COPD and Interstitial Lung Disease
2017

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Chronic Lung Disease

◆ COPD – 8 Cases
  ➢ Morbidity and Prognosis
  ➢ First Inhaler? Second Inhaler? …A Guide
  ➢ COPD vs Asthma
  ➢ Frequent Exacerbations (2) – not just azithro
  ➢ Dyspnea – not always COPD
  ➢ Acute Exacerbation - mgt, prognosis

◆ Interstitial lung disease – 3 Cases
  ➢ Aging Population = more interstitial disease
  ➢ Dying with vs dying of ILD
  ➢ New treatments – that actually work
**COPD – Death and Disability**

- **Top 10 causes of death, US 2010**
  - CJ Murray, *NEJM* 2013; 369:448
  - *Ischemic heart disease*
  - *Stroke*
  - *Lung cancer*
  - *Alzheimers*
  - *COPD*

- **Disability-Adjusted life years (DALYs)**
  - *Ischemic heart disease*
  - *COPD*
  - *Low back pain*
  - *Lung cancer*
  - *Depression*

- **ACOs**
- **CMS: Hospital Compare**
COPD – So What’s New?

- Classic care – early 1990s to the present
  - *Smoking cessation*
  - *Vaccination – flu and pneumovax, PCV 13*
  - *Oxygen, post hospitalization Pulm Rehab*
  - *Inhalers*
    - Bronchodilators
      - Short acting beta-agonists – albuterol
      - Long-acting beta-agonists (LABA) - *Salmeterol, formoterol*
      - Long-acting musc. antagonists (LAMA) - *Tiotropium, aclidinium*
    - Inhaled corticosteroids – if frequent exacerbations

1. **Preventive meds for frequent exacerbations**
   - Guidelines: ACCP/CTS, *Chest* April, 2015; 147:894-942

2. **COPD as a marker for severe comorbidities**
#1 - COPD and Hospice?

- 63 yo woman with severe COPD, FEV1 = 0.8L, on 2 liter/min O2
- Functional status – walking around apt
  - *Desaturates to 70s*
- Every new VNA – phone call re dyspnea
- PCP and case mgr – referral to hospice?
#1: COPD GRADE & Tx per GOLD

**Prognostic Grade: More than FEV1**

- **FEV1** – how fast can you puff?
- **Functional Status** – what can you do?
  - $B = BMI \leq 21$
  - $O = Obstruction – FEV1$
  - $D = Dyspnea – MRC scale$
  - $E = Exercise Capacity$

- **Exacerbations** – How often are you sick?
**COPD: Prognosis**

**BODE Index – Looking and Listening**

63 yo woman, FEV1 = 0.8L, 2 L/min O2

- **Skinny? – BMI** 0 pts
- **Low FEV1? -- Severe Obstruction** 3 pts
- **Breathless? – Dyspnea score** 3 pts
- **Activity limitations? -- Exercise cap.** 3 pts

**SCORE = 9/10 pts**
Common Knowledge: Severe COPD

**GOLD Class 3-4**
- Poor QOL
- High mortality in ICU
- Difficult to extubate
- Mech ventilation is burdensome and feared

- Predictable course
- Die of COPD
Humility re: Severe COPD

**Severe COPD Myths**
- Poor QOL
- High mortality in ICU
- Difficult to extubate
- Mech ventilation is burdensome and feared
- Predictable course
- Die of COPD

**Reality**
- Compared to?
- Lowest mort 20-25%
- Easier than ARDS, etc
- 96% said Yes, Again
  - Wildman MJ (UK ICUs).
- Unpredictable
- 2-3x more CV deaths
COPD Prognostication: Humility

* 63 yo woman, FEV1 0.8L, 2 L/min O2
* Discussion 2003....

* Last appt - 2017: FEV1=0.8L, 3-4L/min O2, SOB walking around apt

> Casanova et al. The Progression of COPD is Heterogeneous: BODE Cohort. Am J Respir Crit Care Med 2011;184: 1015-21

> 75% - no change in FEV1 or BODE score over 12 yr

* Key – No exacerbations - BODE-X Score
#2: 75 yo M, COPD – 1st Inhaler?

**Retired lawyer, likes competitive tennis**

- **Dyspnea, decr FEV1 to 1.4L, 46% pred.**
- **Exacerbations – 0-2 per yr.**

**Inhaled corticosteroid?**

**Long-acting bronchodilator?**

- **LA anti-muscarinic:** Tiotropium, aclidinium
- **LA beta-agonist:** Salmeterol, formoterol
COPD: Grade and Tx per GOLD

I: Mild
- FEV₁/FVC < 0.70
- FEV₁ ≥ 80% predicted

Add short-acting bronchodilator (when needed)

II: Moderate
- FEV₁/FVC < 0.70
- 50% ≤ FEV₁ < 80% predicted

Add regular treatment with one or more long-acting bronchodilators (when needed); Add rehabilitation

Add inhaled glucocorticosteroids if repeated exacerbations

III: Severe
- FEV₁/FVC < 0.70
- 30% ≤ FEV₁ < 50% predicted

Add long term oxygen if chronic respiratory failure. Consider surgical treatments

IV: Very Severe
- FEV₁/FVC < 0.70
- FEV₁ < 30% predicted or FEV₁ < 50% predicted plus chronic respiratory failure

LAMA - Tiotropium, Aclidinium, Umeclidinium

LABA (beta-agonists)
Salmeterol, Formoterol

COPD, not Asthma
#2: 75 yo M, COPD – 1st Inhaler?

- Retired lawyer, tennis
  - Dyspnea, FEV1 46% pred, occ Exac
- Inhaled corticosteroid, Montelukast
- Long-acting bronchodilator –LAMA, LABA
  - LA anti-muscarinic (LAMA): Tiotropium, aclidinium
    - Grade 1C recommendation (ACCP, 2015)
  - LA beta-agonist (LABA): Salmeterol, formoterol

CHEST 2015; 147 (4): 894-942
#3: SOB Despite Bronchodilator

- 61 yo taxi driver, >50 PY, SOB past 1-2 y
- DM, obesity, FH CAD
- Spirometry – FEV1 1.5L, 554% pred
- Meds – Tiotropium, albuterol PRN

Next Steps?

- Budesonide/Formoterol?, Mometasone/Formoterol
- Formoterol?
- Theophylline?
COPD Inhalers: Guide for the Perplexed

- **LAMA, LABA and other strange beasts**

  - **LABA** = long-acting beta-agonist \_\_terol
    - Salmeterol, formoterol, indacaterol, vilanterol, olapaterol

  - **LAMA** = long-acting muscarinic antagonist \_\_ium
    - Tiotropium, aclidinium, umeclidinium

  - **ICS** = Inhaled corticosteroid \_\_one, \_\_ide
    - Fluticasone, budesonide, mometasone
# Hunting Guide: Bronchodilator Inhalers

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#3 - SOB Despite Inhaler

- 61 yo taxi driver, >50 PY, Dyspnea 1-2 yr
- DM, obesity, FH CAD
- Spirometry – FEV1 1.5L, 55% pred
- Meds – Tiotropium, albuterol…..Next??
- FEV1….Functional Status…Flares? - NO
  - Budesonide/Formoterol?, Mometasone/Formoterol
  - LABA – 2d bronchodilator - Formoterol
  - Theophylline? – Not yet
#4 - 60 yo F smoker, allergies

- **Smoking:** 50 pk-yr, to present
- **No childhood allergies** – recently allergic rhinitis, cold-air worsening, nighttime awakenings
- **FEV1 = 1.2L, 52% predicted**
- **Acute Bronchodilator:** 11% increase

- **RX** – LAMA?, LABA?, LABA/Steroid?
**Asthma Vs COPD**

**One Disease?**

- Shortness of breath
- Wheezing episodes
- Rescue treatment with puffers
- Systemic steroids for flare-ups

✓ NO! – Eos vs PMNs, Reversibility
#4 - 60 yo F smoker, allergies

- **Asthma/COPD Overlap Syndr (ACOS)**
  - Obstruction – FEV1 52% pred, >10% incr

- **Asthma**
  - Bronchial hyperresponsiveness – partial
  - Inflammation – Eosinophilic, Allergic
    - Hay fever, Cold air, Nighttime events
  - CBC/Eos = 5%, IgE = 648

- + Smoking == Asthmatic Bronchitis

- RX – LAMA², LABA², LABA/Steroid
**COPD – Recurrent Exacerbations**

- **#5 - 65 yo man w/severe COPD**
  - 7 flares in 2.5 yrs

- **#6 - 73 yo woman w/severe COPD**
  - 5th hospitalization in 10 mos
Exacerbations: Preventable?

◆ #5: 65 yo M undertaker with COPD

- FEV1 = 1.2L, 25% predicted, no O2
- 7 flares/2.5 yrs, despite vaccinations etc
- Meds: Salmeterol/flutic.250/50, Tiotrop
  - Another inhaler?
  - Maintenance prednisone?
  - Prophylactic antibiotics? Vit D?
  - Roflumilast (PDE4 inhibitor) ?
  - Care management/Self-management?
Exacerbations: Preventive antibiotics?

- **Olden** Vintage mgt – rotating antibiotics
- **Azithromycin** – Guidelines *Chest* 4/2015 – Grade 2A
  - 5 RCTs, Largest NEJM- 8/25/11 RCT of azithro 250 mg/d
    - Pts on O2 or prednisone chronically, with at least one exac in past yr
    - Less freq exac (1/4-1/3 less), occurring later
    - **BUT** hearing loss, resistant organisms
    - CV events? --- Baseline EKG

- Considering azithro, other antibx?
- REFER for evaluation of “COPD”
  - *Bronchiectasis*?
  - *Mycobacterium avium (MAC)*? – drug resistance
Recurrent Exac. in COPD

- Maintenance prednisone?---- No!
- A pt who is “steroid-dependent”?
  - Not quite Nevah, but almost
  - Needs referral - Alternative dx?
    - Bronchiectasis, CV disease?
    - Pt confuses steroid-withdrawal w/ flaring disease
- Vitamin D?
  - Multiple studies – Maybe ….in severe deficiency
Recurrent Exac. in 65 y.o. man

- **Roflumilast** (Daliresp) - PDE4 inhibitor
  - **Severe to v. severe COPD**?
  - **Frequent exacerbations**?
  - **Adverse effects**
    - Wt loss (2 kg), GI upset
    - Suicidal ideation, other psychiatric symptoms
    - Drug interactions - P450 CYP3A4 and 1A2
  - **Decr exac, bronchodilation**
  - **Prescribing** – Rarely and carefully
COPD Self-Management? RCTs

- Great Idea – But….
- Glasgow  *BMJ* 2012; 344 – no diff
- VA System *Ann Intern Med.* 2012;156:673
  - Trial halted due to higher mortality- 28 deaths vs 10 in controls

- ACCP/CTS Guidelines April 2015 - Exacerbators
  - Self-management
  - Individual components
  - Specialist Access + Educ’n + Case mgt + Action plan (Grade 1C)
COPD and post-DC Inhaler Use

- Prospective cohort of hospitalized pts w/ COPD

- Patients – Avg age 71, FEV1 50% predicted, 15 meds
  - **Cognitive impairment** – 55% - mild-moderate

- Technique Errors - despite DC prep and score 8/10
  - **Low inspiratory rate, Multiple inhalations per dose**
  - **Despite inhaler proficiency** - 8 on 10 scale (pre-DC)

- 1/3 Low use/ poor techn, 1/3 Use but poor techn
- 1/3 good # and good technique
- Overall adherence – 6% achieved 80% of doses
  - *Dublin – cost not the issue*
COPD – Recurrent Exacerbations

◆ 65 yo undertaker, FEV1 25% pred, 7 flares in the past 2.5 yrs, triple inhalers

➢ COPD and bad prognosis?
➢ COPD + ?
  ♦ Bronchiectasis

➢ Another lung disease?
  ♦ Interstitial lung disease ---- PFTs

➢ CHF or CAD
COPD + Bronchiectasis

- First Question – truly COPD?
  - Smoking history, chronic bronchitis?

- Mod-severe COPD, FEV1 <50% pred
  - Bronchiectasis on CT in half
  - University hospitals, Spain
    - Am J Respir Crit Care Med April 2013; 187:823

- More exacerbations, mortality (2.5x)

- Pseudomonas, H. flu
Approach to Bronchiectasis

- High-resolution CT of chest
- Sputum culture: bacterial, AFB, fungus
- Immunological defect?
  - Quant. Ig, SPEP, subclasses
  - Alpha-1-antitrypsin level
- Cystic fibrosis?
  - CFTR testing, Sweat test
- Misc: Ciliary dysf, RA, ABPA...MAvium
**COPD: Recurrent Exacerbations**

- 65 yo M, 7 flares/2.5 yr
  - COPD + ?
  - Bronchiectasis
    - Chronic productive cough
    - Pseudomonas, H.flu, Strep pneumo
    - IgG v. low, IgG subsets v. low, no response to Pneumovax
  - Mgt – IgG, Azithro, neb Tobra
COPD + Bronchiectasis

- Classical bronchiectasis
  - Purulent, bacterial flares
  - Due to immunodeficiency or idiopathic

- Nodular bronchiectasis
  - Mycobacterium avium (MAC)
  - Higher mortality with macrolide alone

- Azithro prophylaxis – Wrong!
#6: 73 yo F, COPD, 5th hosp/yr

- 73 yo woman admitted for the 5th time in 10 mos with sudden dyspnea, hypoxemia
  - **Severe COPD** (FEV1 0.7L, 34% pred)
  - On home oxygen, 4 lpm
  - Morbid obesity – 64 inches, 265 lbs
  - Sleep apnea
73 yo F w/COPD, 5th hosp/yr

- Typical COPD flare?
  - New cough, sputum, prior URI…..Not always

- Dual diagnosis? COPD +
  - Sleep apnea – rather compliant
  - COPD variant? Bronchiectasis? No sputum, CT neg

- Atypical CHF?
  - Rarely crackles on exam or leg edema or CXR
  - Elevated BNP - not always
  - Key – Pizza boxes

→ No hospitalizations for > 2 yrs
#7: 82 yo M, COPD, incr SOB

- Smoked 50 PY, mild clinical COPD
- New dyspnea over 6 mos
  - Couldn’t do chores, yard work
- Barrel-chested, no distress, faint end-expiratory wheezes
- Meds: Tiotropium
#7: 82 yo M, COPD, incr SOB

- Your choices are:
  1. *Prescribe another MDI*
  2. *Refer to pulmonary rehabilitation*
  3. *Order spirometry*
  4. *Order a stairlift or scooter*
**Spirometry in Primary Care?**

- **Overdiagnosis/Misdiagnosis – other Dx**
  - Life’s not fair – only 30-40% of smokers get COPD
    - *Chest 2006 Oct;130:1129 - Finland*

- **Underdiagnosis - Severe COPD (FEV1 < 50%)**
  - Only half diagnosed by MD
    - *(NHANES, Arch Int Med 2000; 160:1683)*

- **Practice Guidelines – Require spirometry to dx and stage COPD in pts > age 40 w/ resp symp.**
  - *NCQA/HEDIS measure 2006, 2010, Medicare*

- **Reality - New dx COPD, Only 1/3-1/2 have spirom*
Why not Spirometry in Primary Care?

- ¾ of PCPs diagnose COPD clinically

- Focus group of 12 PCPs – Why not spirometry?
  - J COPD 2013; 10: 444

- Newly suspected COPD
  - Confident in history and results of inhaler trial
  - Uncertain utility of spirometry, pulm interpretation

- Pre-existing Dx of COPD
  - Middle aged-elderly, smoker, inhalers….Lots of other problems

- Priority of COPD during visit
  - No A1c equivalent, Monitoring per pt report

- Health system barriers - PFT appt, insurance
  - Most admitted lack of value, not logistics
#7 - 82 yo man w/ COPD, dyspnea

- Smoked 50 PY, mild clinical COPD
- New dyspnea doing yard work
- FEV1 1.2L, 40% pred
- 3 yr ago - FEV1 = 1.2L
- ETT equivocal....
- Cath: Left Main CAD....CABG
COPD: Marker for Cardiovascular Dis

- COPD pts
  - CV Disease: Meta-Analysis, Systematic Rev.
  - 2 to 5 fold higher rates – vs peers in gen pop
    - Ischemic heart disease
    - Arrhythmias
    - Heart failure
  - Death: Twice as likely to die of CV disease as of COPD

GOLD 2011 vs 2007

- **Group B**
  - Mild-mod obstruction
  - Dyspnea – prominent

- Higher mortality than Gp C
  - Despite better FEV1

- Died of CAD, HF and lung cancer

Lange et al (Denmark) Am J Respir Crit Care Med 2012; 186, 975
COPD and Cardiovascular Disease

European Journal of Heart Failure (2012) 14, 348–350
doi:10.1093/eurjhf/hfs022

Chronic obstructive pulmonary disease: a slowly progressive cardiovascular disease masked by its pulmonary effects?

Frans H. Rutten* and Arno W. Hoes

🔹 Beta-blockers in COPD pts with CAD
  ➢ Markedly underprescribed
  ➢ COPD esp on no BB associated with worse outcomes in MI, CAD, CHF
Long term Oxygen

- Standard of Care/ Medicare - 2017
- Resting SpO2 - 88% or lower - 24 hr/d
  - Or cor pulmonale
- Exertional/ Nocturnal desaturation
  - To 88% or lower
  - RX - Portable or bedroom oxygen
O2 for Mild-Mod Hypoxemia?


  - *n* = 734 pts, Stable COPD – avg 18 mo follow up
  - 1. Resting sat 89-93% → 24 hr/d oxygen
  - 2. or Exertional desaturation - Spo2 < 90, <80% for <1 min
    - suppl 02 during exercise and sleep
  - *Usage* – 24 hr/d = 15 hr use, Exercise/night = 11 hr use

◆ Death or Hospitalization, Exac – No difference

◆ QOL scores, distance walked – No difference

◆ How to apply this in real life?
#8: Acute COPD Management

- 67 yo man with >100 pk-yr
- 1\textsuperscript{st} admission 2/03
  - \textit{COPD Exacerbation}
- FEV1 0.65L
  - 23% predicted

- Treatment options?

- What about death and dyspnea?
  - \textit{Will COPD kill him?}
  - \textit{What is his ICU prognosis if he lands in MICU?}
Antibiotics in Acute COPD?

- Hospitalized pts - YES
  - Early antibiotics (cohort study)
    - Rothberg, M. B. et al. JAMA 2010;303:2035-2042.
  - Reduced risk of:
    - Mech ventilation, Mortality, Readmission
  - Regardless of change in sputum
- Decreased mortality (RR=0.87) regardless of sputum increase
Antibiotics in Acute COPD?

- **Outpatients**
  - **Anthonisen criteria**
    - Dyspnea, Sputum amount, Sputum purulence
  - **If Sputum change – YES**
  - **Duration – 5-10 days**
  - **Agent: almost any, exc if MDR risk**

- **Sputum culture – Please!**
  - **Targeting**
    - **Another diagnosis- Bronchiectasis**
      - Pseudomonas, Stenotrophomonas
Systemic Steroids = Targeted

- Mildly ill – mortality risk ~1% - PO predn ok
  - Prednisone = IV steroids?
  - *JAMA* June, 2010: retrospective, acute COPD, non-ICU
    - 92% IV steroids….Matched the 8% treated PO
    - Oral steroids –pts did **No Worse**…99% survival

- Regimen? – 2 wk vs 5 days
  - *JAMA* May, ‘13 - RCT, pred 40 mg/d
    - 14days = 5 days

- Failure/ relapse after 2 wk
  - Not simply “steroid-dependent COPD”
**Systemic Steroids = Targeted**

- **Mildly ill** – mortality risk ~1% - Prednisone

- **Moderate-severely ill** – mortality 10-20% 
  - *IV methylprednisolone* - < 500 mg/d

  - [Am J Respir Crit Care Med 2014.189:1052-1064]
COPD admission: Alternative Dx?

- **Occult PE**
  - *Only if atypical flare – no sputum/infection*
  - *PE-CT - PE in 20-25% of cases*

- **CHF or CAD** – as alternative or simultaneously
  - *BNP or nt-proBNP, Troponin*
    - Every COPD admission
COPD and the Advance Directive

- 67 y. o. man w/ COPD, FEV1 0.6 L

1. You will never get extubated?

2. You may get extubated, but won’t survive hospitalization?

3. You are likely to survive the ICU and hospital, but will need post-acute care?

4. You are likely to survive ICU and be discharged to home?
Humility re: Severe COPD

**Severe COPD Myths**
- Poor QOL
- High mortality in ICU
- Difficult to extubate
- Mech ventilation is burdensome and feared
- Predictable course
- Die of COPD

**Reality**
- Compared to?
- Lowest mort 20-25%
- Easier than ARDS, etc
- 96% said Yes, Again  
  - *Wildman MJ (UK ICUs).*  
- Unpredictable
- 2-3x more CV deaths
The Real Meaning of ICU

- ICU = Intensive Clarification Unit
- Preparation is rare
- Emergent discussions are difficult
- Futility as a basis for limiting care?
  - Prediction at outset is inaccurate
    - Most intubated pts (75-85%) are going to survive
  - 48-72 hr re-evaluation
- Prognosis - Trajectory of disease
  - Frailty, functional status, exacerbations
COPD and the Advance Directive

- 67 y. o. man w/ COPD, FEV1 0.6 L, good ex tolerance, no exacerbations

1. He will never get extubated

2. He may get extubated, but won’t survive hospitalization

3. He is likely to survive the ICU and hospital, but will need post-acute care

4. He survived intubation and the ICU, and was discharged home
COPD and the Advance Directive

- 71 y. o. man w/ COPD, FEV1 0.6 L, 4 yr later, after 3 exac in past yr
- Prognosis Now?
  - **BODE score – high ... + Exacerbations**
  - He survived the ICU and was discharged to rehab
  - Died 6 mos later, 4.5 yrs after the initial discussion
Interstitial Lung Disease

Questions for From the Internist

Elderly patients – not just a referral

- Biopsy vs no biopsy?
- Empiric steroids vs No empiric steroids?
- Death from interstitial disease, or with it?
Interstitial Disease – Cases

- Dyspnea in the dining room

- Crackles on preop exam
  - Is it interstitial disease, and if so, cause?
  - Will he die with the interstitial disease, or of it?

- Dyspnea in a patient with known pulmonary fibrosis
  - It’s interstitial lung disease progression, right?
Dyspnea in the Dining Room

- 84 yo woman referred with dyspnea in the past year
  - Moved back to MA from Fla
  - Subtly increasing shortness of breath, w/ longer recovery, on walking to the dining
  - Mild dry cough
  - Exam – fine crackles lower ½ of chest
  - Spirometry – FVC 85% predicted
  - CT – interstitial disease
  - FVC: 2.5L …..2.3L 2 yr later, more SOB
Interstitial Disease – the other 10-15%

- **Inflammatory disorders**
  - Granulomatous – Sarcoidosis
  - Pneumonia-like – BOOP
    - Bronchiolitis obliterans-organizing pneumonia
  - Vasculitic – Wegener’s (ANCA-associated vasc.)

- **Exposure-related**
  - Drugs
    - Amiodarone
    - Nitrofurantoin
  - Environmental - Hypersensitivity pneumonitis
    - Mold, Pigeons

- (Idiopathic pulmonary fibrosis - UIP etc.)
Biopsy in Interstitial Disease

- Clinical data, chest CT, PFTs
  - **IPF Clinic, only 85-90% accurate**

- Thoracoscopic biopsy - VATS
  - Much better tolerated, mortality <0.5% if done early when well

- Might defer if
  - Multiple other medical problems esp CAD
  - Incidentally detected
ILD Treatment: Do Something?

- Inflammatory element
  - Steroids? Azathioprine? NAC?

- ILD + Pulm HTN
  - PDE5 inhibitors – Sildenafil?
  - Endothelin antagonists – Bosentan?
Interstitial Lung Disease

▸ First – admit ignorance

➢ Pathogenesis
  ▪ Original paradigm - Inflammation
  ▪ Current paradigm – Injury, disordered repair

▸ Treatment: Mainly non-steroidal

➢ GERD mgt – Early-bird eating, Wedge, PPI bid

➢ Observation – many die with ILD, not of it

▸ Targeted agents: pirfenidone, nintedanib?
ILD Treatment: Less is More

◆ Am Thoracic Soc (and others) Guidelines, 2015
  ● Am J Respir Crit Care Med. 192; e3–e19, Jul 15, 2015

  ➤ Steroids? — Azathioprine? — NAC?

  ➤ Strong Recommendation Against Steroids

◆ ILD + Pulm HTN

  ➤ PDE5 inhibitors — Sildenafil?

  ➤ Endothelin antagonists — Bosentan?

  ➤ Strong Recommendation Against Pulm HTN meds

◆ Targeted agents – pirfenidone, nintedanib

  ➤ Conditional Recommendation For
**Pirfenidone - antifibrotic**

- **RCT, NEJM, May 2014**
  - Mild-moderate pulmonary fibrosis (IPF)
    - FVC 50-90% predicted
  - Outcomes
    - Less loss of lung function
    - Lower mortality
      - 3.6 vs 5.1%
  - Adverse effects
    - Photosensitivity
    - GI upset, fatigue
Nintedanib – Tyr kinase inhibitor

- **RCTs, including NEJM, May 2014**
  - **Mild-moderate pulmonary fibrosis (IPF)**
    - FVC $\geq 50\%$ predicted
  - **Outcomes**
    - Less loss of lung fcn $\sim 100$ ml less per yr
    - Not proven - lower mortality (5.5 vs 7.8%), fewer acute exac
  - **Adverse effects**
    - Diarrhea, nausea
Interstitial Lung Disease #2

- 72 yo retired executive
- Preop for prostate resection
- Lung exam – crackles
- CT chest

- PFTs
  - FVC 75% predicted
  - Ex oximetry
    - 96% at rest and after 3 min walking
Interstitial Lung Dis and Age


Prevalence Rate per 100,000

Age, years

18-34: 2.8, 0.0
35-44: 1.1, 5.4
45-54: 11.4, 10.9
55-64: 35.1, 22.6
65-74: 49.1, 36.0
75+: 97.6, 62.2

Men
Women
ILD #3: Known ILD, new SOB

- Retired 82 yo teacher, no exposures
- Distant smoking
- Repeat spirometry
  - $FVC 2.2L$, 65% pred
- Stable for $>2$ yrs
- Now dyspnea for the past 3-6 mos
Interstitial Lung Disease

- Dyspnea in a patient with interstitial disease - not always the crackles....

  - IPF with progression
  - CAD, CHF
  - Pulmonary embolism
  - Pulmonary HTN
  - Lung cancer
81 yo man with ILD, new SOB

- FVC 2.2L, 65% pred.
  - Stable PFT
- Cardiac cath
  - Severe 3 vessel CAD
**Conclusions: COPD**

- **#1 - Hospice for COPD 2003-16? Uncertainty...**
  - $FEV_1 + \text{Functional Status} + \text{Flares}$

- **#2 - 1st inh – Lawyer gets a LAMA**

- **#3 – 2d inh – Cabbie w/LAMA...LABA**

- **#4 – Lady w/ COPD and allergies**
  - *Asthma-COPD Overlap Syndr – only 10%*
Conclusions: COPD

- Recurrent exacerbations - #5 and #6
  - Triple inhalers
    - Inh steroids – low-dose for freq exac, ACOS
  - Caution on azithro, Roflumilast – REFER

- Look for a Cause - Not always COPD?...
  - Another DX - Bronchiectasis, HF, CAD, PE

- Adherence? Inhaler or Placebo?
Conclusions: COPD

- #7 - 82 yo trucker - unable to mow lawn
  - Get w/Guidelines – Spirometry …
  - COPD a marker for CAD – yes to BBs

- #8 - Acute exacerbation
  - Dangerous Myths about COPD
  - Rationale for steroids + antibiotics
  - Consider other causes – PE, CHF, CAD

- #8 ……Four years later --- cp to Patient #1
  - FEV1 + Functional Status + Flares
Conclusions: Interstitial lung disease

- Look for the treatable/reversible alternative diagnoses
  - Vasculitis? BOOP?
  - Hypersensitivity pneumonitis/drugs?
- Recurrent injury
  - GERD mgt, NOT steroids
- Idiopathic pulm fibrosis (IPF)
  - Prostate CA paradigm….in some cases
  - New good treatments – eg pirfenidone
  - ILD w/ dyspnea – could be CV disease?