AHD: Pulmonary Hypertension

Case #1

Ms. Alto is a 48 yo F with a history of depression who presents to your medicine clinic for PCP establishment. She hasn't seen doctor since college but decided to come because her shortness of breath. She been having worsening shortness of breath on exertion for the past 6 months. In the past, she walked up the several flights of steps to her apartment with ease but now she has to stop several times to catch her breath. She has no associated cough, sputum production, or wheezes. She has never smoked.

Home Meds: Sertraline 50 mg qday, PRN ibuprofen

Yes, this is the pulmonary hypertension AHD so it's already on the differential diagnosis. What else is in the differential diagnosis? What further information do you want to know?

Exam:
T: 98 HR: 80 BP: 125/65 RR: 16 Oxygen saturation: 98% on RA Wt: 140 lbs (63.5 kg)
Gen: young Caucasian woman, AAOx3.
HEENT: Moist oral mucosa. No lymphadenopathy
Neck: No JVD, trachea midline
Heart: RRR, normal S1/S2. No murmurs, rubs or gallops.
Lungs: CTAB, no wheezing or crackles noted.
Abd: + BS. Soft. NTND
Ext: Puffy, nonpitting edema of her fingers with associated skin thickening. No LE edema
Neuro: Alert and Oriented to person, place, time and situation. No focal neurologic deficits.

Where do you start with your workup?

What is the most likely diagnosis? What are the common causes of PH and how are they organized?

What are the next steps in her workup?

Ms. Alto gets PFTs, which are normal aside from a mildly decreased DLCO. HRCT of chest is unremarkable aside from an enlarged pulmonary artery. V/Q scan show no ventilation-perfusion mismatch. Serum BNP was mildly elevated.

What is needed to confirm Ms. Alto's diagnosis?

What do you expect it to show? What information you would like to know from the procedure?

What treatment options are available for Ms. Alto?

Given Ms. Alto's new diagnosis of PAH, is there any further recommended counseling?

Assume Ms. Alto had PH related to the another WHO Group, how would you then treat her pulmonary hypertension?

Case #2

Ms. Ivey is a 56 yo F with a history of DM2, OSA on CPAP, PAH on multiple medications, who presents to the ED with worsening SOB with a productive cough. She has a history of progressive, idiopathic PAH that was diagnosed 1.5 years ago via RHC and she is currently being treated with PO ambrisentan, PO tadalafil, and IV epoprostenol (via portable pump and Hickman catheter). At baseline, she is typically comfortable at rest without SOB however, for the past 4 days she has been feeling SOB even while sitting in her chair. Her cough started 5 days ago, is productive with yellow sputum, and she has subjective fevers. She denies sick contacts. She lives at home by herself and has not been hospitalized in the last 6 months.

What are etiologies/things to consider when a patient with a diagnosis of PAH presents with worsening shortness of breath?

What further workup would you like to obtain? Any specific things to look for on exam?

Exam:
T 100.1 P 102 BP 99/66 R 22 Sat 89% RA
GEN: Awake, alert. In mild distress
HEENT: No cervical adenopathy, no JVD, no bruits
CV: Tachycardic, regular rate, normal S1, loud S2, no murmur. No parasternal heave
RESP: labored, inspiratory crackles in the left base. No expiratory wheezing
ABD: Soft, non-tender, non-distended, no ascites
EXT: Trace bilateral edema of LE below ankles, warm distal extremities, palpable pulses
NEURO: AAOx3, no gross motor or sensory deficits
SKIN: Tunneled catheter (Hickman) present on right chest, no pain to palpation of catheter site, no surround erythema or discharge at insertion site. Pump appears to be functioning properly

Labs:
131 | 98 | 18 / 14 \ 11 / 300 Diff: 82% Neut
---------- 131 / 33 \
4 | 22 | 1.1 \
UA: no blood, trace protein, negative LE/Nitrite UPT: negative
BNP: 80
CXR: Prominent pulmonary arteries, LLL opacity with air bronchograms
EKG: Sinus tachycardia, normal axis, no concerning ST-T wave changes

Fill in admission orders below:

Case #3

Ms. Lannister is a 61 yo F with a history of HTN, hypothyroidism, PAH, who presents to the ED with worsening SOB and weight gain. She has a history of idiopathic PAH that was diagnosed 11 months ago via RHC and she is currently being treated with ambrisentan and tadalafil. Ms. Lannister can typically play with her grandchildren with mild SOB, but over the past month she has been getting more easily SOB even with simply walking to the bathroom. In addition, in the past week she has noticed about 12 lb of unintentional weight gain, predominantly in her lower extremities and abdominal swelling. She's having difficulty fitting into her pants. She came to the hospital this morning after she had an episode of palpitations and lightheadness requiring her to lie down after walking from the kitchen to the living room. She denies CP, cough, fever/chills, n/v. She denies any sick contacts.

Exam:
T 98.5 P 112 bpm BP 95/52 R 20 Sat 90% RA
GEN: Awake, alert but fatigued. In distress
HEENT: No cervical adenopathy, + JVD to mandible
CV: Tachycardic, regular rate, normal S1, loud P2, holosystolic murmur loudest at lower left sternal border that becomes louder with inspiration. +Right parasternal heave
RESP: CTAB. No expiratory wheezing
ABD: Soft, NT, +distended. Liver edge palpable 3 cm below costal margin
EXT: 2+ bilateral pitting edema of bL LE above the knees, cool distal extremities
NEURO: AAOx3, no gross motor or sensory deficits

Labs:

130 | 95 | 30 /	8.9 \ 12 / 250	ABG: 7.3/36/59/20/89%
---------- 131	/ 36 \	D-dimer = < 0.5
4.5 | 20 | 1.9 \		Lactate = 2.8
		BNP = 1200

CXR: Enlarged pulmonary vasculature, enlarge right cardiac border, clear lung fields
EKG: Sinus tachycardia, right axis deviation, right atrial enlargement, no ST changes

What is the most likely diagnosis? How can you confirm?

Can you explain the pathophysiology?

Where do you admit Ms. Lannister? What are your basic treatment goals?