The Pulmonary Fibrosis Foundation presents

IPF SUMMIT 2011
FROM BENCH TO BEDSIDE

DECEMBER 3, 2011
7:00 AM–3:05 PM

Marriott Magnificent Mile
Chicago, Illinois

Patients and Caregivers
Dear Friends of the Foundation,

Welcome to the IPF Summit 2011: From Bench to Bedside. This is our inaugural scientific healthcare conference where we are bringing together some of the best and the brightest in the PF community. It is our hope that fostering this type of collaboration will lead to improved diagnosis, better patient care, and help stimulate the research needed to find better treatments and ultimately a cure for IPF. We are pleased to be able to provide sessions for both the professional and patient/caregiver populations. Patient support and education is an important component of our mission, and in order to supply as many patients, caregivers, and family members with this information, we are providing a live webcast for the patient/caregiver sessions.

As many of you know, I have a personal connection to IPF. The Foundation was the brainchild of my father Albert Rose and his brother Mike Rosenzweig, who had been both recently diagnosed with IPF when the Foundation was started. Their sister Claire had died from pulmonary fibrosis a few years earlier, and they both felt a profound need to help find a cure for this devastating disease. My father died in early 2002, but his brother continued to work untiringly and passionately to build the Foundation, fund research, and create a financially viable entity. Over the past decade the Foundation has also significantly expanded its support network and has become a beacon for those afflicted with this deadly disease. I am extremely honored to carry on their legacy as President and Chief Executive Officer for the Foundation.

This Summit is a milestone for the Foundation as we celebrate our tenth anniversary. Many people have worked hard for the past two years to make the Summit come to fruition. I would like to thank the Foundation’s staff and our partners, The France Foundation and National Jewish Health, for what was truly a team effort in making this event a success. Of course, many thanks must go to our “all star” Faculty for sharing with us their knowledge and expertise. Lastly, I would like to thank our generous sponsors, both individual and corporate, for their generosity.

I have been doing quite a bit of traveling during the last few months attending some medical conferences, participating in fundraising events, and engaging in advocacy activities for the Research Enhancement Act. A couple of things have greatly impressed me. First, there is considerable interest in drug development for IPF. The research community is working hard to discover new and successful agents. There are a number of exciting therapies that are in early development while others are working their way through the clinical trial process. Parenthetically, it is critically important for the patient community to participate in clinical trials. This is the only way we can develop new, effective treatments. Lastly, I am continually impressed and motivated by the courage and commitment of the patients and their family members. They are a constant source of inspiration for all of us that are associated with the Foundation, and I assure everyone that we will work tirelessly and passionately to help find a cure for this illness!

Warmest Regards,

Daniel M. Rose, MD
President and Chief Executive Officer
Dear Patients and Caregivers,

It is a true pleasure to welcome you to the *IPF Summit 2011: From Bench to Bedside*. Despite two decades of progress, our understanding of the pathobiology and more importantly our ability to treat idiopathic pulmonary fibrosis (IPF) remains woefully inadequate. Because of this, we felt it important to have a focused, innovative conference to help expand our knowledge and to encourage the development of new treatment options. It is obvious that no one person can answer the big questions in IPF, that is why so many physicians, researchers, patients, family members, and industry representatives are here today; to learn, to collaborate, and to share information.

The *IPF Summit* has been organized by the Pulmonary Fibrosis Foundation in order to provide the most up-to-date material to the medical and research communities, as well as to be a source of information and support for those affected by this disease. This international conference includes a faculty of distinguished experts in the field of pulmonary fibrosis who have put together an outstanding program. While we will not answer all of the questions, here is an opportunity to bring talented and dedicated people together to work towards a single goal: making a difference to those who suffer from IPF.

I am honored and excited to be the Program Chair for the *IPF Summit 2011*, and thank each of you for your attendance and for your willingness to share your experience, knowledge, and expertise. Your engagement and input throughout the *Summit* will help shape the next decade of pulmonary fibrosis research, and with our collective efforts we can improve the future of those affected by this terrible disease.

Sincerely,

Kevin K. Brown, MD  
Professor and Vice Chair, Department of Medicine  
National Jewish Health

Pulmonary Fibrosis Foundation’s Medical Advisory Board, Chair  
*IPF Summit 2011: From Bench to Bedside*, Program Chair
Table of Contents

About the Pulmonary Fibrosis Foundation ................................................................. 1

PFF Research Fund to Cure Pulmonary Fibrosis ......................................................... 6

Meeting Information and Procedures ................................................................. 7

IPF Summit 2013 ........................................................................................................... 9

New Decade, New Reach Tenth Anniversary Dinner ............................................. 10

Agenda .......................................................................................................................... 11

Navigating the Summit: Meeting Space Map .......................................................... 13

Program Faculty ......................................................................................................... 14

Pulmonary Fibrosis Foundation Sponsors .............................................................. 15

Exhibitors ..................................................................................................................... 16

Clinical Session Overview ....................................................................................... 18

Faculty Slides .............................................................................................................. 19
About the Pulmonary Fibrosis Foundation

Mission
The mission of the Pulmonary Fibrosis Foundation (PFF) is to help find a cure for idiopathic pulmonary fibrosis (IPF); advocate for the pulmonary fibrosis community both locally and in Washington, DC; promote disease awareness; and provide a compassionate environment for patients and their families.

History
The Pulmonary Fibrosis Foundation is a 501(c)(3) nonprofit organization that was founded in 2000 by two brothers, Albert Rose and Michael Rosenzweig, PhD. Their sister Claire had died from idiopathic pulmonary fibrosis (IPF), and the brothers were both later diagnosed with the disease. Their vision shaped the PFF to become a leader in the IPF community for research, advocacy, awareness, and support. In February of 2002, sad to say, Albert Rose succumbed to the disease.

Dr. Rosenzweig was the Foundation’s first President and Chief Executive Officer. He worked tirelessly and passionately to build the Foundation, fund research, and create a financially viable entity. He also helped recruit an outstanding Medical Advisory Board, which has provided keen insight and direction.

Unfortunately, due to the progression of his disease, Dr. Rosenzweig retired as President and CEO in March 2009. Daniel M. Rose, MD, the son of Albert Rose and chairman of the Board of Directors, then assumed the positions of President and CEO. Dr. Rose had previously been a practicing cardiothoracic surgeon and Chief of Cardiothoracic Surgery at St. Vincent's Medical Center in Bridgeport, Connecticut, for 19 years. Having had three relatives afflicted with IPF, he brings to the Foundation a family member’s passion and motivation, along with a broad medical background and a profound desire to lead the PFF into its second decade.

Under Dr. Rose’s guidance the PFF has embarked on several ambitious initiatives to:

- Increase funding for IPF research and assist in creating partnerships between the academic research community and the biopharma industry.
- Foster the sharing of information and ideas in the clinical community through the creation of a biennial IPF Summit, beginning in 2011.
- Help create a national pulmonary fibrosis registry.
- Sponsor a series of webinars to more efficiently bring the latest information and research to patients and families.
- Establish a National Affiliate Program, which will increase patient outreach, enhance advocacy, expand disease awareness, and augment our fundraising.
- Continue our commitment to strongly advocate for the IPF community locally and in Washington, DC.
- Aggressively pursue increased public awareness through a series of public service announcements (PSA), social networking, and traditional media exposure.
Board of Directors

Daniel M. Rose, MD*
President and Chief Executive Officer

Joseph Borus, Esq*
Secretary

Thomas E. Hales*
Treasurer

Patti Tuomey, EdD
Chief Operating Officer

*Denotes Executive Committee

Pulmonary Fibrosis Foundation Team

Daniel M. Rose, MD
President and Chief Executive Officer

Patti Tuomey, EdD
Chief Operating Officer

Dolly Kervitsky, CRT, CCRC
Vice President
Patient Relations

Scott Staszak
Associate Vice President
Finance and Information Technology

Cara Schillinger
Associate Vice President
Communications and Marketing

Jennifer Bulandr

Amy Butler

Lyla Conrad

Peter Cremer

Matt Derda

Wendy Escobar

Courtney Firak

Mary Lou Ibadlit

Meredith Mann

Jennifer Mefford

Amanda Miller

Michelle Miller

Susan Murphy

Elizabeth Price

Francisco Rosas

Stephanie Seweryn

Leanne Storch

www.pulmonaryfibrosis.org
Medical Advisory Board

*Indicates Member of the Research Advisory Committee

Kevin K. Brown, MD*
CHAIRMAN
Vice Chair, Department of Medicine
Professor of Medicine
National Jewish Health
Denver, Colorado

David W. Kamp, MD
MEDICAL DIRECTOR
Professor of Medicine
Associate Chief, Pulmonary and Critical Care Division
Northwestern University Medical School
Chicago, Illinois

Jesse Roman, MD*
CHAIRMAN, RESEARCH ADVISORY COMMITTEE
Professor of Medicine
Chairman, Department of Medicine
Pulmonary, Critical Care, and Sleep Disorders Medicine
University of Louisville
Louisville, Kentucky

Marvin I. Schwarz, MD
PAST-CHAIRMAN
Professor of Medicine Division Co-Head
University of Colorado
Denver, Colorado

Timothy S. Blackwell, MD*
Professor of Medicine
Professor of Cancer Biology
Ralph and Lulu Owen Chair in Medicine
Professor of Cell and Developmental Biology
Vanderbilt University Medical Center
Nashville, Tennessee

Jeffrey T. Chapman, MD
Chair, Quality and Patient Safety
Cleveland Clinic Abu Dhabi
Abu Dhabi, United Arab Emirates

Harold R. Collard, MD
Associate Professor of Medicine
Division of Pulmonary and Critical Care Medicine
University of California, San Francisco
San Francisco, California

Rany Condos, MD
Associate Professor of Medicine
School of Medicine at New Bellevue Chest Group
New York University
New York City, New York

Gregory P. Cosgrove, MD
Associate Professor of Medicine
Department of Medicine
Co-Director, Interstitial and Autoimmune Lung Disease Program
National Jewish Health
University of Colorado
Denver, Colorado

Roland M. duBois, MD
Emeritus Professor of Respiratory Medicine
National Heart and Lung Institute
Imperial College
University of London
London, United Kingdom

Christine Kim Garcia, MD, PhD
Associate Professor of Medicine
Mcdermott Center for Human Genetics
Department of Internal Medicine, Division of Pulmonary and Critical Care Medicine
University of Texas Southwestern Medical Center
Dallas, Texas

Susan S. Jacobs, RN, MS
Center for Interstitial Lung Disease
Pulmonary and Critical Care Medicine
Stanford University Medical Center
Stanford, California

Naftali Kaminski, MD*
Professor of Medicine, Pathology, Human Genetics and Computational Biology
Director, Dorothy P. and Richard P. Simmons Center for Interstitial Lung Disease
Director, Lung, Blood and Vascular Center for Genomic Medicine
Pittsburgh, Pennsylvania

www.ipfsummit.org
Medical Advisory Board
*Indicates Member of the Research Advisory Committee

Joseph Lasky, MD*
Professor of Medicine
Section of Pulmonary Diseases
Co-Director, Interstitial Lung Disease Clinic
TCC Program Member
Tulane University
New Orleans, Louisiana

Andrew H. Limper, MD*
Professor of Medicine
Chair, Pulmonary and Critical Care Medicine
Mayo Clinic
Rochester, Minnesota

Kathleen Lindell, PhD, RN
Clinical Nurse Specialist
Director, Quality of Life Program
Division of Pulmonary, Allergy, and Critical Care Medicine
Dorothy P. and Richard P. Simmons Center for Interstitial Lung Disease
University of Pittsburgh
Pittsburgh, Pennsylvania

James E. Loyd, MD*
Professor of Medicine
Director, Familial Primary Pulmonary Arterial Hypertension Registry
Director, Familial Idiopathic Pulmonary Fibrosis Registry
Vanderbilt University Medical Center
Nashville, Tennessee

David Lynch, MD
Professor of Radiology
Co-Director, Division of Radiology
National Jewish Health
Denver, Colorado

Fernando J. Martinez, MD, MS*
Professor, Department of Internal Medicine
Director, Pulmonary Diagnostic Services
University of Michigan
Ann Arbor, Michigan

Imre Noth, MD
Associate Professor of Medicine
Director, Interstitial Lung Disease Program
University of Chicago
Chicago, Illinois

Ralph Panos, MD
Professor of Medicine
VA Pulmonary Section Chief
Associate Clinical Director Pulmonary, Critical Care and Sleep Medicine
University of Cincinnati
Cincinnati, Ohio

Ganesh Raghu, MD
Professor Laboratory Medicine
Division of Pulmonary and Critical Care Medicine
Director, Interstitial Lung Disease / Sarcoid / Pulmonary Fibrosis Program
Medical Director, Lung Transplant Program
University of Washington Medical Center
Seattle, Washington

Glenn D. Rosen, MD
Associate Professor of Medicine
Division Co-Chief, Pulmonary and Critical Care Medicine
Director of theInterstitial Lung Disease Program
Stanford University School of Medicine
Stanford, California

Patricia J. Sime, MD*
Professor of Medicine (Pulmonary and Critical Care), Environmental Medicine, and Oncology
University of Rochester Medical Center
Rochester, New York

David A. Schwartz, MD*
Professor of Medicine
Chairman, Department of Medicine
University of Colorado Medical Center
Denver, Colorado

Moisés Eduardo Selman, MD
Director of Research
Mexican National Institute of Respiratory Diseases
Mexico City, DF Mexico
Medical Advisory Board

*Indicates Member of the Research Advisory Committee

Charlie Strange, MD
Professor of Pulmonary and Critical Care Medicine
Medical University of South Carolina
Charleston, South Carolina

Robert Strieter, MD*
Professor of Medicine
University of Virginia School of Medicine
Charlottesville, Virginia

Jeffrey J. Swigris, DO, MS
Associate Professor of Medicine
National Jewish Health
Denver, Colorado

Janet Talbert, MS, CGC
Certified Genetic Counselor
Director, Familial Pulmonary Fibrosis Genetic Counseling Program
National Jewish Health
Denver, Colorado
PFF Research Fund to Cure Pulmonary Fibrosis

The Pulmonary Fibrosis Foundation recently established the PFF Research Fund to Cure Pulmonary Fibrosis. The goals of the Fund are to support basic research, clinical research, and translational research to help identify new treatments and, ultimately, a cure for PF. In 2012, four recipients will receive awards totaling $200,000.

The PFF Research Advisory Committee, chaired by Jesse Roman, MD, will determine the grant recipients. Applications are scored based on their scientific merit, originality, and responsiveness to the specific purpose of each award category.

**Award Categories**

**Young Investigator Award:** An award of up to $50,000 to be given over a two-year period to encourage young investigators (individuals within five years of completion of their formal training) to maintain and enhance their interest in IPF research during the early stages of their academic career.

**Established Investigator Award:** An award of up to $50,000 to be given over a two-year period to established investigators to explore innovative areas of research that may not yet be eligible for an NIH (or similar) grant.

For details of each award category, including specific criteria for investigator eligibility and an annual calendar of deadlines, please visit www.pulmonaryfibrosis.org/research/PFFgrants.
Meeting Information and Procedures

**Name Badge**
Your name badge is your admittance to activities during the conference; it is color coded to denote the session and room in which you are registered to attend. Please wear your badge for the duration of the conference.

**General Session, Exhibit-Poster Hall, and Meals**
You must have a name badge to enter the sessions, Exhibit-Poster Hall, and to participate in meal functions. The Exhibit-Poster Hall is located in Halsted and is open during the following hours:

- **Thursday, December 1st:** 5:00−7:30 PM
- **Friday, December 2nd:** 7:00 AM−5:45 PM
- **Saturday, December 3rd:** 7:00 AM−2:30 PM

Continental breakfasts, lunch, and breaks are for meeting registrants only.

**Cell Phones and Pagers**
Please be courteous to fellow participants and turn your phones and pagers to silent during the lectures.

**“Together We Will Make a Difference in PF” Message Board and Community Map**
The Message Board is a place for conference attendees to leave inspirational and positive messages. We would like participants to connect and collaborate. The Community Map is a visual representation of how the PF community has assembled at the Summit. Mark your hometown with a color-coded pin that indicates your interest in PF. We want all of you to have an impact and “make a difference”! Both are located near the Registration Desk.

**Project PF Action Center**
Share your Summit experiences via social media and raise PF awareness, ask your representatives to support the PFREA with a few simple clicks, and participate in the Project PF action for the day.

**Oxygen Station**
Oxygen refills will be available during Summit hours on Friday, December 2 and Saturday, December 3 to patients with valid prescriptions and who have made an advanced request. Thank you to Lincare for supplying oxygen.

**Medical Emergencies**
If you are experiencing a medical emergency, please call 911. There is no physician or nurse on site who can legally see or care for a patient with a medical emergency.

**Photography, Filming, and Recording of IPF Summit 2011**
The IPF Summit 2011 will be photographed, videotaped, and/or recorded in its entirety by staff and third party vendors. The Patient session will be webcast live and on-demand. All sessions will be available post-conference on, but not limited to, the Pulmonary Fibrosis Foundation’s website. Crews will be videotaping and taking still photographs of all sessions, meals, and periphery Summit activities. Conference video, still photographs, and quotes may be used and/or repurposed in promotional materials for the PFF and future IPF Summits, including but not limited to the website, print materials, and social media. All attendees will be asked to sign a Release at registration. For those who do not wish to be filmed or photographed, please be sure to get a red name badge holder at registration for identification.
Evaluation Form
Your feedback is very important to us and will help us plan future Summits. Please remember to complete your evaluation form and return it to the registration desk.

Disclaimer
Recording of any session is strictly prohibited. The views of the speakers do not necessarily reflect the views of the presenting, partnering, or endorsing organizations. The Pulmonary Fibrosis Foundation, National Jewish Health, and The France Foundation present this information for educational purposes only. The content is provided solely by faculty who have been selected because of recognized expertise in their field. Participants have the responsibility to ensure that products are used appropriately on the basis of their healthcare provider’s clinical judgment and accepted standards of care. The Pulmonary Fibrosis Foundation, National Jewish Health, and The France Foundation assume no liability for the information herein.

If you have any questions or need assistance, please visit the Registration Desk.
IPF SUMMIT 2013
DECEMBER 5–7, 2013

We’ve set the date, now help us choose the location.

Cast your vote at the registration desk or email summit@pulmonaryfibrosis.org with the city of your choice in the subject line.

To receive information about IPF Summit 2013, or to be placed on the pre-registration list, please email summit@pulmonaryfibrosis.org or call 888-733-6741.

SAVE THE DATE
Don’t miss this inspiring event.

Please join us in commemorating the Pulmonary Fibrosis Foundation’s first ten years of research, advocacy, awareness, and support. As we cross our tenth anniversary, we are poised to conquer challenges, expand our reach, and touch the lives of all those affected by pulmonary fibrosis.

FRIDAY, DECEMBER 2, 2011

6:30pm RECEPTION
7:30pm DINNER
8:00pm PROGRAM

TICKETS $150 • BUSINESS OR COCKTAIL ATTIRE

EVENT CO-CHAIRS
Former Congressman Brian Baird, Ph.D.
Julie Halston, award-winning Broadway and TV actress and comedienne
Ralph Howard, anchor for the Howard Stern Show, Howard 100 News
Senator Mark Kirk

EVENT EMCEE
Nesita Kwan, reporter and anchor at NBC 5 Chicago News

To purchase tickets please visit the registration desk.

Proceeds will benefit the Pulmonary Fibrosis Foundation’s mission to help find a cure for idiopathic pulmonary fibrosis (IPF), advocate for the pulmonary fibrosis community both locally and in Washington, D.C., promote disease awareness, and provide a compassionate environment for patients and their families.

NEW DECADE
NEW REACH

Together we’ll make a difference in pulmonary fibrosis
Agenda

SATURDAY, DECEMBER 3, 2011

CLINICAL SESSIONS
PATIENTS AND CAREGIVERS
2011 Patient Update on Pulmonary Fibrosis

7:00–7:45 AM Registration and Continental Breakfast

7:45–8:00 AM Introduction
Daniel M. Rose, MD

8:00–8:30 AM What is Pulmonary Fibrosis and What are the Causes?
Gregory P. Cosgrove, MD

8:30–9:00 AM What are the Symptoms of Pulmonary Fibrosis and What are the Treatment Options?
Susan S. Jacobs, RN, MS

9:00–9:30 AM Lung Transplantation
Kenneth R. McCurry, MD

9:30–10:00 AM Familial Pulmonary Fibrosis and Genetic Counseling for Pulmonary Fibrosis
Janet Talbert, MS, CGC

10:00–10:15 AM Visit Exhibits and View Posters

10:15–10:45 AM Pulmonary Rehabilitation for Pulmonary Fibrosis
Brenda Crowe, CRT

10:45–11:10 AM Navigating Medicare and Health Insurance
Miranda G. James, Esq

11:10–11:40 AM The What, Why, and How of Clinical Trials
Gail G. Weinmann, MD

11:40 AM – NOON Advocacy
Former US Representative Brian Baird, MS, PhD and Mark Shreve

NOON – 12:15 PM Visit Exhibits and View Posters

12:15–1:15 PM LUNCH Roundtable Discussions with the Experts
Table 1: PF Specialist - Jeffrey J. Swigris, DO, MS
Table 2: PF Specialist - Gregory P. Cosgrove, MD
Table 3: Lung Transplant Specialists - Kenneth R. McCurry, MD
Table 4: Genetic Counseling - Janet Talbert, MS, CGC
Table 5: Advocacy - Former US Representative Brian Baird, MS, PhD and Mark Shreve
Table 6: Navigating Insurance - Miranda G. James, Esq
Visit Exhibits and View Posters

How to Establish a Support Group and the Benefits of Participation
Wendi R. Mason, MSN, ACNP-BC

Palliative and Hospice Care for Pulmonary Fibrosis Patients
Bronwyn Long, DNP, MBA, RN

Living with Pulmonary Fibrosis and How to Improve Quality of Life
Kathleen Lindell, PhD, RN

Session Wrap-up
Navigating the Summit: Meeting Space Map
SATURDAY, DECEMBER 3, 2011

Marriott Magnificent Mile, 4th Floor

Registration
Halsted (1)

Breakfast and Lunch
Waveland (2)

Breaks
Halsted (1)

Oxygen Refills
Waveland (2)

Speaker Ready (Faculty and Staff Only)
Grace (3)

Sessions for Physicians, RNs, Researchers, and Allied Health Professionals
Marriott Ballroom – Main Floor Seating (4)
Sheffield – Live Feed Seating (5)
Waveland – Open Seating (2)

Sessions for Patients and Caregivers
Avenue Ballroom – Main Floor Seating (6)
State – Live Feed Seating (7)

Exhibit Hall
Halsted (1)

Poster Hall
Halsted (1)
Program Faculty

Brian Baird, MS, PhD  
Former US Representative to Congress  
Vancouver, Washington

Gregory P. Cosgrove, MD  
Associate Professor of Medicine  
Department of Medicine  
Co-Director, Interstitial and Autoimmune Lung Disease Program  
National Jewish Health  
University of Colorado  
Denver, Colorado

Brenda Crowe, CRT  
Manager, Pulmonary Rehabilitation  
Exempla Lutheran Medical Center  
Wheat Ridge, Colorado

Susan S. Jacobs, RN, MS  
Center for Interstitial Lung Disease  
Pulmonary and Critical Care Medicine  
Stanford University Medical Center  
Stanford, California

Miranda G. James, Esq  
Caring Voice Coalition  
Mechanicsville, Virginia

Dolly Kervitsky, CRT, CCRC  
Vice President, Patient Relations  
Pulmonary Fibrosis Foundation  
Chicago, Illinois

Kathleen Lindell, PhD, RN  
Clinical Nurse Specialist  
Director, Quality of Life Program  
Division of Pulmonary, Allergy, and Critical Care Medicine  
Dorothy P. and Richard P. Simmons Center for Interstitial Lung Disease  
University of Pittsburgh  
Pittsburgh, Pennsylvania

Bronwyn Long, DNP, MBA, RN  
Division of Oncology  
Lung Cancer Center  
National Jewish Health  
Denver, Colorado

Wendi R. Mason, MSN, ACNP-BC  
Nurse Practitioner  
IPF Program Director  
Department of Pulmonary Medicine  
Vanderbilt University Medical Center  
Nashville, Tennessee

Kenneth R. McCurry, MD  
Surgical Director, Lung and Heart-Lung Transplantation Program  
Department of Thoracic and Cardiovascular Surgery  
Cleveland Clinic  
Cleveland, Ohio

Mark Shreve  
Pulmonary Fibrosis Foundation  
Chicago, Illinois

Jeffrey J. Swigris, DO, MS  
Associate Professor of Medicine  
National Jewish Health  
Denver, Colorado

Janet Talbert, MS, CGC  
Certified Genetic Counselor  
Director, Familial Pulmonary Fibrosis Genetic Counseling Program  
National Jewish Health  
Denver, Colorado

Gail G. Weinmann, MD  
National Heart, Lung, and Blood Institute  
Bethesda, Maryland
Pulmonary Fibrosis Foundation Sponsors

The Pulmonary Fibrosis Foundation is grateful to our generous sponsors for helping us fulfill our mission. Their support helps us assist the entire PF community in so many ways. Thank you to our sponsors for assisting us in funding research, advocating on behalf of the PF community, raising awareness and providing education, and playing a critical role in the lives of thousands of patients and families with whom we connect each year. Together we will win the fight against pulmonary fibrosis.

See Summit website for current list.

**EVEREST**
The Bean Family
Mr. and Mrs. Thomas E. Hales

Hogan Lovells
Mr. and Mrs. Charles P. McQuaid

**KILIMANJARO**
Daniel M. Rose, MD

**RAINIER**
Booz Allen Hamilton
Phyllis N. Demont
Jenny H. Krauss and Otto F. Krauss Charitable Foundation Trust

**SHASTA**
Jennifer A. Galvin, MD

**INTERMUNE**
Daniel M. Rose, MD

**McKINLEY**

**PROMEDIOR**

**Pulmonary Fibrosis Foundation Sponsors**

Phyllis N. Demont
Jenny H. Krauss and Otto F. Krauss Charitable Foundation Trust

Anonymous
Julia Willis O’Connor
Pacific Northwest Friends of the Pulmonary Fibrosis Foundation
Exhibitors

Boehringer Ingelheim Pharmaceuticals, Inc
Ridgefield, Connecticut

Boehringer Ingelheim Pharmaceuticals, Inc, the US subsidiary of Boehringer Ingelheim, headquartered in Germany, operates globally in 50 countries with more than 41,500 employees. The company is committed to researching, developing, manufacturing and marketing novel products of high therapeutic value for human and veterinary medicine. For more information please visit http://us.boehringer-ingelheim.com.

Horizon Hospice & Palliative Care
Chicago, Illinois

Horizon Hospice and Palliative Care, Chicago’s first hospice, is a not-for-profit, community-based organization whose mission is to provide comfort for the dying, to preserve dignity at the end of life for all in need of services and to educate the community. Horizon delivers high-quality hospice and palliative care to adults and children throughout Metropolitan Chicago, neighboring counties such as Lake, DuPage and Will.

Ikaria
Hampton, New Jersey

Ikaria is positioned to develop and deliver innovative therapeutics and interventions to meet the needs of critically ill patients. It is our aim to be an indispensable partner to clinicians, providing help and critical care just when patients need it the most. Being part of this lifeline, our desire is to have an impact on the lives of critically ill patients in the hospital and ICU settings. Delivering effective and efficient therapies is our way of serving and supporting these patients.

InterMune
Brisbane, California

InterMune is a biotechnology company focused on the research, development, and commercialization of innovative therapies in pulmonology and fibrotic diseases. In pulmonology, we are focused on therapies for the treatment of idiopathic pulmonary fibrosis (IPF), a progressive and fatal lung disease.

Lincare
Chicago, Illinois

Lincare is dedicated to the service and care our patients, customers, and physicians. Providing the highest level in quality and standards throughout the industry. We are committed to everyone that we deliver and educate through our home care services. We respond to the needs of all patients individually with oxygen systems most appropriate for their condition and lifestyles.
Mayo Clinic
Rochester, Minnesota

Mayo Clinic is the first and largest integrated group practice in the world. Doctors from every medical specialty work together to care for patients. The entire staff at Mayo Clinic is joined by the philosophy "the needs of the patient come first."

National Jewish Health
Denver, Colorado

Our mission since 1899 is to heal, discover, and to educate as a preeminent healthcare institution. We serve by providing the best integrated and innovative care for patients and their families; by understanding and finding cures for the diseases we research; and by educating and training the next generation of healthcare professionals to be leaders in medicine and science.

National Jewish Health, the #1 hospital for respiratory care in America, treats patients from all over the country and conducts innovative and groundbreaking research to improve health worldwide.

Pulmonary Fibrosis Foundation
Chicago, Illinois

The mission of the Pulmonary Fibrosis Foundation (PFF) is to help find a cure for idiopathic pulmonary fibrosis (IPF), advocate for the pulmonary fibrosis community both locally and in Washington, DC, promote disease awareness, and provide a compassionate environment for patients and their families.
Clinical Session Overview

2011 Patient Update on Pulmonary Fibrosis

What is Pulmonary Fibrosis and What are the Causes?
Gregory P. Cosgrove, MD

What are the Symptoms of Pulmonary Fibrosis and What are the Treatment Options?
Susan S. Jacobs, RN, MS

Lung Transplantation
Kenneth R. McCurry, MD

Familial Pulmonary Fibrosis and Genetic Counseling for Pulmonary Fibrosis
Janet Talbert, MS, CGC

Pulmonary Rehabilitation for Pulmonary Fibrosis
Brenda Crowe, CRT

Navigating Medicare and Health Insurance
Miranda G. James, Esq

The What, Why, and How of Clinical Trials
Gail G. Weinmann, MD

Advocacy
Former US Representative Brian Baird, MS, PhD and Mark Shreve

Roundtable Discussions with the Experts
Table 1: PF Specialist – Jeffrey J. Swigris, DO, MS
Table 2: PF Specialist – Gregory P. Cosgrove, MD
Table 3: Lung Transplant Specialists – Kenneth R. McCurry, MD
Table 4: Genetic Counseling – Janet Talbert, MS, CGC
Table 5: Advocacy – Former US Representative Brian Baird, MS, PhD and Mark Shreve
Table 6: Navigating Insurance – Miranda G. James, Esq

How to Establish a Support Group and the Benefits of Participation
Wendi R. Mason, MSN, ACNP-BC

Palliative and Hospice Care for Pulmonary Fibrosis Patients
Bronwyn Long, DNP, MBA, RN

Living with Pulmonary Fibrosis and How to Improve Quality of Life
Kathleen Lindell, PhD, RN
What is Pulmonary Fibrosis and What are the Causes?

GREGORY P. COSGROVE, MD
Associate Professor of Medicine
Department of Medicine
Co-Director, Interstitial and Autoimmune Lung Disease Program
National Jewish Health
University of Colorado
Denver, Colorado
What is Pulmonary Fibrosis and What are the Causes?

Gregory P. Cosgrove, MD

Notes
Notes
What are the Symptoms of Pulmonary Fibrosis and What are the Treatment Options?

SUSAN S. JACOBS, RN, MS

Center for Interstitial Lung Disease
Pulmonary and Critical Care Medicine
Stanford University Medical Center
Stanford, California
What are the Symptoms of Pulmonary Fibrosis and What are the Treatment Options?
Susan S. Jacobs, RN, MS

Challenges of PF Treatment
- PF is difficult to diagnose, especially IPF
- Research for effective drugs is ongoing
- Successful symptom management requires a multi-faceted approach

Goals of Talk
1. Provide a brief overview of drug management of pulmonary fibrosis and its symptoms
2. Review specific symptoms of shortness of breath, cough, and fatigue and identify strategies to minimize them

Treatment of PF Focuses on Both Quantity and Quality of Life
- **Quantity**
  - Medications
  - Oxygen
  - IPF exacerbations
  - Drug research
  - Lung transplant
  - "Comorbidities"
- **Quality**
  - Physical comfort
  - Strength and energy
  - Social connectedness
  - Emotional support
  - Spirituality
  - Mood

Disease and Symptom Management of IPF

Classification of Interstitial Lung Diseases
Pharmacologic Treatment of ILD

<table>
<thead>
<tr>
<th>Drug Options</th>
<th>Exposure-Related (Hypersensitivity Reactions)</th>
<th>Autoimmune</th>
<th>Sarcoidosis</th>
<th>Idiopathic Pulmonary Fibrosis (IPF)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Steroids</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td></td>
</tr>
<tr>
<td>Azathioprine</td>
<td>X</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mycophenolate Mofetil (CellCept)</td>
<td>X</td>
<td>X</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cyclophosphamide (Cytoxan)</td>
<td>X</td>
<td>X</td>
<td></td>
<td></td>
</tr>
<tr>
<td>IV cyclophosphamide</td>
<td></td>
<td></td>
<td></td>
<td><strong>X</strong></td>
</tr>
<tr>
<td>Methotrexate (Plaxanad)</td>
<td>X</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pulmonary HTN drugs (Inhalers, Thalidomide)</td>
<td>X</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Oxygen</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Investigational Drug - Clinical Trial</td>
<td>X</td>
<td></td>
<td></td>
<td>X</td>
</tr>
</tbody>
</table>

Goals of Pharmacologic Treatment

- **Slow** the progression of pulmonary fibrosis
  - Monitor breathing tests (forced vital capacity, diffusion capacity) every 3-6 months.
  - Monitor resting and exercise oxygen saturations.
  - Monitor symptoms – dyspnea and cough severity may predict survival.
- If no response to treatment, change or stop treatment. Refer for lung transplant if appropriate, or offer participation in clinical research trial.

Treatment of Comorbidities

- Gastroesophageal reflux – GERD
- Pulmonary hypertension
- Obstructive sleep apnea
- COPD/emphysema
- Cardiac problems

Disease and Symptom Management

- **Drug therapy**
  - Non-drug therapy
  - Comorbidities
  - Preventive care
- **Knowledge**
  - Values & Preferences
  - Advanced Care Planning
- **Symptom-Centered Management**
  - Dyspnea
  - Cough
  - Fatigue/Deconditioning
  - Depression and Anxiety

“How Does IPF Affect Your Life?”

Results of Interviews with 20 IPF Patients

- Symptoms: cough, shortness of breath
- IPF therapy: oxygen, side effects of meds
- Sleep: disturbed sleep
- Exhaustion: lack of energy, fatigue
- Forethought: need to always plan ahead
- Employment: finances, work, security
The Pulmonary Fibrosis Foundation presents

IPF SUMMIT 2011
FROM BENCH TO BEDSIDE

“How Does IPF Affect Your Life?”
Results of Interviews with 20 IPF Patients (cont)

• Dependence: need to rely on others
• Family: impact on family relationships
• Sexual relations: limitations on activity
• Socialization/Leisure: social isolation
• Mental and spiritual: fear, worry
• Mortality: feelings about death

Symptom Management

• Cough
• Dyspnea/anxiety
• Decreased mobility/isolation
• Fatigue/exhaustion
• Depression

The Cough

“What Causes the Cough in ILD?”

• The pulling or stretching of fibrotic lung tissue stimulates release of substances in the lung that trigger cough
• The cough receptors in airways of patients with ILD are up-regulated compared to normal airways, i.e., more sensitive
• Reflux/aspiration?
• We really don’t know for sure

“If I Could Just Get Rid of the Cough”

• Exhausted
• Unable to do activities
• Dizziness
• Rib fractures
• Sleep interruption
• Can’t phone, talk, sing, laugh
• Decreased socialization
• Change in lifestyle

More on Cough

• Cough is noted by over 80% of patients with IPF
• Cough is usually present by the time we first see a patient with IPF
• There is a constant urge to cough, but it is not relieved by coughing
Objective Cough Frequency in Idiopathic Pulmonary Fibrosis

- **Goal:** To measure cough rates in patients with IPF and determine the association between cough frequency and quality of life, and also see how the severity of cough correlates to the severity of disease.
- **Methods:** 19 patients with IPF underwent breathing tests, 24-hour cough recordings, quality of life questionnaires, and cough severity scores.

How to Manage the Cough

- First step is to eliminate the top 3 causes of cough for most people:
  - Rule out asthma
  - Aggressively treat and prevent GERD
  - Eliminate post-nasal drip

Patient Tips on Managing Cough

- Try lozenges, honey and lemon, hot water
- Avoid irritants, triggers
- ↑ oxygen during coughing as needed
- Hypnosis
- Yoga/relaxation techniques
- It is difficult to treat

Objective Cough Frequency in Idiopathic Pulmonary Fibrosis

- Cough rates averaged 14.6/hr day and 1.9/hr night.
- The greater the cough frequency, the worse the patients' rating of their cough-related quality of life.
- There was no correlation between pulmonary function tests and cough frequency, except for total lung volume.
- Patients' estimation of cough severity correlated well with the cough counter device's results.

Medications to Treat Cough

- Inhaled steroids (Advair® (combination), QVar®)
- Guaifenesin (Mucinex®)
- Benzonatate (Tessalon Perles®)
- Oral steroids (Prednisone)
- Nebulized anesthetics (Lidocaine)
- Opiates (Codeine)
- Experimental: baclofen, gabapentin, thalidomide, tramadol, interferon alpha

Shortness of Breath: What Causes It?

- Increased 'work' to breathe due to stiffness of the fibrosed or scarred lung tissue
- Possible receptors in lung that sense pressure and stretch of the lung tissue
- Low oxygen levels
- Deconditioning
- Contributing factors, such as anxiety and depression
Managing Shortness of Breath

- Exercise
- Fan/cold air
- Relaxation
- Distraction
- Yoga
- Oxygen
- Opiates

Impact of Exercise on SOB

- Builds endurance
- Strengthens muscles
- "Desensitizes" you to SOB
- Improves mood
- Facilitates independence, travel, and socialization
- Maintains ideal weight
- Decreases anxiety, panic
- Does not change PFTs

Exercise Stops the Downward Spiral of Dyspnea - Anxiety - Decreased Activity

Exercise as a Strategy to Treat Dyspnea, Fatigue, Anxiety, & Depression

- Regular, planned exercise can improve endurance, shortness of breath, and quality of life
- Deconditioning can be more limiting than the level of disease reflected on breathing tests
- Exercise benefits are also a result of desensitization to SOB as well as motivation
- Adequate oxygenation during exercise remains a challenge for many patients with ILD
- Exercise has demonstrated more consistent benefits for decreasing SOB than IPF clinical drug trials

Exercise as a Treatment for IPF Symptoms: Research Results

<table>
<thead>
<tr>
<th>Study</th>
<th>Sample</th>
<th>∆ 6MWT, m</th>
<th>Dyspnea</th>
<th>QOL</th>
</tr>
</thead>
<tbody>
<tr>
<td>Swigris 2011</td>
<td>21</td>
<td>202</td>
<td>Fatigue</td>
<td>Improved</td>
</tr>
<tr>
<td>Sath 2010</td>
<td>11</td>
<td>107</td>
<td>Improved</td>
<td>NA</td>
</tr>
<tr>
<td>Fennema et al 2009</td>
<td>99</td>
<td>56</td>
<td>Improved</td>
<td>NA</td>
</tr>
<tr>
<td>Holland 2008 (RCT)</td>
<td>57</td>
<td>36</td>
<td>Improved</td>
<td>Improved</td>
</tr>
<tr>
<td>Nishiyama 2008 (RCT)</td>
<td>28</td>
<td>46</td>
<td>No Change</td>
<td>Improved</td>
</tr>
<tr>
<td>Jasztal 2006</td>
<td>31</td>
<td>NA</td>
<td>Improved</td>
<td>Improved</td>
</tr>
<tr>
<td>Naj 2006</td>
<td>28</td>
<td>NA</td>
<td>Improved</td>
<td>Improved</td>
</tr>
</tbody>
</table>

Oxygen as a Treatment of Shortness of Breath

- The need for oxygen is determined by measuring oxygen saturation during rest, exercise, and sleep – not by how you "feel"
- You may experience shortness of breath but have normal oxygen saturations (above 90%)

OR

- You may not feel shortness of breath and have low oxygen saturations
Oxygen as a Treatment of Shortness of Breath (cont)

- Portability of your oxygen system will be determined by how high a liter flow you need.
- Portable oxygen concentrators (POCs) and pulse or conserving devices may not provide enough oxygen flow to keep your saturations above 90% with exertion (3L/min on pulse ≠ 3L/min on contin)
- It is important to have your oxygen saturations checked with rest and exercise at regular intervals as your needs may increase over time.

Summary: Treating IPF and Its Symptoms

- Research is ongoing to identify effective drugs to treat IPF and PF
- Use multiple strategies for SOB, cough, and fatigue
- Learn as much as you can about pulmonary fibrosis, your medications, and oxygen therapy
- Enroll in a Pulmonary Rehabilitation program and continue to exercise after you finish
- Connect with others for support (ILD Support Groups) and meaningful emotional and social interactions

Thank You!
Lung Transplantation

KENNETH R. McCURRY, MD
Surgical Director, Lung and Heart-Lung Transplantation Program
Department of Thoracic and Cardiovascular Surgery
Cleveland Clinic
Cleveland, Ohio
Notes
Familial Pulmonary Fibrosis and Genetic Counseling for Pulmonary Fibrosis

JANET TALBERT, MS, CGC
Certified Genetic Counselor
Director, Familial Pulmonary Fibrosis Genetic Counseling Program
National Jewish Health
Denver, Colorado
Familial Pulmonary Fibrosis and Genetic Counseling for Pulmonary Fibrosis
Janet Talbert, MS, CGC

Agenda
- Causes of Pulmonary Fibrosis
- Idiopathic Interstitial Pneumonias
- Genetics and IPF
- Familial Pulmonary Fibrosis
- Genetic Counseling for Pulmonary Fibrosis
- Research

Pulmonary Fibrosis
- Causes
  - Environmental/Occupational
  - Autoimmune
  - Drug-induced
  - Idiopathic
- Idiopathic
  - Exclusion of known causes
  - Idiopathic interstitial pneumonias

Idiopathic Interstitial Pneumonias (IIPs)
- Distinct classifications
  - Idiopathic Pulmonary Fibrosis (IPF)
  - Nonspecific Interstitial Pneumonia (NSIP)
  - Cryptogenic Organizing Pneumonia (COP)
  - Respiratory Bronchiolitis-Interstitial Lung Disease (RB-ILD)
  - Desquamative Interstitial Pneumonia (DIP)
  - Lymphocytic Interstitial Pneumonia (LIP)
  - Acute Interstitial Pneumonia (AIP)
- IPF most common
  - Up to 60% of cases of IIP
  - Associated with a pathological lesion known as
    Usual Interstitial Pneumonia (UIP)

Identification of Genes in IPF
- Define causative factors
- Improve understanding of pathogenesis
- Suggest potential treatment options
- Several lines of evidence for a genetic basis

Sporadic IPF Pedigree
Could IPF Be Genetic?

- Familial clustering
- Twin studies
  - Raised together
  - Raised apart
- Transmitted with other known rare genetic conditions
- Now associated with mutations in several genes

Familial Pulmonary Fibrosis Pedigrees

Familial Interstitial Pneumonia

- Also known as Familial Pulmonary Fibrosis or “FPF”
- Two diagnoses consistent with IIP
- Exact percentage of FPF cases unknown
- Reports of FPF in IPF cases
  - UK 0.5–2.2%\(^1\)
  - Finland 3.3–5.7%\(^2\)

Familial Interstitial Pneumonia

- Percentage likely underestimated
- Newer reports
  - Vanderbilt Lung Transplant Center\(^1\)
  - 10% of cases of IPF reported family history
  - Netherlands ILD clinic\(^2\)
  - 10% of IIP cases reported family history
  - National Institute of Respiratory Diseases in Mexico\(^3\)
  - 20% of IPF cases reported family history

Genes for Pulmonary Fibrosis

- Human genome
  - ~25,000 genes
  - Little more than ½ have known function
- FPF\(^1\)
  - Five genes associated
- Sporadic IPF\(^1\)
  - Variants in genes involved in
    - Immune system
    - Protein molecules
    - Various enzymes and proteins
  - 3 of 5 FPF genes associated with sporadic cases\(^2\)


The Pulmonary Fibrosis Foundation presents

**IPF SUMMIT 2011**
FROM BENCH TO BEDSIDE

---

**Genes Associated with FPF**

- SFTP\(\text{C}\)\(^1\) and SFTP\(\text{A}\)\(^2\)
  - Surfactant genes
  - Important in lung function
- TERT and TERC\(^3,4\)
  - Involved in DNA replication
  - Longevity and stability of cells during cell division
- ELMOD\(\text{2}\)\(^5\)
  - Implicated in antiviral response
  - Candidate gene from Finnish population

---

**FPF vs. IPF**

<table>
<thead>
<tr>
<th></th>
<th>FPF</th>
<th>IPF</th>
</tr>
</thead>
<tbody>
<tr>
<td>SFTPA(\text{C})(^1)</td>
<td>~3%*</td>
<td>Less than 1%</td>
</tr>
<tr>
<td>TERT(^1)</td>
<td>~16%</td>
<td>~5%</td>
</tr>
<tr>
<td>TERC(^1)</td>
<td>~1%</td>
<td>n/a</td>
</tr>
<tr>
<td>ELMOD(\text{2})</td>
<td>Unknown</td>
<td>Unknown</td>
</tr>
</tbody>
</table>

*SFTP\(\text{C}\) mutations were recently reported in 25% of FPF cases in a Dutch cohort!*

---

**MUC5B**

- Discovered through FPF research
- Variant by a single letter in the genetic code
  - AA\(\text{T}\)CT \(\rightarrow\) AA\(\text{T}\)CT
  - Single nucleotide polymorphism “SNP”
- Associated with IPF and FPF
- Also found in 19% of healthy subjects

<table>
<thead>
<tr>
<th></th>
<th>FPF</th>
<th>IPF</th>
<th>Healthy Controls</th>
</tr>
</thead>
<tbody>
<tr>
<td>MUC5B</td>
<td>59%*</td>
<td>67%*</td>
<td>19%*</td>
</tr>
</tbody>
</table>

*Estimated Carrier Frequency in IPF, FPF, and control populations from study*

---

**Genes for Pulmonary Fibrosis**

- Majority remain unknown or under investigation
- Complex disease
  - Genetic predisposition
  - Environmental triggers
- More research = more genes to discover
- Lead to therapies and treatment

---

**Genetic Counseling for IPF/FPF**

- Evidence of genetic involvement
- More genes will be discovered
- More tests will be on the market
- Need for physicians and patients to understand role of genetics in diagnosis
What Is Genetic Counseling?

- Genetic counseling is the process of helping people understand and adapt to the medical, psychological, and familial implications of genetic contributions to disease

Genetic Counseling

- Calculate risks of occurrence or recurrence
- Education
  - Inheritance
  - Management
  - Genetic testing
  - Resources
  - Research

Familial Pulmonary Fibrosis

- 2+ IPF/IP cases per family
- Concerns for IPF
  - Personal risk
  - Risk to offspring
  - Risk to other family member
  - Reproductive decisions
- Genetic testing

Sporadic IPF

- Only one case in family
- Concern for siblings or offspring
- Risk to family members
- Genetic testing

Genetic Testing for IPF

- Genetic testing
  - Complex, limited to a few genes
  - Best to test diagnosed family members first
  - Currently expected to pick up only
    - >15% of patients with IPF
    - >1-3% of patients with PF
  - 85 to 97% won’t have a definitive answer
- MUC5B genetic testing
  - Launch January 2012
  - National Jewish Health

Genetic Testing

- Clinical testing can be expensive
  - $$$ for initial test on affected family member
  - ~$ for other family members once mutation is known
  - May or may not be covered by insurance
- Test results can be complex
- Risks and benefits of testing
- Testing not usually recommended
- Genetic counseling recommended for any genetic testing
The Pulmonary Fibrosis Foundation presents
IPF SUMMIT 2011
FROM BENCH TO BEDSIDE

Familial Pulmonary Fibrosis
Genetic Counseling Service

- Telephone-based Genetic Counseling
- Located at National Jewish Health
- Staffed by Board-Certified Genetic Counselor
- Supported by Pulmonary Fibrosis Foundation
- Toll-free number
National Jewish Health
Janet Talbert, MS, CGC
1-800-423-8891 ext. 1097

Familial Pulmonary Fibrosis
Genetic Counseling Service

- Provide medical information and support
- Assist individuals in understanding how heredity may contribute to the diagnosis
- Assist individuals in making informed decisions for genetic testing
- Provide referral to medical care if needed

Research in Pulmonary Fibrosis

- Familial Pulmonary Fibrosis Study
- 2+ family members with pulmonary fibrosis
- Enrollment
  - National Jewish Health
    - Contact: Janet Talbert 1-800-423-8891 ext. 1022
  - Vanderbilt University
    - Contact: Cheryi Markin 1-888-896-1550
  - Duke University
    - Contact: Raven Kidd 1-877-587-4411

Research in Pulmonary Fibrosis

National Institutes of Health
Clinical trials website
www.clinicaltrials.gov

References

Pulmonary Rehabilitation for Pulmonary Fibrosis

BRENDA CROWE, CRT
Manager, Pulmonary Rehabilitation
Exempla Lutheran Medical Center
Wheat Ridge, Colorado
Pulmonary Rehabilitation for Pulmonary Fibrosis
Brenda Crowe, CRT

Objectives
- Definition of Pulmonary Rehabilitation (PR)
- Criteria for referral to Pulmonary Rehab
- Benefits of PR to the patients and their families
- Measurable outcomes

Definition
- Pulmonary rehabilitation is a multidisciplinary program of care for patients with chronic respiratory impairment that is individually tailored and designed to optimize physical and social performance

Referral Criteria
- Patient who has been diagnosed with COPD or non-COPD
- Patient should have symptomatic clinically stable respiratory disease of at least a moderate degree
- Metrics
  - FEV₁ ≤ 85% of predicted
  - FVC ≤ 65% of predicted
  - DlCO ≤ 65%
  - Resting or exertional hypoxemia ≤ 90%

Referral Criteria
- Dyspnea /fatigue chronic respiratory symptoms
- Impaired quality of life, reduction in physical activity
- Psychosocial issues, anxiety/depression related to lung disease
- Non-smoker or attempting to quit smoking

Diagnosis of...
- COPD
- Emphysema, alpha 1
- Chronic bronchitis
- Non-COPD
- ILD
- Pulmonary fibrosis

www.pulmonaryfibrosis.org
**Medicare Guidelines**

- Physician referral
- PFT (within the last 3 months)
- Exercise Ox (within the last 3 months)
- FEV₁/FVC – DL₆₅ CO ≤ 65%
- History and physical

**Getting Started**

- Initial patient assessment
- Medical history and physical exam
- Assessment of
  - ADLs, exercise
  - Nutrition
  - Education
  - Psychosocial state
- Goal development

**Goals**

- Improve knowledge of disease process
- Correct use of medications/oxygen
- Breathing/pacing techniques
- Recognition of signs and symptoms of infection/exacerbation
- Collaborative self-management
- Community resources

**Disease Management Classes**

- Breathing retraining
- Medications
- Bronchial hygiene
- Signs and symptoms of respiratory infections/exacerbations
- Oxygen
- Traveling with O₂
- ADLs
- Nutrition
- Counseling
Physical Conditioning

- Upper and lower extremity endurance training
- Strength training
- Duration, frequency, mode, and intensity
- Goals
  - Increased tolerance for dyspnea
  - Improved appetite
  - Increased tolerance for exercise
  - Improved quality of life

Goals

- Improve knowledge of disease process
- Correct use of medications/oxygen
- Breathing/pacing techniques
- Recognition of signs and symptoms of infection/exacerbation
- Collaborative self-management
- Community resources

Outcomes

- Reduction in symptoms (dyspnea, fatigue)
- Increased exercise performance
- Enhanced ability to perform ADLs
- Improved health-related quality of life
- Improvement in symptoms of anxiety and depression
- Reduced hospitalizations and the use of medical resources

References

- American Association for Respiratory Care
- American Lung Association
- American Thoracic Society
- AACVPR Guidelines for Pulmonary Rehabilitation
- Living Well with Chronic Lung Disease
- Medicare Guidelines for Pulmonary Rehabilitation

Dedicated to...

Phil Konsella
IPF patient and founder of
Wheat Ridge
Pulmonary Fibrosis Support Group
Navigating Medicare and Health Insurance

MIRANDA G. JAMES, Esq
Caring Voice Coalition
Mechanicsville, Virginia
Navigating Medicare and Health Insurance
Miranda G. James, Esq

What to Expect
- Overview of CVC
- COBRA
- Individual Plans
- Governmental Programs
- Pre-Existing Condition Plan
- High-Risk Pool
- Helpful Links

Caring Voice Coalition, Inc.'s (CVC) Insurance Education & Counseling
- Our Mission
  - CVC is dedicated to helping individuals and families affected by serious, chronic illnesses. We offer outreach and support that directly benefit the patient community

Caring Voice Coalition, Inc.'s (CVC) Insurance Education & Counseling
- Why We Help
  - CVC established its Insurance Education and Counseling ("IEC") program for two reasons
    - Because we know that adequate health insurance coverage is essential for persons with chronic illnesses
    - Because we know that for those with health insurance, coverage issues can be complex and difficult to resolve

COBRA
- Continuation of Health Coverage – COBRA
  - The Consolidated Omnibus Budget Reconciliation Act (COBRA) gives workers and their families who lose their health benefits the right to choose to continue group health benefits provided by their group health plan for limited periods of time under certain circumstances

COBRA
- Who is entitled to benefits under COBRA?
  - There are three elements to qualifying for COBRA benefits. COBRA establishes specific criteria for plans, qualified beneficiaries, and qualifying events
**COBRA**

- To continue benefits under COBRA, you must be a qualified beneficiary and have a qualifying event
  - Qualified beneficiary: an individual who, on the day before the qualifying event, is an enrollee of a group health plan as either a qualified employee or a qualified dependent
  - Qualifying event: an event that usually (without COBRA) would result in loss of health insurance coverage

---

**COBRA**

- Qualifying Event Types
  - Termination of employment
  - Voluntary or involuntary
  - Reduction in hours
  - Full or part-time
  - Eligibility for Medicare
  - Divorce or legal separation
  - Loss of dependent child status
  - Death

---

**COBRA**

- The type of qualifying event determines who the qualifying beneficiaries may be and how long they are eligible to continue benefits under COBRA

<table>
<thead>
<tr>
<th>Qualifying Event</th>
<th>Beneficiary</th>
<th>Eligibility Period</th>
</tr>
</thead>
<tbody>
<tr>
<td>Termination, Reduced Hours</td>
<td>Employee, spouse, dependent child</td>
<td>18 months</td>
</tr>
<tr>
<td>Employee entitled to Medicare, Divorce or legal separation, Death of covered employee</td>
<td>Spouse or dependent child</td>
<td>36 months</td>
</tr>
<tr>
<td>Loss of dependent child status</td>
<td>Dependent child</td>
<td>36 months</td>
</tr>
</tbody>
</table>

---

**Mini-COBRA**

- State COBRA expansion programs extend coverage to employees in firms with fewer than 20 workers who are not covered by COBRA, the federal law
- The following link shows each state and if they offer a Mini-COBRA plan and what the terms are

---

**COBRA and Convertible Coverage**

- When a group health plan offers participants the option of moving into individual coverage after group coverage is lost, they must do the same for COBRA participants as well

---

**COBRA**

Questions or personal experiences with COBRA?
<table>
<thead>
<tr>
<th>Individual Plans</th>
<th>Individual Plans</th>
</tr>
</thead>
</table>
| • Secure independent coverage through an individual plan | • HIPAA (Health Insurance Portability and Accountability Act)  
  - [http://www.dol.gov/dol/topic/health-plans/portability.htm](http://www.dol.gov/dol/topic/health-plans/portability.htm)  
  - When you're buying individual health insurance, HIPAA eligibility gives you greater protections than you would otherwise have under state law |

<table>
<thead>
<tr>
<th>Individual Plans</th>
<th>Government Programs</th>
</tr>
</thead>
</table>
| Questions or personal experiences with Individual Plans? | • Medicaid  
  - Medicaid is a state-administered program and each state sets its own guidelines regarding eligibility and services  
  - Supplemental Security Insurance (SSI) |

<table>
<thead>
<tr>
<th>Government Programs</th>
<th>Government Programs</th>
</tr>
</thead>
</table>
| • Medicare  
  - Eligibility  
  - Social Security Disability Insurance  
  - Part D Plans  
  - Advantage Plans | Questions or personal experiences with Governmental Programs? |
Pre-Existing Condition Insurance Plan

- The Pre-Existing Condition Insurance Plan makes health insurance available to people who have had a problem getting insurance due to a pre-existing condition
- https://pcip.gov/

Pre-Existing Condition Insurance Plan

- Covers a broad range of health benefits, including primary and specialty care, hospital care, and prescription drugs
- Does not charge you a higher premium just because of your medical condition
- Does not base eligibility on income

Pre-Existing Condition Insurance Plan

- Eligibility
  - Citizen or national of the United States or reside in the US legally
  - Must have been without coverage for at least the last 6 months
  - Must have a pre-existing condition or have been denied health coverage because of your health condition

Pre-Existing Condition Insurance Plan

- July 2011 PCIP Program Changes: NEW RATES
  - As of July 1, 2011, some PCIP enrollees in the federally-administered plan will see a reduction in their monthly premium
  - Premiums vary depending on the state you live in

High-Risk Pools

- State-sponsored risk pools are aimed at helping the small slice of Americans who can afford to buy health insurance, but who are denied affordable health insurance coverage by insurance carriers because of a pre-existing condition
- http://www.naschip.org/states_pools.htm

Other Helpful Links

- Caring Voice Coalition – http://www.caringvoice.org/
- Social Security – http://www.ssa.gov/
- HealthInsuranceInfo – http://www.healthinsuranceinfo.net/
Notes
The What, Why, and How of Clinical Trials

GAIL G. WEINMANN, MD
National Heart, Lung, and Blood Institute
Bethesda, Maryland
The What, Why, and How of Clinical Trials
Gail G. Weinmann, MD

What Is a Clinical Trial?

A Clinical Trial is...
- A controlled experiment to answer specific diagnostic and therapeutic questions in humans

Why Do Clinical Trials?

Decision

Bled 8 pints in 9 hours
Doctors noted that he “... appeared physically calm” before his death

If the Doctors Had the Results of a Trial...

“...It had been so arranged, that... each of us had one-third of the whole. The sick were indiscriminately received, and were attended as nearly as possible with the same care and accommodated with the same comforts...”

Neither Mr. Anderson nor I ever once employed the lancet. He lost two, I four cases; whilst out of the other third [treated with bloodletting by the third surgeon] thirty-five patients died.”

Why Do Clinical Trials? Because...

Surfactant treatment in premature newborns
Drugs for pulmonary hypertension

Clinical trials provide evidence for decisions on prevention, diagnosis, and treatment.

www.pulmonaryfibrosis.org
The Pulmonary Fibrosis Foundation presents

IPF SUMMIT 2011
FROM BENCH TO BEDSIDE

Trials Changing Your Care Today

Trial Quality Key Elements

Important Research Question
Robust Trial Design
Careful Oversight
Dissemination
Impact on Practice

How Clinical Trials? Different Phase Trial for Different Purposes

Pre-Clinical Phase
Clinical Phases

Pre-Clinical
Phase I
Phase II
Phase III
Phase IV

Pre-Clinical
• Sub-cellular
• Cellular
• Animal

Clinical
• Dosing
• Safety
• Efficacy
• Toxicity
• Pharmacokinetics
• Pharmacodynamics
• Advanced outcome

Number of Subjects

What Is a Randomized Controlled Trial?

randomize

intervention
open label

Analyse by treatment assigned, not received

Why Placebo Controlled? Why Meaningful Outcomes?

Why are some studies placebo-controlled, i.e., one of the treatments is an inactive substance?

Why should studies have measures that patients care about?

Treatment of PVCs After a Heart Attack

• The frequency of PVCs increase after a heart attack
• The number of PVCs correlate with death after a heart attack
• Doctors can suppress PVCs

The Trial Results

**Results:**
The placebo group had better survival

**Conclusion:**
"... need for more placebo controlled clinical trials...with mortality endpoints."

---

**Therapies Must Be Proven:**
Heart Attack and the Occluded Artery

---

**Observational Findings**

---

**When Researchers Wanted to Do a Trial, They Ran Into a Problem...**

Many doctors were so convinced of the value of this procedure...that they thought it would be unethical to assign patients to the control group, which would get all the best medicines for this condition but not the artery-reopening procedure.

But the researchers persisted, with heavy support from the National Heart, Lung, and Blood Institute. After four years of work examining 2,166 patients, they came to an unexpected conclusion....

---

**The Findings and Response**

---

**Other Modern Examples of Bloodletting**

- Postmenopausal hormone replacement (cancer, strokes, general youth)
- Bone marrow transplantation for breast cancer (higher death rate)
- Vitamins to prevent cancer/CVD (failed)
- Oxygen for premature infants (too much causes blindness)
The Pulmonary Fibrosis Foundation presents

IPF SUMMIT 2011
FROM BENCH TO BEDSIDE

PANTHER

PANTHER-IPF

- Prednisone-Azathioprine-N-acetylcysteine: A Trial That Evaluates Responses in IPF

- Testing two accepted treatments for IPF against placebo

83 Years After Hamilton’s Trial on Bloodletting...

“During the last decades we have certainly bled too little.”

William Osler, MD

What Happens to Trial Results?

Primary Endpoint: Change in FVC at 60 Weeks

Thank you
Notes
Advocacy

BRIAN BAIRD, MS, PhD
Former US Representative to Congress
Vancouver, Washington

and

MARK SHREVE
Pulmonary Fibrosis Foundation
Chicago, Illinois
Advocacy
Former US Representative Brian Baird, MS, PhD and Mark Shreve

Advocacy
Brian Baird, MS, PhD
Former US Representative to Congress
Vancouver, Washington

Mark Shreve
Pulmonary Fibrosis Foundation
Chicago, Illinois

National Advocacy at the PFF
- “Advocating for the PF Community Locally and in Washington”
- Raise awareness of PF in Congress and within federal agencies
- Represent the impact of PF on patients
- Express the need for increased clinical research $$
- Provide PF community perspective on policy/legislation

Patient Advocacy
- Helping patients identify the resources they need – educational materials, support groups, centers of excellence, specialist referrals, lung transplant referrals
- Insurance and financial hardship issues via Caring Voice Coalition

Organizational Advocacy
- Congress: PFREA, NIH, Medicare/Medicaid, federal budget
- SSA: Social Security Disability Insurance – “Compassionate Allowances” Program
- CMS – Center for Medicare/Medicaid Services: improve access to care; coverage policy
- NIH – increased funding for PF and related disorders, appropriations language

Partnerships
- National IPF Awareness Week
- Support for Genetic Alliance Advocacy Day
- Endorsement of ATS PAR and national policy initiatives

2011 Legislative Initiatives
- January–March 2011 – H.R. 1 Full Year Continuing Appropriations Act
- June 2011 – Pulmonary Fibrosis Research Enhancement Act (PFREA) (S. 1350, H.R. 2505)
- NIH – “Report Language” in Final NIH Appropriations Bill
Historical Legislative Issues

- H.C.R. 176: S.236 (2005-07) - Introduced by Charlie Norwood (R-GA); precursor to PFREA
- The Medicare Improvements for Patients and Providers Act of 2008 (S. 3101/H.R. 6331)
  - Repealed 36-month transfer of ownership of O2 equip
  - Created national coverage policy for cardio/pulm rehab
- Ending the Medicare Disability Waiting Period Act (2006-10: S.700)
  - Eliminate 24-month waiting period policy for additional life-threatening conditions - like PF
- Stem Cell Research Enhancement Act (2005-08)

The Pulmonary Fibrosis Research Enhancement Act

- S. 1350 – Introduced by Senators Coons (D-DE) and Kirk (R-IL)
  - Assigned to Health, Education, Labor and Pensions (HELP) Committee with 5 co-sponsors
- H.R. 2505 – Introduced by Representatives Paulson (R-MN), Baldwin (D-WI), and Markey (D-MA)
  - Assigned to Energy and Commerce (E&C) with 26 co-sponsors

What Would PFREA Accomplish?

- Establish the first National PF Registry
- Establish a National PF Advisory Board
- Call for a national PF education and awareness plan
- Encourage the NIH to expand, intensify, and coordinate its activities with respect to research on PF

What You Can Do to Help

- Call/write/email your Members of Congress (www.house.gov/writerep/)
- Schedule an in-district meeting with your Member
- Spread the word through online (Facebook, Twitter, emails, etc.)
- Ask friends & family to contact their Members
- Visit the PFF Web site to learn about PFREA, view sample letters, and contact your Members
- Call the PFF at (888) 733-6741
Roundtable Discussions with the Experts

Table 1: PF Specialist – Jeffrey J. Swigris, DO, MS
Table 2: PF Specialist – Gregory P. Cosgrove, MD
Table 3: Lung Transplant Specialists – Kenneth R. McCurry, MD
Table 4: Genetic Counseling – Janet Talbert, MS, CGC
Table 5: Advocacy – Former US Representative Brian Baird, MS, PhD and Mark Shreve
Table 6: Navigating Insurance – Miranda G. James, Esq
Roundtable Discussions with the Experts

Notes

____________________________________________________________________

____________________________________________________________________

____________________________________________________________________

____________________________________________________________________

____________________________________________________________________

____________________________________________________________________

____________________________________________________________________

____________________________________________________________________

____________________________________________________________________

____________________________________________________________________
Notes
How to Establish a Support Group and the Benefits of Participation

WENDI R. MASON, MSN, ACNP-BC
Nurse Practitioner
IPF Program Director
Department of Pulmonary Medicine
Vanderbilt University Medical Center
Nashville, Tennessee
How to Establish a Support Group and the Benefits of Participation

Wendi R. Mason, MSN, ACNP-BC

How to Establish

• Look for someone/institution with an interest in PF
  – A pulmonary medicine provider group
  – A pulmonary rehabilitation group
  – A hospital

How to Establish

• Determine extent of patient population
  – Are there 5 patients or 50 patients?
  – Is it feasible for 6? 15? 25?

How to Establish

• Determine and set up meeting place and meeting times
  – Library conference room, rehab facility conference room, provider’s office conference room, local school cafeteria, others?
  – Quarterly vs. monthly meetings; afternoon vs. evenings

How to Establish

• Determine how to capture patient interest
  – Speakers and topics
    ➢ Speakers may include PF providers, transplant providers, pulmonary rehab specialists, psychologists, respiratory technicians, oxygen companies, and others
    ➢ Topics may include “PF 101”; “Familial PF”; “Transplant Work-up, Wait, and Action”; “The Importance of Pulmonary Rehab”; “Chronic Diseases and Depression”; “The Relationship of GERD and PF”; “Updates in Research” and more
  – Food!

• Ask your provider/pulmonary rehab program to contact PF patients via:
  – Mail
  – Email or electronic communication

• Include in this letter your name and contact information, the meeting location, time, and topic of discussion. Encourage patients to bring family members or friends
Benefits of Participation

“Camaraderie”

“Channel your energy into something positive!”

“Support from people who are going through what you are...or have already been down the path you are preparing to travel.”

Benefits of Participation

“Encouraging!”

“You learn that you are not alone.”

“Good information for us ‘new folks’.”

“Allows me to keep up with research!”

Benefits of Participation

“Lunch!”

“Caregivers get a lot from it.”

So...

What are you waiting for?

Questions??
Wendi Mason
615-343-7068
wendi.mason@vanderbilt.edu
Palliative and Hospice Care for Pulmonary Fibrosis Patients

BRONWYN LONG, DNP, MBA, RN
Division of Oncology
Lung Cancer Center
National Jewish Health
Denver, Colorado
Palliative and Hospice Care for Pulmonary Fibrosis Patients
Bronwyn Long, DNP, MBA, RN

Objectives
- Describe key elements of palliative care
- Distinguish between palliative care and hospice
- Illustrate how palliative care can benefit pulmonary fibrosis patients

Pulmonary Fibrosis and Palliative Care
- Transitions can be rapid
- Sudden change of status

Life Cycle
- Well Role
- Sick Role
- Dying Role

Old Model of Care: “Over the Wall”

Curative

Diagnosis

Hospice

Death

Modified WHO Model

Curative  
Palliative  
Hospice  
Bereavement

Diagnosis  
Death

Hospice and Palliative Care

• All hospice care is palliative
• Not all palliative care is hospice

Palliative Care

GOALS OF CARE

- Pain and symptom management
- Advance care planning/advance directives
- Psychological support
- Spiritual support
- Quality of life
- Hospice

Comfort

• Physical
• Emotional
• Social
• Intellectual
• Spiritual

Disease and Palliative Care

• Simultaneous care
  - Curative care
    - Surgery, chemotherapy, radiation therapy
  - Palliative care
    - Symptom management, advance care planning, goals of care, counseling and support, quality of life
• Hospice care
  - Transition to end of life
    - Comfort, symptom management, life review, counseling and support, bereavement

Research: Simultaneous Care

• Examination of effect of introducing palliative care early after diagnosis to patients with metastatic NSCLC, n = 151
• Experimental group: early palliative care integrated with standard cancer care
• Control group: standard cancer care only
• Functional Assessment of Cancer Therapy-Lung scale (FACT-L)
• Results: early palliative care led to
  - Improvements in quality of life and mood
  - Less aggressive care at end-of-life
  - Longer survival by 3 months

The Pulmonary Fibrosis Foundation presents
IPF SUMMIT 2011
FROM BENCH TO BEDSIDE

Palliative Care Toolkit

- Symptom management/physical comfort
- Advance directives

Symptom Management

- Pain
- Fatigue, insomnia
- Nausea, vomiting
- Constipation, diarrhea
- Edema
- Dyspnea
- Cough
- Hemoptysis
- Fever
- Anxiety, depression
- Grief, loss
- Adjustment to illness
- Family issues
- Delirium, dementia
- Anorexia, cachexia, dysphagia
- Pruritus
- Hydration, thirst, nutrition
- Spiritual/Cultural Context

Advance Directives

- Five Wishes®
- Medical Orders for Scope of Treatment (MOST)
- Physician Orders for Life-Sustaining Treatment (POLST) Forms

Five Wishes®

- Wish 1: Surrogate/MDOA
- Wish 2: Medical Treatments
- Wish 3: Comfort
- Wish 4: Care
- Wish 5: Messages

MOST/POLST

Interdisciplinary Palliative Care Team

- Physician: board-certified in hospice and palliative medicine subspecialty
- Nurse: board-certified CHPN/ACHPN
- Social Worker: ACE course
- Chaplain: ACE course
- Additional team members
  - Pharmacist
  - Nutritionist
  - Psychologist
Taking Control

- The Four Things:
  - “Please forgive me.”
  - “I forgive you.”
  - “Thank you.”
  - “I love you.”


Questions?

Resources

- Hospice and Palliative Nurses Association (HPNA): www.hpna.org
  - Patients/Family teaching sheets on symptom management and general topics:
- National Hospice and Palliative Care Organization (NHPCO):
  www.nhpco.org
  - Caring Connections website:
    http://www.nhpco.org/dl/pages/index.cfm?pageID=3754
- American Hospice Foundation: http://www.americanhospice.org/
  - Hospice publications:
- International Association for Hospice and Palliative Care (IAHPC):
  www.hospicecare.com
  - Resources for patients and relatives:

References

- MD Anderson Cancer Center. 15th Annual Symposium on Pain and Palliative Care Conference Proceedings. October 7, 8, 2006, Houston, TX.
Notes
Living with Pulmonary Fibrosis
and How to Improve Quality of Life

KATHERLEEN LINDELL, PhD, RN
Clinical Nurse Specialist
Director, Quality of Life Program
Division of Pulmonary, Allergy, and Critical Care Medicine
Dorothy P. and Richard P. Simmons Center for Interstitial Lung Disease
University of Pittsburgh
Pittsburgh, Pennsylvania
Living with Pulmonary Fibrosis and How to Improve Quality of Life

Kathleen Lindell, PhD, RN

What’s Happening in the Lungs

- Problem is scarring at the area of gas exchange
- Leads to shortness of breath, cough, and fatigue
- Eventually leads to need for supplemental oxygen

What’s Happening with You?

- How are you and your caregivers coping?
- Are you sure of your diagnosis?
- Do you have a treatment plan?
- Have you been seen at a center that specializes in the care of patients with PF?
- Are you living “Your Normal” Life?

Where We’ve Come in the Last 10 Years

- More support groups
- More specialty centers
- More emphasis on research
- More clinical trials
- More lung transplants for patients with PF
- More awareness…but still need MORE!

How to Live with Pulmonary Fibrosis

- Symptom management
- Optimization of oxygen therapy
- Pulmonary rehabilitation
- Early referral for lung transplantation
- Support systems
- Participating in research
- Psychological counseling services

Dealing with the Elephant in the Room!

Palliative Care’s Place in the Course of Illness

[Diagram showing the progression of illness with stages for diagnosis, life-prolonging therapy, palliative care, Medicare, hospice benefit, and death]
Getting On With Your Life

- Acknowledging the difficulties
- Dealing with your feelings
- Having realistic expectations
- Lung transplant and clinical trials
  - Are they an option for you?
- Making the most of what you have
  - We should all have a bucket list!
- “Saving your energy for the GOOD STUFF!”
Notes