness.....	Marilyn Dirr
<i>Pathlight</i> Editors.....	Teresa Knazik, then Mark & Pat Murphy*
Publications & Membership.....	Teresa Knazik
Public Affairs & Volunteer Coordinator.....	Bonnie Dukart
Volunteer coordinator.....	Gloria Blodgett

As committees and their chairs were in constant flux, this list may not be comprehensive.

*Patricia Murphy died that year. It's sad to mention deaths, but such passings are noted here because it is the nature of this dreadful disease that makes PHA necessary, and that motivates us to find a cure. To balance its "Passages" column, early *Pathlights* also listed members who were celebrating birthdays that quarter.

Support Group Activity

Support groups now existed in Chicago, Florida (two), New Jersey/New York, California, Minnesota (two), North Georgia, Texas, Virginia, and the Chesapeake area.

Learning from NORD

In April 25, 1994, a NORD meeting in Washington, D.C. focused on health care reform and on how to manage a not-for-profit organization. Representatives from UPAPH attended at their own expense.

A NORD survey found that persons with a rare disease have:

- An average 5-year delay from onset of symptoms to diagnosis
- Two-thirds of the dollars spent on diagnostic tests are wasted
- 30 percent of patients must travel at least 10 miles for treatment
- Services must be obtained at least 10 times per year
- 48.5 percent of diagnosed patients are satisfied with their care
- 40 percent thought that switching physicians would be too much of a burden

The survey also identified the following patient problems:

- Getting and keeping insurance
- Exclusions in insurance policies
- Costs
- Concerns that specialists/tertiary care physicians will be hard to access
- Funding for research will dwindle
- Comprehensive, teaching medical centers will not receive funding

Major health care reform concerns identified by NORD were: 1) universal, mandatory, non-discriminating insurance coverage; 2) access/referral to experts of rare diseases; 3) never losing insurance coverage; and 4) access to orphan drugs, which were expected to be expensive. Concerns expressed at the NORD meeting typified the PH patient.

Attendees were updated on how the Orphan Drug Act Amendment of 1994 was working its way through Congress. Abby Meyer, who is with NORD, says that the most important part of the Act was to get drug companies interested in looking for drugs to treat rare diseases when they could not expect to make a profit from such drugs. After the NORD meeting,

attendees went to Capitol Hill to lobby for passage of this amendment. They were successful, and the incentives of tax benefits and a longer patent time in fact did produce more activity in "orphan drugs."

Pathlight

Credibility and integrity has always been foremost in any publication of UPAPH/PHA. The following is an example. In one edition of *Pathlight*, Mark forgot to include the disclaimer. He had shipped the *Pathlights* in addressed envelopes to Florida, UPAPH's designated not-for-profit mailing site, where Dorothy and Harry Olson discovered the omission. Mark reprinted one page of *Pathlight* and shipped it and new labels to the Olsons, who inserted the new page, put labels on new envelopes, and sorted the mailing according to postal regulations. As Dorothy said, "it was important to get it right."

The winter issue of *Pathlight* ran a story on the National Registry of Primary Pulmonary Hypertension in Families (see below).

Teresa Knazik, who played such an important role in founding UPAPH and *Pathlight*, and who had won the Outstanding Patient award at the conference just two months earlier, died August 27, 1994. This is what Bob Knazik wrote about his late wife for this PHA history—it is a moving story that will resonate with both PH patients and doctors:

Teresa Knazik, PHA and *Pathlight* Pioneer



Teresa Knazik

She was a little dizzy. Teresa had been running in little circles in the backyard with our 4-year-old son, Carl, and she felt breathless. She told me she really needed to get into better shape. We were in Clovis, New Mexico, where I was stationed at Cannon AFB in 1985. Teresa and I had met in the Air Force at Maxwell AFB in Montgomery, Alabama in 1980. We were married in May of 1980, and Carl was born in March of 1981. Teresa left the military in 1984, and I had retrained into a more interesting job.

During the summer of 1985, Teresa, Carl, and I went to visit her relatives in Florida. One day we went to the

beach and walked 3 or 4 miles in the sand. When we were leaving, she simply fell down and passed out. She regained consciousness a few seconds later, and woke with a loud scream. She said she dreamt that a monster was after her. Turns out, one was...

The doctors were baffled. Some thought she had a mental illness, since they could find nothing wrong with her. After being seen by a parade of puzzled doctors, a young internist listened to her heart and suspected that she had a serious problem. She was sent to the Air Force's main hospital, Wilford Hall in San Antonio, Texas, where she was examined by their best cardiologists and underwent many tests. They confirmed the young doctor's suspicions, and told her she had something called pulmonary hypertension. The doctors gave us a copy of the only known study of pulmonary hypertension done at the time, with 167 patients and a "dreadful" prognosis. We left the hospital and drove half way home, then stopped for the night at a motel. Teresa was tired and went to sleep. After a little while she came out from the bedroom, sat in my lap, and started crying. She was 30 years old and very much afraid of dying.

Teresa went to the William Beaumont Army Medical Center in San Antonio for further testing and evaluation. It was there that our relationship with doctors changed.

Few doctors back then had examined a patient with PPH. Teresa became more of an exhibit than a patient. One day they put her on a gurney in the hall outside a classroom. A class of interns then walked by, each of them listening to her heart and lungs. I understood that it was important they learn about this disease, but I also felt it was important that Teresa be treated like the human being she was. We stopped regarding doctors as somehow above mere lay people like ourselves and started regarding them as closer to people mechanics. They learn stuff, get experience, and then take a good guess, no more. They make mistakes, hopefully learn from them, and go on, just like the rest of us.

I was given a hardship reassignment to Homestead AFB in Miami in the winter of 1986, so Teresa and I would be closer to her family. As we packed for the moving truck, Teresa's mother passed away in Florida. Teresa had been very close to her mother. The shock of her mother's death pushed her into near mental collapse. We arrived at Homestead AFB, and things did not get much better; she actually contemplated ending her suffering.

Her new pulmonologist, Tahir Ahmed, worked at Mt. Sinai in Miami Beach, and he took a real interest in Teresa's disease. He put her on a drug that seemed to stop the progression of the PPH. But just as importantly, he gave her hope. Hope, that concept that had been drained from her over the past 18 months, was now hers again, and with it, a renewed sense of purpose. Dr. Ahmed didn't see any reason why Teresa couldn't live a decade or

more, and who knew what would be discovered in the next few years? The summer of 1987 was the start Teresa's journey back from despair. The journey would be hard, but there would be new friends, many, many smiles and laughs along the way, the promise of new medications and procedures, and there would be great accomplishments. There would be *Pathlight*.

Teresa died in the fall of 1994. The night before the operation that would try to keep her failing heart beating, I lay with her on her hospital bed and I cried while she comforted me. It would ever after always be clear to me which of us proved to be the stronger person. She had faced those early demons and conquered them, shutting them behind her as she walked into the light. PH may have eventually taken Teresa's physical life, but she lives on still in PHA.

Before she died, Teresa found someone to replace her as *Pathlight's* editor, Mark Taylor Murphy. (As noted above, Mark's wife, Pat, had died of PPH shortly before Teresa did.)

On the Medical Front

List of PH Physicians. In 1994 UPAPH published a list of physicians who said they had experience treating PH patients. Most physicians still either had never heard of the disease or were ignorant of treatment possibilities, and it was important for patients to find clued-in doctors.

Scientific Advisory Board Developments

The second SAB of 30 members was appointed by the UPAPH Board.

Appointments were for two-year terms. New members are in bold type.

Gary J. Alexander, MD (University of Utah School of Medicine)

David Badesch, MD (University of Colorado)

Robyn J. Barst, MD (Columbia/Presbyterian Babies Hospital)

Bruce H. Brundage, MD (UCLA School of Medicine)

Linda Clayton, Pharm D (Burroughs Wellcome Co.)

James W. Crow, PhD (Burroughs Wellcome Co.)

Alfred P. Fishman, MD (University of Pennsylvania Medical Center)

Valentine Fuster, MD, PhD (Harvard Medical School)

Frank Gold, MD (Peoria, IL)

Timothy J. Higenbottam, MD (Papworth Hospital, England)

Lisa Kaufmann, RN (University of Illinois, Chicago)

Jillian Kirkpatrick, RN (Columbia/Presbyterian Babies Hospital)

David Langleben, MD (Dept. of Medicine, Montreal, Canada)

Walker A. Long, MD (University of North Carolina)
James E. Loyd, MD (Vanderbilt University)
Michael D. McGoon, MD (Mayo Clinic, Rochester, MN)
Kenneth M. Moser, MD (University of California, San Diego)
John H. Newman, MD (St. Thomas Hospital)
Harold Palevsky, MD (University of Pennsylvania Medical Center)
Stuart Rich, MD (University of Illinois, Chicago)
Lewis Rubin, MD (University of Maryland)
Robert Safford, MD, PhD (Mayo Clinic)
Cathy J. Severson, RN (Mayo Clinic)
Warren R. Summer, MD (Louisiana State University Medical Center)
Victor E. Tapson, MD (Duke University Medical Center)
Carol E. Vreim, PhD (National Institutes of Health)
Dianne L. Zwicke, MD (University of Wisconsin)

“National Registry of PPH in Families” Begins

In about six percent of PPH cases, the disease affects more than one member of a family. The symptoms, prognosis, and response to therapy are similar to those of nonfamilial (“sporadic”) idiopathic pulmonary arterial hypertension. Although PPH can be inherited from a male or female parent, the inherited form strikes roughly twice as many females. It is a dominant trait, but many persons who get the gene do not develop PPH.

Dr. James Loyd and Dr. John Newman at Vanderbilt University (Nashville, TN) had worked with PPH families since 1980, and had located 13 families that carried the disease. With financial support from the NIH they began The National Registry of Primary Pulmonary Hypertension in Families. Their goal was to learn more about the “PPH gene” by enrolling as many families as they could find who had members willing to share their medical histories and give blood samples. They also hoped to learn more about how the disease is passed along in the families. Links among families would be searched for using a computer database. Lisa Wheeler was (and is) the Registry Coordinator. She is fiercely protective of families’ privacy.

In an effort coordinated with the Vanderbilt Registry, two other physicians, Greg Elliott and Gary Alexander (University of Utah School of Medicine) assembled a registry of U.S. patients with *sporadic* PPH.

As mentioned, many UPAPH patients who attended the Stone Mountain conference contributed blood and histories to this effort. By discovering previously unknown familial ties, the doctors learned that some apparently sporadic PH was actually familial.

In the years beyond the scope of this history, this research would lead to the discovery of the PPH gene(s) on the bone morphogenetic protein receptor 2, a gene associated with cell growth and development. Knowing the gene, and learning how it works, will help scientists to develop treatments and perhaps a cure for PH. By 2005, over 80 families were participating in the familial PPH study. Families will be actively enrolled at least through 2008. To join the study or to get information on genetic testing, contact James Loyd, MD, or Lisa Wheeler at 1-800-288-0378; lisa.wheeler@vanderbilt.edu. Dr. Jane Morse and her colleagues at Columbia University in New York City also continue to study the genetics of PAH and provide genetic counseling and testing. For further information contact Jane Morse, MD, at 1-212-305-5916 or Robyn J. Barst, MD, at 1-212-305-4436. Dr. Greg Elliott and his colleagues in Utah are also still enrolling patients in their studies of sporadic PH and can also provide information on genetic testing. For information on the current status of their research contact Jon Schmidt at the LDS Hospital in Salt Lake City, at 1-801-408-1875.

1995

HELPING TO GET FLOLAN APPROVED

In 1995 UPAPH worked with Burroughs Wellcome to get FDA approval for its drug epoprostenol (Flolan), established a telephone support line, began publication of *Persistent Voices*, and acquired its first full-time volunteer. In the spring of 1995, UPAPH had 411 members. The number grew to 545 by winter.

Participating in the February 23 FDA Hearing on Flolan

PH patients continued to die at an alarming rate, and UPAPH's immediate top priority was to get the FDA to approve Flolan as a treatment. Knowing the powerful impact of patient stories, Judy Simpson wanted to have PPH patients at the FDA hearing to testify about their experiences with the drug. She phoned Dr. Stuart Rich and Dr. Lewis Rubin for advice and both initially responded negatively to her idea. As researchers, they believed Flolan had to stand on its own merits.

Several of Dr. Rubin's patients agreed to testify. One was LuAnne Washburn, a young woman who had participated in the first clinical trial at the University of Maryland (they didn't use pumps then--the medication was delivered by a 50 cc syringe with a spring). A patient of Dr. Stuart Rich's also testified-- Connie Schweitzer, the vivacious teenager on Flolan who had performed at the 1994 conference. UPAPH prepared a required pre-hearing registration for LuAnne and Connie, including an outline of what they would say, and flew the young women to Bethesda, MD, where the hearing was held.

The hearing opened on Thursday, February 23. Burroughs Wellcome and physician investigators presented volumes of Flolan clinical trial data. Dr. Rubin then asked to have LuAnne and Connie speak. They overwhelmed the crowd with their journeys. LuAnne told how she was able to go back to

work full time, and Connie told how she was able to fully participate in high school and even do acrobatic dancing. Both women answered questions about how they dealt with the Flolan delivery system.

The Flolan data showed remarkable improvement in PPH patients using the drug. On Friday morning, the hearing concluded with a recommendation that the FDA approve Flolan. UPAPH later learned that the complex delivery system had greatly concerned the panel. LuAnne and Connie played an important part in convincing them that the clumsy delivery method was no “big deal” when it brought a life-saving drug to them.

The FDA staff raised another concern that might have derailed Flolan: they wondered if Burroughs Wellcome had designed their research in a fraudulent manner so that the sickest patients did not get the drug, thus making those who did get the drug look even healthier. Basically, they just couldn’t believe that Flolan could really do so much to help very sick PH patients. There was no proof of any intent to deceive, however, and Dr. Greg Elliott, who was testifying as an expert, argued successfully to the cardio-renal FDA advisory board that his, and other doctors’ experience showed that the drug really did help their patients and should be approved.

While still in the D.C. area, LuAnne, Connie, Bonnie and Dr. Gary Dukart, and Ed and Judy Simpson met with Stephen Groft, Pharmacy Doctor, and director of the newly instituted Office of Rare Diseases in his office at the NIH. It was Dr. Groft’s first meeting with any patients “on the pump.” They spoke with him about PPH and Flolan. Dr. Groft became, and remains, a supporter of UPAPH/PHA.

In September 1995, the FDA granted Flolan its final approval.

Transitioning from Trials to In-Home Use of Flolan

The next step was to get Flolan to the patients who needed it. UPAPH sent a questionnaire to its members to learn more about their needs and

situations. One hundred and thirty-five patients responded. (There were overlapping responses.) This is what we learned:

What is Your Source of Healthcare Funding?

<u>Source</u>	<u>Number of Patients</u>
Insurance	86
Disability	46
No Insurance or Disability	3
Personal Resources	34
Insurance plus own resources	0

Can you work?

No	78
Yes	38
Part-time	5
No response	2
Children	12

During the transition from the clinical trials of Flolan to its FDA approval and commercial availability in September 1995, Burroughs Wellcome offered uninterrupted access to therapy (including both Flolan and its related supplies) for patients who had participated in the trial, including those in the control group, regardless of their ability to pay. Prior to FDA approval, some other PPH patients were put on Flolan under a compassionate use plan. Judy Simpson appointed a Flolan Patient Advisory Committee to work with Burroughs Wellcome in planning the transition. Linda Carr, Bonnie Dukart, Pat Paton, Susie Richardson, Connie Schweitzer, Judy Simpson, and Jerry Wojciechowski made up the committee. This group met on April 8, in Research Triangle Park, NC. Burroughs Wellcome gave an honorarium of \$3,000 to this committee, which was donated to UPAPH.

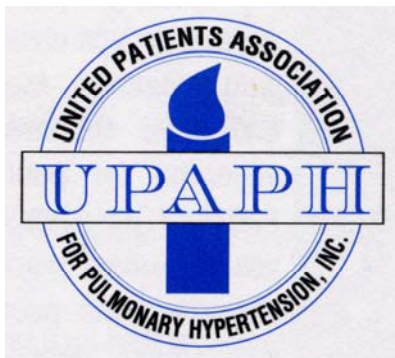
After FDA approval, Burroughs told Judy that they would give Flolan to any PPH (they specifically said PPH) patient who needed the drug, regardless of their ability to pay. To the best of her recollection, she thinks that Glaxo

Wellcome also honored this policy for PPH patients. (Glaxo was taking over Burroughs Wellcome—the new company would be called Glaxo Wellcome.)

On April 7, 1995, Judy spoke to three home health agencies interested in providing services to Flolan patients once the drug went on the market. Glaxo Wellcome decided that Quantum Health Resources would provide home health services and have exclusive distribution rights for Flolan. Quantum agreed to provide a 24-hour, toll-free telephone contact service that would be known as Flolan Lifeline Services. All PPH patients who needed Flolan would receive the drug (which could cost \$120,000 per year) regardless of their ability to pay.

New Logo

The Cooney-Waters group designed a new logo for UPAPH:



Patient-to-Patient Helpline Set Up

A big step forward came when, early in the year, UPAPH obtained an 800-telephone number, 1-800-74UPAPH (1-800-748-7274). For the first year, Suzie Richardson, stepmother of a teen with PH, answered the line on her home telephone. Soon, PH patients themselves would answer the line. The same number is still being used today.

Support Group Activity

In another new use of telephones to connect PH patients, a support group phone meeting was held via conference call. There were now 20 support groups for PH in the U.S., in 15 states. UPAPH sent each support group leader a set of tapes of the 1994 conference.

First Full-Time Volunteer

Julie Paton Hendry became the first full-time UPAPH volunteer. She worked out of her home in Speedway, Indiana. UPAPH bought a computer and a standing fax machine for her use. The UPAPH post office box was moved to Speedway. UPAPH was then the only PH patient group anywhere in the world, and more letters from abroad began to arrive. Julie and other UPAPH volunteers reported a total of 5,577 hours of service in 1995. (Julie's passing was noted a few years later, in the Summer 1998 issue of *Pathlight*.)

Networking with Other Organizations

Ed Simpson was elected to the NORD Board in the summer. In September, Bonnie Dukart and Ed and Judy Simpson attended the NORD meeting. And late in the year, Judy Simpson went to the Chicago O'Hare Airport to meet with Glaxo representatives. She was reassured that Flolan services would be continued in a seamless fashion.

Persistent Voices Joins Pathlight

The first issue of *Persistent Voices*, edited by patient Sue Kelly, was published, and mailed along with *Pathlight*. Two issues a year were planned. The aim of this more literary cousin to *Pathlight* was to allow patients/families to share their stories, essays, and poetry. It was, and continues to be, inspiring.

Mark Murphy set up an Internet address: *Pathlight1@aol.com*, which was announced in the newsletter. The suggested UPAPH donation was \$15 a year, but patients could receive the newsletter regardless of their ability to contribute.

The winter issue of *Pathlight* expressed concern about a bill proposed by Senator Pryor that would permit manufacturers of generic drugs to bring their products to market before the patents on pioneering companies' products expired. This would have undermined the patent rights in the General Agreement on Tariffs and Trade (GATT) that implemented strong intellectual property protection in 123 countries. UPAPH believed this would undermine the development of life-saving drugs like epoprostenol/Flolan, and also hoped that drugs better than Flolan would be developed. (Of course, many PH patients used expensive Coumadin (warfarin) and ProCardia XL (nifedipine) because generic versions were not then readily available to all, so the issue cut both ways.)

Board Activities in 1995

The officers in 1995 were:

President.....	Judy Simpson
Vice President.....	Bonnie Dukart
Recording Secretary.....	Dorothy Olson
Treasurer.....	Jerry Paton
Assistant Treasurer.....	Carol Wilson (appt. late in year)
<i>Pathlight</i> Editor.....	Mark Taylor Murphy
Corresponding Secretary.....	LuAnne Washburn

Directors-at-Large were:

Eleanor Bails	Gary Dukart
Harry Olson	Pat Paton
LuAnne Washburn	Peggie Wessinger
Dena Giddens	

The January 14 meeting was held at the Patons' home in Florida. Three people from Cooney Waters, Inc., were present, who were sent by Burroughs Wellcome to videotape interviews to use at the launch of Flolan.

While this shuffling was going on, the treasury remained guarded and the yearend balance was \$24,957.66. The Dauna Bauer Foundation, which donated \$3,000 in unrestricted funds in 1995, continued to be an important source of financial support.

Dena Giddens, a feisty redhead from Texas whose husband, Gary, had familial PPH, was elected to the Board during a conference call Board meeting in April of 1995.

Snapshot of Committee Chairs in 1995

Advisory Board Liason.....	LuAnne Washburn
Creative Writing.....	Sue Kelly
Children with PH/PPH.....	Linda Carr
Donations.....	Michelle Dawson
Librarian.....	Debra Cane
Mail.....	Julie Hendry
Membership.....	Sally Atkins
Membership List Coordinator.....	Eleanor Bails
Networking.....	Pat Paton
Organ Donor Awareness.....	Marilyn Dirr
<i>Pathlight</i> Editor.....	Mark Taylor Murphy
Public Affairs.....	Bonnie Dukart
Volunteer Coordinator.....	Julie Hendry

Martine Rothblatt, founder of the PPH Cure Foundation and the parent of a child with PPH, had contacted UPAPH in her effort to raise money for PPH research. Her request to use UPAPH's membership list for solicitation was denied by the Board, but it was agreed that information about the PPH Cure Foundation would be placed in *Pathlight*.

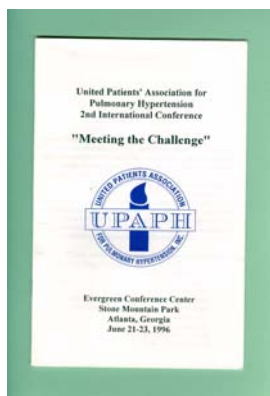
After much searching, UPAPH decided to buy an insurance policy that covered general liability for "counseling" by Board members and volunteers.

The Board also decided where and when to hold the second conference the following year.

1996

A SECOND CONFERENCE & THE DIET PILL SCANDAL LOOMS

Second International Conference



The highlight of 1996 was the second international UPAPH conference, held June 21-23 at the Evergreen Conference Resort, Stone Mountain, Georgia. As Ed Simpson said at the end of the conference, UPAPH conferences provided a “new model of healthcare education for the 21st Century, where patients, their families, physicians, researchers, and healthcare service providers meet on common ground in a cooperative, interactive format to learn from each other for the betterment of all.”

Ed Simpson and Bonnie Dukart co-chaired this conference, which had the theme “Meeting the Challenge.” The medical community gave input as to which topics should be covered in the sessions. There were 325 attendees, including physicians from Canada, England, and France. Of the attendees, 42 were professionals and 130 were patients. Some physicians brought their wives and families.

During the conference, patients eagerly ran (or wheeled) from session to session soaking up all they could about new treatments and ways to cope with the disease. They renewed friendships made at the previous conference and memorized the names and faces of top researchers (more than one patient commented that PH docs tended to be better looking than average) and buttonholed them with their questions. Some Flolan patients learned how to go swimming (carefully) while their doctors looked the other way.

Research at the conference. Physician/researchers again gathered blood samples and histories from patients and their families. A physician remarked to a patient, "It's hard enough to have a meeting of three or four renowned experts of PH at any one time--how was UPAPH able to get so many experts together for the Stone Mountain conference?" The patient replied, "No one ever said we couldn't."

Awards presented:

Outstanding PH Citizen.....**Dorothy Olson & Pat Paton**

Outstanding Physician.....**Dr. Lewis Rubin**

Costs. The total Conference bill came to \$114,713.00; UPAPH paid about \$9,000 of this from its treasury. The rest was covered by grants, donations, registration/conference fees, and (once again) a generous donation from the Evergreen Conference facility. Glaxo Wellcome, the major sponsor, provided \$25,000 in addition to a travel grant of \$10,000. Bonnie Dukart divided the latter into 30 individual patient travel grants. All patients received a discounted registration fee of \$110, and one significant other also had a reduced rate. Quantum helped provide oxygen for patients and Quantum nurses provided services during the conference. Charles Lybrook, Pat Paton's cousin, again printed the conference program as a donation to PHA. Conference sessions (excluding support group meetings) were again videotaped and the tapes were made available either for purchase (\$400 for a complete set) or from UPAPH's library.

Hints of the Coming Fen-Phen/Redux Scandal

Shortly before the conference, the U.S. FDA had approved dexfenfluramine for extended use by persons hoping to lose weight. Several doctors at the conference expressed alarm at this and at the growing use of related diet pills. Francois Brenot, MD, Hospital Antoine Beclere, Clamart, France, asked to speak to attendees. He warned listeners about the new

diet drug dexfenfluramine/Redux. (A similar generic drug containing fenfluramine and phentermine [a stimulant], called “fen-phen,” was also becoming very popular.) Sr. Brenot warned that a drug that was very similar to these had been removed from the market in France due to an increased incidence of PH in persons taking it.

Because at that point there was inadequate research to validate Dr. Brenot’s warning, UPAPH would have become vulnerable to lawsuits by the manufacturers of the diet pills if it had then issued an official statement saying the drugs could cause PH. However, those at the conference heeded the warning and spread the word.

Dr. Stuart Rich (who sat on PHA’s SAB), was also at the forefront of issuing warnings about the new diet pills. In fact, his efforts to stop the FDA’s approval of Redux thrust him into the national spotlight. In 1995, Dr. Rich served on the FDA advisory committee on whether Redux should be approved. He argued convincingly that it wasn’t safe. When it looked as if the committee was going to vote the drug unsafe, a very peculiar thing happened. The director of the division tabled the vote and postponed it to another day when none of the committee consultants were present. The drug was narrowly approved.

After this happened, the “Today” show had Stuart on as their guest. He talked about the paper that he and others had written on the European experience with diet pills. That paper was the lead article in *The New England Journal of Medicine* in August 1996. And then something even more peculiar than the FDA approval occurred: Dr. Rich says that he got a call from an executive at American Home Products, the maker of Redux. The executive warned him that if he continued to talk to the media about their new drug, it would be very dangerous for him and “bad things would happen.”

Well, Dr. Rich was a man with a wife and four children. He stopped giving special interviews to the press. But he didn't stop talking, lecturing, researching, or publishing on the matter. He and his colleagues, many of them members of UPAPH's SAB, organized an independent safety study of Redux, *paying for it out of their own pockets*. American Home Products, needless to say, wasn't happy about that, because they had no control over the collection of data. This study was a reason that the drug company finally agreed to withdraw Redux from the market.

In the summer 1966 issue of *Pathlight*, Dr. Rich's article on "PH and Diet Drugs" told readers about the link found in Europe, in 1967, between very similar diet pills (aminorex) and pulmonary arterial hypertension and warned that "exposure to fenfluramine and dexfenfluramine increased the relative risk [in Europe] of developing PPH, with the risk highest in people who used the drugs for greater than three months." In October, UPAPH presented a draft of the "UPAPH Official Statement on Dexfenfluramine." At the end of the year it was under review by the SAB before being published. (The makers of the pills did not yank them from the U.S. market until September 1997, after many users had already suffered adverse consequences in the form of heart-valve problems and PH. It is estimated that 1 in every 20,000 users developed PH. Six million people in the U.S. took the pills.)

Board Activities in 1996

The officers in 1996 were:

President.....	Judy Simpson, then Bonnie Dukart
Vice President.....	Bonnie Dukart, then Judy Simpson
Recording Secretary.....	Dorothy Olson, then Gary Dukart
Treasurer.....	Jerry Paton
Assistant Treasurer.....	Carol Wilson
<i>Pathlight</i> Editor.....	Mark Taylor Murphy
Corresponding Secretary.....	LuAnne Washburn

Directors-at-Large were:

Eleanor Bails	Gary Dukart
Dena Giddons	Harry Olson
Pat Paton	Ed Simpson
Peggie Wessinger	

(Bruce Brundage, MD, joined the Board later; in September, Linda Carr, Diane Adkins, and David Gunn were elected to the Board.)

In January, the Board again met at the Paton's home in Florida. Throughout 1996, Board members still were paying their own expenses to meetings. A lot of little matters were attended to. It was decided, for example, that Board members would begin serving staggered two-year terms.

An important decision was to stop basing the Patient-to-Patient Helpline in Susie Richardson's home, and instead to move it about to the homes of volunteer patients and family members knowledgeable about PPH/PH. Training would be provided for these volunteers. As far as PHA knows, having a helpline staffed by volunteer patients is unique to our organization. Bonnie Dukart arranged for the Helpline to use Totaltel as a server. Totaltel agreed to return 10 percent of revenues to UPAPH for those who would personally use their services, an arrangement that provided a very modest amount of revenue.

It was reported that an average of 31 new patient packets were being mailed out each month at a cost of \$4.83 each.

At the June Board meeting following the conference, Judy Simpson resigned as President of UPAPH. The Simpsons believed that bringing in new leadership, rather than letting UPAPH become a Simpson dynasty, would best serve the organization. The Board elected Bonnie Dukart as the new president.

Bonnie Dukart Brought Innovation and Professionalism to PHA



Bonnie Dukart was a person of great intelligence and courage. She was diagnosed with PPH in 1982 when she was 25 years old. For some years after her diagnosis she continued to pursue a career in banking, and was promoted to vice president of a large New York bank. When such work was no longer possible for her to do, she threw her considerable talents, brains, and remaining energy into helping UPAPH achieve its goals. She had learned of UPAPH over the fledgling Internet and volunteered. At the start she answered the Patient-to-Patient Helpline and did some public relations work. Later she co-chaired two international conferences and served as president. She raised money and volunteered for a clinical trail.

Bonnie was also a wife and mother, a daughter to her own parents, and a dear friend to many. For nineteen years after her diagnosis she lived a rich and happy life. She died in January 2001. Her ideas continue to shape PHA.

From the dedication of the Second Edition of
Pulmonary Hypertension: A Patient's Survival Guide.

UPAPH had begun to refine its goals. Its purpose statement in 1996 read:

The purpose of the United Patients Association for Pulmonary Hypertension, Inc. (UPAPH) is to provide fellowship and educational support to patients with pulmonary hypertension and primary pulmonary hypertension, their families, and physicians. The Association shall provide information to patients, families, physicians, researchers, and the public at large on issues pertinent to the disorders, such as current research and findings, early detection, resource organizations, organ transplantation, and support networks through the ongoing distribution of knowledge by means of periodic publications, symposia, and regional support group meetings.

Snapshot of (Informal) Committee Chairs in 1996

Children with PH/PPH.....	Linda Carr
Creative Writing.....	Sue Kelly
Donation Thank-Yous.....	Michelle Dawson
Librarian.....	Deborah Cane
Mail.....	Julie Hendry
Membership.....	Sally Atkin
Membership List Coordinator.....	Eleanor Bails
Networking.....	Pat Paton
Organ Donor Awareness.....	Marilyn Dirr
Public Affairs.....	Diane Adkins, then Bonnie Dukart
SAB Liason.....	LuAnne Washburn
Volunteer coordinator.....	David Gunn

Later in the year, Marcie Long and Dena Gibbons headed up fund-raising efforts, and David and Pat Gunn handled new-patient packets.

Finances and Membership. At the end of 1996, UPAPH reported total revenue of \$142, 827.62, and total expenses that were about \$6,000 less than this. At the September Board meeting it was reported that the average membership renewal was a \$35 donation, and income from TotalTel was \$50 a month. At the end of 1966, UPAPH had 630 patient/family members, 130 professional members, and 13 organization members, for a total of 773 members. There were members in 45 states and several from Australia and Canada.

Scientific Advisory Board

When the Scientific Advisory Board (SAB) met at the 1996 UPAPH conference, it elected Bruce Brundage as its first chair. The SAB requested that their chair have a seat on the UPAPH board to improve communications between doctors and the rest of the organization. This sensible request was granted. (Years later, the SAB would change its name to the Scientific Leadership Council [SLC] and would have three representatives on the board: the SLC chair, vice-chair, and immediate past chair.) Dr. Brundage would remain chair of the SAB until 2001.

Other pioneering PH physicians also joined the SAB early on. The following list includes only doctor/researchers who were both on the SAB during its very early years, and who went on to make quite a splash in the field. In the early years, the SAB was quite inclusive, and not all doctors who were on the Board later became leaders in the field. We've included photos whenever we could find one (this was a volunteer almost zero-budget project).

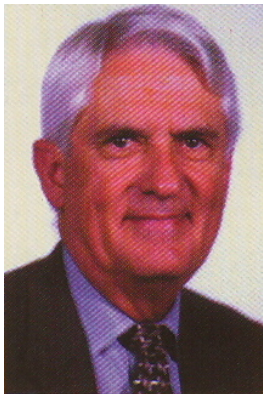


David Badesch. Clinical director of the Pulmonary Hypertension Center at the University of Colorado Health Sciences Center, Dr. Badesch is widely recognized as an outstanding researcher in the field of pulmonary hypertension. Much of his work has focused on vascular remodeling—on what happens at the cellular level in the walls of small arteries. He has led or participated in many such studies, and also in the development of such PH medicines such as iloprost, epoprostenol, and endothelin receptor antagonists.

Robyn J. Barst. Dr. Barst was captain of the cheerleading squad at her high school and once wanted to be a fashion designer, but barely missed out on getting a scholarship to Pratt. She had to settle for becoming a doctor (something her PH patients are deeply grateful for).



A pediatric cardiologist by training, she has been director of the Pulmonary Hypertension Center at New York Presbyterian Hospital since 1987. She is also the present chair of PHA's SLC. The first PH patient she saw, a teenage girl, died awaiting lung transplantation, an event that devastated Dr. Barst. She boned up on PPH and learned that there was then no treatment for it. Colleagues told her the disease was so rare that she'd never see another case. To say they were wrong is a gross understatement: the team she leads now serves at least 800 patients of all ages. She has coauthored countless important papers on PH, many of them focusing on treatments for children, treatments that she was instrumental in developing. "Today," she says, "we can guarantee that we can make patients feel better and live longer. However, although we have come a long way, we still have a long road ahead; we must--and we will—determine how to cure and prevent this deadly disease."



Bruce Brundage. Dr. Brundage is now more or less retired and enjoying life in Bend, Oregon, but he is one of the most respected figures in the annals of pulmonary hypertension and has a reputation as a man of integrity. It was in the late 1970s that Dr. Brundage began treating PPH patients. As this history shows, few doctors back then were even aware of the disease. During his long and distinguished career, he was associated with virtually every development in the treatment of PH. Along with PH pioneer Dr. Stuart Rich he co-authored the first paper to definitely show that calcium channel blockers could decrease pulmonary artery pressures. He sat on the steering committee of the National Registry, and was chief of the Department of Cardiology at Harbor-UCLA Medical Center. He served on the boards of the Los Angeles Heart Association and the American Board of Internal Medicine. His contributions to the clinical trial of epoprostenol were enormous, and he was

a coauthor of the resulting paper. He was the first to suggest that PHA have a medical arm, and thus became the first chair of PHA's enhanced SAB. Later he served as chair of PHA's Board of Directors and volunteered to "go anywhere and talk to anyone" about PHA to raise money for its mission. Dr. Brundage also served as the medical consultant for Gail Boyer Hayes' first two editions of PHA's *Pulmonary Hypertension: A Patient's Survival Guide*.

Greg Elliott. Dr. Elliott is the clinical director of the Pulmonary Hypertension center at LDS Hospital and professor of medicine at the University of Utah. His first experience with PH was with patients who had developed it secondary to acute lung injury. Vasodilators didn't help them. Dr. Elliott was interested in vasodilators and looked for patients the drugs would help, such as PPH patients. He became director for the Utah center for the NIH Registry, and got actively involved with other doctors who shared his interest in PH. His work soon began to focus on the genetics of PH. His patients, he says, are his real heroes and they have inspired him to focus his work, to explore new therapies, and to seek the cause of PH. He was instrumental in getting Flolan approved (see 1995). At the very first PHA conference he collected histories and samples for research into the genetic basis of PH. As a result, he discovered some common ancestors among patients who were thought to have sporadic PH (a hypothesis first put forward by Dr. John Newman). He succeeded Dr. Bruce Brundage as chair of SAB/SLC and enlarged it and formalized its process of succession and set up many committees, including those on education, research, and publications.

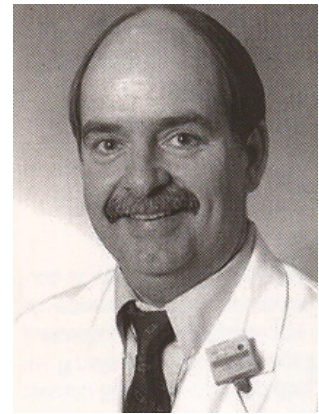


**Greg Elliott, MD (center)
with Mike McGoon, MD
(left) and PH patient and
Persistent Voices editor
Jerry Wojciechowski**



Tim Higenbottam. In 1994, a female PPH patient was awaiting a heart transplant at Papworth Hospital in Great Britain. Dr. Higenbottam infused her with a prostacyclin analog and saved her life. He wrote a case report on it, which was published. It was the first report on the use of this type of drug on a PH patient. Dr. Higenbottam is now a science advisor to the pharmaceutical firm AstraZeneca in Leicestershire, UK.

Jim Loyd. It was in Dr. Loyd's last year of his residency at Vanderbilt that he encountered a 30-year-old woman with PPH. Her family history contained many mysterious early deaths. The prevailing view then was that genes played almost no role in PH. This didn't seem right to him. Eventually he uncovered 22 PPH patients in the woman's family. When **John Newman**, MD, arrived at Vanderbilt, he was already interested in PH, and he and Dr. Loyd located other PPH families and did research on inheritance patterns that pointed to a PH gene. Other researchers joined the hunt, and in 2000 the first report appeared of the "PPH gene"—BMPR-2, which means mutations on the bone morphogenetic protein receptor on the long arm of chromosome 2.



David Langleben. Dr. Langleben is chief of the Department of Cardiology at busy Jewish General Hospital in Montreal, Canada, and an associate professor of medicine at McGill University. His particular area of expertise is pulmonary hypertension. His group has made significant contributions to the understanding of the endothelium (the innermost layer of the tiny blood vessels that get blocked in PH) and to the pathobiology of PH. Other interests are to develop

better medicines to treat PH and the epidemiology of the disease. He founded the first pulmonary hypertension clinic in Canada, the Centre for Pulmonary Vascular Disease.

Walker Long. A winner of PHA's Outstanding Physician Award, he helped develop the original Flolan trial when he was at Wellcome Research Laboratories (1983-1993). He gave early participants in the Flolan clinical trials his private telephone number and told them to call him day or night with questions. Today he is Research Professor of Pediatrics in the School of Medicine at the University of North Carolina at Chapel Hill. One of his current interests is telemedicine and digital innovations in health care.



Michael McGoon. As a former chair of PHA's SLC, Dr. McGoon has done much to help turn the attention of doctors and researchers away from using just vasodilators to developing and using more effective medicines. He graduated from the Johns Hopkins School of Medicine and did his residency at the Mayo Clinic, in Rochester, Minnesota, where he is now a professor of medicine. In fact, Dr. McGoon has lived in Rochester since he was six years old, and has the typical modesty of people from that part of the country. It was the complexity of PH, its impact on patients' lives, and the lack of effective treatment that led him to focus on the disease. He also says that he was fortunate to have worked with some of the great vascular biology researchers during his early years, including Ron Vlietstra, MD, and Dr. Paul Vanhoutte.

Jane Morse. Now a professor emerita of clinical medicine at Columbia University, Dr. Morse was a key part of the team there that, along with researchers from Vanderbilt, found the "PPH gene." Beyond just finding the gene, they have learned



much about how it contributes to the pathobiology of PH, with a focus on familial PAH, sporadic PAH, and PH associated with connective tissue diseases, HIV, and congenital heart problems. A rheumatologist by training, Dr. Morse, like many of the other physicians on this list, has frequently spoken at UPAPH/PHA conferences.

Kenneth Moser. Before he died of lung cancer, Dr. Moser, who worked at the University of California San Diego, made great contributions to the field by developing the leading program in the nation for thromboendarterectomies—a treatment for those PH patients with operable blockages that were causing their PH. He was also a principal investigator for the NIH Registry. In 1998 a Kenneth Moser Memorial Award for Outstanding Contributions to the Medical Community was given at the PHA conference.



John Newman. He and Dr. Loyd, both at Vanderbilt University, researched the pedigrees of families with PH, and their work ultimately contributed greatly to the discovery of the “PH gene.” (A team at Columbia located the gene at about the same time.) He also put forth the notion, later proven correct, that many cases of PH that were thought to be sporadic were actually inherited. For more, see 1994.

Harold Palevsky. Dr. Palevsky worked with Dr. Fishman at the University of Pennsylvania Medical Center, and is the model of what a caring PH doctor should be. Today he is chief of the Pulmonary, Allergy and Critical Care Division at the University of Pennsylvania Medical Center where he heads the Pulmonary Vascular Disease Program.



Lewis Rubin. Dr. Rubin had a bizarre experience near the start of his career. It was during his residency at Duke that he treated a man with advanced PH with vasodilators. After a couple years, the man developed severe anemia and kidney

failure, and died. No one was surprised; all PH patients died back then. About two months later the North Carolina State Board of Investigations called Dr. Rubin and told him they were exhuming the body. It turned out that the patient's wife had poisoned him with arsenic so that she could profit by selling some land they owned. In addition to achievements noted elsewhere in this history, Dr. Rubin was deeply involved in the first clinical trials of epoprostenol. He now has over 20 years experience in treating PH patients, has coauthored many research papers on PH, and led the effort to form the first American College of Chest Physicians guidelines for PH (and to update them in 2004). In 1996 he won PHA's Outstanding Physician award. And at the 1998 PHA conference he was presented the Kenneth Moser, MD, Memorial Award for Contributions to the PH Medical Community. Formerly with the University of Maryland, Baltimore, he is now a professor of medicine at the University of California, San Diego. Like most PH docs, he often goes to extraordinary lengths for his patients. When a patient in Istanbul could not go to California for a checkup, Dr. Rubin went to Istanbul to see him.

Stuart Rich. One of the first doctors to dedicate his career to the treatment of PH patients, Dr. Rich was a pioneer in the use of calcium channel blockers and warfarin, an outspoken critic of the diet pills Redux and fen-phen, a participant in many important clinical trials, and the author of groundbreaking papers on PH, many of which appeared in top medical journals. He was affiliated with the University of Illinois, Chicago for 16 years, and then moved to Rush University for 8 years. In 1998, when he was at the Rush Heart Institute in Chicago, Dr. Rich was awarded PHA's Outstanding Physician award. Today he is with the University of Chicago (where he says he's likely to stay put). Their Center for Pulmonary Hypertension is the largest PH referral center in the U.S. (currently following over 1,200 patients). They do genomic and



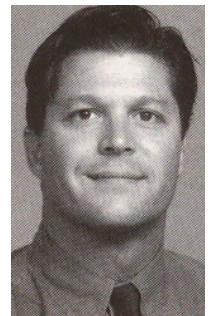
proteomic research, epidemiology, diagnostic studies, and work on developing new therapies for all forms of PH. A recent million-dollar grant from the dean of the medical school will allow them to investigate creative treatments for PH that industry would find too risky. Dr. Rich also devotes 30 percent of his time to work as an internal consultant for United Therapeutics. He notes that this allows him to pursue new therapies with the support of the company, such as the pill form of prostacyclin they are currently working on.



Cathy Severson. An RN, BSN, Kathy works with Dr. Michael McGoon at the Mayo Clinic. She is an excellent nurse, a patient advocate, and has contributed much to *Pulmonary Hypertension: A Patient's Survival Guide*. In 1998 she won PHA's Outstanding Medical Professional award for service above and beyond the call of duty. However, she is so Minnesota modest that you

can't even pry such information out of her.

Victor Tapson. A pulmonary hypertension specialist at Duke University, Dr. Tapson has participated in many clinical trials and served as the first editor of PHA's medical journal, *Advances in Pulmonary Hypertension*.



Dianne L. Zwicke. Board certified in both cardiovascular diseases and internal medicine, Dr. Zwicke is a Clinical Associate Professor at the University of Wisconsin at Madison. She is also associated with Heart Care Associates in Milwaukee, WI. Dr. Zwicke has

participated in clinical trials and published extensively in professional literature. Her sub-specialty areas are pulmonary hypertension, high-risk cardiac O.B., congestive heart failure, and heart/lung transplant.

Scientific Advisory Board Members in 1996

Chairman: Bruce H. Brundage, MD (UCLA School of Medicine)

(Names of new members listed below are in boldface type.)

Gary J. Alexander, MD (University of Utah School of Medicine)
David Badesch, MD (University of Colorado)
Robyn J. Barst, MD (Columbia/Presbyterian Babies Hospital)
Linda Clayton, Pharm D (Glaxo Wellcome Co.)
C. Gregory Elliott, MD (LDS Hospital, University of Utah)
Alfred P. Fishman, MD (University of Pennsylvania Medical Center)
Valentine Fuster, MD, PhD (Mt. Sinai Hospital, New York, NY)
Frank Gold, MD (Peoria, IL)
Timothy J. Higenbottam, MD (Sheffield, England)
Mieke Jobsis (Glaxo Wellcome Co.)
David Langleben, MD (Dept. of Medicine, Montreal, Canada)
E. Clinton Lawrence, MD (Emory Clinic)
Walker A. Long, MD (University of North Carolina)
James E. Loyd, MD (Vanderbilt University)
Michael D. McGoon, MD (Mayo Clinic, Rochester, MN)
Jane H. Morse, MD (Columbia University)
Kenneth M. Moser, MD (University of California, San Diego)
John H. Newman, MD (St. Thomas Hospital)
Harold Palevsky, MD (University of Pennsylvania Medical Center)
Stuart Rich, MD (Rush Presbyterian, Chicago)
Lewis Rubin, MD (University of Maryland)
Robert Safford, MD, PhD (Mayo Clinic)
Cathy J. Severson, RN (Mayo Clinic)
Warren R. Summer, MD (Louisiana State University Medical Center)
Victor E. Tapson, MD (Duke University Medical Center)
Beth Vogel, RN (Maine Medical Center)
Carol E. Vreim, PhD (National Institutes of Health)
Dianne L. Zwicke, MD (University of Wisconsin)

First Paid Staff Member

In spite of its many volunteers, UPAPH had grown to the point where it was necessary for Bonnie Dukart to hire a part-time secretary, Glenda Ellis. Glenda was UPAPH's first paid employee.

PH Ribbon



Dena Giddens designed PH awareness ribbons for UPAPH. The color is periwinkle, a gray-blue like the lips and nails of some PH patients prior to treatment! The ribbons could be purchased as either a ribbon with a gold metal heart stickpin, or as a die cast

lapel pin. They cost \$3 each. Periwinkle thus became the “official” ribbon color of PHA; no other group could use our color.

On the Research Front

A new clinical study on using epoprostenol (Flolan) to treat PH associated with the scleroderma spectrum of diseases was announced. Because many patients have PH secondary to one of the connective tissue diseases, this study was of great interest to UPAPH members.

Dr. Jim Loyd, Vanderbilt University, gave the UPAPH Board an update on the National Registry of Primary Pulmonary Hypertension Registry in Families. Fifty-three families that had more than one family member with PPH were then being studied. The increased number of families provided a more adequate base to work with for locating the PH gene. UPAPH was instrumental in helping to locate families with more than one PH patient.

Speaking Out on Capitol Hill

In February, Judy Simpson testified before a Senate Judiciary Committee hearing. She spoke in opposition to the Pryor-Chaffee bill, S1191, to amend the GATT agreement. This bill would have allowed companies that made generic copies of drugs to get their drugs ready for market before the patents on their brand-name equivalents expired. Senators Hatch, Chair, Feinstein, Simon, Kennedy, Leahy, Heflin, DeWine, Grassley, Specter, Pryor, Chaffee and Faircloth were present. Judy explained how patients with rare diseases need continued Research and Development (R and D) of new drugs. She stressed the need for protection for pharmaceutical companies to have their full span of patent protection in order to have the funds to make such investments. It took about \$500 million to bring a new drug, like Flolan, to the FDA for approval, she told the Committee. Flolan would never pay for itself, she said, so people with rare diseases must rely on profits a pharmaceutical company makes on high-volume drugs to support R and

D. (U.S. pharmaceutical companies claimed that they were returning 14 percent of their profits to R and D; the companies said this was the highest percentage of profit returns to R and D of any industry in the world.) Therefore, UPAPH opposed a shortening of the patent period for pharmaceuticals. (Future consequences of laws can be tricky to predict. A search of bills passed by the Senate during this period could find no record of this bill reaching the floor for a vote. It will be interesting to see how PHA members feel on this issue when someone wants to market a generic version of sildenafil [Viagra]). By 2005 it was known that some "orphan" drugs for rare diseases can be great profit centers for companies.) Judy paid for all of her expenses for this trip to Washington, DC, out of her own pocket.

On another front on Capitol Hill, Martine Rothblatt (PPH Cure Foundation founder) succeeded in getting PPH into the fiscal year 1997 Senate Appropriations Committee Report Language for NHLBI:

Primary pulmonary hypertension (PPH) is a progressive fatal disease whose mostly female victims are of all ages and races, involves deadly deterioration of the heart and lungs, and literally robs these people of their very life's breath. The Committee urges the Institute to expand its support of research devoted specifically to PPH gene therapy, basic research, and clinical trials of promising pharmaceuticals.

In response to the congressional mandate, the NHLBI Division of Lung Diseases soon issued a Program Announcement, "Cellular and Molecular Mechanisms of PPH." Researchers were invited to apply for grants.

Between 1998 and 2004, the NHLBI portfolio grew from under \$8 million to over \$27 million. This increase benefited PH researchers. -

Networking with Other Organizations

In September, Diane Adkins, Bonnie Dukart, Dena Giddens, and Ed and Judy Simpson attended a NORD Board meeting, all at their own

expense. Issues discussed at the meeting included: 1) ways to provide more financial support for rare disease research, 2) ethical considerations in gene therapy, and 3) how to increase the development of orphan drugs. (Glaxo Wellcome was honored by NORD and received the Corporate Leadership Award for their development of the orphan drug Flolan.) After the NORD meeting, the UPAPH group went on to meet with Dr. Stephen Groft of NIH's Office of Rare Diseases.

Judy Simpson was appointed to the NIH National Heart, Lung and Blood Institute (NHLBI) Advisory Council (NHLBAC) as a patient advocate. This was a big deal. Here is the NHLBI website description of what the Council does:

The National Heart, Lung, and Blood Advisory Council advises the Secretary of DHHS, the Assistant Secretary for Health, the Director of National Institutes of Health, and the Director of the National Heart, Lung, and Blood Institute on matters relating to the cause, prevention, diagnosis, and treatment of heart, blood vessel, lung, and blood diseases; the use of blood and blood products and the management of blood resources; and on sleep disorders. The Council considers applications for research and research training grants and cooperative agreements and recommends funding for those applications that show promise of making valuable contributions to human knowledge. The Council may also make recommendations to the Director, NHLBI, respecting research conducted at the Institute. The Council meets four times a year--winter, spring, and two meetings in the fall.

Everybody but Judy had a long string of initials after his or her name (Judy *could* have had RN, Ed.S.). Twelve of the members were leading representatives of the health and scientific disciplines, and the Secretary appointed six from the general public. Such non-credentialed luminaries as Frank Sinatra and Arthur Ashe have also been appointed to the Council. Frank, however, handled it his own way and never showed up, and Arthur opted out after a few meets. Judy, on the other hand, raised awareness of PH among an important set of policy makers.

Meanwhile, Bonnie Dukart sat on the Board of the PPH Cure Foundation. The Foundation raised over \$550,000 for PPH research, UPAPH members were asked to help identify potential donors of \$10,000 or more. The Foundation was organized in 1995 by Martine Rothblatt, JD, PhD, MBA. Martine is such a fascinating person, and her life has had such a positive impact on the lives of many PH patients, that she deserves some pixels here. Although she had no medical training, after her daughter's diagnosis with PPH in 1994, Martine quickly learned a great deal about the disease. She had more than a bit of money from her careers as a lawyer and from launching several satellite communications companies. So in 1996 she founded the biotechnology company United Therapeutics. It is the company that makes treprostinil (Remodulin). Martine had talked Burroughs Wellcome (now GlaxoSmithKline) into granting her the rights to the drug after the big firm had given up on it because they thought the market was too small. (Because Martine's daughter is a responder to calcium channel blockers, she does not use Remodulin.) Martine also put a lot of her own money into the PPH Cure Foundation, which awards millions of dollars in grants to PH researchers.

If you Google "Martine Rothblatt" you will learn that she has authored books on genomics, xenotransplantation, and gender freedom (Martine underwent sex-reassignment surgery in the early 1990s and changed her name from Martin to Martine). Her latest book is *Two Stars for Peace: The Case for Using U.S. Statehood to Achieve Lasting Peace in the Middle East*. In this book she proposes creating two new states: Israel and Palestine. This is clearly a person who can think outside the box.

Publications in 1996 Get the Word Out

- Dr. J. Robyn Barst et al. published "A Comparison of Continuous Intravenous Epoprostenol (Prostacyclin) With Conventional Therapy For Primary Pulmonary Hypertension" in *The New England Journal of Medicine*, February 1, 1996, 334:296-301. Through a grant, Glaxo Wellcome had 10,000 reprints made for UPAPH for distribution to patients and physicians.
- The spring issue of *Pathlight* was sponsored by Quantum Health Resources and explained in detail how this Flolan provider would work with patients needing the drug.
- NIH/NHLBI revised and reprinted its PPH pamphlet.
- The Mayo Clinic donated 100 of their publications on PH to be used in new member packets until the revised NHLBI pamphlets arrived.
- Glaxo Wellcome paid for 18,000 new UPAPH brochures that were produced by the Cooney-Waters Group.

1997

UPAPH BECOMES PHA AND GETS A WEBSITE

As UPAPH's new president, Bonnie Dukart brought innovative ideas to the organization and polished its professional appearance. The rapid growth of the Internet and its use offered an opportunity to reach far more PH patients. It was agreed that the name "UPAPH" was somewhat hard to remember and would be difficult to find in a search on the Internet. Austin Carr, an attorney and father of PH patient Hannah, volunteered to do the paperwork to change the name from the United Patients Association for Pulmonary Hypertension (UPAPH) to the Pulmonary Hypertension Association (PHA). Our logo was modified accordingly.

Website Begun

The UPAPH/PHA website was launched January 8, 1997. It was the work of Michael and Kathy Szczepkowski. Kathy had PH secondary to scleroderma, and Michael was an engineer who specialized in knowledge engineering and user-interface design. (Sadly, Kathy died the following year.) Michael chaired a new Internet Committee, which was appointed to work on the website. The brother of PH patient Jan Travioli offered free space for the site. PHA registered its domain name: <http://www.phassociation.org/>. Carol Aserinsky, a PH patient, later arranged for a professional website designer to redesign the site pro bono. Later in 1997, Michael et al. improved the website and included a threaded message board, a navigational site map, and site-searching capabilities.

Diet Pill Tragedy

In January, UPAPH's statement warning of the dangers of using dexfenfluramine was approved by SAB members Dr. Stuart Rich and Dr. Lewis Rubin. It was sent to support group leaders, area directors, and the Patient-to-Patient Helpline volunteers. It was not until September that the makers of the dangerous diet pills withdrew them from the market.

Leadership Conference

An issue of *Pathlight* in 1997 filled an entire page with a list—in a small font size--of PHA volunteers. Coordinating the efforts of an exploding number of volunteers was challenging. So early in June, a leadership conference was held in Oakbrook, IL, for the Board, committee chairs, and support group leaders. There were 46 participants. Co-chairs were Ed Simpson and Susie Richardson. Glaxo Wellcome and Olsten Health Services were corporate sponsors. (Quantum had morphed into Olsten, which later became Gentiva Health Services and then Accredo Therapeutics. This is an oversimplification of this entity's complicated genealogy.)

Livingston Francis, a professional facilitator, helped keep the meeting on track. An organizational chart of PHA that assigned the supervision of the various committee chairs and liaisons to particular officers was proposed, a new mission statement worked on, and strategic objectives and a vision for the future discussed. Nancy Davis, who worked for Olsten, led a session for the Patient-to-Patient Helpline volunteers, and Craig Mears, also with Olsten, led a support-group-leader workshop. The sessions looked at structure and responsibilities and set goals. These ideas for new areas of focus for PHA were suggested: thinking on an international scale, children with PH, providing up-to-date information on the diet pill issue, separating fact from myths about medications and treatments, facilitating the formation of more support groups, using the Internet as a resource to locate materials, using

members as a resource for research (via questionnaires, at the conferences, etc), and compiling a list of philanthropic organizations. A list was made of tasks to be done, the persons responsible, and due dates. Most goals set at the Leadership Conference were met as of the start of the following year.

First National PH/PPH Awareness Week

PHA orchestrated the first national PH/PPH Awareness Week, held November 2-8, 1997. Members and supporters were urged to wear their PH ribbons, to schedule support group meetings or social gatherings, and to try to drum up some publicity for UPAPH from their local media outlets.

The Week was declared a rousing success, although its results seem modest by today's standards, when events such as a concert at the Kennedy Center featuring the Secretary of State at the piano are held to raise PH awareness. But the first Awareness Week was a good start. Quite a few PH doctors spoke at support group meetings and several newspapers ran stories on PH, most of them focusing on individual patients.

Betty Lou and Jerry Wojciechowski: PH Runs in the Family and So Does Helping Others



A major force behind Awareness Week was (and is) PHA's vice president for support, Betty Lou Wojciechowski (shortened to "Wojo" by her friends). Betty Lou is a force of nature. Seldom seen without a smile on her face and moving at warp drive speed, she and her family have contributed to UPAPH/PHA in a great many ways. Her husband, Jerry, was diagnosed with PPH in 1994, and two of their sons have died of PH (two other children survive). The family's DNA is in labs around the world where research is being done on the genetics of IPAHA and familial PAH. As Betty Lou says, the Wojos do "anything we can do to further treatment and find a cure." Their home base is Mission Viejo, California, but Betty Lou and Jerry can pop up anywhere they are needed, even in Canada. They formed a support group at UCLA-Harbor, and published the newsletter "Lung and Times" that was sent to over 120 PH patients in California. Jerry is presently the editor of *Persistent Voices*.

The first Awareness Week has a special meaning for the Wojos, because it was just before that, on October 28, that they lost a second child (24-year-old Michael) to familial PAH. Betty Lou says: "His funeral service was held on November first, and we took some comfort in the fact that even in his death, he reached over a thousand people with PH awareness; each attendee at the service received a PHA brochure and envelope, and we passed out many, many pins. There was a brief explanation of the disease and a request for donations to PHA in Michael's name on the back of the service outline, and the minister announced that this was the first day of Pulmonary Hypertension Awareness Month."

Families like the Wojciechowski's are the driving spirit behind PHA.

Medical and SAB Developments

A conference on the "Mechanics of Proliferation and Obliterative Vascular Diseases: Insights from the Pulmonary and Systemic Circulations" was sponsored on September 2-3 in Bethesda, MD, by the Foundation for PH, the PPH Cure Foundation, and Lung RX. The meeting was chaired by Dr. Alfred Fishman and brought together cardiologists and pulmonologists (specializing in both right and left heart problems). Gary and Bonnie Dukart, Armond and Carol Aserinsky, and Ed and Judy Simpson attended this meeting at their own expense. It was an exciting event that raised the prospect of joint research projects.

In July 1997, the Brenot Memorial Symposium on the Pathogenesis of Primary Pulmonary Hypertension was held in Corsica. The papers presented there were published in *Chest* 1998 144 (3 Supp): 183S-247S.

The NHLBI continued to seek grant applications for "Cellular and Molecular Mechanisms of PPH." A few came in, but progress was slow. PHA notified PH doctors/researchers of the availability of the grants.

The PPH Research Foundation (as mentioned earlier, it was founded by Gabriel Miyara, a patient, and was based in Marina del Rey, CA) offered a

grant to Dr. Lewis Rubin. (In 2000, PHA and the Primary Pulmonary Hypertension Research Foundation completed negotiations that resulted in the merger of PPHRF into PHA. As a consequence, the Gabrielle Miyara and Rachel Hoyt Research Fellowship was first presented at the 2000 PHA International Conference.)

The PPH Cure Foundation announced that it was supporting research on treprostinil (Remodulin). This prostacyclin analogue was of interest because, unlike epoprostenol (Flolan), it would be stable at room temperature and could be injected into the fat under a patient's skin. This would eliminate much of the danger of infection that was associated with epoprostenol. It would also require a smaller pump than epoprostenol.

Sixty-five families were now included in the National Registry for Familial PPH.

Scientific Advisory Board Members in 1997 & 1998

Chairman: Bruce H. Brundage, MD (UCLA School of Medicine)
(Names of new members below are in boldface type.)

David Badesch, MD (University of Colorado, Denver, CO)
Robyn J. Barst, MD (Columbia/Presbyterian Babies Hospital, New York, NY)
Richard Channick, MD (UC San Diego, CA)
C. Gregory Elliott, MD (LDS Hospital, University of Utah, Salt Lake City, UT)
Alfred P. Fishman, MD (University of Penn. Medical Center, Philadelphia, PA)
Adaani Frost, MD (Baylor College of Medicine, Houston, TX)
Timothy J. Higenbottam, MD (University of Sheffield, Sheffield, UK)
Mieke Jobsis (Glaxo Wellcome, Research Triangle Park, NC)
Abby Krichman, RRT (Duke University Medical Center, Durham, NC)
David Langleben, MD (Jewish General Hospital, Montreal, Quebec, Canada)
James E. Loyd, MD (Vanderbilt University, Nashville, TN)
Michael D. McGoon, MD (Mayo Clinic, Rochester, MN)
John H. Newman, MD (Nashville VA Hospital, Nashville, TN)
Harold Palevsky, MD (University of Penn. Medical Center, Philadelphia, PA)
Stuart Rich, MD (Rush Heart Institute, Chicago, IL)
Lewis Rubin, MD (University of Maryland School of Medicine, Baltimore, MD)
Cathy J. Severson, RN (Mayo Clinic, Rochester, MN)
Victor E. Tapson, MD (Duke University Medical Center, Durham, NC)
Beth Vogel, RN (Maine Medical Center, Portland, ME)
Carol E. Vreim, PhD (National Institutes of Health, Bethesda, MD)
Dianne L. Zwicke, MD (University of Wisconsin, Milwaukee, WI)

Free Legal Help for PH Patients Becomes Available

ACCESS (Advocating for Chronic Conditions, Entitlements, and Social Services) was founded in 1991 by the late Rachel Warner to help families coping with hemophilia to obtain the state, federal, and private sector (insurance) to which they were rightfully and legally entitled. Smart lawyers who can guide patients through the maze of medical insurance are on the staff ACCESS. Quantum Health Resources provided this service, which was expanded to include a few other chronic conditions, including PH. It was (and is) not necessary for a PH patient to be a client of Quantum or its successors (Olsten Health Services, Gentiva Health Services, and Accredo Therapeutics) to get this valuable help.

Board Activities in 1997

The officers in 1997 were:

President.....	Bonnie Dukart
Vice President.....	Judy Simpson, RN, Ed.S.
2 nd Vice President.....	Betty Lou Wojciechowski
Secretary.....	Gary Dukart,MD
Treasurer.....	Jerry Paton
Assistant Treasurer.....	..Carol Wilson

Directors-at-Large were:

Diane Adkins	Bruce Brundage, MD	Linda Carr
Dena Giddons	David Gunn	Dorothy Olson
Susie Richardson	Ed Simpson	

In January, the UPAPH Board met at the home of Leonard Dukart in Delray Beach, Florida. Attending (at their own expense) were Diane Adkins, Gloria Blodgett, Linda and Austin Carr, Bonnie and Gary Dukart, Angie Eldam, Dena Giddons, David and Pat Gunn, Pat Paton, Susie Richardson, Judy and Ed Simpson, Barbara and Vern Smith, and Betty Lou Wojciechowski. The bylaws were updated and it was decided to set up terms on the Board on a three-year rotation.

Plans were made for a national support group week and for a guidebook about the disease for patients with PH. After some skillful coaxing by Bonnie, Gail Boyer Hayes, a PH patient and former journalist, had agreed to author and edit the guide. Pat Gunn, the vivacious and charming wife of patient David Gunn, agreed to chair the 1998 UPAPH Conference, to be held in Dallas, TX. When David Gunn was diagnosed with PPH in 1993, he and Pat were ranchers in western Texas. He finished the rounding up and sheep sheering before going to Chicago to see Dr. Stuart Rich. Calcium channel blockers didn't work for David. Both he and Pat decided not to live in fear, and as is evident from material above, they both did a great deal of valuable volunteer work for PHA, often in leadership positions. David later tried Flolan, but it could not keep him alive, and he died in July 1999.

Not long after the January meeting, Mark Murphy resigned from the Board and Susie Richardson joined it.

Snapshot of Committee Chairs in 1997

Caregivers.....	Dena Giddens
Creative Writing (Persistent Voices)...	Sue Kelly
Children with PH/PPH.....	Linda Carr
Librarian.....	Debra Cane
Mail & Donations.....	Julie Hendry
Membership.....	Sally Maddox
Membership List Coordinator.....	Brad Belliston/Eleanor Bails, then Bonnie Dukart
Networking.....	Pat Paton
Organ Donor Awareness.....	Sharren Yamron
Organizational Liaisons	
NORD Board	Ed Simpson
NIH Advisory Board.....	Judy Simpson
<i>Pathlight</i> Editors.....	Jan Travioli, Gary Dukart, Victor Tapson (MD) (Medical Editor)
Public Affairs.....	Diane Adkins, then Shirley Craig
Volunteer Coordinator.....	Kathy Szczepkowski
Web Site Coordinator.....	Michael Szczepkowski
Web Site Guest Book.....	Kathy Levitt
800 Number Coordinator.....	..Kathy Windhorn

In addition to the committee chairs, there were a great many other volunteers by 1997: support group leaders, those helping to organize the next conference, patients answering the Patient-to-Patient Helpline, etc.

The volunteer in charge of membership, Sally Maddox, was diagnosed with PPH about the time UPAPH was being formed, when she was being evaluated for a transplant at St. Louis. In addition to later volunteering as a state coordinator for Georgia and helping out at conferences, she presently serves on PHA's Board. After being diagnosed Sally went to college (while raising a stepson she gained when she married) and earned a specialist degree in mathematics education. Her PhD now almost in hand, she is an inspiration to every PH patient.

In June a Board meeting was held in Chicago, immediately following the Leadership Conference. Although Board members were still paying their own expenses to Board meetings, this meeting was an exception because it was attached to the Leadership Conference and was covered by a grant.

Requests had been received for a special issue of *Persistent Voices* dealing with diet-drug-induced PH. The Board approved \$1,000 to cover the expense. PHA's organizational structure was modified according to the proposal from the Leadership Conference. Dena Giddens reported on fund-raising ideas she had collected. There were the usual suggestions such as bake sales, and garage sales, and unusual ones such as a dog wash.

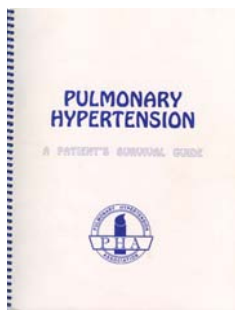
The PPH Action Coalition, a group interested in legislation and advocacy, and coordinated by Sandy Silbermann of Maryland, had begun to work separately from the PPH Cure Foundation. The Coalition lobbied hard for things like getting Congress to push NIH to spend more money on PPH. The Action Coalition was invited to become a part of PHA with an annual budget of \$400. They accepted.

By the end of 1997, there was \$82,948.12 in PHA's treasury. At the start of the year UPAPH/PHA's membership was 800; by year's end it was 1,250.

1998

FIRST *PATIENT'S SURVIVAL GUIDE* PUBLISHED

Pulmonary Hypertension: A Patient's Survival Guide



**First Edition
(1998): 123 pp**

The Internet age was just dawning and most PH patients still did not have access to it. Even those who did lacked the sort of in-depth, accurate information they needed to cope with their illness and make intelligent decisions about which treatment options to discuss with their doctors. Both PH and its treatments were exceedingly complex. It is quite impossible for a patient with such an illness to remember everything a doctor tells him or her during an office visit, and equally impossible for a doctor to be comprehensive in the time allotted. PHA decided, therefore, that patients needed a comprehensive reference book they could turn to for reassurance and for guidance.

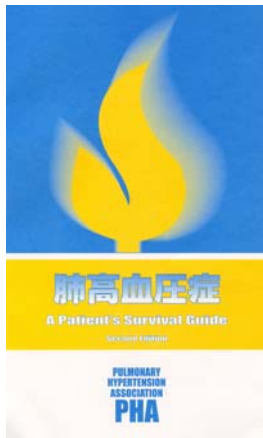
PPH patient and writer/editor Gail Boyer Hayes (Seattle, WA) wrote the 123-page first edition of *Pulmonary Hypertension: A Patient's Survival Guide*, which PHA published in 1998. Before she was diagnosed with PPH at the age of 40, Gail had been working as a lawyer in California, Colorado, and Washington, DC. And before that, she had been a book reviewer, magazine and newspaper reporter, short-story writer, and television talk show hostess. For the six years following her decision to write the *Survival Guide*, the *Guide* and its updates consumed nearly all of her free time. The medical consultant



**Gail Boyer
Hayes**

was the distinguished Dr. Bruce Brundage. Andrea Rich (the wife of Dr. Stuart Rich) did the medical illustrations. Other PH patients, doctors, nurses, and family members reviewed the book prior to publication to make sure it answered the right questions and explained things in language a layperson could understand. Chapters included: What is PH?; So How Do I Know It's Really PH?; What Causes PH?; Treating PH; Tell Me Doc, How Long Do I Have?; Children and PH; Living with PH, and Tedious Paperwork and Legal Matters. Cost of the 123-page *Guide* to PHA members was \$10.00.

Gail was concerned that the *Guide* not be slanted to curry the favor of any commercial interest. Therefore, she did not turn over the copyright to PHA until she was assured of this. (It turned out not to be an issue; PHA also wanted a *Guide* that patients could trust.)



Japanese version of Second Edition (2001), 189 pp in Japanese, 215 pp in English

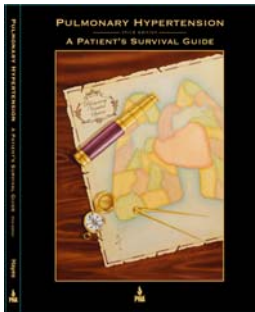
The *Patient's Survival Guide* was an immediate hit and flew out the door. Olsten provided PHA with \$1,500 to include a copy in the new patient packets. Many doctors bought multiple copies to give out to their patients.

PH patient Barbara Smith and her husband Vern, residents of Odessa, Florida, who had built up a chain of plumbing, cable, air conditioning, and electrical businesses, mailed the *Survival Guides* and paid for postage out of their own pocket. Barbara was already familiar with PPH before she was diagnosed in 1995,

because both her sister Rachel, and her daughter Angela, had died of the disease. When Angela died, she was pregnant with her third child. After Barbara became too ill to handle the mailings herself, the Smiths continued to pay for them. For at least 5 years they also mailed out membership packets, new patient packets, PH pins and cards, and other



Barbara Smith



**The Third
Edition (2004):
286 pp**

materials, and paid the postage on them. Few patients realized the generosity that made their low-cost orders possible. Barbara once said, "I couldn't figure out why I was still alive after losing my daughter and my sister. The only reason I can think of is to help other people." Barbara survived until October 2005.

Third International PHA Conference and Birth of PH Resource Network

Pat Gunn chaired this conference, which was held June 21-23, at the DFW Lakes Hilton Hotel in Dallas, TX. The theme was "Hope Through Education," and about 500 people attended. Support groups representing 17 categories met. Banners made by support groups from around the nation decorated the walls. Fifty-four professional presenters were listed in the program. New sessions on liver/PH and scleroderma/PH were included.

A group of nurses who worked with PH patients met at the conference and made plans to start the Pulmonary Hypertension Resource Network (PHRN). PHRN membership is open to any medical professional, nurse, respiratory therapist, pharmacist, medical technician, etc., who is employed to care for PH patients.

As at all PHA conferences, there was a lot of laughter. Doctors lightened their presentations with jokes, and dozens of patients proved that it is possible to do Texas line dancing while "on the pump."

Awards presented:

- **Kenneth Moser, MD**, Memorial Award for Contributions to the PH Medical Community-- Dr. Lewis Rubin (University of Maryland, Baltimore)
- Outstanding Physician—**Dr. Stuart Rich** (Rush Heart Institute, Chicago, IL)
- Outstanding Medical Professional--Cathy **Severson** (Mayo Clinic), for service above and beyond the call of duty

- Teresa Knazik Memorial Award for a PH patient who has given the most to PHA during the previous two years-- **Jan Travioli** (*right*) for her work on *Pathlight* and the PHA Board. Jan was the editor of *Pathlight* from 1996 to 2000. A skilled jewelry designer, her necklaces and bracelets are treasured by many, and she designed the first PHA awareness bracelet in sterling and amethyst. When she died from PH in 2004 she made a last gift to all PH families—she donated her body for research.
- Outstanding caregiver--**Harry Olson** for his devoted care to his wife Dorothy Olson and for his service on the Board
- The Judy and Ed Simpson Family Member Award for family members who have provided special services to PHA - **Judy and Ed Simpson** for everything they had done as recounted in this history.



The videotapes, although they were professionally made, were difficult to hear and/or understand. The tapes, such as they were, were again available for free borrowing through the PHA Library. It was decided to make only audiotapes of future conferences.

Conference evaluation forms were distributed during the conference, and the results showed a high level of satisfaction with the event. Three-quarters of participants learned of the conference from *Pathlight* or PHA's website; others heard about it from health care professionals and support groups. As would often be the case, there were complaints about hot weather and suggestions made that another time of year be selected for conferences. But it was decided, as usual, that the weather problem was outweighed by the need to meet when school is out, when prices are lower, and when medical professionals have more free time.

New Fundraising Efforts

At the June Board meeting, it was decided that PHA and the PPH Cure Foundation should work together on fund raising for research. PHA was to receive 5 percent of monies raised up to \$100,000. PPH Cure was interested in very large donations from persons located through PHA. It was planned that Bonnie Dukart would continue to

have a seat on the PPH Cure Foundation Board, and a person from the PPH Cure Foundation Board would take a seat on the PHA Board. This arrangement didn't work out, and the agreement soon ended.

Also at the June meeting, the Board discussed trying to raise money in smaller amounts for PHA to earmark for research. This was a new and exciting step for PHA.

During the Texas conference, the first fund-raising "Hoops and Hops for PPH" was held at Rachel Gildea's school and raised over \$16,000.00 for the PPH Cure Foundation for research. This was the largest amount yet raised by volunteers. Students got pledges to participate in the event during gym class, choosing to either shoot basketball hoops or jump rope for an allotted time period. A dozen other such events were scheduled.

Support Group Activity and Awareness Week '98

By the start of the year, there were about 35 PH support groups, and the need to provide them with more information and guidance was clear. Many ideas were bandied about, but mostly left unresolved. The Board did vote not to have local chapters, due to the individual state requirements and costs involved. Because of PHA's tax status, individual PHA support groups could not open their own bank accounts (although large groups might have petty cash accounts). The PHA Board decided to handle matters through reimbursements for submitted bills. Marcie Long had succeeded in finding state coordinators for most states. One of their duties was to write welcoming letters to new PHA members in their states.

The Omaha, Nebraska, support group sent out letters about PH to 119 cardiologists and pulmonologists in their area. In Oklahoma, a group set up an information table at a local hospital. Chicago published and sold 455 cookbooks. In Salt Lake City, Barbie and Brad Belliston sold raffle tickets for

a “quillow,” a combination quilt/pillow. Many support groups met during the week and some new ones formed.

Internet Activity

Carol and Armond Aserinski chaired the Internet Committee and reported that the PHA WEB site was averaging 19 visits per day. During one week in February, however, the site had 7,000 visits from the U.S. and nine other countries. The message board was the most popular feature. A password-protected message board for medical professionals was also ready to be tested. Michael Szczepkowski hoped that medical centers would be encouraged to prepare web pages that could be linked to the PHA site.

On the Medical Front

PHA began making a determined effort to reach out to physicians who were not PH specialists. PHA literature was available at the American Academy of Family Physicians’ fall meeting, and ways were sought for PHA to have a presence at the American College of Cardiology and the American Thoracic Society meetings. Interest began to gel around the idea of preparing materials on PH aimed at medical professionals.

The World Symposium on PPH was held in Evian, France on September 7, 1998. It was the first such gathering since 1973 and did a lot to integrate what was happening around the world on various PH fronts. Sixty-nine physicians/scientists attended this meeting, which was sponsored by the World Health Organization. Committees worked on pathology, pathobiology, genetics, risk factors and associated conditions, diagnosis and assessments, and medical treatment. The current understanding was that idiopathic PH required both a genetic predisposition plus exposure to some environmental factor to “trigger” it. A loose consensus was reached on what drugs and toxins might act as a trigger. A definite link was found to aminorex (European diet pills), fenfluramine, dexfenfluramine, and toxic rapeseed oil.

A very likely link was found to amphetamines and contaminated L-tryptophan. A possible link existed to cocaine and chemotherapeutic agents. And the following were thought by most doctors to probably not be directly linked to PH: antidepressants, birth control pills, estrogen therapy, and cigarette smoking.

Also at the meeting, a reclassification of PH was recommended. (This reclassification would occur at the next World Symposium on PPH in Venice in 2003, which is beyond the scope of this history. For readers who may be confused by some of the terms used, here is a nutshell version of the changes made at the 2003 Symposium: the term primary pulmonary hypertension [PPH], which just meant unexplained PH, was put under the category of pulmonary arterial hypertension [PAH]; PAH was then divided into three categories: idiopathic PAH [IPAH], familial PAH [FPAH], and PH associated with other diseases [APAH]. Other classes of PH included pulmonary venous hypertension, PH associated with hypoxia, PH due to chronic and/or embolic disease, and a miscellaneous category.)

S.D. Lee et al. reported in the *Journal of Clinical Investigation* that while 77 percent of endothelial cells in plexiform lesions from four patients with PPH were monoclonal (originating from a single cell), all lesions from four patients with PH secondary to another disease were polyclonal (originating from multiple cells).

M. Humbert et al. reported in the *European Respiratory Journal* that growth factors such as platelet-derived growth factor might play a part in the initiation and/or progression of PPH.

John V. Conte et al., writing in the *Journal of Heart & Lung Transplantation*, concluded that "...prostacyclin therapy is an effective means of delaying, possibly indefinitely, the need for lung transplant in patients with PPH, and that excellent results can be obtained when lung transplant is performed after prostacyclin therapy. Consideration should be given to

initiating prostacyclin therapy in all patients whose conditions do not respond to conventional therapy before proceeding with transplantation."

In the *Journal of the American College of Cardiology*, M.J. Ricciardi et al. wrote that: "The pulmonary vascular response to inhaled NO accurately predicts the acute hemodynamic response to nifedipine in PPH, and a positive response to NO is associated with a safe nifedipine trial....a trial of nifedipine in NO nonresponders appears unwarranted and potentially dangerous."

Stuart Rich et al. published a case series study in the *Journal of American College of Cardiology* of PPH patients with cardiac outputs that were too high. By carefully reducing their dose of epoprostenol/Flolan they were able to lower the patients' cardiac outputs without any significant change in their pulmonary artery pressures.

A.L. Churnock et al. wrote in the *American Journal of Medical Science* about their retrospective review of 41 PPH patients. They found that nine of 40 patients (22.5 percent) had hypothyroidism, which is a much greater incidence than in the general population. They suggested that PPH patients be regularly screened for this disease, particularly if their symptoms suddenly grow worse.

Norbert Voelkel's group at the University of Colorado did exciting work on the role of a critical enzyme, PG12-synthase (PG12 is prostacyclin) and concluded that it plays a big role in modifying the response of lung blood vessels to a chronic lack of oxygen. Such work might point the way to future gene therapy, since PH patients don't appear to make enough prostacyclin.

In a multi-center study not yet complete, PH doctors were finding that epoprostenol appeared to benefit not only PPH patients, but also those with PAH secondary to another disease (APAH), such as a connective tissue disease or congenital heart disease.

A joint Scientific Advisory Board and PHA Board meeting was held at the June conference. Topics discussed were: the *Patient's Survival Guide*, the website, the name change from UPAPH to PHA and bylaw revisions, and PHA's list of doctors treating PH patients. The SAB suggested that PHA not get into recommending doctors because that would be a credentialing process best done by other organizations. It was decided to keep publishing names, however, along with a disclaimer saying that the names on the list were self-reported and that PHA was not endorsing any doctors or treatment centers.

PH physicians expressed interest in forming a consortium. The PHA Board was willing to provide financial support, if necessary. (It took a long time for this idea to become reality, but Dr. Dave Badesch finally submitted a proposal for the SLC Consortium to NIH in 2005.) The NHLBI was spending \$10.8 million annually for PH research.

Board Activities in 1998

The officers in 1998 were:

President	Bonnie Dukart
Vice President, Administration.....	David Gunn
Vice President, Advocacy.....	Ed Simpson
Vice President, Education.....	Michael Szczepkowski
Vice President, Support.....	Judy Simpson, RN, Ed.S.
Secretary.....	Gary Dukart,MD
Treasurer.....	Jerry Paton
Assistant Treasurers.....	..Carol Wilson. Pat Paton

Directors-at-Large were:

Bruce Brundage, MD	Linda Carr
Pat Gunn	Dorothy Olson
Jan Travioli	Kathy Windhorn

The terms of officers were one year, but it was hoped that persons would remain in their position for two years.

At the start of 1998, PHA's savings account balance was \$55,000. At the end of the year (after a successful international conference) PHA was still nearly \$18,000 ahead. Brad Belliston arranged with major credit card companies for PHA to begin accepting credit card payments and donations. There would be small monthly charges for this ability.

Board members agreed that the rotation of volunteers answering the Patient-to-Patient Helpline would be two weeks instead of one month. The line was averaging 60-80 calls a month.

Future goals were set for PHA including advocacy, public relations, organ-donor awareness and transplantation, and working with members of Congress for the benefit of PH patients. The Board again discussed the need to pay more attention to PH secondary to other diseases, and to reach out more to such patients.

The PHA organizational chart was updated. The final product had 33 boxes and assigned each volunteer project to a particular officer. It looked impressive hung on a wall.

On the advice of attorneys, the corporate structure of PHA had to be revised to be consistent with the growing (1,000+ members) organization. A second parallel corporation under PHA, Inc., handled by attorney/PHA member Austin Carr, would have new articles of incorporation, bylaws, and a tax exemption status as a 501(c)(3). Operations under UPAPH would be phased over into PHA, Inc. This switchover was completed by mid-year, but UPAPH continued to exist as sort of a ghost organization (with its own Board of Directors elected as well as the Board of Trustees for PHA) until PHA received the official tax determination letter the following year. PHA remained incorporated in the state of Florida.

PHA's mailing address was changed from Speedway, Indiana, to P.O. Box 463, Ambler, Pennsylvania 19002, to be near the home of president Bonnie Dukart.

In a move that was to have a profound impact on PHA, Ed Simpson, chair of the search committee for a part-time executive director, wrote a job description and placed the ad in *The Chronicle of Philanthropy* and *The NonProfit Times*. Plans were for this person to begin work in January 1999 as a part-time (20 hours weekly) employee. PHA's first real office would be located for the convenience of the new executive director. Ed conducted telephone interviews with candidates who applied for the position. Three finalists were selected, and they were interviewed by Ed, Bonnie Dukart, and Michael Szczepkowski.

By the fall of 1998, PHA membership had climbed to 1,416.

Snapshot of Committee Chairs in 1998

Caregivers Committee.....	Dena Giddens, then Armond Aserinsky
Creative Writing (<i>Persistent Voices</i>)	Sue Kelly
Children with PH/PPH	Linda Carr
Librarian	Debra Cane, then Kathy Windhorn
Mail & Donations	Julie Hendry and Lenna West, then Glenda Ellis, Selma Smith, and Lenna West
Membership Statistics	Sally Maddox
Organ Donor Awareness	Sharon Yamron
Organizational Liasons	
NORD Board of Directors	Ed Simpson, Ed.D.
NIH Advisory Board on Heart, Lung, & Blood	Judy Simpson, RN, Ed.S.
<i>Pathlight</i> Editors	Jan Travioli, Gary Dukart and Victor Tapson (Medical Editor)
Public Affairs.....	Shirley Craig, then Maribeth McCarthy
Volunteer Coordinator	Kathy Szczepkowski
Web Site Coordinators.....	Carol and Armond Aserinsky
Web Site Guest Book Coordinator	Kathy Levitt, then Shirley Craig, then Candy Bleifer
800 Number Coordinator	Kathy Windhorn
Year 2000 PHA Conference Coordinator	Pat Gunn

1999

PHA HIRES AN EXECUTIVE DIRECTOR, & GOLF RAISES A LOT OF GREEN FOR RESEARCH

The year began with a bang, with Rino Aldrighetti reporting for work as PHA's part-time Executive Director on January 1. Also in January, PHA added support for medical research to its mission, and Jack Stibbs began an incredibly successful fund-raising campaign. In early March 1999, a conflict-of-interest issue arose over a Board member, and led to several resignations.

Rino Aldrighetti Takes the Helm



Rino Aldrighetti had previously been executive director of the National Family Farm Coalition, executive secretary of the National Volunteer Organizations Active in Disaster (an umbrella organization with 31 national member organizations and a multitude of state, county, and municipal chapters), development director for the National Council for Adoption, and associate director of development and public relations for the National Shrine of Immaculate Conception (the largest Catholic church in the Western Hemisphere). As a part of such work he had learned how to raise considerable sums of money, deal with the press, and write news stories and proposals. In addition to this experience with nonprofit groups, he had been active in his community of Takoma Park, Maryland, as a grass-roots activist in matters such as public safety.

A tall man with a commanding, but friendly, presence, Rino would encourage PHA to take some risks, risks that paid off and resulted in a rapid expansion of the organization's budget and usefulness. (Later, Rino's title would change to President and he would work not 20 hours a week, but more than full time.) Rino was instructed to look for office space in Silver Spring, Maryland, close to his home in Takoma Park. Once the office was established, he hired a part-time office worker, Dorothy Bradley.

At the January PHA Board Meeting Major Fundraising Events Are Planned

Rino was introduced to the PHA Board at the January meeting, which was held in Orlando, FL. President Bonnie Dukart opened the meeting and reported a 20 percent increase in membership, and that PHA was now accepting money for research that would be held in a separate research account. She suggested it was time for PHA to get involved in raising funds for research. Jack Stibbs, a lawyer living in Woodlands, TX, offered to chair the major fundraising events. (Jack and Marcia Stibbs are parents of a girl with PH.) Donna Gildea had previously agreed to help with other fundraisers.



**Attorney and
"PH Dad"
Jack Stibbs**

This new emphasis raised over \$100,000 during the first year of activity. The first Houston golf tournament that Jack held for PHA on April 22 in Houston netted \$86,000 for the PHA Research Fund. Smaller efforts were also successful, such as the Chicago support group's Walk for a Cure in September, during PH Awareness Week. As of September 1999 PHA had \$98,000 in its restricted research fund. (During the year 2000, a total of \$450,000 was raised for this priority and the research effort began to take on greater shape and definition. Over the years, the golf tournaments Jack organized would rise *over a million dollars* for research on PH!)

When the slate of new officers was approved during the meeting, Michael Szczepkowski, a cognitive engineer with expertise in knowledge engineering and user-interface design, assumed the chairmanship/presidency. Snowbound, Michael was attending the meeting by phone. He laid out his vision for PHA, which included a need to reach out more aggressively to those with PH secondary to other diseases, and to identify physicians who worked with such patients and to draw both them and their patients into PHA.

The Patient-to-Patient Helpline was then taking 80-90 calls a month, the Board learned, and Armond Aserinski reported, also by phone, that the PHA Internet Committee had 13 members and that over 6,000 people from 35 countries had visited PHA's website.

There was a discussion about similarities and differences of "advocacy" and "education." For the present, it was decided, advocacy should include legislation to help people buy health insurance, to encourage Medicare to pay for more than two years of anti-rejection drugs following an organ transplant, and to encourage NIH to continue to fund PH research.

Bruce Brundage raised the question of whether there should be a formal scientific session for physicians at the 2000 conference, and whether a physicians' group should be established as an adjunct to PHA. The SAB was split over the wisdom of the first idea, but a majority of the SAB supported the latter idea, and a subcommittee of the Board was formed to look into it. Dr. Brundage also noted that the diagnostic classifications of PPH were changing and that the term "PPH" may need to be scrapped (see the discussion of the World Symposium on PPH in Evian, which is in the "On the Medical Front" section in the 1998 chapter).

Bonnie Dukart was thanked by all for the exceptionally good job she had done as President of PHA. She took PHA to a new, more professional

level. (After retiring as President, Bonnie's PAH worsened. She died in January 2001, shortly after undergoing a lung transplant.)

Schism

On March 3, 1999, Michael Szczepkowski tendered his resignation as president of the PHA Board because of a perceived conflict of interest concerning Board Member Gary Dukart, MD. The conflict surfaced when an anonymous person posted a message on PHA's website message board accusing Gary, an employee of Wyeth/Ayerst, of having a special interest in the production of the diet pill Redux. (American Home Products, the maker of Redux, changed its name to Wyeth in 2002.) Another allegation was that PHA had dragged its organizational feet in getting out the word on the dangers of dexfenfluramine-containing diet pills (the implication was that Gary and Bonnie Dukart should have known that Redux could cause PAH and should have warned the PH community of this in a timelier manner).

An emergency PHA Board meeting conference call was held March 5. On the call were Rino Aldrighetti, Ed and Judy Simpson, Pat and Jerry Paton, Kathy Windhorn, Lenna West, Jan Travioli, Dorothy and Harry Olson, Carol Wilson, David and Pat Gunn, Dr. Bruce Brundage, Jack Stibbs, Bonnie and Dr. Gary Dukart, Linda Carr, Carol Aserinsky, Sally Maddox, Barbara Smith, and Lora Hindsley.

The Board unanimously accepted Mike's resignation as president. (On March 9, Ed Simpson was selected as the replacement interim president; his term would end the following January.) Later in the meeting, it was sensibly suggested that PHA should have a vice president (or president-elect) in line and ready to take the president's place should a need again arise.

Gary offered to resign his seat on the Board if the Board so desired. Because Board members believed Gary was faultless, they did not accept his resignation. Nevertheless, a fact-finding committee was set up to investigate the charges. Ed Simpson chaired the committee, which included

Dr. Bruce Brundage (a physician), Jack Stibbs (an attorney), and non-Board member Maribeth McCarthy. (Maribeth was Support Group Chair and later Public Relations Chair.) Carol Aserinsky suggested that an outside evaluator (someone not on the Board) also be appointed, but no action was taken on this suggestion.

The committee's report, issued in April, would find that Gary Dukart was not involved with clinical studies or marketing plans involving Redux while he worked for Wyeth, nor was he ever involved in any work with fenfluramine. He had always been upfront in his relations with PHA and its SAB about what could be perceived as a conflict of interest and neither Gary nor Bonnie made any attempt to influence anyone about the safety of diet pills. "Both of the Dukarts provided leadership in getting the SAB to assist in preparing an official position statement as quickly as possible without putting the organization at risk of liability," the committee concluded. In January 1996, attorneys and the SAB had warned PHA that it must not go too far out on a limb before clear scientific evidence was published. But the Report noted how PHA had nevertheless issued very early warnings of the probable danger of the drug.

Rino urged the Board to continue with its work, vision, and goals so that PHA could move forward. He charged the Board to 1) find trust in each other, 2) to come together to find common ground on which to meet and work for PHA, and 3) to put our faith in the process, not in personalities. Each Board member had the opportunity to express his or her thoughts on recent happenings.

Message Board policies were discussed. It was decided to have a disclaimer added to the PHA website message board to safeguard against liability for incorrect medical information, because messages were not removed from the message board unless they had nothing to do with PH. It

was further agreed that PHA should look at the policies of other message boards, and develop a plan for how to handle highly charged issues.

Conflict-of-interest issues were discussed. It was generally acknowledged that PHA needed to develop a conflict-of-interest statement for Board members to fill out and sign.

The "Fact-Finding Report of Alleged Conflict of Interest and Diet Drug Information Suppression" was presented to the Board on April 1. The Report, 41 pages in length, "indicated there was no conflict of interest on the part of any PHA Trustee and that PHA in fact did inform subscribers of PHA about diet drugs and their possible association with PPH." Gary Dukart was not involved with clinical studies or marketing plans involving Redux while he worked for Wyeth, the Report found, nor was he ever involved in any work with fenfluramine. He had always been upfront in his relations with PHA and its SAB about what could be perceived as a conflict of interest. Furthermore, neither Gary nor Bonnie had made any attempt to influence anyone about the safety of diet pills. "Both of the Dukarts provided leadership in getting the SAB to assist in preparing an official position statement as quickly as possible without putting the organization at risk of liability," the committee concluded.

With regard to PHA's actions as an organization, the Report said: "Rather than suppressing information about Redux, as alleged, PHA published several articles, documents, references and an official statement warning about the use of Redux and incidents of pulmonary hypertension as early as 1996." Any PHA member wanting a copy of the Report could get one from the central office.

Armond Aserinsky was not satisfied with the findings and on April 4 he resigned as Chair of the Internet Committee. His wife, Carol Aserinsky (who had developed PH after using Redux) resigned as a member of the Board of Trustees. Armond and Michael would start their own PH website, called

PHCentral, which began operating midyear and would become a fine Internet resource for PH patients, caregivers, and medical professionals. In August, PHA recreated its own website and message board with a new server.

In summary, PHA learned a lot from the painful schism experience. Most importantly, it was clear that greater attention must be paid to potential conflicts of interest, and that formal conflict-of-interest statements should be sought from Board members and established procedures followed when a conflict existed. (It is common for charitable board to have such conflicts; this is why bylaws contain procedures for dealing with them.) PHA also rethought its website policies, and recognized the need to have a president-elect ready to step in and fill a vacant slot.

June Board Meeting

At the June PHA Board meeting, held at the Park Hotel in Charlotte, North Carolina, the new conflict-of-interest statement was read:

I agree to absent myself, not take part in the discussion, or vote on any measure covered in the PHA Board meeting June 11-12, 1999, in which I, or my spouse has a financial involvement or is associated through employment. I also agree to hold in confidence the proceedings of this Board meeting, other than information intended for the public record by the Board.

The statement was approved and signed by all Board members. Ever since, it has been PHA policy to have Board members regularly sign similar statements.

The "Fact-finding Report on Alleged Conflict of Interest and Diet Drug Information Suppression" was approved by all Board members except Bonnie Dukart, who abstained. (Gary Dukart, who had resigned despite the Board's vote of confidence, was not present at the meeting.)

The following trivia were revealed at the June meeting: 600 thank-you notes were sent during the period from January to May, and 932 Patient-to-Patient Helpline calls were answered in the same period (that worked out to 186+/month or 6+/day). As of June 20, 1999, PHA had 1,613 patient/family

members, 210 medical professional members, and 14 organizational members, making a grand total of 1,837 members.

The Board learned that banking for PHA Support Groups must go through the PHA treasury to protect the groups' 501(C)(3) status (as well as PHA's).

By now, Board members no longer had to pay their own way to meetings, although some donated the cost of their airfare, hotel, etc.

September Board Meeting

In September, the Board met via a conference call. Rino now had two part-time employees, central office expenses were expected to be \$52,000, and membership stood at 1,837. There were 1,613 patient/family members, 210 medical professional members, and 14 organization members. Hearing all this, call participants said, "WOW!"

At Jack Stibbs suggestion, a Committee of Ten was launched. Persons who gave \$1,000 would be named as members of the Committee of Ten. (This "committee" has proved to be oversubscribed every year.) Another fundraiser, "Art From the Heart" notes and holiday cards designed by talented children from PH families, were offered for sale. They also proved popular, although it took a long time to pay off the printing costs.

Board Officers and Trustees at the End of 1999

(The alert reader will note that in 1999 Board members began calling themselves Trustees instead of Directors.)

President.....	Ed Simpson
Executive Director.....	Rino Aldrighetti (ex officio member)
Vice President.....	Judy Simpson
Admin. Vice President, Advocacy.....	Jack Stibbs
Admin. Vice President, Education.....	Sally Maddox
Admin. Vice President, Fund Development.....	Bonnie Dukart
Admin. Vice President, Support.....	Lora Hindsley
Secretary.....	Pat Gunn
Treasurer.....	Jerry Paton
Assistant Treasurers.....	Carol Wilson, Pat Paton

Trustees-at-Large:

Bruce Brundage, MD
Dorothy Olson
Jan Travioli
Kathy Windhorn

Linda Carr
Barbara Smith
Lenna West

Snapshot of Committee Chairs in 1999

Creative Writing (*Persistent Voices*) Sue Kelly
Children with PH/PPH Linda Carr
Librarian Kathy Windhorn
Mail & Donations Selma Smith, and Lenna West
Membership Statistics Sally Maddox
New Member Packets Brad Belliston
Organ Donor Awareness Sharon Yamron
Organizational Liasons
 NORD Board of Directors Ed Simpson, Ed.D.
 NIH Advisory Board on Heart, Lung, & Blood ... Judy Simpson, RN, Ed.S.
Pathlight Jan Travioli, Editor;
 Shirley Craig, Staff Writer;
 Victor Tapson, Medical Editor
Public Affairs Maribeth McCarthy
State Coordinator Marcie Long
Web Site Coordinator Armond Aserinsky
Web Site Guest Book Coordinator Candy Bleifer
800 Number Coordinator Kathy Windhorn
Year 2000 PHA Conference Coordinator Pat Gunn

Reaching Out to Patients

California PH support groups held a PH Forum in San Diego June 4-6th, 1999, that over 85 persons attended. Dr. Bruce Brundage, Dr. Ronald Oudiz, and Dr. Richard Channick were among the speakers.

Plans were made for the fourth International PHA Conference, to be held June 23-25, 2000, at the Wyndham Hotel in Chicago. Pat Gunn would again take on the mammoth job of being Chairman of the Conference Committee.

In 1999, with the help of her brother (also a PPH patient), husband, and parents, JoAnne Sperando-Schmidt formed the Long Island, NY, PHA support group, which quickly grew to become one of PHA's largest support groups. JoAnne was the third person in her family to be diagnosed with PPH.

(She began serving on the PHA Board in 2001 and has since contributed to PHA in myriad ways.)

PH Awareness Week '99

Awareness Week was moved to a more "climate friendly" time of year, and was held from September 18 through 26 (to include two weekends). There were several news stories featuring PH patients, and at least one new support group was formed. The Chicago group held a BBQ and picnic in a park that 75 people attended. Jim Wilson coordinated the first Dallas/Ft. Worth "Cure PH" golf tournament , which raised \$15,000. The fall issue of *Pathlight* focused on these and many other support group activities.

On the Research/Medical Front

PH Resource Network (PHRN) finally became an official branch of PHA. The objective of PHRN is to build a network among non-MD medical professionals who work with PH patients. (Membership has grown steadily, and is expected to exceed 250 by the end of 2005.)

PH medicine developments. In 1999, the Food and Drug Administration approved nitric oxide, a gas produced by the body that can relax constricted blood vessels, for use in full-term babies with persistent pulmonary hypertension of the newborn, a life-threatening disease.

In the summer of 1999, treprostinil (Remodulin, a.k.a. UT-15) Phase III clinical trials were announced. Treprostinil is a chemically stable "cousin" of epoprostenol that does not have to be mixed each day or kept cold. It can be delivered by a smaller pump than epoprostenol/Flolan requires and is pumped into fat under the skin (usually on one's tummy). A total of 448 patients would be enrolled in the treprostinil trials at 40 major medical centers.

TheraCom became a provider of epoprostenol/Flolan.

Robyn J. Barst, MD, et al. reviewed 13 years of experience with vasodilator therapy for PPH in children in *Circulation*. The authors concluded that such therapy improves survival in children at least as well as (and perhaps better than) it does in adults, and that categorizing children as “acute responders” (to NO, for example) and “nonresponders” helps define treatment and survival.

Sean Gaine, MD, PhD (Mater Misericordiae Hospital in Dublin, Ireland) became author of the “Research Corner” section of *Pathlight*. He reported on the encouraging results when epoprostenol was used on patients with diseases other than PPH, such as portopulmonary hypertension and PH secondary to connective tissue diseases.

Erika Berman Rosenzweig, MD, et al. published results of a study treating patients with PH due to congenital heart defects with long-term use of epoprostenol (Flolan). Despite the lack of a quick response to the drug, the patients slowly improved over the course of a year. This study was important in helping such patients get insurance coverage.

Strong Relations with NHLBI Are Maintained

The National Heart, Lung and Blood Institute (NHLBI) is the division within the National Institutes of Health that relates most directly to pulmonary hypertension. In FY 1998 and 1999 PHA focused on making the listing of pulmonary hypertension a priority of the NHLBI. The NHLBI spent \$12 million for PH research and training in 1999.

Also in 1999, the NHLBI invited representatives from 68 patient interest organizations dealing with heart, lung, blood, or sleep disorders to attend a meeting in Bethesda. Judy Simpson helped organize this meeting, and she and Rino Aldrighetti represented PHA at the meeting. The representatives learned about NHLBI services, told the NHLBI of their needs, and attendees discussed ways to coordinate their activities in areas of common interest.

The meeting has become an annual event, and PHA always sends representatives.

With support from the office of Congressman Kevin P. Brady (R - Texas), PHA was given the opportunity to meet with Dr. Claude Lenfant, NHLBI. So in July, in Bethesda, MD, Bonnie Dukart, Dr. Bruce Brundage, Jack Stibbs, Rino Aldrighetti, and Judy Simpson met with both Dr. Lenfant and Dr. Carol Vreim. NHLBI proposed that PHA enter into a memorandum of understanding with NHLBI to explore the possibilities of joint projects, including research funding. (PHA and NHLBI eventually formalized an agreement that established a partnership for joint funding of an annual career development award program in PH.)

Scientific Advisory Board Members in 1999

Chairman: Bruce H. Brundage, MD (UCLA School of Medicine)
(Names of new members, below, are in boldface type.)

David Badesch, MD (University of Colorado, Denver, CO)
Robyn J. Barst, MD (Columbia/Presbyterian Babies Hospital, New York, NY)
Joy Beckman, RN, MSN (University of California-Harborside, Torrance, CA)
C. Gregory Elliott, MD (LDS Hospital, University of Utah, Salt Lake City, UT)
Alfred P. Fishman, MD (University of Pennsylvania Medical Center, Philadelphia, PA)
Adaani Frost, MD (Baylor College of Medicine, Houston, TX)
Sean Gaine, MD (Johns Hopkins University Medical Center, Baltimore, MD)
Abby Krichman, RRT (Duke University Medical Center, Durham, NC)
David Langleben, MD (Jewish General Hospital, Montreal, Quebec, Canada)
James E. Loyd, MD (Vanderbilt University, Nashville, TN)
Michael D. McGoon, MD (Mayo Clinic, Rochester, MN)
John H. Newman, MD (Nashville VA Hospital, Nashville, TN)
Harold Palevsky, MD (University of Pennsylvania Medical Center, Philadelphia, PA)
Stuart Rich, MD (Rush Heart Institute, Chicago, IL)
Ivan Robbins, MD (Vanderbilt University, Nashville, TN)
Lewis Rubin, MD (University of California, San Diego, CA)
Cathy J. Severson, RN (Mayo Clinic, Rochester, MN)
Carmen Skurdal, RN, BSN (Olsten Health Services, Edgewater, MD)
Victor E. Tapson, MD (Duke University Medical Center, Durham, NC)
Carol E. Vreim, PhD (National Institutes of Health, Bethesda, MD)

2000 AND BEYOND

PHA has continued its dramatic growth in effectiveness and size. A great deal of more recent information can be found on PHA's website, including old issues of *Pathlight* and of our medical journal, *Advances in Pulmonary Hypertension*, as well as annual reports prepared by the PHA President or Chair of the Board.