

Part I

Study Questions

1. A 59-year-old man with a history of pulmonary embolism diagnosed 2 years ago and treated with warfarin for 6 months is evaluated for progressive dyspnea and bilateral lower extremity edema. His physical examination is notable for a prominent P2 and a tricuspid regurgitant murmur, as well as 2+ bilateral lower extremity edema. Radionuclide V/Q scanning revealed absent flow to the right lower lobe (RLL) and right middle lobe (RML; Figure 1-A). Right-sided heart catheterization data are given (Table 1-A.)

Pulmonary angiography shows a pouch defect with complete occlusion of RLL and RML arteries. No other cause for pulmonary hypertension is uncovered by his evaluation. The patient's pulmonary hypertension is categorized as World Health Organization Functional Class II.

Which of the following is most appropriate therapy for this patient?

- A. Balloon pulmonary angioplasty
- B. IV alteplase
- C. Oral riociguat
- D. Surgical pulmonary thromboendarterectomy

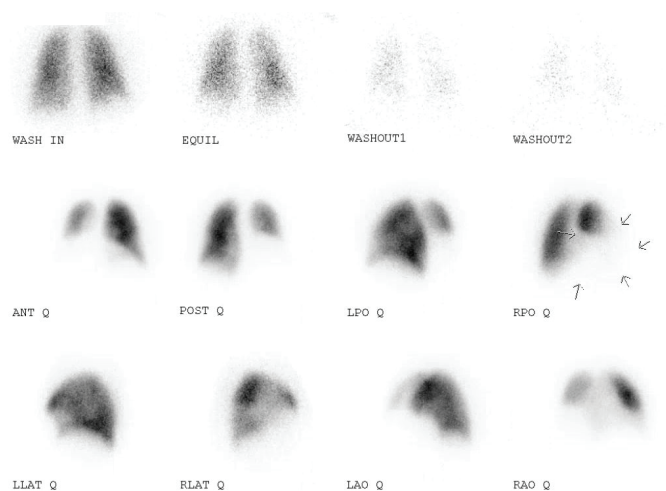


Figure 1-A V/Q scan showing absent blood flow to the right middle and lower lobes (arrows).

Table 1-A Right-Sided Heart Catheterization Data

Right atrium	16 mmHg
Right ventricle	67/5 mmHg
Pulmonary artery	69/32 mmHg (mean 47 mmHg)
Pulmonary artery wedge pressure	15 mmHg
Fick cardiac output (index)	5.37 L/min (2.36 L min/m ²)
Pulmonary vascular resistance	402 dynes • sec • cm ⁻⁵

2. A 50-year-old, previously healthy man is admitted to the ICU secondary to progressive acute respiratory failure due to sepsis of pulmonary origin. He requires low tidal volume ventilatory support because of high peak and plateau pressures. Which of the following is most likely to occur in this patient on a protective ventilatory strategy?

- A. Increased lung endothelial and epithelial injury
- B. Increased release of proinflammatory cytokines from alveolar macrophages and the lung epithelium
- C. Increased alveolar fluid clearance through the active transport of sodium and chloride across the alveolar epithelium
- D. Increased fibrin deposition

3. A 48-year-old woman with a longstanding history of severe asthma is referred to your specialty asthma clinic regarding suitability for bronchial thermoplasty. She is currently symptomatic and has had eight exacerbations in the past year requiring courses of oral corticosteroids, has been hospitalized three times for asthma exacerbations, and has required mechanical ventilatory support on one occasion. Her current maintenance therapy includes combination inhaled corticosteroid/long-acting β_2 -agonist (ICS/LABA), inhaled long-acting muscarinic antagonist (LAMA), and leukotriene antagonist therapy. She has excellent inhaler technique and is adherent to the prescribed therapies. She has received two different biologic agents in the past 2 years; they were discontinued by her regular pulmonologist because of lack of efficacy. A chest radiograph confirms no significant abnormalities except for hyperinflation. Spirometry testing results are shown in Figure 3-A.

From the information provided, what is your assessment of this patient's suitability regarding bronchial thermoplasty?

- A. She is an ideal candidate because of the severity of the asthma, frequency of exacerbations, and hospitalization for life-threatening asthma exacerbations.
- B. She may be a suitable candidate pending the results of additional investigations.
- C. She is not an ideal candidate because maintenance therapy has not yet been maximized.
- D. She is not an ideal candidate but may be suitable if bronchial thermoplasty is performed within an approved clinical trial.

	Predicted	Pre-Bronchodilator	% Predicted	LLN-ULN	Post-Bronchodilator	% Predicted	% Change
FVC [L]	3.47	3.26	94	(3.0 - 4.4)	3.32	96	2
FEV ₁ [L]	2.65	1.01	38	(2.3 - 3.5)	1.03	39	2
FEV ₁ /FVC	0.76	0.31		(0.71 - 0.90)	0.31		

Figure 3-A LLN = lower limit of normal; ULN = upper limit of normal.

4. A 19-year-old woman is diagnosed with acute promyelocytic leukemia and therapy is initiated. Five days later, the patient develops fever, peripheral edema, pulmonary opacities, hypoxemia, respiratory distress, hypotension, acute kidney injury, and elevation of transaminase levels.

Which of the following drugs was included in her regimen?

- A. Arsenic trioxide
- B. Cytarabine
- C. Daunorubicin
- D. Etoposide

5. Which of the following patients would be most appropriate for tunneled pleural catheter placement?

- A. A 45-year-old man with adenocarcinoma of unknown primary origin with known lymphangitic spread, with dyspnea, and a large, primarily unilateral malignant effusion. The marked dyspnea was not relieved in the past by a therapeutic thoracentesis resulting in complete drainage.
- B. A 60-year-old man with a slowly resolving right-sided anaerobic empyema receiving a prolonged antibiotic course and thoracostomy tube drainage.
- C. A 50-year-old homeless man with lymphoma who is receiving chemotherapy and has dyspnea and a large, recurrent, unilateral malignant effusion with dyspnea relieved by thoracentesis in the past.
- D. A 60-year-old woman with metastatic breast cancer with dyspnea and a moderate-sized, unilateral malignant effusion. She has had dyspnea relief with thoracentesis in the past, although there is a residual trapped lung.

6. Which of the following is the most appropriate indication for noninvasive ventilation using an adaptive servo ventilator?

- A. Central sleep apnea associated with Duchenne muscular dystrophy
- B. Cheyne-Stokes respiration associated with a cardiac ejection fraction of 25%
- C. Hypercapnia in a patient with obesity hypoventilation syndrome
- D. Treatment-emergent periodic breathing persistent after 3 months of CPAP

7. A 44-year-old woman is seen for evaluation of chronic cough for the past 24 years. She has had extensive allergy testing and her symptoms have been unresponsive to bronchodilators. Because of progressive breathlessness, she was referred for a pulmonary evaluation. Her cough is dry and nonproductive. She experiences paroxysms of “coughing fits” with gagging. Her cough is more pronounced at night, increases with temperature shifts, inhaled irritants, and deep breathing. Associated symptoms include intermittent flushing and concomitant watery diarrhea that she noticed over the past year.

Pulmonary function testing results are shown in Figure 7-A. A posteroanterior chest radiograph was obtained, followed by CT imaging of the chest (Figure 7-B and Figure 7-C).

To define the radiographic abnormalities, a surgical lung biopsy was performed (Figure 7-D). As seen in the images, the histologic samples reveal hyperinflated parenchyma with numerous small carcinoid tumorlets that are free of cytologic atypia, increased mitosis activity, or necrosis. Nests of neuroendocrine cells are also visible in the subepithelial compartment of bronchioles. The alveolated parenchyma is hyperinflated, with patchy, mild, chronic cellular bronchiolitis along with other bronchioles, showing subepithelial scarring and obliteration consistent with a component of constrictive or obliterative bronchiolitis. No granulomas are present. No lymphoid hyperplasia is identified.

Which of the following should be communicated to the patient regarding her condition?

- A. The condition is a smoking-related lung disease.
- B. This is a premalignant condition in which neuroendocrine cell proliferation is more pronounced than normal.
- C. Men are more commonly affected than women.
- D. Bronchoscopy with transbronchial biopsy provides a confident diagnosis in most patients.

	2006	2012	2016
TLC, L (% Predicted)	4.84 (96)	4.99 (100)	4.65 (93)
RV, L (% Predicted)	1.95 (118)	2.34 (137)	1.93 (112)
FVC, L (% Predicted)	2.87 (80)	2.65 (73)	2.72 (75)
FEV-1, L (% Predicted)	1.72 (62)	1.58 (54)	1.65 (57)
FEV-1/FVC	60	60	60
DLCO, ml/min/mg Hg (% Predicted)	18.2 (75)	18.2 (76)	19.79 (80)

Figure 7-A Serial pulmonary function testing results.

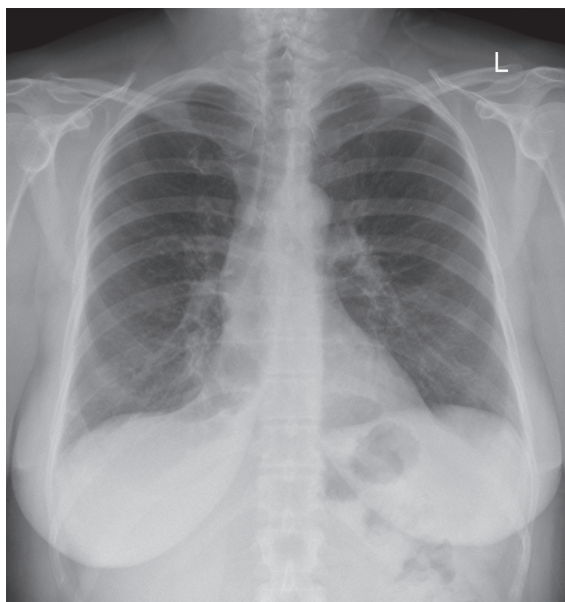


Figure 7-B Posteroanterior chest radiograph.



Figure 7-C CT imaging of the chest.

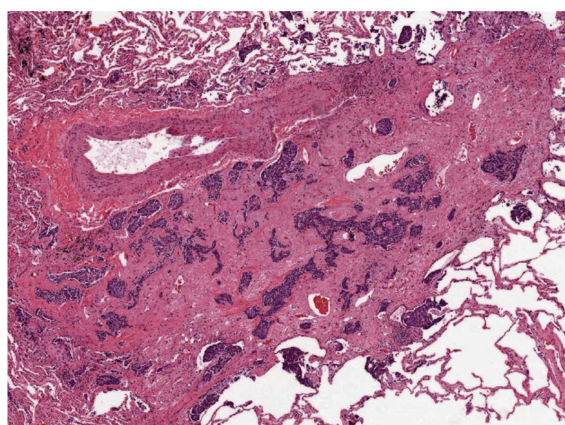


Figure 7-D Hematoxylin-eosin stain ($\times 5$ magnification) of right lower lobe wedge resection.