



Education. Support. Hope.

INTRODUCTION

HOW TO USE THIS KIT

The purpose of this kit is to provide you with the tools you need to create, manage and grow a support group or self-help group for patients with pulmonary fibrosis (PF) and related interstitial lung diseases.

Written by the Coalition for Pulmonary Fibrosis and members of its Scientific and Strategic Advisory Boards, the ***PF Support Group Coordinator's Kit*** offers guidance and tips on organization, finding a meeting location, communicating with support group members and information on topics and issues of interest to PF patients and their friends, families and caregivers.

Any and all information included in this binder is to be used, photocopied, and distributed to help in your efforts to educate each other and share your experiences of living with PF. We encourage you to use this as your workbook and to share it with other members of your group.

The kit is divided into three sections: Organization and Management; Suggested Topics; and Resources.

Organization and Management Information

The first three sections of this kit include tips and information on managing a PF support group with a limited budget. The advice included in this kit is designed to help the volunteer or group of volunteers with little support or resources – get started.

Suggested Topics

This section of the kit provides a list of suggested topics and speakers that could be used by a support group for an education-focused session. The support group setting is an excellent opportunity to present and discuss issues critical to the health, outlook and lifestyle of PF patients.

This section also includes several topic outlines and handouts. These outlines have been prepared to ease the preparation work of the presenter. For example, if your support group is interested in learning more about the nutrition concerns, you can simplify planning by sharing the outline *Keeping Healthy*, with a dietitian or nutrition specialist you would like to invite as a guest speaker for this educational session.

Resources

A collection of handouts and helpful materials to share with your support group members is included at the end of this kit. The CPF will continue to provide information and access to new materials as they become available. Please check our website at www.coalitionforpf.org for updates. The CPF also encourages you to let us know about any other useful articles, booklets or handouts so that other support groups can use them as resources, too.



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GETTING STARTED

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Why a Support Group?

Working together is key to dealing with life's many challenges, and learning to deal with a recent diagnosis, new medical treatment or an uncertain future is no exception.

Pulmonary Fibrosis (PF) is a serious and life-threatening lung disorder that is difficult to diagnose and treat, and there remains little understanding around why the disease occurs or how to stop it. Receiving a diagnosis of PF, or another equally devastating disease, can leave patients and their loved ones feeling lost, disconnected and uninformed.

Support groups can provide PF patients and their loved ones with a much-needed network of people who have had similar experiences. A support system of peers as well as medical experts can inform, comfort and guide patients in dealing with their diagnosis.

As is to be expected with a diagnosis of a little-known and life-threatening disorder, some patients are full of questions, "what if" and a desire to find a light at the end of the tunnel. For others, a sense of defeat and hopelessness consumes them as they struggle with the harsh reality of their diagnosis.

Regardless of a patient's outlook or stage of disease progression, PF support groups can begin and sustain dialogues that are as critical to the healing and disease management process as medical treatment.

Learning from Others

Starting a support group is a big job, so before you get going, be sure to do a little bit of research – you don't need to reinvent the wheel to establish a successful support group. *Before you start planning, see what others are already doing in your area and check the listing of current CPF support groups at www.coalitionforpf.org. Contact information for support group leaders is included in most listings, so feel free to reach out to those leaders for information or advice.* You can find other existing support groups by calling local hospitals, lung associations or by searching their websites.

The Support Group Leadership Team

A few people and even sometimes one person can serve as a support group team. If you can cultivate others to help, the ongoing tasks of the support group will be easier for everyone.

Membership Coordinator

The membership coordinator's role would be to gather, organize and maintain current membership records including each member's contact information. (See sample contact list *Facilitating A Support Group.*)

Programming Coordinator

The programming coordinator's responsibilities include managing all logistics related to each meeting (including selecting the location, time, refreshments, room set up, etc.), meeting format and guest speaker scheduling. The programming coordinator position is best done by you, if time permits, as you will be facilitating the meeting (See *Suggested Topics for Education Session* for more information on inviting speakers). It is a good idea to send a thank you note to presenters, telling them you appreciated their time and insight; it is a simple gesture that may encourage them to come back next time they are asked.

Publicity Coordinator

The publicity coordinator's job is to promote the meetings by way of newsletter, community calendar postings, flyer distributions, hospital and pulmonary rehabilitation clinic flyers, online promotion (especially through social media like Facebook and Twitter).

Librarian

The librarian oversees all credible PF related information resources that are to be made available for member review at each meeting.

Medical Advisors

Every support group should include support group advisors – specialists who are accessible to the group for questions and insight from a clinical perspective. They do not need to attend each meeting, but should be available to attend occasionally and to serve as a reliable resource for group members. Support group advisors could include pulmonologists, registered nurses who specialize in pulmonary care, registered respiratory therapists, registered dietitians, hospice workers, grief counselors and social workers.

Identifying the Needs of Your Group

Before you initiate a support group, it is important to find out as much as possible about who they are and what they are looking for in a support group setting.

One way to learn more about your potential membership is to *conduct a simple survey to determine what people are interested in gaining from attending our group*. (See sample interest survey at the end of this section). Do they want to have an opportunity to share with and learn from other patients who are battling PF? Do they want more access to healthcare professionals who can teach them more about their disease? Are they interested in learning about emerging treatment options?

Also, it is a good idea to find out where they currently go for resources, their doctor's name and how long they have had PF. Ask if they have attended another support group and what they are looking for from yours.

As you move forward, *be sure to stay in touch with the needs of your group members*. Periodically ask them about their needs and whether or not those needs are being met. If something is lacking, ask them what they think can be done to help better meet their needs.

Other Considerations

After you have the results from your interest survey, you should be able to identify what direction you will take with your support group. Knowing what your members are interested in will help you in selecting a name for your group.

The group's name should be easy to remember and one that is fitting with the nature and goals of your group. Some suggested names include: *PF Support Group of (insert name of your community)*, *PF Patient Support Network*, *PF Support Group*, and *Living with PF*.

Support Group Interest Survey

Please provide the following information to help us in our efforts to meet your needs in this support group.

Date: _____

Name: _____

Doctor's Name/Hospital Affiliation: _____

1. What are you most interested in gaining/learning from attending this group?
2. Why did you decide to join this group?
3. What is your connection to PF (are you a patient, family member, caregiver)? If you are a patient, how long have you had PF?
4. Where do you go to find information on PF?
5. Have you attending another support group?
6. Please share any additional comments, questions or concerns.

Talk with others who have successfully started a support group. Refer to the CPF website for contact names of others who have set up support groups for PF (www.coalitionforpf.org). Ask them about the methods they utilized to make the group work. Find out what worked best and what posed problems to help you avoid similar issues. Many communities have “Better Breather” or “Easy Breather” support groups for patients with lung disorders. Your local American Lung Association (ALA) office can provide you with information. Most communities have cancer support groups, as well, that could offer some guidance.

If possible, consider attending one or two support group meetings that are similar to the one you are starting to get an idea of how they operate. After visiting, you can borrow what you consider to be their best techniques for use in your group. It is a good idea to call the support group leader prior to attending a meeting to let him/her know you would like to visit their group.

What is the Role of a Support Group Coordinator?

As a support group leader/coordinator, it is easy to feel overwhelmed by the amount of work required to start or maintain an effective support group. However, you shouldn't feel as if the weight of this entire project is on your shoulders alone. As a coordinator, it is your job to make sure things get done, but not necessarily to do everything by yourself.

Often, the most effective support group coordinators ask others who are interested in a support group to help in its planning and implementation. These individuals can be medical professionals, family members of PF patients, or patients attending the group. Sharing leadership roles and rotating the responsibilities is important so no one feels overburdened or “burned out”. Some of the CPF's most successful support group leaders partner with a local hospital, pulmonary rehabilitation clinic, or local physician's office to provide support and some management of their meetings.

Contact the CPF once you've got the details of your support group set and are planning your first (or other meetings). We can help promote your group on our website and through emails directly to patients who live in your area, as well as through our national newsletter. Call us at 1-888-222-8541 or contact us via email at info@coalitionforpf.org.



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Planning a Support Group

Planning a Support Group

Selecting a Location

The support group location should be carefully selected with the needs of your members in mind. When choosing your meeting place, be sure to consider its accessibility and atmosphere.

Keep in mind that many of your attendees may be seniors and some may be dependent on supplemental oxygen. That said, accessibility is key to ensuring anyone interested in attending is physically able to do so.

The location should:

- have adequate parking near the meeting area
- be close to Public Transportation
- have wheelchair access
- have adequate restroom facilities

Equally important is the atmosphere of your meeting place. Look for a location that fosters comfort, privacy, and security. A busy restaurant or coffee shop, for example, is not an ideal location because they do not allow your members to openly discuss private issues as freely as they might in a private setting.

The location you select should be neutral, centrally located and free of charge. Some examples of possible meeting locations include hospital meeting rooms, church halls, senior centers or independent living centers. Also, you might consider contacting your local chapter of the American Lung Association to inquire about its meeting space or tips on meeting facilities in the area.

Deciding When to Meet

Now that you have given some thought to where you would like to hold your support group meetings, it is time to consider when and how often you should meet. While many existing support groups meet once a quarter, once a month and sometimes more often, to help maintain interest and focus, you will need to decide with your group what works best for you, especially at the beginning.

As with selecting a location, it is important to maintain consistency for the scheduled time of your meeting to minimize the possibility of confusion. You should strive to hold meetings on the same day/time each month (i.e. the first Tuesday evening of every

month). Also, take into consideration the needs of your members. Given that some of them may work and others may be seniors, it is a good idea to schedule the meetings when best for your members to travel. If you are not able to hold the meetings during the week, consider having them on Saturday mornings.

Finally, you will need to decide how long your meetings will last. Depending on what you intend to cover and the interest expressed by your members, you should plan to schedule meetings accordingly. If you plan to coordinate an education session with a presenter, the meeting should last between 1.5 – 2 hours. If you are gathering for an open discussion or time of sharing, the meeting may only need to last 1 – 1.5 hours. *As a rule of thumb, 2-hour meetings should be considered the maximum length of time.*

Always be considerate of your group members' time by starting and ending each meeting on time.

Developing the Program

What you do in your meetings should reflect the needs and interests of your group's members. Based on the information you gather in the interest survey, you will need to decipher what and how much should be covered in the meetings. Determine if members are more interested in support and open discussions or information or both.

If your membership tends to lean more toward support, plan your meetings with a heavy emphasis on group sharing by inviting members to talk openly about their experiences. Consider inviting special guests who might be able to help them professionally, including a counselor or social worker.

For groups that are more interested in learning about the disease, utilize the topic outlines included in this kit for tips and ideas on educational sessions about PF. Also, schedule guest speakers who are qualified to answer complicated medical questions about PF, including pulmonologists, respiratory nurses and therapists, registered dietitians, or social workers who have experience with lung disease patients. Review the CPF contact list in the *Resources* section of this kit to see if there is a CPF Board member in your area for a possible guest appearance (you can contact the CPF for assistance in involving these experts). Additionally, the *Resources* section includes various educational handouts and sources for more information on PF.

Remember that even the best-laid plan is not always the right fit. What may seem to be appropriate at the first meeting may not be of interest to members down the road.

It is important to continually check in with your members to see if they are satisfied with the group and what other needs they want to have met. You should be open to adapting to the needs of the group so it helps them best learn how to live with PF.

Getting the Word Out

Now that you are ready to start a support group, you need to let people know what you are up to. Publicity is crucial to initiating a support group and it is important to utilize all available media. Included in this section are a number of templates to serve as a guide for your publicity campaign. Feel free to use them in entirety or in part for the creation of your own communication tools.

Start by creating a flyer for distribution to local hospital pulmonary rehabilitation clinics, senior centers and doctor's offices (see sample at the end of this section). Because of patient confidentiality issues, you will not be able to obtain a mailing list of PF patients from a doctor or clinic. Instead, deliver the flyers personally or by mail. Follow up with your contacts by way of personal visits or phone calls to ensure the flyers are posted or distributed to patients. Call your local hospital's marketing/communications department or public affairs department to inquire about placing the information in their patient newsletters.

A great way to communicate with your group and potential group members is through a regular newsletter. (see sample newsletter at the end of this section). Keep members up-to-date on the group's activities and programs and let them know about future meetings. Make the publication something worthwhile for your audience by including current information about PF. You can gather information for your newsletter from the News & Events section of the CPF's website (www.coalitionforpf.org), the CPF's patient brochure and the CPF's newsletter (both on the CPF website) and your local lung associations. You can also be updated on news by registering your email address and the addresses of your support group members (with their permissions) with the CPF through the website or by emailing info@coalitionforpf.org. It's free to join the CPF and all newsletters and information are also free.

To make things easy for you and your group, plan to write and distribute this newsletter publication monthly or quarterly. You may do so in hard copy, but email is the most inexpensive and quick option. Call local hospitals, pulmonary rehabilitation centers or pulmonologists' offices to inquire about an in-kind donation of postage or copying.

Another way to publicize your group's activities is through community calendar postings (see sample letter for community calendar listings at the end of this section). Postings in newspaper, television and radio community calendar listings are free of charge and have the potential of reaching a large audience. Once you have drafted your request letter, compile a list of your area's newspaper, television and radio stations. Call the media outlets on your list and ask for the name of the public affairs director or whoever is in charge of community calendar postings and be sure to get their contact information.

Keep in mind that most media outlets require a four-to-six week period from the time the letter is received to placement, so it is important to plan ahead. After the request letters are sent, it is appropriate to make a round of follow-up calls to ensure the letter was received and to see if it is under consideration for their calendar listings.

Finally, be sure to register your support group with the CPF by emailing us to let us know the details at info@coalitionforpf.org or call us at 1-888-222-8541. This will not only keep you on the CPF's distribution list for updated educational materials and resources, but it will allow the CPF to refer PF patients and family members from your area to your support group. The CPF will also post your support group to our website and will send out emails to patients in your area letting them know about your meeting (just email the CPF at info@coalitionforpf.org at least two weeks in advance with all of the details for the meeting).

Membership in the CPF is open to all individuals and organizations interested in promoting awareness of pulmonary fibrosis and in helping to advance pulmonary fibrosis education, patient support, treatment, and research. No membership fee is required, though donations are encouraged.

Membership Benefits Include:

- E-mail updates on breaking pulmonary fibrosis news
- Access to peer-reviewed educational materials authored by our world-class scientific advisory board
- Access to the latest information on clinical trials
- A free subscription to the CPF Action Alert Quarterly newsletter
- Information on local education and awareness events
- Opportunities to become involved in education and awareness efforts in your community
- Recognition as a CPF partner (organizations only) in all public information and materials



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FACILITATING A SUPPORT GROUP

Facilitating a Support Group

Create a Supportive Environment

Think of facilitating a support group as hosting a warm gathering at your home. You want your guests to immediately feel welcome and comfortable, and their participation to be as convenient as possible.

There are a number of ways to involve members in discussion, and to make them feel safe and comfortable in a new environment. The *"The How to Create an Encouraging Environment"* guide (at the end of this section) offers ten tips for ensuring that your group's sessions flow smoothly.

Be an Effective Facilitator

Facilitating the work of a support group can be a very rewarding experience. One rule of thumb is to be flexible –avoid having an agenda that is too rigid or fixed. Your group will quickly let you know what topics they want to learn about or what issues they need to work on; your job will be to help guide them along the way in order to meet their needs.

Identify and Recruit Presenters

Guest speakers add variety and expertise to your support group sessions. While it is not necessary to invite a guest to present a topic at every session, someone such as a pulmonologist or registered dietitian can bring better understanding to difficult-to-answer questions and pressing concerns.

When inviting a guest speaker to present to your group, first consider what you want to learn from him/her, then use your local resources and contacts to secure the speaker. Examples of topics, resources and contacts are included in the *Suggested Topics for Education Sessions* section of this kit.

Speakers or resources can easily be found within your community. A few places to start include:

Your Local Institutions – pulmonary rehabilitation centers, churches, hospital speakers bureaus, occupational rehabilitation centers

Your Contacts – registered nurses, doctors who specialize in pulmonary care, registered dietitians, social workers with hospitals, medical librarians, senior/disability representatives, PF patient and/or their friends or family members

The CPF – don't forget to check with the CPF to determine if one of its Advisory Board members or one of the Board of Directors are in your area and available to come to your meeting. A list of the CPF board members is available on the CPF website at www.coalitionforpf.org. You can contact the CPF offices to assist in contacting one of these experts or seeking pulmonologists in your area.

Once you have identified the appropriate person to speak at your support group meeting, *be sure to give your guest ample notice, a clear definition of what his/her role is and what you hope your group will learn.*

Also, be sure to indicate how long he/she should be prepared to speak and don't forget to save time for a question and answer session at the end.

Member Contact List

Collecting current contact information from your group members is important to staying in touch with them. Email is especially important as it is an easy way to stay in touch! Sometimes asking for personal contact information, however, can be a sensitive issue, so be sure to indicate the list will not be sold or shared with anyone outside of the group. If your group would like to be updated on information regarding PF, ask for their permission to have them added to the CPF mailing list and become a CPF member. It's free and it's the best way to stay updated and informed. Email your list, once your have member approval, to info@coalitionforpf.org.

The member contact list should be made available to all members and should include necessary contact information like name, phone number and email address. You, as facilitator, can own the list or, if the entire group agrees, you can distribute copies of the list to the group. Be sure to ask about the best way to get in touch with each member – phone, mail or email.

A template for your support group's contact list is provided at the end of this section.



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Suggested Topics for Education Sessions

Suggested Topics for Education Sessions

Suggested Topics for Possible speakers

For many of your support group meetings, you may want to invite guests or medical experts to lead a discussion on topics relating to the treatment and management of PF.

As was mentioned in the previous section of this kit, *Facilitating a Support Group*, guests can add new depth to topics and can be found in your own backyard.

Below is a list of possible topics (for one or more support group sessions) and suggested healthcare professionals or education advisors for each of those topics. Several topic outlines and handouts are included in this kit for you to offer to your guest as a potential guide or tool to help him/her design a presentation.

Remember to use your resources and friends to help identify the right presenter for the topics of greatest interest to your group.

What is PF?

Possible speakers: A respiratory nurse, therapist or pulmonologist involved with interstitial lung disease is best to address this topic. A doctor who is involved in research or at a teaching hospital can offer a good overview of the disease, as well as its history. (See the presentation outlines: *What is PF? and Emerging Treatments and Therapies*, at the end of this section.)

Oxygen Use

Possible speakers: A home care respiratory therapist, respiratory nurse, or an oxygen supplier representative is recommended for this topic.

Traveling with PF

Possible speakers: A respiratory therapist or pulmonary rehabilitation specialist can provide excellent advice on this topic. There are also travel groups that specialize in traveling with oxygen (perform an Internet search for currently available resources on the subject), such as Medical travel (www.medicaltravel.org or 1-800-778-7953) and The Oxygen Traveler (www.theoxygentraveler.com or 1-800-308-2503), as well as www.access-able.com or 303-232-2979.

Managing Breathlessness and Cough

Possible speakers: Respiratory nurses are the best qualified speakers. The American College of Chest Physicians has a publication called *Cough* that can help your group understand coughs by disease (See presentation outline *Managing Shortness of Breath and Cough*, at the end of this section).

How to Maximize the Benefits of Exercise

Possible speakers: Pulmonary Rehabilitation specialist, occupational therapists or exercise physiologist who has experience with chronic lung disease patients.

Understanding your Medications

Possible speakers: A pulmonary nurse can provide guidance, as well as a pulmonologist or doctor from a hospital pharmacy can be excellent facilitators for this discussion. Ask a pharmacist to visit your support group and help patients by reviewing their current medications to determine if they are all necessary and if there are any potential harmful drug interactions. (See presentation outline, *Medications*, at the end of this section.)

Using and Finding Local Support

Possible speakers: Social worker from local hospital or representative from local lung association chapter. Hospitals often have speakers bureaus, including respiratory specialists.

Test Results: What do they Mean?

Possible speakers: A pulmonologist or radiologist can provide the best detail on lab tests and lung scans; pulmonary rehabilitation specialists are helpful for pulmonary function tests.

Energy Conservation

Possible speakers: An occupational therapist at a hospital is the best resource for physical energy conservation. Also, consider discussing environmental energy conservation; some companies offer discounts for people with disabilities, including oxygen use.

Keeping Healthy:

Possible speakers: A Registered dietitian and/or a respiratory nurse (See presentation outline *Keeping Healthy* at the end of this section for more detail).

Finding Medical Information on the Internet:

Possible speakers: Education specialists and computer experts at your local college, library or lung association chapter can provide guidance, and in some cases, provide support group members with access to computers.

Session Outline #1:

1. Definition of PF:

Pulmonary Fibrosis (PF) is a lung disorder characterized by a progressive scarring – known as fibrosis -- and deterioration of the lungs, which slowly robs its victims of their ability to breathe. Approximately 128,000 Americans suffer from PF, and there is currently no known cause or cure. An estimated 48,000 new cases are diagnosed each year. PF is difficult to diagnose and an estimated two-thirds of patients die within five years of diagnosis. Sometimes PF can be linked to a particular cause, such as certain environmental exposures, chemotherapy or radiation therapy, residual infection, or autoimmune diseases such as scleroderma or rheumatoid arthritis. However, in many instances, no known cause can be established. When this is the case, it is called idiopathic pulmonary fibrosis (IPF).

2. Interstitial Lung Diseases:

PF is one of approximately 200 related diseases known as interstitial lung diseases (ILDs). PF is one of the most common forms of ILDs – an estimated 128,000 people have PF in the U.S. (for a complete list of ILDs, refer to the *Interstitial Lung Disease, III Edition* in the *Resources* section).

3. Confusion in the Name

Until the last decade or so, many ILDs and/or idiopathic interstitial pneumonias were grouped and identified as PF. A newer and specific clinical definition of PF now separates it from other conditions including: desquamative interstitial pneumonia, respiratory bronchitis interstitial lung disease, acute interstitial pneumonia, nonspecific interstitial pneumonia and cryptogenic organizing pneumonia. All of these diseases are characterized by pulmonary fibrosis and inflammation, making diagnosis difficult.

4. Other Disorders with Pulmonary Fibrosis

Other lung disorders are also characterized by pulmonary fibrosis, however their causes are known and in some cases, effective treatment is available. These disorders include sarcoidosis, scleroderma, systemic lupus and rheumatoid arthritis.

5. What PF does to the Lungs

Although what causes PF is unknown, the result is clear. An abnormal and exaggerated healing process produces fibrous scar tissue. The scarred tissue causes the lung's tiny air sacs, alveoli, to thicken and stiffen, making it difficult to process oxygen. The scarring is progressive.

6. Challenges of Diagnosis

PF is a diagnosis of exclusion – many other diseases with similar characteristics and symptoms must be ruled out first. New standards call for a 'multiple disciplinary diagnosis' that relies on the expertise of a pulmonologist, radiologist and pathologist. For a firm diagnosis, some cases require a lung biopsy.

Suggested Handouts for Education Session #1. *What is PF?:*

- CPF booklet, *Let's Talk About Pulmonary Fibrosis*, a free brochure with comprehensive information on PF, oxygen management and lung transplant designed for patients, families and caregivers; available on the CPF website (www.coalitionforpf.org) or hard copies may be ordered by emailing info@coalitionforpf.org or calling 1-888-222-8541.
- Diagnosis Explanation Sheet (taken from CPF booklet, *A Reporter's Guide to PF* included in *Resource* section of kit
- *Understanding Interstitial Lung Disease: A guide to Managing Your Lung Condition*" booklet published by Krames, the StayWell Company. Copies can be purchased by calling 1-800-333-3032 or via their website at www.krames.com.
- American Thoracic Society _____

Session Outline #2: PF Patient Evaluation and Management

This education session was prepared by Robert Strieter, MD, Henry B. Mulholland Professor of Medicine, University of Virginia School of Medicine, Charlottesville, VA.

The process that a pulmonary physician will use to determine if a patient has idiopathic PF or if the PF may be related to an underlying specific cause, such as a rheumatological disorder, is as follows:

1. Medical History and Exam

The patient will undergo an extensive history and physical examination. This process highlights whether the patient fits a specific age group; the duration of symptoms, the magnitude of shortness of breath in relation to activities of daily living; medication use; family history of relatives having PF; occupational and exposure history; and whether on physical examination, for example, there are the presence of skin lesions, arthritis, crackles on auscultation (listening with a stethoscope), and clubbing of the digits.

2 Laboratory Tests

The patient will undergo a number of laboratory tests to assess their white blood count, differential, and platelet count; chemistry profile to measure their electrolytes, kidney and liver function; serological evaluation to rule in or out an associated rheumatological disorder; and urine analysis.

3. X-Rays

The patient will undergo a plain chest x-ray, or the patient may immediately undergo a high resolution CT of the chest to better visualize the lung tissue.

4. Pulmonary Function Testing

The patient will undergo pulmonary function testing that will evaluate their lung volumes, airway mechanics, and gas exchange. Under certain circumstances, some pulmonary physicians will want to evaluate a patient's lung compliance; oxygenation during sleep, rest, and with exercise using a treadmill or bicycle ergometer.

5. Lung Biopsy

The patient may be requested to undergo a bronchoscopy of their airway/lungs that will include bronchoalveolar lavage and biopsy of their lung tissue. This is usually done as an outpatient. Under certain circumstances, the patient may be requested to undergo an open lung biopsy that will require hospitalization and is usually performed by a thoracic surgeon.

6. Management

Once the diagnosis is consistent with PF and not related to an underlying rheumatological disorder, the patient's pulmonary physician will discuss the available therapeutic options. Once the therapeutic option is decided upon, then the patient is

followed in a longitudinal manner. These visits may be as often as once per month or as long as every 3 to 6 months. The clinic visits are often associated with the assessment of the following:

- The magnitude and change in shortness of breath in relation to activities of daily living (this could include sexual activity), and whether their quality of life has been altered over time.
- Adverse effects of any medications, including potential infectious disease complications related to the medications that the patient may be taking at the time.
- Pulmonary Function Testing that will evaluate their lung volumes, airway mechanics, and gas exchange. Under certain circumstances some pulmonary physicians will want to evaluate the patient's lung compliance, oxygenation during sleep, rest and with exercise using a treadmill or bicycle ergometer.
- A repeat chest x-ray or high resolution CT of the chest may be requested. This is usually performed if there is evidence of a significant change that has occurred with one or all of the above parameters.
- Laboratory tests may be requested depending on the medication prescribed to treat the patient's PF. For example, the patient may be requested to obtain white blood count, differential and platelet count; chemistry profile to measure their electrolytes, kidney and liver function; and urine analysis.

Session Outline #3: Keeping Healthy

1. Healthy Eating

It is important to eat a well-balanced diet, including meat, bread and plenty of fruits and vegetables. A healthy diet helps to support your body and keeps up your strength. However, there are certain foods that may cause you additional discomfort due to gas (i.e. beans, spicy foods). Consider eating smaller, more frequent meals during the course of your day. Many patients find it easier to breathe when their stomach isn't completely full. If you have any specific nutritional concerns, talk to your doctor, nurse or a registered dietitian.

1. Regular Exercise

Try some moderate exercise, such as walking or riding a stationary bicycle. If you're already exercising, keep up your regular workout. This helps you maintain strength and lung function. Be sure to talk to your doctor before starting a new exercise program. Those who need to can generally use supplemental oxygen during this kind of activity.

2. Pulmonary Rehabilitation

Consider enrolling in a pulmonary rehabilitation program to help increase your strength, learn breathing techniques and expand your social support network. Ask your doctor or nurse for more details about pulmonary rehabilitation programs in your area.

3. Supplemental Oxygen

Your doctor may have prescribed supplemental oxygen, which can provide your body with the oxygen it needs but your lungs can no longer supply. Some patients fear they will become addicted to oxygen, but this is not true. Supplemental oxygen can help you feel less breathless and more energetic. There are risks associated with not getting enough oxygen when you need it such as risk for developing pulmonary hypertension. Discuss your oxygen needs with your healthcare provider to make certain you have the correct amount of oxygen support when you need it.

4. Taking medication

Be sure to carefully take all of the prescribed medications as prescribed by your doctor. Be sure to be open and honest with your doctor about all of your medications – prescriptions and over-the-counter – to help avoid unnecessary adverse reactions. If you notice anything unusual about how you are feeling or how your medicines are working, call your doctor right away.

5. Smoking Cessation

If you smoke, it is extremely important that you stop as soon as possible, especially if you are on supplemental oxygen. Smoking not only causes harm to your lungs, but smoking near oxygen poses a dangerous explosive hazard. Ask your doctor or nurse about smoking cessation programs and products that can help you overcome the dangerous habit.

Suggested Handouts for Education Session #3, *Keeping Healthy*:

- *Let's Talk about Pulmonary Fibrosis*, a free CPF booklet (available at www.coalitionforpf.org for download or to order copies, call (888) 222-8541.
- American Dietetic Association food guide pyramid; see www.eatright.org for specific recommendations on eating healthy or for information on dietitians.
- For help with smoking cessation, visit the Surgeon General's website at: <http://www.surgeongeneral.gov/tobacco> for specific information or find information at <http://www.jointogether.org/resources/2009/sclc-catalogue-of-tools.html> for information provided by the Smoking Cessation Leadership Center or call them at (415) 502-8880.

Session Outline #4: Managing Shortness of Breath and Cough

Prepared by Susan Jacobs, RN, MS, Director of Clinical Research, Stanford University Medical Center, Stanford, CA

The patient with PF lives with two dominating symptoms that impact their daily social and functional activities: shortness of breath and a dry, hacking cough. An understanding of strategies to manage breathlessness and cough assists patients and their families to cope with this chronic lung disease

1. Shortness of Breath (also called dyspnea)/fatigue

The shortness of breath in interstitial lung disease is a result of increased lung stiffness and the resulting increased work to get the air in and out of the lungs. Even with a normal arterial oxygen level, the brain senses it is putting out a great deal of effort or work for very little return in the way of tidal volume (the volume of gas inhaled and exhaled in one respiratory cycle).

Breathlessness in PF can be profound despite normal oxygen levels because of this increased work and the sensation of uncomfortable, difficult breathing. It is important for the patient to know that even though oxygen saturations are normal, he/she may still feel extremely breathless because of the amount of work it takes to inflate stiff, non-compliant lungs. Therefore, breathlessness is not necessarily an indicator for the need of supplemental oxygen. However, if the patient's oxygen levels are low (below PaO₂ of 60mmHg or below 90% saturation) the increased respiratory drive of hypoxemia (a low level of oxygen in your blood) further increases the patient's sensation of uncomfortable breathing.

A regular exercise program may help decrease breathlessness by reversing the downward spiral of breathlessness/inactivity that can lead to significant deconditioning and worsened dyspnea. Avoiding activities where one become short of breath leads to a lifestyle of social isolation and physical inactivity. A regular exercise program can increase strength, flexibility, and endurance as well as that it can improve mood, and enhance sleep.

Strategies to be included in helping patient manage their breathlessness include avoiding infections, stopping smoking, maintaining an ideal body weight, eating small frequent meals, and learning energy conservation techniques.

Managing shortness of breath is most effective if multiple tactics are employed. In addition, explaining the mechanisms of breathlessness in PF is the first step in helping patients understand how various interventions may work.

Tips:

- Stop smoking to decrease irritation to the airways, loss of lung function and increased susceptibility to infections
- Learn controlled breathing techniques to slow your breathing rate and relax breathing muscles. Shortness of breath becomes worse when you tense up all of your accessory chest muscles and panic
- Use positions to relax chest muscles such as resting elbows on the table, or elevating them slightly as on the back of a chair
- Maintain ideal body weight. Being underweight can affect your immune response and being overweight puts an additional load on your heart and lungs
- EXERCISE REGULARLY (aerobic, strength, stretch) to:
 - Strengthen muscles
 - Build endurance
 - “desensitize” yourself to breathlessness
 - Improve your mood
 - Enable you to get through your day more easily
 - Listen to music or watch TV or do anything to distract yourself during the exercise to make it go by faster and easier
 - Use oxygen if prescribed
 - Ask your physician for a referral to a local pulmonary rehabilitation program that will provide you with structured education and exercise sessions along with others who have chronic lung disease
- Avoid large meals; a distended stomach can push up on your diaphragm adding to a sense of breathlessness
- Pace yourself to conserve energy for the fun stuff! Break large tasks such as meal preparation or a house project into small steps that can be spread out over a few days

2. Cough

The cough experience by patients with PF is thought to be the result of interstitial lung receptors sensing inflammation and scarring surrounding the alveoli. The cough is dry and hacking and can worsen dyspnea, cause desaturation, and sometimes result in pre-syncopal episodes (near fainting). Patients describe profound dyspnea and panic along

with these episodes of coughing. The cough is difficult to treat and may often be the single most bothersome complaint of the patient.

Oral or inhaled prednisone may or may not provide any relief. Use of narcotics such as codeine preparations may suppress the cough as well as provide some relief of dyspnea. However, the decision to begin the use of narcotics is one that is made after all other alternatives have been tried. Benzonatate (Teesalon Preles) is an oral cough suppressant that directly acts on the brain's cough center, the medulla. Other alternative remedies used by patients include using various throat lozenges or sipping honey and lemon in hot water.

Patients should be instructed to avoid known irritants that may trigger cough such as fireplaces, BBQs, aerosol sprays, etc. If the patient is known to desaturate during these episodes, increasing oxygen flow rates may help prevent other side effects of dizziness and increased breathlessness (consult your physician before increasing oxygen flow).

- Interstitial nerve receptors may trigger dry hacking cough in patients with fibrosis
- Cough can be difficult to treat
- Medication options:
 1. Prednisone: oral or inhaled? May or may not be helpful
 2. Codeine: a narcotic cough suppressant which may be helpful temporarily or at night. Side effects of drowsiness and constipation can be a problem
 3. Tessalon Perles: another type of cough suppressant with fewer side effects; effectiveness varies
 4. Honey and Lemon in hot water
 5. Lozenges: try different ones, some may help
- Avoid irritants: fireplaces, BBQs, sprays, etc.
- Increase oxygen during cough spells if needed

3. Anxiety and Panic with Shortness of Breath (SOB)

Panic control or relaxation techniques are also critical for patients to practice when they are not experiencing shortness of breath.

- Learn relaxation techniques – listen to tapes, use headphones
- Learn when SOB occurs and ways to keep it under control so you can PREVENT situations that trigger panic
- Remember that SOB will only worsen if you get anxious or tense
- Have a positive attitude – think about what you *can* do

Session Outline #5: Medications

Physicians generally follow standards of care intended to improve symptoms and hopefully slow the progression of the disease. In 2001, a panel of experts sponsored by the American Thoracic Society and the European Respiratory Society recommended that if therapy was given, it should consist of a trial of corticosteroids and an immunosuppressive agent (e.g. azathioprine or cyclophosphamide). Recent evidence has suggested that adding a third therapy called N-acetylcysteine (NAC) to this regimen may be beneficial. *It is important to recognize, however, that there are no definitive studies showing that this treatment approach is effective, and there is no consensus regarding the use of this approach in the pulmonary community. For a complete listing of active clinical trials for Pulmonary Fibrosis, please visit www.coalitionforpf.org.*

Importantly, these therapies can cause side effects-some minor and some more serious. The potential risks and benefits of therapy should be discussed with your physician in detail. Be sure to contact your doctor or nurse if you have any negative reactions to any medications you have been prescribed.

In all cases, doctors should consider referring their patients to a clinical trial to gain access to experimental treatments and also refer patients for lung transplantation evaluation.

ARE THERE ANY NEW TREATMENTS ON THE HORIZON?

Researchers are developing and testing a variety of new ways to treat Pulmonary Fibrosis. These approaches target the various steps in the disease process. Therapies under investigation include:

- Antifibrotic or anti-fibrogenic agents to suppress the scarring process
- Antioxidants (such as N-acetylcysteine-NAC) to prevent damage to lung tissue
- Endothelin antagonists (such as certain medications for pulmonary arterial hypertension (PAH))
- Monoclonal antibodies to inhibit "bad" cytokines (protein growth factors, such as TGF-beta, TNF-Alpha, or CTGF) that activate inflammation.

There are several emerging therapies currently in clinical trials for Pulmonary Fibrosis patients. Contact the CPF or visit www.coalitionforpf.org for a complete up-to-date listing of active investigational research. The National Institutes of Health (NIH) also offers information at www.clinicaltrials.gov for those interested in identifying Pulmonary Fibrosis-related research at medical centers across the United States. It represents the most up-to-date research information for patients including clinical trial sites, patient inclusion/exclusion criteria, and contact information for each trial.

As with any course of care, please consult your physician to determine what current treatment options, including participation in clinical trials, may be appropriate for you.

ARE YOU A PATIENT WITH A FAMILIAL FORM OF PULMONARY FIBROSIS?

The Coalition for Pulmonary Fibrosis (CPF) and National Jewish Medical and Research Center partnered to launch the first genetic counseling program for patients and families affected by FPF. The telephonic counseling program is operated by National Jewish, and funded by CPF. An estimated 10-15 percent of Pulmonary Fibrosis patients have a form of the disease that runs in families, known as familial pulmonary fibrosis (FPF).

The genetic counseling program provides a qualified genetic counselor, who has expertise in FPF, to discuss by phone various issues surrounding FPF. These can include preparation for and interpretation of genetic tests, and various life decisions, such as having children and planning for the future. Experts recommend talking to a counselor prior to having any genetic tests, so that people are prepared to learn the results.

For further information on the FPF Genetic Counseling Program or to speak with a genetic counselor, call (800) 423-8891, ext. 1097.

LUNG TRANSPLANTATION:

What Every Patient with Pulmonary Fibrosis (PF) Should Know

For certain individuals (usually 65 years old or younger, although program criteria may differ between transplant centers), lung transplantation may also be an appropriate therapy. Lung transplantation can both extend life and enhance the quality of life.

Pulmonary Fibrosis patients should discuss the possibility of lung transplantation as a treatment option with your physician as soon as possible during the course of their care.

The need for transplant evaluation early in a Pulmonary Fibrosis patient's disease course stems from the potentially long waiting time on the transplant list combined with the progressive nature of Pulmonary Fibrosis. According to United Network for Organ Sharing (UNOS), approximately 30% of Pulmonary Fibrosis patients currently listed for transplantation will succumb to their disease prior to transplantation.

WHAT IS A LUNG TRANSPLANT?

Lung transplantation is the placement of one (unilateral) or both lungs (bilateral) from a deceased donor into a recipient with end-stage lung disease. The donor lung must be matched for blood type and size to that of the recipient. The decision to

transplant one or both lungs is dependent upon the extent of disease and the results of pre-transplant testing.

In a single lung transplant, an incision is made on the side of the chest. The patient's old lung is removed and the new lung is then sewn in, connecting the blood vessels to the lung and from the lung to the heart and the main airway. At the end of the operation, the ribs are brought back together and the incision is closed with layers of stitches. Chest tubes are inserted to drain air, fluid and blood out of the chest for several days to allow the lungs to remain fully re-expanded.

In a double lung transplant, an incision is usually made side-to-side across the middle of the chest just under the nipple line and both chest cavities are entered between the ribs. The lung on one side is removed and the new lung sewn in place. The opposite lung is then removed and the second new lung sewn in place. At the end of the operation, the ribs are brought back together and the incision is closed in layers with stitches.

Controversy exists regarding single versus bilateral lung transplantation for certain lung diseases. Patient outcomes for single or bilateral transplantation should be discussed on an individual basis with the transplant center, in the context of their specific experience performing the procedure.

TALK TO YOUR DOCTOR ABOUT LUNG TRANSPLANTATION - TODAY!

It is important to understand that pulmonary fibrosis is a progressive disease for which there is no cure, and that lung transplantation is the only treatment option shown to extend survival for patients with Pulmonary Fibrosis. The American College of Chest Physicians (ACCP) provides the following initial questions that patients should ask their physician and/or transplant center:

- Is transplantation my best option for treatment of my condition?
- What are organ and patient survival rates at this institution for the type of transplant I will need?
- How many transplants of this type are done every year at this institution? How many by my physicians and surgeons? How long have they been doing this type of transplant surgery?
- What costs of transplantation and rehabilitation are covered by my insurance? What out-of-pocket costs will I have to pay?
- Am I likely to get a donor lung more quickly if I get wait-listed at more than one medical center, in different parts of the country?

Lung transplantation may be a viable option for patients with Pulmonary Fibrosis, but don't wait too long to become educated about the details of this procedure. Become proactive and find out more from your physician. This is a commitment you need to make to yourself - today.

Because of the progressive nature of Pulmonary Fibrosis, it is critical for you to discuss lung transplant with your doctor as early as possible to determine if you are a candidate.



Education. Support. Hope.

Resources

Coalition for Pulmonary Fibrosis Contact Information

CPF

Suite F, #227
1659 Branham Lane
San Jose, CA 95118-5226
(888) 222-8541
info@coalitionforpf.org

Visit us on the web at
www.coalitionforpf.org
And on Facebook!

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Frequently Asked Questions

Does the CPF provide information regarding Pulmonary Fibrosis specialists for initial treatment or second opinions?

It's not surprising that many patients ask about getting a second or third opinion. The CPF can easily connect patients with specialists across the country as well as those closest to where they live. The CPF has relationships with PF specialists nationwide. Some of the best doctors in the world are also part of the CPF's Board of Directors and Scientific Advisory Board, which are listed at www.coalitionforpf.org/cpf/boardofdirectors.

What is Idiopathic Pulmonary Fibrosis?

Idiopathic pulmonary fibrosis (PF) is a debilitating disease-marked by progressive scarring of the lungs-that gradually interferes with a person's ability to breathe. Pulmonary Fibrosis belongs to a family of approximately 100 related diseases, called interstitial lung diseases (ILDs) that have similar characteristics and can result in scarring. The lung scarring, a condition typical of these disorders, is referred to as pulmonary fibrosis (PF).

What caused my pulmonary fibrosis?

Sometimes Pulmonary Fibrosis can be linked to a particular cause, such as certain environmental exposures, chemotherapy or radiation therapy, residual infection, or autoimmune diseases such as scleroderma or rheumatoid arthritis. However, in many instances, no known cause can be established. When this is the case, it is called idiopathic pulmonary fibrosis or IPF.

Is Pulmonary Fibrosis the same as Cystic Fibrosis?

No. Cystic Fibrosis is a genetic disease that causes the body to produce abnormally thick, sticky mucus, due to the faulty transport of sodium and chloride (salt) within cells lining organs such as the lungs and pancreas, to their outer surfaces. It primarily affects children. Pulmonary Fibrosis is a completely different clinical diagnosis.

What if my doctor called my condition by another name?

Your doctor may have referred to your condition by one of a number of names that are sometimes used to describe Pulmonary Fibrosis. This confusion may be due, in part, to similarities between Pulmonary Fibrosis and the other forms of interstitial lung disease. Only since 2001 has Pulmonary Fibrosis been recognized as a distinct clinical disorder, meaning that specific clinical criteria were developed to determine a diagnosis. Your physician combines clinical information derived from a medical evaluation and certain diagnostic tests to diagnose Pulmonary Fibrosis according to these criteria.

How many people are suffering from Pulmonary Fibrosis?

Approximately 128,000 people in the United States have Pulmonary Fibrosis, and prevalence is on the rise with an estimated 48,000 new cases developing each year. Prevalence has increased 156% since 2000, when it was thought that only 50,000 were affected.

How does Pulmonary Fibrosis affect a person?

Pulmonary Fibrosis hinders a person's ability to take in oxygen. It causes shortness of breath and is usually associated with a persistent dry cough. The disease progresses over time, leading to an increase in lung scarring and a worsening of symptoms. Unfortunately, Pulmonary Fibrosis is ultimately disabling and can be fatal. The median survival rate is just over three years.

Is Pulmonary Fibrosis genetic?

Pulmonary Fibrosis appears to be genetic in as many as 15% of all Pulmonary Fibrosis cases. Many scientists believe research involving these families may hold the key to understanding the causes of Pulmonary Fibrosis and may lead to future treatments or a cure. A large familial Pulmonary Fibrosis research study is underway at National Jewish Health, Duke University and Vanderbilt University Medical Center (a joint study) to further study the genetic underpinnings of Pulmonary Fibrosis that may be found in families like this. Recent research has identified two genetic mutations that are associated with familial pulmonary fibrosis, and tests for those mutations have recently become available to the public.

If you have two or more cases of Pulmonary Fibrosis diagnoses in your family, you may want to get involved. The CPF and National Jewish Health established the first Genetic Counseling program for families affected by pulmonary fibrosis. For further information on the Familial Pulmonary Fibrosis Genetic Counseling Program, to speak with a genetic counselor, or to learn how your family can participate in this landmark study of Pulmonary Fibrosis, please call 1-800-423-8891, ext. 1022.

What causes Pulmonary Fibrosis?

While the cause of Pulmonary Fibrosis remains a mystery, what researchers do know is that Pulmonary Fibrosis involves changes in the lung's normal healing process. Pulmonary Fibrosis' chronic cycle of injury leads to an exaggerated or uncontrolled healing response that, over time, produces fibrous scar tissue. This scarring, in turn, causes the lung's tiny air sacs, called alveoli, to thicken and stiffen-rendering the lungs less able to function and provide the body with the oxygen it needs.

Why does the lung scarring take place?

Exactly what sets this abnormal tissue-repair process in motion is unclear. The body's own immune response appears to play a major role. Researchers are investigating a number of potential risk factors that may make a person more likely to develop Pulmonary Fibrosis. These risks may include:

- Certain occupational exposures (e.g. metal or wood dust, pesticides)
- Cigarette smoking
- Use of anti-depressant medications
- Asbestos exposure
- Viral infection
- A family history of pulmonary fibrosis
- Acid reflux Disease or GERD

What are the symptoms?

Symptoms of Pulmonary Fibrosis usually have a gradual onset and may include:

- Shortness of breath during or after physical activity
- Spasmodic, dry cough
- Weight loss
- Fatigue

Patients may also develop a buildup of tissue in their fingertips. This condition is called clubbing.

How is Pulmonary Fibrosis diagnosed?

Because Pulmonary Fibrosis has symptoms and an underlying pattern of scarring similar to those of other lung disorders, it can be difficult to diagnose. In fact, in most cases, identifying PF is usually a matter of ruling out other diseases. Also, until recently, the medical community had no agreed-upon standards for the diagnosis of Pulmonary Fibrosis. Consequently, other related diseases may have been mistakenly classified as Pulmonary Fibrosis. With new diagnostic standards now in place, the recognition and management of PF should be substantially improved.

How is Pulmonary Fibrosis diagnosed?

Because Pulmonary Fibrosis has symptoms and an underlying pattern of scarring similar to those of other lung disorders, it can be difficult to diagnose. In fact, in most cases, identifying PF is usually a matter of ruling out other diseases. Also, until recently, the medical community had no agreed-upon standards for the diagnosis of Pulmonary Fibrosis. Consequently, other related diseases may have been mistakenly classified as Pulmonary Fibrosis. With new diagnostic standards now in place, the recognition and management of PF should be substantially improved.

What types of test will my doctor administer to determine if I have Pulmonary Fibrosis?

To diagnose Pulmonary Fibrosis, your doctor will take a complete medical history and perform a thorough physical examination. During this exam, the doctor will use a stethoscope to listen to your chest to try to determine if your lungs produce any abnormal sounds when you breathe. He or she may then order one or more of the following diagnostic tests or procedures.

Chest Imaging	Use of radiologic machines to take, pictures (x-ray or CT scan) of the lungs	To view lung structures look for scar tissue, and assess patterns of scarring
Pulmonary function test	A test using a device with a mouthpiece to measure a patient's breathing capacity	To measure the degree of impairment in lung function
Arterial blood test	A measurement of oxygen levels in blood taken from an artery	To determine how well the lungs are performing vital gas exchange
Exercise Test	A test in which the patient is monitored while using a treadmill or stationary bicycle	To measure how well the lungs and heart respond to physical activity
Bronchoalveolar lavage (BAL)	A "lung-washing" procedure conducted through a flexible tube (bronchoscope) inserted into the airways through the nose or mouth; fluid (salt water) is injected into the lungs and then removed for	To examine cells and look for signs of inflammation in the lungs

inspection

Lung biopsy

A procedure in which a tissue sample is obtained through a bronchoscope (see BAL, above) or by means of a small surgical incision between the ribs (open-lung biopsy)

To obtain a sample of lung tissue for direct examination

What may happen as Pulmonary Fibrosis progresses?

Pulmonary Fibrosis affects each person differently and progresses at varying rates. Generally, the patient's respiratory symptoms become worse over time. Activities (such as walking or climbing stairs) become more difficult. In addition:

- The patient may require supplemental oxygen
- Advanced Pulmonary Fibrosis makes it difficult for a person to fight infection
- Pulmonary Fibrosis causes a lack of oxygen in the blood. This condition (called hypoxemia) puts a strain on the heart and on the blood vessels in the lungs, and may lead to high blood pressure in the lungs (pulmonary hypertension).
- Pulmonary Fibrosis has also been associated with these potentially life-threatening conditions: heart attack, respiratory failure, stroke, blood clot in the lungs (pulmonary embolism), lung infection, and lung cancer.

How is Pulmonary Fibrosis treated?

Physicians generally follow standards of care intended to improve symptoms and hopefully slow the progression of the disease. In 2001, a panel of experts sponsored by the American Thoracic Society and the European Respiratory Society recommended that if therapy was given, it should consist of a trial of corticosteroids and an immunosuppressive agent (e.g. azathioprine or cyclophosphamide). Recent evidence has suggested that adding a third therapy called N-acetylcysteine (NAC) to this regimen may be beneficial. It is important to recognize, however, that there are no definitive studies showing that this treatment approach is effective, and there is no consensus regarding the use of this approach in the pulmonary community. For a complete listing of active clinical trials for Pulmonary Fibrosis, please visit www.coalitionforpf.org.

Importantly, these therapies can cause side effects—some minor and some more serious. The potential risks and benefits of therapy should be discussed with your physician in detail. Be sure to contact your doctor or nurse if you have any negative reactions to any medications you have been prescribed.

In all cases, doctors should consider referring their patients to a clinical trial to gain access to

experimental treatments and also refer patients for lung transplantation evaluation.

Is a lung transplant an option?

In some cases, doctors may consider lung transplantation as a life-saving treatment option for Pulmonary Fibrosis. Please visit the Lung Transplantation section of our Web page for comprehensive information about lung transplantation to learn more. This procedure is most often performed in patients under 60 years of age whose Pulmonary Fibrosis has not responded to other treatments, but eligibility criteria vary between medical centers, which is why it is critical to discuss this potential treatment with your doctor as soon as possible.

What can I do?

If you have been diagnosed with Pulmonary Fibrosis, there are a number of things you can do to take part in your own treatment and help yourself stay healthy. Caregivers may also be interested in the following information, to assist a family member diagnosed with Pulmonary Fibrosis:

- Call your doctor or nurse with any questions about your condition or its treatment. If you notice anything unusual about how you are feeling or how your medicines are working, call your doctor right away.
- Take all prescribed medications as instructed by your doctor.
- Get your influenza (flu) vaccine every year, and also make sure that your pneumococcal vaccine (Pneumovax) is up to date.
- If you smoke, it is very important that you stop as soon as possible. Ask your doctor or nurse about smoking cessation programs and products that can help.
- Your doctor may have prescribed supplemental oxygen, which can provide your body with the oxygen it needs but your lungs can no longer supply. Some patients fear that they will become addicted to oxygen, but this is not true. Supplemental oxygen can help you feel less breathless and more energetic, while protecting your heart and other vital organs.
- Consult your physician about enrolling in a pulmonary rehabilitation or respiratory therapy program to help increase your strength, learn breathing techniques, and expand your social support network. Ask your doctor or nurse for more details. Many patients report improved breathing and quality of life after adding education and exercise to their treatment. Please note you can participate in a respiratory therapy program even if you are currently taking supplemental oxygen.
- Eat a well-balanced diet to maintain in ideal body weight. This helps support your body and keeps up your strength. Discuss any special nutritional concerns with your doctor or nurse.
- Consider eating smaller, more frequent meals during the course of your day. Many patients find it easier to breathe when their stomach isn't completely full.
- Talk about Pulmonary Fibrosis: Get the word out to raise awareness about the disease. Tell your friends, colleagues and local newspapers about your experience – your story can make a difference. Speak and write to members of Congress about Pulmonary Fibrosis and join the CPF in its work to increase Congressional attention to

the disease.

Are there any new treatments on the horizon?

Researchers are developing and testing a variety of new ways to treat Pulmonary Fibrosis. These approaches target the various steps in the disease process. Investigational therapies currently in clinical trials include:

- Antifibrotic or anti-fibrogenic agents to suppress the scarring process
- Antioxidants (such as N-acetylcysteine-NAC) to prevent damage to lung tissue
- Endothelin antagonists (such as certain medications for pulmonary arterial hypertension (PAH))
- Monoclonal antibodies to inhibit “bad” cytokines (protein growth factors, such as TGF-beta, TNF-Alpha, VEGF, or CTGF) that activate inflammation.

A complete list of all clinical trials and research in the U.S. is available by visiting www.clinicaltrials.gov and seeing the update listing for clinical trials on the CPF website at www.coalitionforpf.org.

Where do I turn if I have questions about these trials or want to be involved in a study?

If you have any questions about experimental treatments or ongoing clinical trials, ask your doctor, contact the CPF at info@coalitionforpf.org, or visit www.coalitionforpf.org/cpf_research_clinical.php for an up-to-date listing of active studies, locations, and study-specific contact information.

Once I understand more about Pulmonary Fibrosis, how else can the CPF help?

We are a resource for patients and families across the entire continuum of care for their life with Pulmonary Fibrosis. We educate patients on all of the possible treatment options and clinical trials available. We educate patients about lung transplantation as a potential treatment option. We can also refer them to the best transplant centers, specializing in Pulmonary Fibrosis, in the country, and refer prospective transplant candidates to patients who've received transplants and can share their experience and offer support.

Patients are also educated on the importance of other standards of care associated with PF such as pulmonary rehabilitation, oxygen management, nutrition, and support groups. We are available to discuss end of life issues with patients and their families and can help with any situation that arises during the natural course of their condition.

Our goal is to empower patients with information and education so they can regain a sense of control while facing a disease that has seemingly taken away their control. We talk about our experiences and help others who are active in the search for answers. The CPF has found many friends in those who care about changing the future of Pulmonary Fibrosis. They gain healing and strength from doing this.

such as taking part in our national advocacy campaign, holding a fundraiser, or helping to raise public awareness.

ADDITIONAL RESOURCES FOR PULMONARY REHABILITATION AND OXYGEN MANAGEMENT

American Association of Cardiovascular and Pulmonary Rehabilitation (AACVPR)

www.aacvpr.org

401 North Michigan Avenue, Suite 2200

Chicago, IL 60611

(312) 321-5146

aacvpr@sba.com

American Association of Respiratory Care (AARC)

www.aarc.org

9425 N. MacArthur Blvd. Suite 100

Irving, TX 75063-4706

(972) 243-2272

info@aarc.org

Breathin' Easy

www.breathineasy.com

4848 Massive Peak Way

Antioch, CA 94531-8310

(925) 891-5017

info@breathineasy.com

National Home Oxygen Patients Association

www.Homeoxygen.org

5454 Wisconsin Avenue, Suite 1270

Chevy Chase, MD 20815-6920

RESOURCES FOR TRANSPLANT CANDIDATES AND RECIPIENTS

- Coalition for Pulmonary Fibrosis - www.coalitionforpf.org
- United Network for Organ Sharing (UNOS) - www.unos.org
- U.S. Department of Health and Human Services Organ Donation - www.organdonor.gov
- Second Wind Lung Transplant Association, Inc. - www.2ndwind.org
- American Lung Association - www2.lungusa.org
- Transplant Speakers International, Inc. - www.transplant-speakers.org
- Children's Organ Transplant Association, Inc. - www.cota.org
- National Foundation for Transplants - www.transplants.org
- National Transplant Assistance Fund - www.transplantfund.org

- Medicare Rights Center - www.medicarerights.org

Glossary

This glossary may be useful to you as you learn more about IPF.

Alveoli

Very small air sacs found in the lungs. Click here to see how the alveolar tissue changes with IPF.

ambrisentan (Leitaris)

an endothelin receptor antagonist (ERA) manufactured by Gilead, and currently FDA approved for the treatment of pulmonary arterial hypertension (PAH), an exceedingly prevalent complication for IPF patients as their disease progresses. Currently in Phase III clinical trials as a potential therapy for Pulmonary Fibrosis (January, 2009)

Antifibrotic/antifibrogenic agents

Experimental medications used to suppress the scarring process associated with IPF.

Antioxidants

Experimental medications that may prevent or reverse lung tissue damage caused by IPF.

Azathioprine

A medication commonly prescribed for IPF patients that can suppress the activity of the immune system and reduce inflammation.

Bronchoalveolar lavage (BAL)

A diagnostic technique in which fluid is instilled into the lungs and removed for examination.

Clubbing

A buildup of tissue in the fingertips (or sometimes the toes). Clubbing is a sign of advanced IPF. Click here to see what

Collagen

Proteins found in tendons, bones, and connective tissue. Scar tissue found in the lungs of patients with IPF also contains

Computed tomography (CT or CAT scan) or high-resolution computed tomography (HRCT)

A type of x-ray for which a computer is used to construct a three-dimensional image from a series of cross-sectional images. The procedure generates multiple pictures of your lungs in layers (slices) from the top (at your shoulders) to the bottom (just above your waist). This test requires that you lie on a table while the pictures are taken, holding your breath and letting it out as instructed.

Corticosteroids

Medications used to suppress the activity of the immune system and reduce inflammation. The most commonly prescribed corticosteroid is prednisone.

Cyclophosphamide

A medication commonly prescribed for IPF patients that can suppress the activity of the immune system and reduce inflammation.

Cytokines

Molecules produced by the immune system. A cytokine imbalance may be the cause of IPF.

D^LCO

Stands for the diffusing capacity of the lung for carbon monoxide (CO), the test used to determine this parameter. DLCO is the extent to which oxygen passes from the air sacs of the lungs into the blood.

Dyspnea

Shortness of breath or labored breathing, usually associated with physical exertion.

Epidemiology

The incidence, distribution, and control of a disease in a population.

Etiology

The cause(s) of a disease.

Familial

Tending to occur in more members of a family than would be expected by chance alone.

Fibrosis

An abnormal scarring of body tissue.

Forced Vital capacity (FVC)

The maximum volume of air that a person can exhale after maximum inhalation. It can also be the maximum volume of air that a person can inhale after maximum exhalation. A person's vital capacity can be measured by a spirometer which can be a wet or regular spirometer. In combination with other physiological measurements, the vital capacity can help make a diagnosis of underlying lung disease.

Forced Expiratory Volume (FEV or FEV1)

The volume of air that can be forced out taking a deep breath, an important measure of pulmonary function. The forced expiratory volume in the first second is the FEV1.

Histopathology

Tissue changes (e.g., scarring of the lungs) that accompany a disease and are recognized

by microscopic examination.

Hypoxia

A deficiency of oxygen in the blood

Hypertension

High blood pressure.

Hypoxemia

A lack of oxygen in the blood.

Hypoxia

A lack of oxygen in the tissues of the body.

Idiopathic

Arising spontaneously or from an obscure or unknown cause.

Idiopathic pulmonary fibrosis (IPF)

A disease characterized by progressive scarring (fibrosis) and deterioration of the lungs.

Interferon gamma-1b

A regulatory cytokine that has antifibrotic and antifibrogenic effects and may regulate macrophage, fibroblast, and mast cell function; inhibit a variety of neutrophil-derived cytokines; and modify the balance of Th1 and Th2 cells in the lung.

Interstitial lung disease (ILD)

A general term for the approximately 200 disorders characterized by inflammation and scarring (fibrosis) of the lungs' interstitium. IPF is an example of an interstitial lung disease.

Interstitialium

The tissue layers between the lungs' air sacs (alveoli) and blood vessels.

Lung biopsy

A procedure in which a tissue sample is obtained through a flexible tube or by means of a small surgical incision between the ribs.

Lung transplant

Replacement of a lung or lungs with donor organ(s).

Monoclonal antibodies

Experimental medications for the treatment of IPF. They may inhibit "bad" cytokines.

Pathogenesis

The mode of origin or development of a disease.

Pathology

The abnormalities that characterize a particular disease.

Phase I trial

The first phase of drug testing in humans. It usually involves 20 to 100 subjects and focuses on safety.

Phase II trial

The second phase of drug testing in humans. It involves up to several hundred patients, lasts as long as two years, and focuses on safety and effectiveness.

Phase III trial

The third phase of drug testing in humans. It involves several hundred to several thousand patients, often lasts several years, and focuses on safety, dosage, and effectiveness.

Pirfenidone

An orally active small molecule drug manufactured by InterMune, Inc. that inhibits collagen synthesis, down-regulates profibrotic cytokines, inhibits TNF-alpha synthesis and decreases fibroblast proliferation. Recently completed Phase III clinical trial and is being reviewed by FDA during 2009 (January, 2009)

Prednisone

The most commonly prescribed therapy for IPF. Prednisone is a corticosteroid that can suppress the activity of the immune system and reduce inflammation.

Prognosis

The prospect for survival and recovery from a disease.

Pulmonary

Having to do with the lungs.

Pulmonary Arterial Hypertension (PAH)

is increased pressure in the pulmonary arteries. These arteries carry blood from your heart to your lungs to pick up oxygen. PH causes symptoms such as shortness of breath during routine activity (for example, climbing two flights of stairs), tiredness, chest pain, and a racing heartbeat. As the disease worsens, its symptoms may limit all physical activity.

Pulmonary embolism

A blood clot in the lungs.

Pulmonary fibrosis

Thickening and scarring of the lungs, specifically the pulmonary interstitium.

Pulmonary hypertension

High blood pressure in the lungs.

sildenafil (Viagra)

a vasodilator manufactured by Pfizer, Inc., that is currently FDA approved for the treatment of erectile dysfunction, and being investigated as a possible treatment for Pulmonary Fibrosis by several medical centers in independent clinical trials (January, 2009)

spirometer

an instrument for measuring the volume of air entering and leaving the lungs. Used as a preliminary test for early detection of lung disorders

trepostinil (Remodulin)

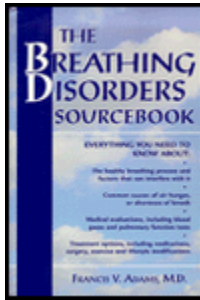
a vasodilator manufactured by United Therapeutics that is administered by IV pump. Currently FDA approved for the treatment of pulmonary arterial hypertension (PAH), and is currently in Phase I clinical trials as a potential therapy for Pulmonary Fibrosis (January, 2009)

Usual interstitial pneumonia (UIP)

A condition indicated by specific changes in the lung tissue. Finding UIP in a lung biopsy strongly points to a diagnosis of IPF (although other criteria must be met).

Recommended Reading

The following books offer information on lung disorders, as well as techniques to improve your breathing and helpful advice on quality-of-life issues. .



The Breathing Disorders Sourcebook

By Francis V. Adams, MD

Lowell House, November 1998



Shortness of Breath: A Guide to Better Living and Breathing

By Andrew L. Ries, et al

Mosby-Year Book, Inc., September 2000



The Lung Transplantation Handbook

By Karen A. Couture

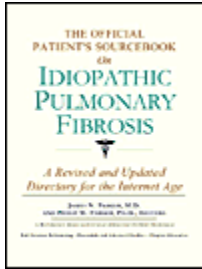
Trafford Publishing, 2001

To purchase this book, [click here](#).



Coping with Prednisone

by Eugenia Zukerman and Julie R. Ingelfinger, M. D.
New York: St. Martin's Griffin, 1998



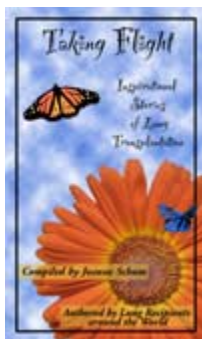
The Official Patient's SourceBook on Idiopathic Pulmonary Fibrosis

James N. Parker & Philip Parker
ICON Health Publications, June 2002



Share the Care: How to Organize a Group to Care for Someone Who Is Seriously Ill

Cappy Capossela & Sheila Warnock
Simon & Schuster, Sept. 1995



Taking Flight - Inspirational Stories of Lung Transplantation

Compiled by Joanne Schum

Authored by lung recipients around the world

October, 2003

To purchase, please visit <http://www.trafford.com/robots/02-0497.html> or <http://www.trafford.com/>



Adventures of an Oxy-Phile

Thomas L. Petty, MD

May, 2004

Publisher

Web Sites and Online Resources

Medical Associations/Societies

American Association of Cardiovascular and Pulmonary Rehabilitation
www.aacvpr.org

American College of Chest Physicians
www.chestnet.org

American Thoracic Society (ATS)
www.thoracic.org

Government Agencies

National Heart, Lung, and Blood Institute (NHLBI)
www.nhlbi.nih.gov

National Institutes of Health (Medline)
www.nlm.nih.gov/medlineplus/pulmonaryfibrosis.html (pulmonary fibrosis information)

National Library of Medicine
www.nlm.nih.gov/medlineplus/druginformation.html (drug information)

Clinical Study Results - Voluntary database of clinical trial results
www.clinicalstudyresults.org

Food and Drug Administration (FDA)
www.fda.gov

Treatment and Research Institutions

Centerwatch - www.centerwatch.com - An international listing of clinical research trials, and physicians and medical centers performing clinical research

University of Alabama at Birmingham
<http://www.health.uab.edu/> (search for "IPF")

Univ. Colorado Health Sciences Center- Dept. of Pulmonary & Critical Care Medicine
<http://www.coloradopulmonarycriticalcare.org/>

Duke University
www.fpf.duke.edu

Emory Center for the Treatment and Study of Interstitial Lung Disease
<http://www.emoryhealthcare.org/departments/lung/index.html>

Inova Fairfax Hospital
General info: www.inova.org/inovapublic.srt/ifh/index.jsp
Lung Transplant info: www.inova.org/lung

University of Iowa (Virtual Hospital)
www.vh.org/adult/patient/internalmedicine/interstitiallung/index.html

Univ. of Miami Medical Center Dept. of Pulmonary & Critical Care Medicine
<http://www.med.miami.edu/med/pulmonarymed/>

National Jewish Medical and Research Center
www.njc.org/diseases/dt10.html

University of Pennsylvania Medical Center- Lung Center
<http://www.uphs.upenn.edu/lungctr/>

University of Pittsburgh Medical Center
<http://ipf.upmc.com>

UC-San Francisco/San Francisco General Hospital
<http://pulmonary.ucsf.edu/>

Univ. of Washington Dept. of Pulmonary & Critical Care Medicine
<http://depts.washington.edu/pulmcc/>

University of Minnesota Center for Advanced Lung Disease
<http://www.cald.umn.edu/>

Vanderbilt University Medical Center
<http://www.mc.vanderbilt.edu/root/vumc.php?site=ipfcenter&doc=6516>

Patient Education and Support Organizations

Pulmonary Fibrosis Friends Online Forum
<http://health.groups.yahoo.com/group/PFF/>

American Lung Association

www.lungusa.org/diseases/pulmfibrosis.html

American Lung Association of California

www.californialung.org

[Breathin' Easy](#)

<http://www.oxygen4travel.com/>

Chronic Lung Disease Forum

www.cheshire-med.com/programs/pulrehab/forum/cldforum.html

Daily Strength.org Online IPF Forum

http://dailystrength.org/discussions/Pulmonary_Fibrosis/messages

Huff 'n Puff Online Lung Disease Forum

<http://www.huff-n-puff.net/newforum/>

MDLinx.com

<http://www.pulmonologylinx.com>

Needy Meds.com

www.needymeds.com

The Pulmonary Paper

www.pulmonarypaper.org

Young Lung Online Support

Online Support: www.geocities.com/younglungz

Email Group: www.topica.com/lists/younglung

Resources for Patient Assistance RX Programs and Discount Card Programs

BenefitsCheckUp

www.benefitsCheckUp.org

Free, online screening tool to more than 240 RX saving programs.

Volunteers in Health Care:

(877) 844-8442

Guide to Patient Assistance Programs.

NeedyMeds:

www.needymeds.com

Directory of Patient Assistance Programs

Pharmaceutical Research and Manufacturers of America (PhRMA):

www.Phrma.org

Directory of Patient Assistance Programs

RxHope:

www.rxhope.org

Information and application forms for Patient Assistance Programs.

Health Resources and Services:

(888) ASH-HRSA (888) 275-4722

Referral source for community health centers that may offer RX assistance.

Local Area Agencies on Aging:

www.eldercare.gov or (800) 677-1116.

Provides assistance to patients 65 and older.

Finding a Doctor

American Medical Association (AMA Physician Select)

www.ama-assn.org

Lung Transplant Information and Support

Second Wind Lung Transplant Association

www.2ndwind.org

United Network for Organ Sharing (UNOS)

www.unos.org

Caregiver Support Organizations

Caregiving.com

www.caregiving.com

Well Spouse Foundation

www.wellspouse.org

Inova Fairfax Hospital

<http://www.inova.org/inovapublic.srt/transplant/index.jsp>

End-of-Life Support Organizations

Aging With Dignity

www.agingwithdignity.org

American Hospice and Palliative Care Organization

www.nhpo.org

Patient Assistance for Medical Air Transportation

National Patient Travel Center

www.patienttravel.org - or 800-296-1217

Nonprofit organization assisting financially-needy patients with financial assistance to travel for specialized medical evaluations, diagnosis or treatment that may require medical air transportation.

24-hour toll free 800-296-1217.

International Organizations

European Respiratory Society (ERS)

<http://dev.ersnet.org/>

4, Ave Sainte-Luce

CH-1003, Lausanne

Switzerland

Tel: +41 21 213 01 01

Fax: +41 21 213 01 00

info@ersnet.org

Irish Lung Fibrosis Association

www.ilfa.ie

4 Trafalger Trc.

Monkstown, CO. Dublin, Ireland

jegan@matter.ie.ilfa.com