Breath Easy: Conquering ILD in the ICU with Confidence and Care

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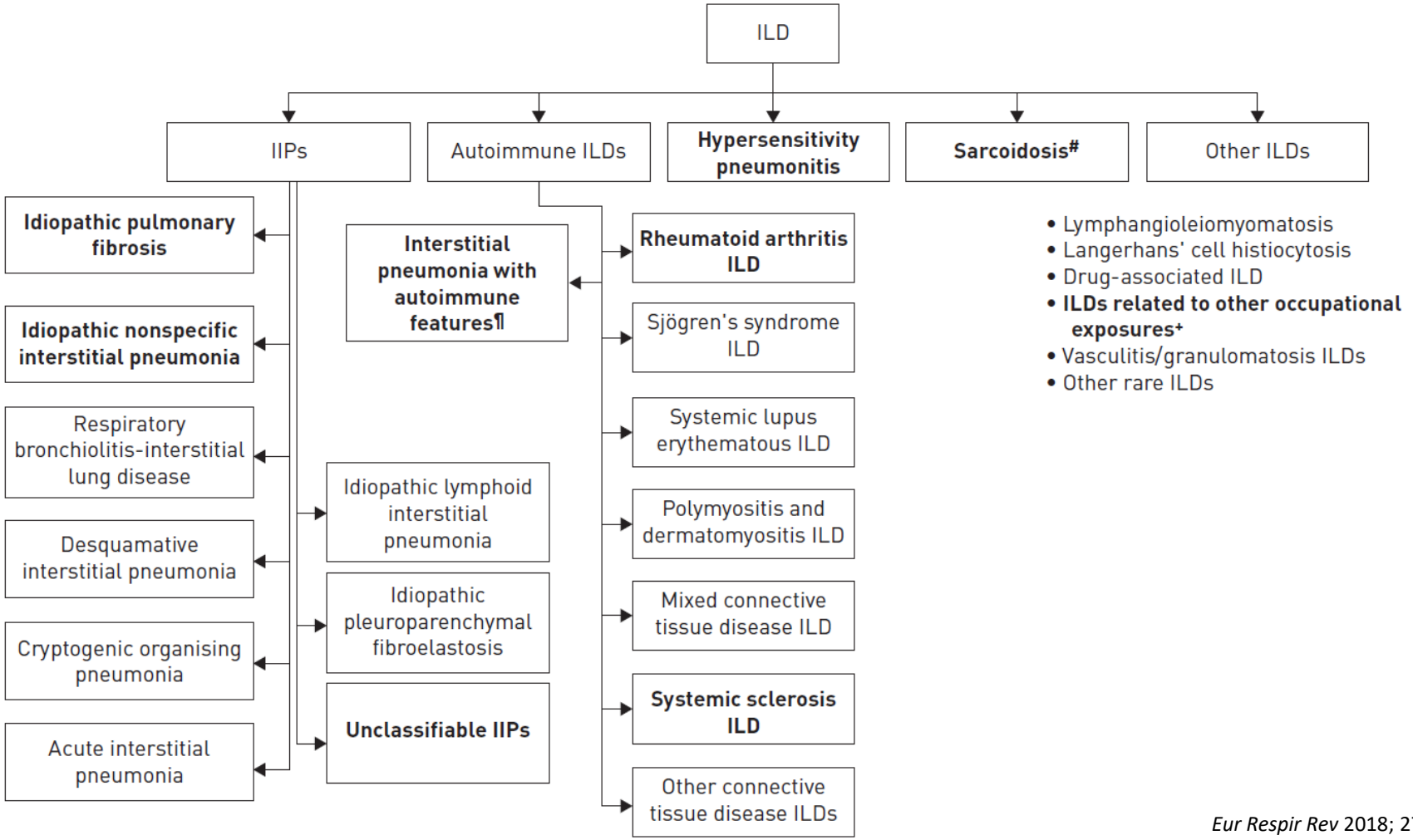
I have no actual or potential conflicts of interest in relation to this presentation.

Objectives

- Provide context for how interstitial lung diseases (ILD) are organized
- Discuss the key components of the ILD evaluation
- Review the essentials of disease management in the intensive care unit (ICU)

Why the confusion? ILD hypernym



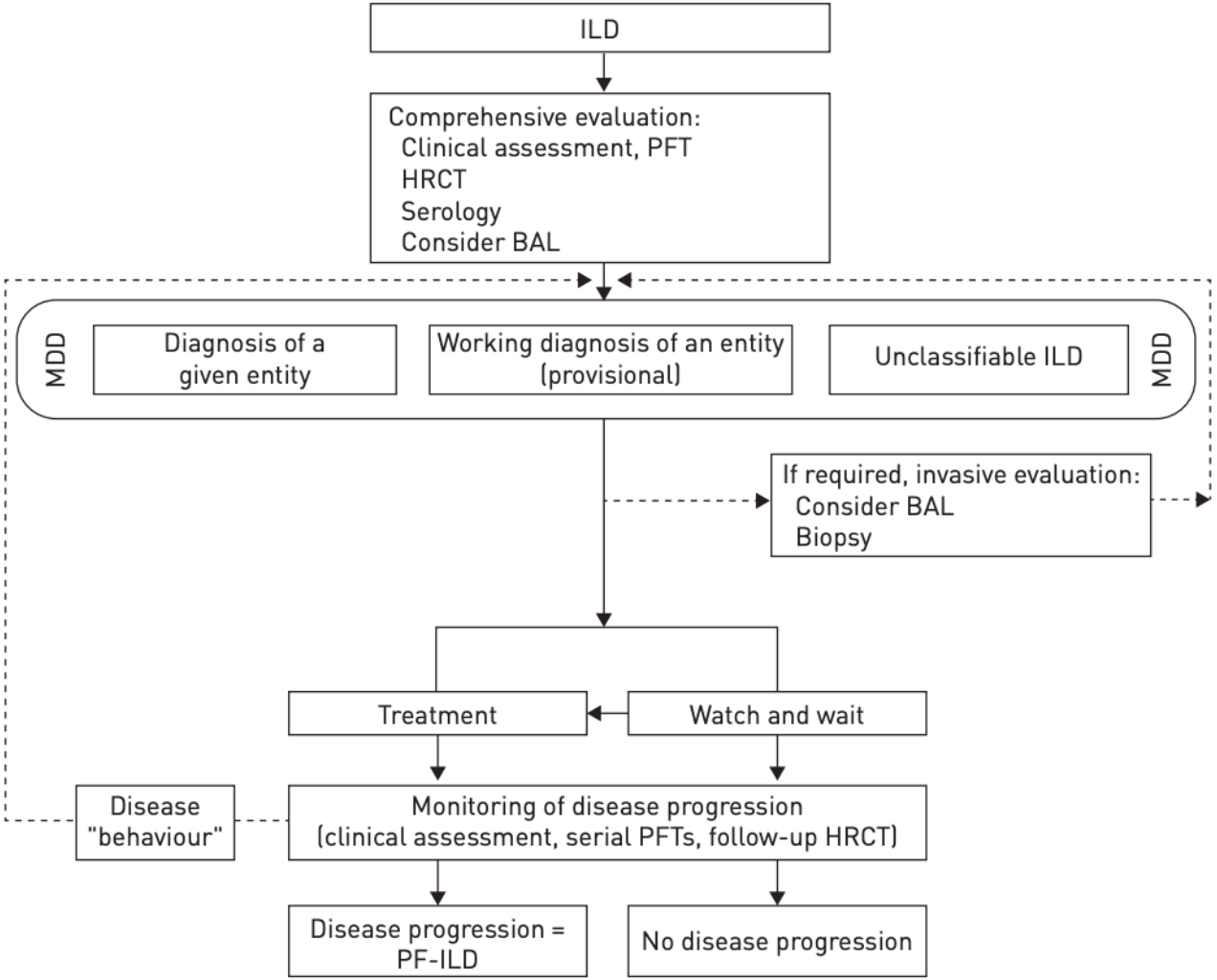
Eur Respir Rev 2018; 27: 180076

Spectrum of disease

<u>Inflammatory Predominant ILDs</u>
Autoimmune ILD (most)
Hypersensitivity Pneumonitis (early)
Cryptogenic Organizing Pneumonia
Drug-induced ILD

<u>Fibrotic Predominant ILDs</u>
UIP due to Scleroderma or RA
Hypersensitivity pneumonitis (late)
Idiopathic pulmonary fibrosis
Asbestosis

Justin Oldham, MD. *Interstitial Lung Disease: An Overview PPT*


ILD
Comprehensive evaluation:
Clinical assessment, PFT
HRCT
Serology
Consider BAL
MDD
Diagnosis of a given entity
Working diagnosis of an entity (provisional)
Unclassifiable ILD
MDD
If required, invasive evaluation:
Consider BAL
Biopsy
Treatment
Watch and wait
Disease "behaviour"
Monitoring of disease progression (clinical assessment, serial PFTs, follow-up HRCT)
Disease progression = PF-ILD
No disease progression

ILD diagnosis gold standard: Multidisciplinary Discussion

Case Presentation

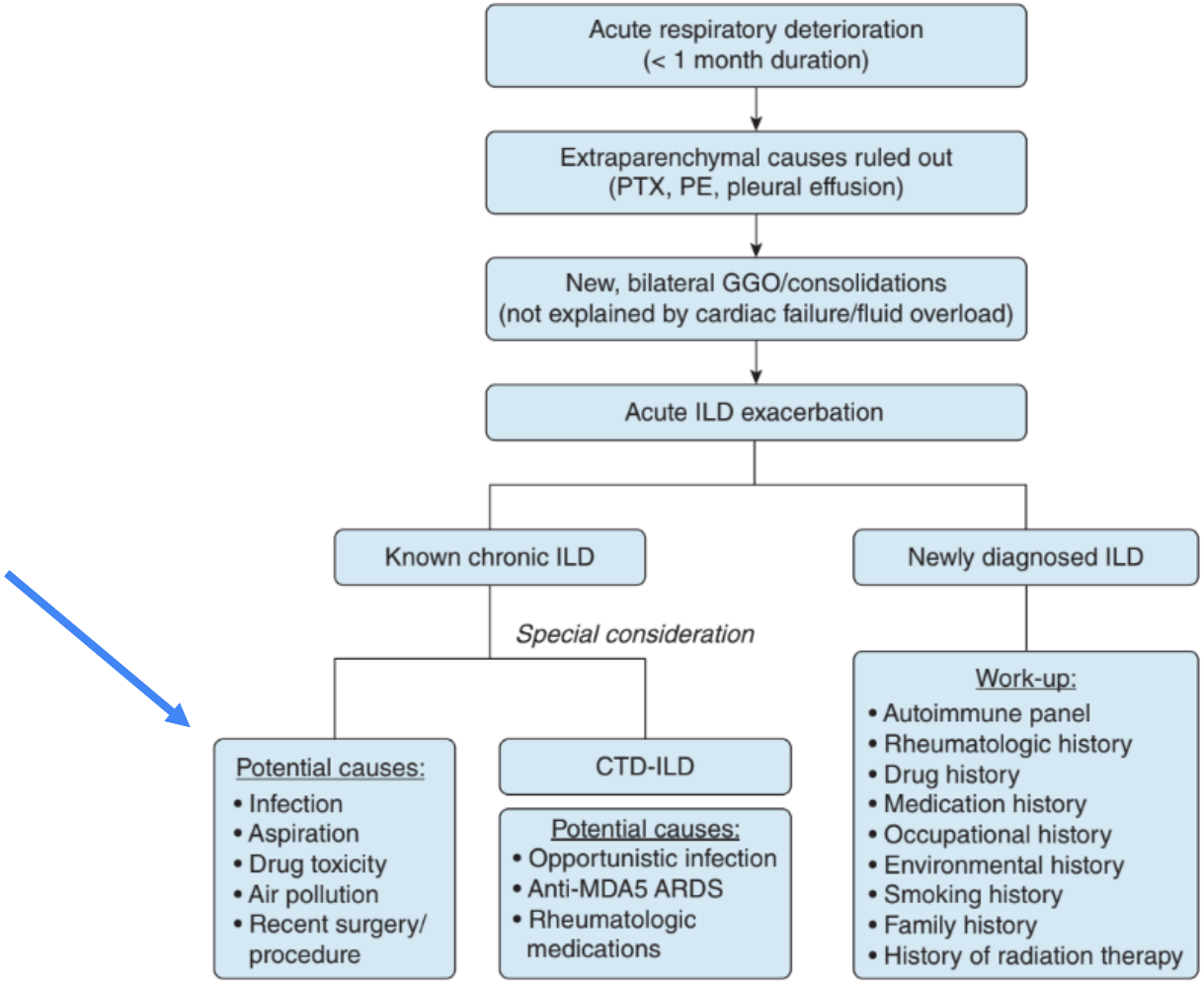
- 32-year-old woman, never smoker, with history of scleroderma on mycophenolate mofetil presents to your ED for dyspnea
- SpO2 75% on room air and initially placed on 10 liters open oxygen delivery system
- Due to respiratory distress + PaO2 50 on 100% high-flow nasal cannula, she is intubated and admitted to the ICU

Why are these cases so challenging?

- History matters: infection and ARDS vs acute ILD exacerbation
- Why are they so hypoxemic and what are some treatment options?
- Why is their lung compliance so terrible?
- How do support these patients on the ventilator with such poor oxygenation, dead space ventilation, and terrible compliance?
- If we don’t know the ILD type should we consider lung biopsy?
- What medication options are potentially beneficial?
- Who are reasonable ECMO candidates?
- What is the prognosis for ILD patients who receive invasive respiratory support?

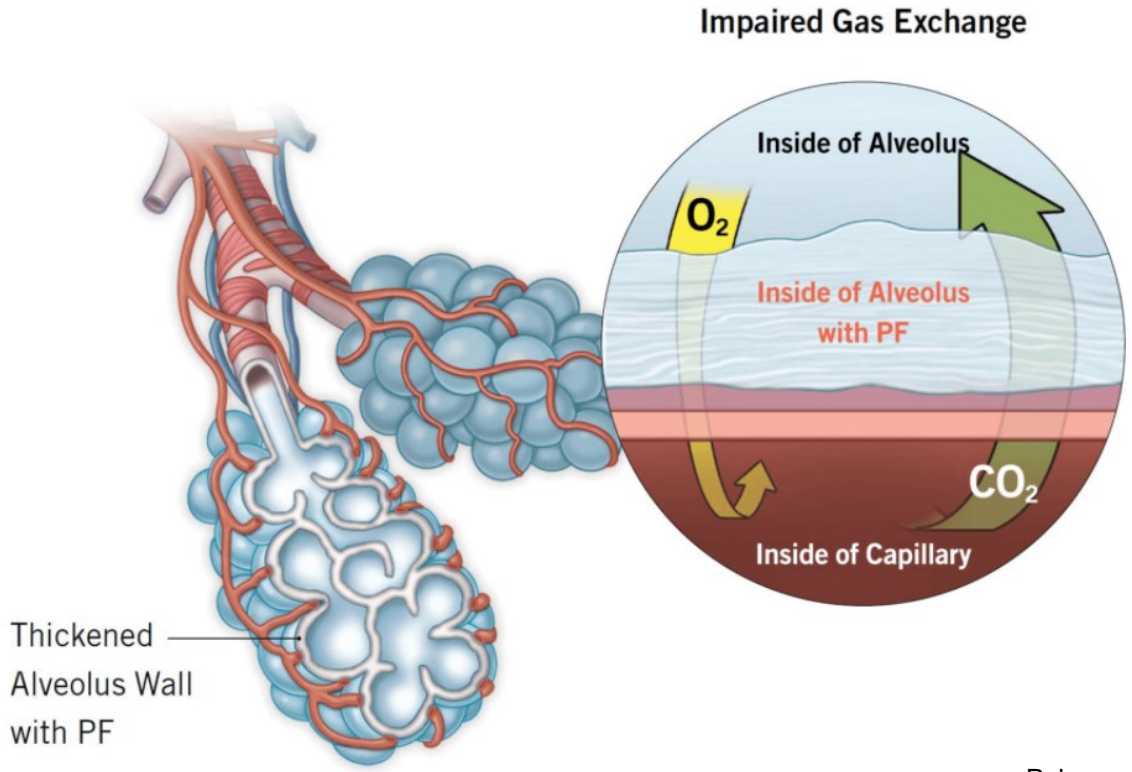
Infection and/or ARDS vs ILD exacerbation, history matters

- Co-morbidities
- Immunocompromised host?
- New medications?
- Dyspnea progression
- Cough changes
- New exposures (air conditioning cleaning, new pets, hobbies)



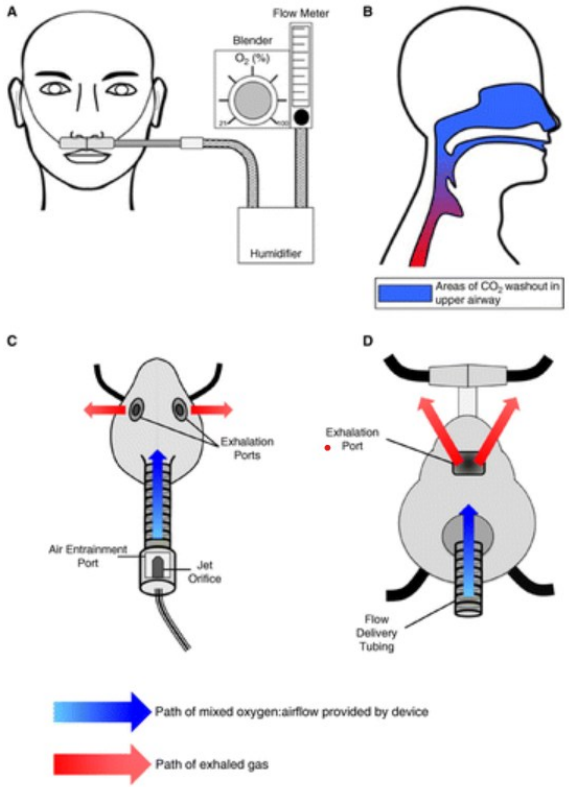
CHEST Critical Care 2023; 1(3):100020

Acute on chronic hypoxemic respiratory failure: Why?



Pulmonary Fibrosis Foundation
https://www.pulmonaryfibrosis.org/life-with-pf/about-pf

High-flow nasal cannula (HFNC)



- Physiologic benefits compared with conventional low-flow oxygenation
  1. *Improved oxygenation*
  2. *Decreased anatomic dead space owing to washout of the upper airway*
  3. *Generation of positive tracheal airway pressure*
  4. *Improved work of breathing*
  5. *Better secretion clearance*
  6. *Superior comfort*

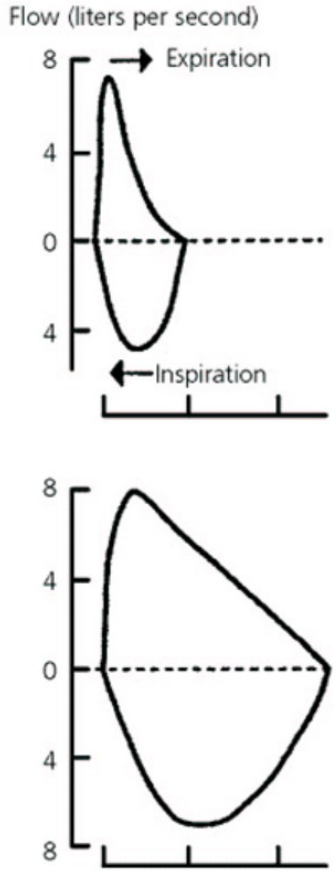
AnnalsATS 2018. 15(2):145-155

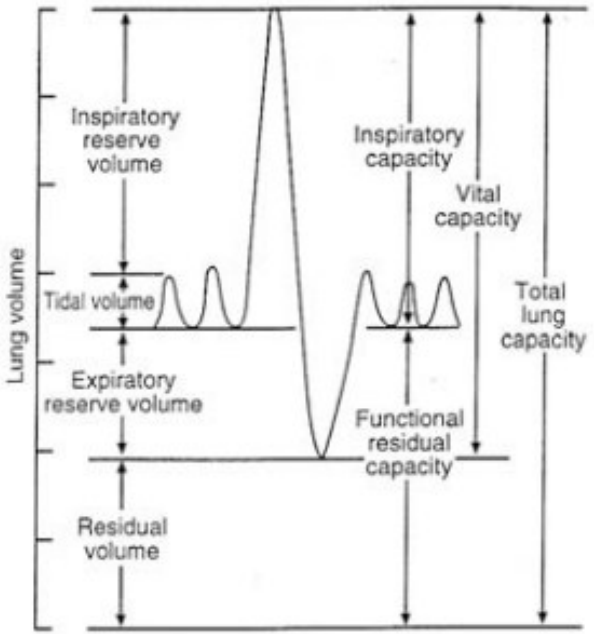
Case presentation

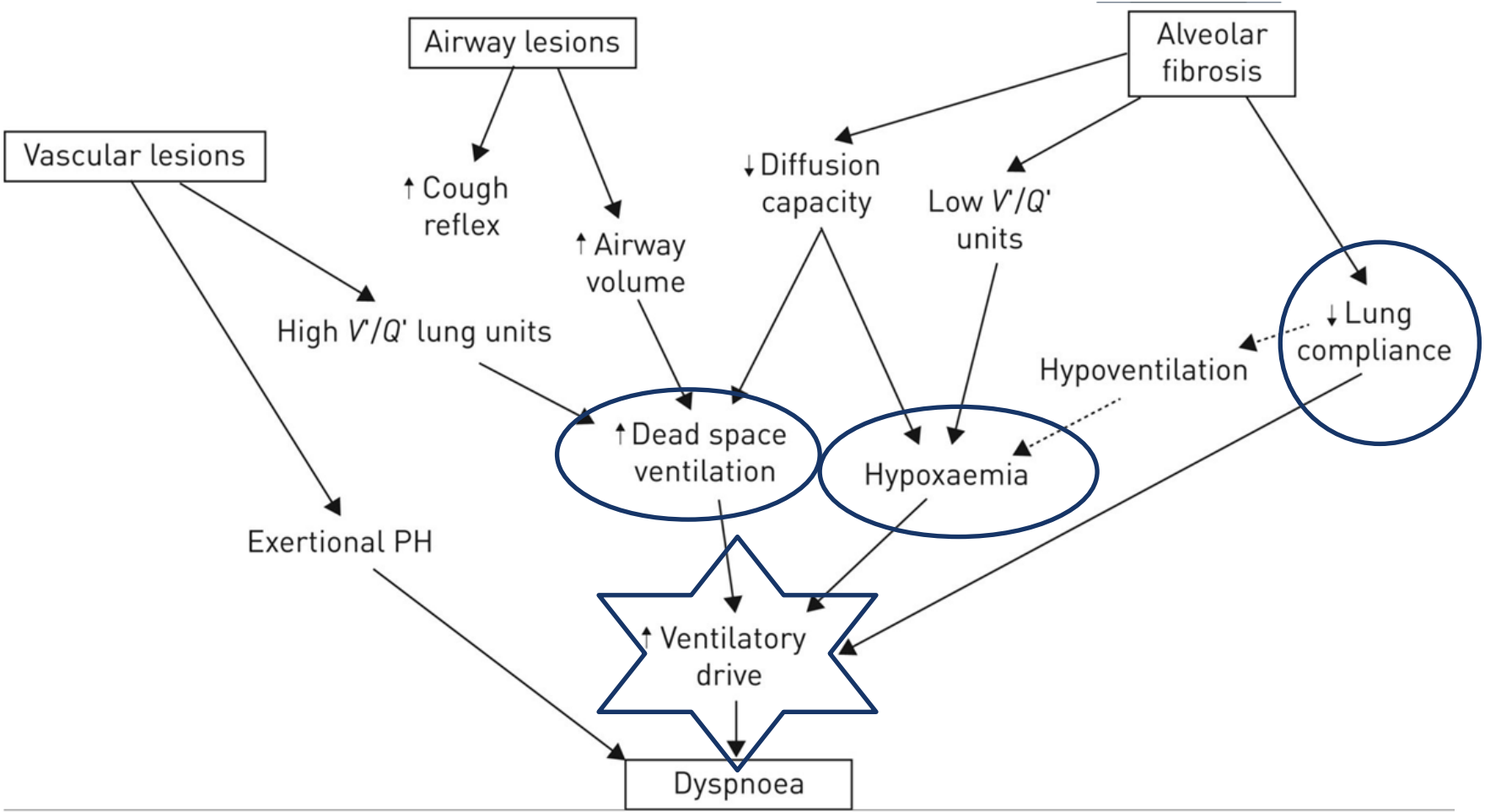
- Volume AC Vt 7 mL/kg/IBW + RR 14 + PEEP 8 + FiO2 80% → ABG 7.30/52/65
- Plateau pressure = 50!

The plateau pressure is 50…why? And why so dyspneic?

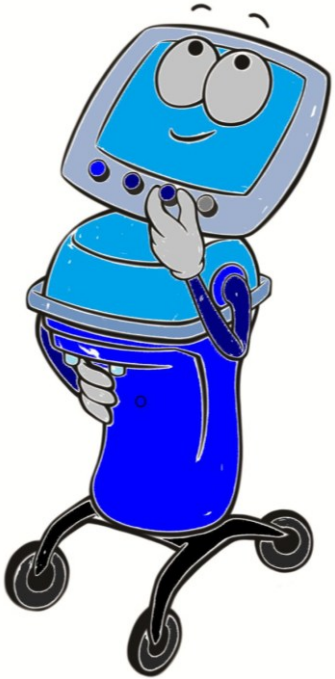
- Loss of surfactant
- Alterations in lung extracellular matrix






Airway lesions
Alveolar fibrosis
Vascular lesions
↑ Cough reflex
↓ Diffusion capacity
Low V'/Q' units
↑ Airway volume
↓ Lung compliance
High V'/Q' lung units
Hypoventilation
↑ Dead space ventilation
Hypoxaemia
Exertional PH
↑ Ventilatory drive
Dyspnoea

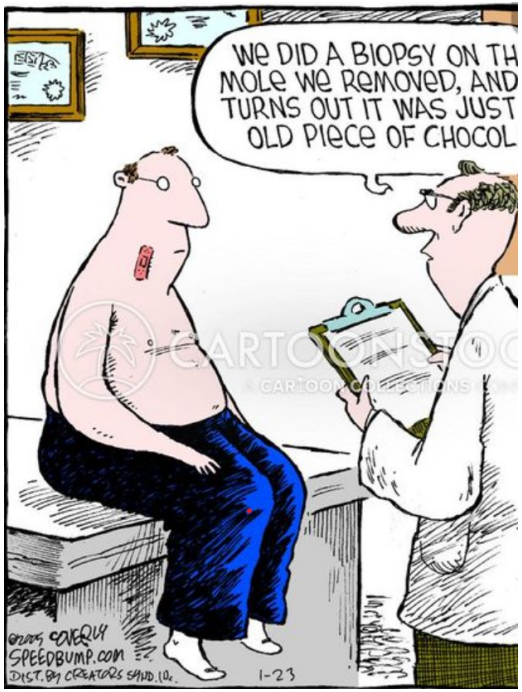
Ventilator strategies for acute exacerbations of ILD



- Titrate PEEP to driving pressure (plateau pressure – PEEP) to minimize alveolar hyperinflation
- Low-tidal ventilation does not apply, either does prone positioning
- Wean FiO2 and invasive respiratory support as tolerated
- May benefit from neuromuscular blockade for patient experiencing hypoxemia 2/2 dyssynchrony
- Agree immunosuppression + diuresis

Lung biopsy, when should we consider this?

- High-resolution has largely replaced surgical lung biopsy in the ambulatory setting
- In the ICU setting, 1/3 of patients had complications of persistent air leak
    - Hospital mortality 54%, how bad do we really need to know?



What pharmacologic options are available to treat acute exacerbation of ILD?

- At minimum…prednisone dose (0.5 – 1.0 mg/kg/IBW) daily
- Consider pulse dose steroids (500 to 1,000 mg daily x 3 days) for most patients who require ICU level of care
- Other considerations include rituximab as rescue therapy; plasmapheresis has a role for cases of diffuse alveolar hemorrhage and MDA-5 dermatomyositis

ECMO candidacy?

- *Are they candidates for lung transplant?* If YES than YES

How bad is it? Short- and long-term mortality is high (30-day mortality 78%; 6-month mortality 96%) in patients with ILD receiving mechanical ventilation

Case presentation

- Patient received pulse dose steroids
- Successfully extubated, discharged on prednisone 1 mg/kg/IBW for 30 days, mycophenolate mofetil, and PJP prophylaxis
- Advised she be seen for lung transplant evaluation

Key points and some…

Most cases of undiagnosed ILD and hypoxemic respiratory failure are responsive to aggressive pharmacologic therapy

Defend the PaO2 until the medications begin to work

If possible, consider HFNC use for as long as clinically tolerated

Review CT imaging with a thoracic radiologist if possible

Dry lungs are happy lungs

If a candidate for lung transplant, they are candidates for ECMO

MDA-5 dermatomyositis is the worst of them all!

SN Comp Clin Med 2022. 5(18)

Thank you for the opportunity to present this topic, any questions?