

# 56 year old woman with dyspnea

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# Medical history

- Social history:
  - cook, early retirement because of fibromyalgia, ex smoker 15 pack years, denies more than "normal" alcohol consumption
- Family history:
  - 2 brothers and both parents with coronary heart disease
- Past medical history:
  - fibromyalgia, psoriasis, hypothyroidism

# 2007

- exertional dyspnea for 6 months

- pulmonary scintigraphy (gp):
  - V/P mismatch of the left upper lobe
- low values of protein c + s and antithrombin III
- diagnosed with pulmonary embolism
  
- CT abdomen:
  - suggested liver cirrhosis with portovenous collaterals
  
- arterial blood gas:  $po_2$  11,2 kPa (11,0-14,0),  $pco_2$  4,26 kPa (4,70-6,00)
- ECG normal, normal chest x-ray, normal spirometry

# What now?

# The liver

- extensive evaluation:
  - clinical examination
  - biochemistry neg (albumin in lower normal range)
- liver biopsy:
  - no cirrhosis, but mild inflammation, early bridging fibrosis, mild steatosis and mild iron deposition
- gastroscopy: no varices
  
- gastroenterologist: biochemistry incl. AFP, ultrasound and clinical evaluation every 6-12 months

**Dyspnea, lung emboli,  
liver cirrhosis of unknown  
cause**

# 2008

worsening exertional dyspnea and also chest pain

- arterial blood gas:
  - $po_2$  10,2 kPa (11,0-14,0),  $pco_2$  3,79 kPa (4,70-6,00)
  
- cardiologist:
  - coronary angiography neg.
  - ECHO: neg
  
- pulmonologist:
  - no findings suggesting a pulmonary disease as a cause of the dyspnea

# 2009

- dyspnea on minimal exertion
- scheduled follow-up, cardiologist:
  - no dyspnea at rest
  - normal echo, still no tricuspidal pulmonary valve insufficiency – can't estimate PAP



Differential diagnosis?  
What to do next?

# 2010

## - another echo

- patient complains of dyspnea when getting dressed, "can't do anything at home"
- normal echo
- arterial blood gas: po<sub>2</sub> 8,39 (11-14), pco<sub>2</sub> 3,69 (4,7-6,0)

# What now?

# Admitted

- CT thorax + abdomen (portovenous p.):
  - no sign of pulmonary fibrosis
  - AV malformation in left lower lobe and between the hepatic artery and the portal vein?
  - new CT liver (three phases) and angio of the pulmonary artery: no sign of AVM.

# Right sided cardiac catheterization:

- no sign of any shunt
- borderline elevation of PCW-pressure at rest, significantly elevated with exertion

- new echo with agitated salt water:
- bubbles seen repeatedly on left side after 5-6 cycles: **intrapulmonary shunt**

# Diagnosis:

Hepatopulmonary syndrome

-summer 2010 : liver tx

-gradually feeling better, now no supplemental oxygen

- last control normal blood gas at rest, almost no bubbles at contrast

-stopped taking warfarin after liver tx