RareDiseaseLive

Idiopathic Pulmonary Fibrosis: Take Control with Knowledge

ILD Collaborative

PLATFORM HEALTH

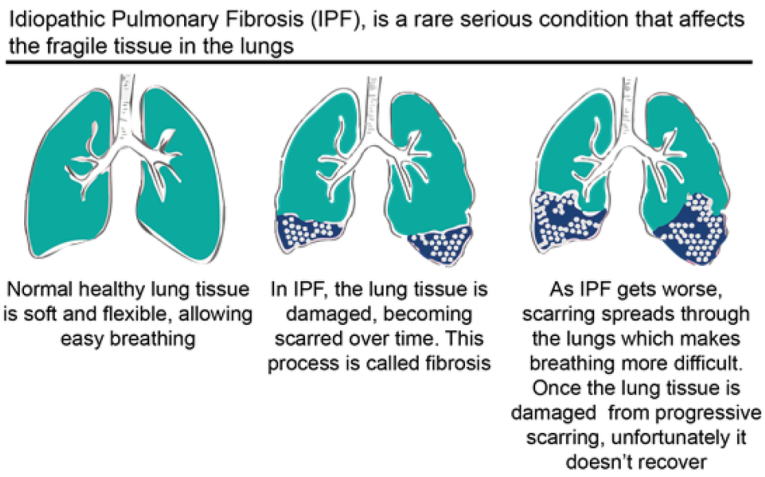
Today's Program

- **Part 1:** Understanding the Diagnosis
- **Part 2:** Options for Treatment
- **Part 3:** Patient and Caregiver Support

Part 1:
Understanding the Diagnosis

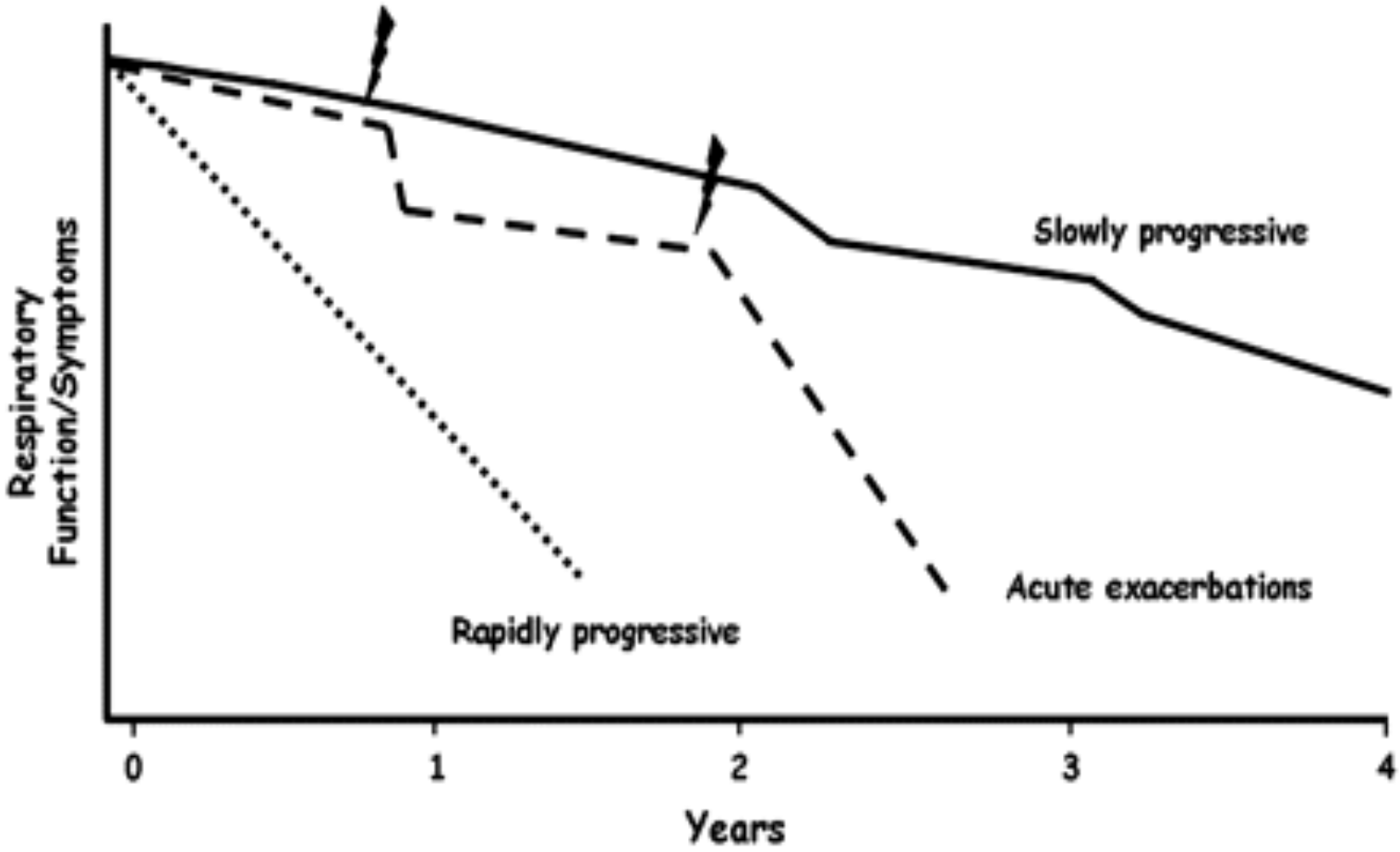
Idiopathic Pulmonary Fibrosis (IPF)

- Scarring of the lungs of unknown cause
- As IPF progresses, it prohibits the lungs from moving oxygen into the bloodstream
- One of the many Interstitial Lung Diseases (ILDs)
- Considered a diagnosis of exclusion – must exclude a number of other possibilities before diagnosis can be made



https://guide.eu-ipff.org/what-is-idiopathic-pulmonary-fibrosis/

Clinical Course of IPF



Walter N et al. Proc Am Thorac Soc. Vol 3. 330–338, 2006

What was most challenging for you, or your loved ones, at the time of diagnosis?

Select one response

A. Struggling to understand and explain IPF

B. Learning about life expectancy with IPF

C. Feeling isolated and unsupported to cope with and manage the diagnosis of IPF

D. Concern for the impact the disease would have upon family

E. All of the above

Radiology, Pathology and Physiology of IPF

- Chest CT (CAT scan)
  - Looking for pattern of abnormalities called *Usual Interstitial Pneumonia (UIP)*
  - 4 classifications: Definite; Probable; Indeterminate; Alternative diagnosis
- Lung biopsy (VATS)
  - Looking for pattern of abnormalities called *Usual Interstitial Pneumonia*
  - Similar classification as CT
- Pulmonary Function Tests (PFTs)
  - Restrictive defects (low lung volumes), diffusing capacity abnormalities (compromised ability to absorb O_2)

Respiratory Symptoms

- Shortness of breath
- Cough
- Chest congestion

How to Come to a Diagnosis of IPF

- Diagnosis is made when a cause of the fibrosis cannot be identified, and there is no underlying illness with which fibrosis can be associated (auto-immune conditions)

AND

- The CT, PFTs, biopsy all support IPF

Challenges at Time of Diagnosis

- Making sense of often unheard of disease
- Accepting diagnosis before being able to move forward
- Communicating medical issues to others
- Learning to live with physical constraints
- Facing realities of advanced care planning

Genetics of IPF

- Patients often have concerns for family members becoming affected
- Majority of testing being done in research settings
- Registry of IPF related genetic testing options:
  - https://www.ncbi.nlm.nih.gov/gtr/all/tests/?term=C1800706&filter=testtype:clinical

Patient Story

ASK THE PANEL questions about your IPF diagnosis

Part 2:
Options for Treatment

After diagnosis of IPF, what was the first treatment you, or your loved one, received?

Select one response

A. IPF drugs (OFEV® or Esbriet®)

B. Pulmonary rehabilitation

C. Oxygen therapy

D. Enrollment in a clinical trial

E. Haven’t/hasn’t started treatment

Treatment Options

- Two oral medications, Esbriet (pirfenidone) and Ofev (nintedanib)
- Both slow the loss of lung function by about 50%
- Initial challenges are identifying which to chose and when to start
- Some patients/physicians chose to start immediately after diagnosis made, others take watchful waiting approach

Treatment Options

- Strategies to introduce medications
  - Consider slowing dose escalation, increase dose only when medication is being well tolerated

Treatment Options – pirfenidone (Esbriet)

- Standard dosing titration schedule:
  - 1 capsule/tab three times a day x 1 week then
  - 2 capsules/tabs three times a day x 1 week then
  - Three 267mg capsules/tablets three times a day (or one 801mg tab three times a day)
- Symptom driven alternative schedule – only increase dose when tolerance achieved
  - Start 1 capsule/tab three times a day
  - If tolerable after 1-3 weeks, increase to 2 capsules/ tabs three times a day
  - If tolerable after 1-3 weeks, increase to 3 capsules/ tabs three times a day

Treatment Options – pirfenidone (Esbriet)

- Can also consider increasing by 1 capsule/ tab per week, i.e. increase from a total of 3 a day to 4, then 4 a day to 5, etc.
- If side effects develop, return to the dose at which there were minimal or no side effects – once symptoms resolve, start a slower dose escalation

Treatment Options – nintedanib (Ofev)

- Standard dosing titration schedule:
  - No up titration, start at 1 capsule twice daily
- Symptom driven alternative schedule – only increase dose when tolerance is achieved, i.e. start with 1 capsule, if tolerating, increase to 1 capsule twice daily after 1-2 weeks
- If side effects develop, return to dose at which there were minimal/ no side effects – slow dose escalation

Treatment Options

- Surveillance
  - Monthly appointments for the first 3-6 months which include lab work– liver function tests (LFTs)

Treatment Adherence – Managing Side Effects: Gastrointestinal

- Pirfenidone: nausea, upset stomach, acid reflux, vomiting, loss of appetite, diarrhea
  - Dose escalation strategies as already discussed
  - Taking the 3 capsules/tablets at separate times during a meal
  - PPI (e.g. omeprazole), H2-blocker (e.g. famotidine, ranitidine)
  - Pro-motility agents
  - Anti-nausea medications
- Nintedanib: diarrhea, nausea, vomiting, anorexia
  - Loperamide (Imodium) – can be used as needed or scheduled routinely 1-2 times a day (2-16mg daily)

Treatment Adherence – Managing Side Effects: Rash

Pirfenidone: important to determine whether photosensitivity vs. drug eruption

- To avoid photosensitivity – sunscreen, sun protective clothing
- Drug eruption not related to sun exposure typically requires permanent discontinuation of the medication

Treatment Adherence – Managing Side Effects: Fatigue and Loss of Energy

- Maintain good posture
- Avoid unnecessary tasks
- Organize your activities
- Reorganize your closets and shelves
- Keep duplicates of frequently used items

Treatment Adherence – Managing Side Effects: Fatigue and Loss of Energy

- Cook on Sunday for the entire week
- Invest in a rolling utility cart or four-wheeled walker
- Ask for help
- Get a disability plate or placard

Treatment Options – When to Change Medications

- Significantly elevated LFTs
- Intolerable/unmanageable side effects
  - If the second medication tried is worse, can return to first if safe to do so (if GI symptoms were predominant reason to change initially)
- Drug rash
  - Non-photosensitivity rash, or if photosensitivity rash and sun simply cannot be avoided
- Lack of efficacy – hard to define, consider if FVC consistently falling > 200mL/yr while on treatment

Other Illnesses or Complications of the Disease That Deserve Attention

- GERD – acid reflux
- Pulmonary hypertension
- Cardiovascular disease
- Lung cancer

Symptom Management - Strategies to Manage Shortness of Breath

- Control your breathing
- Maintain good posture
- Practice relaxation techniques
- Supplemental oxygen
- Pulmonary rehabilitation

Symptom Management – Strategies to Manage Cough

- Common alternative causes to consider:
  - Post-nasal drip from inflammation/ irritation of nose and/or sinuses can be either allergic or non-allergic in nature
  - Asthma / allergies
  - Esophageal reflux (acid reflux, GERD)
  - Smoking, COPD – chronic bronchitis
  - Medications (ACE inhibitors)

Symptom Management – Strategies to Manage Cough

- Medications:
  - Over the counter:
    - Dextromethorphan (max dose 120mg/day)
  - Prescription:
    - Opiates - codeine, hydrocodone
    - Benzonatate
    - Prednisone (may relieve congestion)
  - Nebulized options:
    - Saline
    - Hypertonic saline
    - Ipratropium / albuterol
    - Lidocaine
    - Morphine / hydromorphone

Acute Exacerbations

- Significant worsening of respiratory symptoms, oxygen needs, CT over a course of days to a small number of weeks, not explained fully by an alternative diagnosis
- Provoked (triggered) or Unprovoked (idiopathic)
- Provoked AEs typically triggered by some stress placed upon the lungs (pneumonia, mechanical ventilation, infection elsewhere with/without sepsis, drug toxicity, aspiration event)

Acute Exacerbations

- Prevention:
  - Maintain vaccinations
  - Hand washing / avoid sick contacts
  - Manage GERD appropriately
  - Avoid airborne irritants / pollutants
  - Avoid lung toxic medications
  - Pirfenidone / nintedanib
- Treatment
  - High flow O2 > non-invasive ventilation > mechanical ventilation
  - Steroids, empiric antibiotics, diuresis when appropriate
- If transplant is being considered, maintain lines of communication with transplant team

Following Disease Progression

- Evolution of symptoms
- Pulmonary function test and 6 minute walk test results
- Oxygen needs
- CT findings

Patient Story

ASK THE PANEL questions about your options for treatment

Part 3:
Patient and Caregiver Support

What about IPF concerns you most?

Select one response

A. Understanding treatments and/or trials available to me

B. The uncertainty of the prognosis

C. Preparing for a time when I will lose my independence

D. Effect of shortness of breath and cough upon my daily activities

E. Deciding on whether to pursue lung transplantation

Non-Pharmacological Treatment – Oxygen Therapy

- Criteria for starting O2
- Pulsed vs. continuous flow
- Conserving devices
- Nasal cannulas- finding the right fit
- Using a national or local carrier

Non-Pharmacological Treatment – Oxygen Therapy

- Importance of Using Supplemental Oxygen for those who require it
  - Everyone needs sufficient oxygen to support exertion or exercise
  - Low oxygen levels places a strain on the heart that over time can lead to elevated pressures in the heart and lungs. This may eventually cause the pump action of the heart to be less efficient
  - Low oxygen level can cause fatigue and limit exercise capacity
  - Low oxygen levels can also decrease one's ability to think clearly

Non-Pharmacological Treatment – Pulmonary Rehabilitation

- Exercise program that occurs in a safe, monitored environment while learning more about how to manage your condition
- Common goals:
  - Reverse/minimize the effects of deconditioning
  - Improve strength and flexibility of peripheral muscles (arms and legs)
  - Improve stability of core muscles (trunk and abdominals)
  - Improve level of fitness/aerobic capacity
  - Learn pacing strategies and acclimate to shortness of breath

Non-Pharmacological Treatment – Pulmonary Rehabilitation

- Pulmonary rehabilitation is run by a team of physical therapists, respiratory therapists, nurses, physicians, nutritionists, etc.
- Initial Assessment:
  - Establish a baseline level of function
  - Assess posture, strength, range of motion, flexibility, balance, sensation, and walking pattern
  - 6 minute walk test to assess aerobic capacity – repeated his at the end of rehab to gauge improvement

Non-Pharmacological Treatment – Pulmonary Rehabilitation

- Structure of the Program:
  - Frequency: 3-5x/week
  - Intensity: Find a “sustainable challenge” by reaching into reserve capacity and working at a level beyond most daily activities.
  - Type: need to utilize a type of exercise that is meaningful to your daily function/life demands
  - Time: need to try to achieve 20 minutes or more of aerobic type exercise; cumulative intervals are fine, keeping rest periods short. This is a progressive increase in time, working toward continuous exercise, if possible

Non-Pharmacological Treatment – Pulmonary Rehabilitation

- What happens when the program is over?
  - During rehab, an exercise program for home is developed, it is essential to continue after the structured program ends
  - Gains start to dwindle right away if you stop exercising, but all is not lost….you just have to resume exercise and work towards building up your endurance again
  - If you stop exercising for several weeks, you will have to start again at a much lower intensity or perhaps in intervals, and work your way back up
  - Some pulmonary rehab centers offer maintenance programs on site

Transplant Evaluation – Key Issues

- Deciding whether to pursue
- Timing of transplant
- What's involved in the evaluation process
- Strategies for staying fit during later stages
- Pre and post-transplant support groups

Goals of Care

- Alleviating symptoms like cough and SOB
- Managing medications' side effects
- Being active and eating healthy diet
- Avoiding colds and infections
- Utilizing supplemental oxygen appropriately
- Support with advanced care planning and palliative care resources

Goals of Care

- Advanced Care Planning
  - Living Will
  - Establishing a Support Network/Care Team
- Palliative Care
  - Specialized medical care focused on pain and symptom control, communication/coordination, emotional support, family/caregiver support
  - Appropriate at any stage of illness, used in parallel with other therapies
  - Goal: Wellness, improved quality of life
- Hospice
  - A palliative care approach at the end of life
  - Focus is attending to medical, social, psychological and spiritual needs of patients and families at the end of life

Join a Local/Virtual Support Group

- Finding peer-to-peer connection & support
- Offering answers from personal experiences
- Learning & sharing about living with IPF
- Advocating for holistic care
- Bringing hope to the next person
- Appreciating every day as a victory

Support Care Resources

- Interstitial Lung Disease (ILD) Collaborative
  - A patient-physician collaborative for understanding, management, and treatment of Interstitial Lung Diseases.
  - http://www.ildcollaborative.org/about
- Pulmonary Fibrosis Foundation
  - Connect with a local support group and medical center in the treatment of pulmonary fibrosis around the United States.
  - http://www.pulmonaryfibrosis.org/life-with-pf/find-medical-care?show=support
- American Lung Association
  - Offers free online communities with peer-to-peer support for those Living with Pulmonary Fibrosis and those Caring for Pulmonary Fibrosis.
  - http://www.lung.org/support-and-community/online-support-communities/

Patient Story

ASK THE PANEL any questions, including resources for ongoing support

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Thank you

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