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Welcome to the ATS 2018 International Conference in San Diego, CA!

On behalf of the Education Committee of the American Thoracic Society (ATS), I would like to welcome you to the 2018 ATS International Conference! With more than 13,000 attendees from more than 100 countries, ATS 2018 is one of the largest gatherings of pulmonary, critical care, and sleep medicine professionals in the world.

In an effort to help busy clinicians with the increasing demands of recertification, the ATS Education Committee has provided 2018 conference attendees the opportunity to earn up to 44.5 American Board of Internal Medicine (ABIM) Maintenance of Certification (MOC) Medical Knowledge Points and 10 American Board of Pediatrics (ABP) Part 2 MOC Self-Assessment points through the Adult and Pediatric Core Curriculum modules and 20 additional symposia. All MOC modules are free for attendees with full conference registration for up to two months post-conference. The Society is excited to be able to offer MOC opportunities in conjunction with the clinical and scientific content that attendees have come to expect from the International Conference. For those not seeking MOC points, the questions support learning in key clinical areas. A list of symposia that are eligible for MOC points appears on the next page and eligible symposia are labeled in the Final Program.

This MOC Study Guide is intended to provide you with essential information needed to earn MOC points at ATS 2018. This Study Guide is intended as a resource only and cannot be submitted for credit. In order to obtain ABIM Medical Knowledge Points or ABP Part 2 Points, you must complete the post-test exams online at https://www.xpressreg.net/register/thor0518/evaluations.asp.

If you have any questions about the ATS MOC portfolio, please contact an ATS staff member at MOC@thoracic.org.

Once more, welcome to ATS 2018!

Debra Boyer, MD, MPHE
Chair, ATS Education Committee
About MOC at ATS 2018

Earn MOC at ATS 2018

The ATS Education and the International Conference Committees have collaborated on a plan where 20 symposia as well as the Adult and Pediatric Core Curriculum symposia are eligible for MOC. Under the plan, ATS 2018 attendees will be able to earn up to a total of 44.5 ABIM MOC points and 10 ABP Part 2 MOC points. The symposia will cover adult pulmonary, critical care, and sleep medicine as well as pediatric pulmonary symposia. The list of Adult and Pediatric symposia eligible for MOC at the conference is included in this study guide.

Successful completion of this CME activity, which includes participation in the evaluation component, enables the participant to earn up to 44.5 MOC in the American Board of Internal Medicine’s (ABIM) Maintenance of Certification (MOC) program. It is the CME activity provider’s responsibility to submit participant completion information to ACCME for the purpose of granting ABIM MOC credit.

Successful completion of this CME activity, which includes participation in the activity, with individual assessments of the participant and feedback to the participant, enables the participant to earn 10 MOC points in the American Board of Pediatrics’ (ABP) Maintenance of Certification (MOC) program. It is the CME activity provider’s responsibility to submit participant completion information to ACCME for the purpose of granting ABP MOC credit.

How to Earn MOC at ATS 2018

• **Before ATS 2018**: Take the pre-test for each core curriculum session and MOC eligible symposium you want to attend. Pre-test can be completed until Saturday, May 19, 2018 (midnight PST). Check back in early May for links to the pre-test.
• **During ATS 2018**: Attend any or all of the MOC symposia you are interested in.
• **After ATS 2018**: Take the post-session test. All the tests will be available at on Wednesday, May 23, 2018, 4 pm PST and attendees can take the tests at no cost through July 31, 2018. Please note: audience response during a session does not count at the post-test.

How to Earn CME at ATS 2018

To Claim CME click here [http://conference.thoracic.org/program/cme-nursing-ce.php](http://conference.thoracic.org/program/cme-nursing-ce.php) to generate your CME certificate for your participation in this, and other CME sessions you attended. You will be asked to complete an evaluation for each session attended and generate a certificate. This will not automatically transfer your MOC points.

There are two separate pathways for claiming MOC and CME. In order to claim both CME and MOC you will need to follow steps for both. You must pass the MOC post-test to earn MOC and complete the CME evaluation to claim CME. Claiming one will not automatically transfer the other.
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PCC1: Pediatric Core Curriculum: Cystic Fibrosis

**Question 1**

A 2-week old female infant presents to clinic for evaluation following an abnormal CF newborn screen (elevated IRT and one mutation of F508del). She was born at 39 weeks’ gestation with birth weight of 3.4 kg. She had a meconium stool on the first day of life. She is exclusively breastfeeding and is gaining weight; however, she remains 5% below her birth weight. There is no family history of cystic fibrosis.

What is the best next test in evaluation for this patient?

A. Fecal Elastase  
B. Nasal Potential Difference  
C. Repeat Newborn Screen  
D. Expanded CFTR Genetic Analysis  
E. Sweat Chloride

**Question 2**

A 30-year-old man and 29-year-old woman are undergoing testing for infertility concerns. The man was found to have absence of the vas deferens with normal spermatogenesis. His urologist recommended work up for cystic fibrosis, including sweat chloride and genetic analysis for CFTR mutations. His sweat chloride was 35 mmol/L, but his genetic analysis showed one mutation in the CFTR gene. The patient was surprised and noted that he has never had any problems with respiratory infections or chronic cough and no gastrointestinal complaints. Genetic counseling was offered to this couple since they plan to proceed with assisted reproductive technology.

What is this patient’s diagnosis?

A. Cystic Fibrosis  
B. CF Transmembrane Conductance Regulator-related Disorder  
C. Atypical Cystic Fibrosis  
D. CF Transmembrane Conductance Regulator-related Metabolic Syndrome or CF Screen Positive, Inconclusive Diagnosis  
E. Delayed Cystic Fibrosis

**Question 3**

A 4 month-old baby presents to clinic for repeat sweat chloride testing. He was initially identified with a positive CF newborn screening. Immune reactive trypsinogen level was elevated and DNA testing on the screening revealed 2 mutations, F508del and D1152H. Initial sweat testing at 2 weeks of age was quantity not sufficient. He is followed in clinic and continues to gain weight well. Repeat sweat testing at 4 months of age is 39 mmol/L.

Which of the following is the most accurate diagnosis for this patient?

A. Cystic Fibrosis  
B. CF Transmembrane Conductance Regulator-related Metabolic Syndrome or CF Screen Positive, Inconclusive Diagnosis  
C. CF Transmembrane Conductance Regulator-related Disorder  
D. Not enough information to determine a diagnosis at this time  
E. No CF or CFTR-related Diagnoses
Question 4
A 10-year-old boy with cystic fibrosis and stable nutritional status presents to his quarterly CF clinic appointment. His surveillance respiratory culture at this visit returns positive for the first time with 1 strain of Pseudomonas aeruginosa (Pa). The strain is non-mucoid and sensitive to ciprofloxacin, gentamicin, tobramycin, amikacin, and aztreonam. He is otherwise well-appearing with no respiratory complaints. His FEV1% predicted on spirometry was 108% predicted (at his baseline).

Which of the following choices is the next best step in the management of initial positive Pa respiratory culture?
A. Start no treatment until at least 2 positive cultures in the previous 12 months; follow-up respiratory culture in 3 months
B. Initiate inhaled tobramycin for 28 days; follow-up respiratory culture after treatment completion
C. Initiate oral ciprofloxacin for 14 days; follow-up respiratory culture after treatment completion
D. Initiate intravenous tobramycin for 14 days; follow up respiratory culture after treatment completion
E. Initiate inhaled tobramycin for 56 days; follow-up respiratory culture after treatment completion

Question 5
You are seeing a 3-year-old girl with pancreatic insufficient cystic fibrosis. She has no respiratory symptoms. Her weight-for-age has been at the 5th percentile. Her body mass index (BMI) is at the 10th percentile. She has gained 3g/day over the last 3 months. Her parents are concerned she is a picky eater.

Which of the following statements according to the 2016 CF Foundation Preschool Clinical Practice Guidelines regarding management of nutritional risk is TRUE?
A. Regular assessment of mealtime behavior challenges and family stress is recommended at each clinic visit
B. Inpatient admission is recommended in the initial tier for management of nutritional risk
C. Specialty consultation with gastroenterology is recommended in the initial tier for management of nutritional risk
D. Weight-for-age of preschoolers with CF is recommended to be maintained > 50th percentile
E. Recommended clinic follow up for a toddler in nutritional risk is 12 weeks and close communication with the primary care provider

Question 6
A 17-year-old girl with cystic fibrosis (F508 del homozygous), pancreatic insufficiency, and chronic endobronchial infection with Pseudomonas aeruginosa, presents to CF clinic for her quarterly appointment. Her current medications are dornase once daily, hypertonic saline 7%, albuterol, an oral contraceptive pill, azithromycin, fat soluble vitamin supplements, and pancreatic enzymes. The family has expressed interest in starting lumacaftor-ivacaftor.

Which of the following statements regarding initiation and monitoring of lumacaftor-ivacaftor is NOT recommended?
A. Baseline liver function testing prior to initiation, every 3 months during the first year of use, and annually after
B. Baseline eye exam to evaluate for cataracts before initiation, and during treatment
C. Discuss alternative birth control options and menstrual irregularities during treatment
D. Baseline complete blood cell count and differential, every 3 months during the first year of use, and annually after
E. Discuss any new medications and herbal supplements with the CF team while taking lumacaftor-ivacaftor

A81: Pediatric Year in Review

Question 1
A 15-week old infant girl has just been diagnosed with Spinal Muscular Atrophy type I. She has 2 copies of SMN 2 and first became symptomatic at 8 weeks of age. She is currently requiring long-term ventilation. You are seeing this family in your clinic to review her respiratory status. The parents ask about treatments for their daughter.
Based on the current evidence and clinical milieu, which of the following best describes the treatment options and the potential outcomes for their daughter?

A. They can expect their daughter to achieve normal motor milestones and to be weaned off ventilation within 13 months of starting nusinersen therapy.
B. Gene therapy is clinically available and is the optimal treatment for their daughter because it requires a single intrathecal injection as compared to nusinersen therapy which requires repeated injections.
C. The likelihood of their daughter having improvement in her motor milestones and being weaned off ventilation after 13 months is greater on nusinersen therapy as compared to not being on therapy.
D. Salbutamol is clinically available and is the optimal treatment for their daughter because it has been shown to result in greater improvement in muscle strength than other existing therapies.
E. The likelihood of their daughter having improvement in her motor milestones after 13 months is greater on nusinersen therapy as compared to not being on therapy. However, it is not possible for her to be weaned off ventilation once it has started.

**Question 2**

A 6-year old boy has just been diagnosed with Duchenne Muscular Dystrophy. You are meeting the family for the first time. The parents ask you about treatment for their son.

Which of the following treatments has been shown to improve survival in patients with Duchenne Muscular Dystrophy?

A. Atalauren
B. Nusinersen
C. Eteplirsen
D. Deflazocort
E. Drisapersen

**Question 3**

The application of microbiome methods to chronic lung infections, such as cystic fibrosis lung disease, has identified many microbes in respiratory samples not usually identified by routine culture of the same samples. In CF, studies using microbiome methods have identified some general features of how respiratory specimen microbiology evolves over time within patients.

Which of the following describes some general findings from microbiome studies of CF lung disease?

A. The number of microbial species detected in CF respiratory samples increases as patients get more antibiotics.
B. The overall microbial diversity in CF respiratory samples tends to decrease from childhood to end-stage disease.
C. Fungi are more abundant at end-stage than are bacteria.
D. The early respiratory sample microbiota are dominated by Pseudomonas aeruginosa, after which Burkholderia species supplant it.
E. The microbiota of CF respiratory samples are the same as from people without CF.

**Question 4**

You are considering starting a biologic agent in a 15-year old atopic patient with severe steroid-dependent asthma.

Which of the following statements is true?

A. There are two biologic agents that are approved for use in the US for children of this age.
B. Obtaining a sputum cell count is important in determining what biologic agent to use
C. Omalizumab is ineffective in children with an IgE >700 IU/ml
D. Clinical trials of omalizumab have shown that it is effective in allowing patients to decrease their maintenance oral steroid dose.
E. Clinical trials of mepolizumab have shown that it is effective in allowing patients to decrease their maintenance oral steroid dose.
A1: Clinical Year in Review 1: ILD, Pulmonary Rehabilitation, Lung Transplantation, Interventional Pulmonology

Question 1
A 75-year old woman presents with progressive dry cough and dyspnea on exertion. She is a lifetime non-smoker, there are no environmental/occupational exposures of note and no signs or symptoms of connective tissue disease. Her high resolution computed tomography (HRCT) scan of the chest demonstrates a ‘possible UIP’ pattern. The differential diagnosis includes idiopathic pulmonary fibrosis, but she is deemed too high risk to undergo surgical lung biopsy.

What combination of variables should you consider when applying the ‘UIP score’ to estimate the positive predictive value of a ‘possible UIP’ pattern for underlying histopathological UIP?
A. Sex, smoking history, radiographic presence of honeycombing
B. Age, duration of symptoms, baseline lung function
C. Age, sex, HRCT traction bronchiectasis score
D. Age, sex, family history of pulmonary fibrosis
E. Exposure history, mosaic attenuation or air-trapping on HRCT

Question 2
A 63-year old man with a history of COPD and a forced expiratory volume in one second (FEV1) of 34% predicted is referred for an outpatient eight-week supervised pulmonary rehabilitation program. He has suffered two exacerbations in the past year, one requiring hospitalization. The last exacerbation was four months ago. His modified MRC dyspnea score is 3. His self-reported physical activity level comprises 5 minutes of walking at self-selected pace, once a week.

Which of the following is the most likely outcome to improve with pulmonary rehabilitation?
A. Time spent in at least moderate intensity physical activity
B. Exercise capacity
C. Hospitalization rate
D. Daily step count
E. FEV1

Question 3
A 67-year old man with very severe COPD has noticed worsening breathlessness over the last year. He quit smoking 3 years ago. Lung function shows an FEV1 0.8L (28%) FVC 1.8L with a residual volume of 210% predicted. CT showed heterogenous emphysema and intact lobar fissures. The patient was referred for consideration of endobronchial valve therapy.

Which one of the following statements are correct:
A. Assessment of collateral ventilation with Chartis is not required
B. Endobronchial valves are preferred in the presence of paraseptal emphysema
C. Valves should only be considered when 6 minute walk distance is <150m
D. Endobronchial valves are more likely to be placed in the left lung as the oblique fissure is more likely to be intact on the left
E. The risk of pneumothorax after endobronchial valve placement is approximately 5%

Question 4
A 24-year old male with end stage cystic fibrosis underwent a bilateral sequential lung transplant with a 50-year old male donor with a 28-pack year smoking history who died of complications due to a stroke. Within 72 hours post-transplant, the patient was noted to have severe primary graft dysfunction Grade 3 requiring extracorporeal membrane oxygenation (ECMO) support.

This patient is at increased risk for the development of which of the following?
A. Acute cellular rejection
B. Neurocognitive decline
C. Chronic Lung Allograft Rejection (CLAD) – bronchiolitis obliterans type (BOS)
D. Chronic Lung Allograft Rejection (CLAD) – restrictive allograft syndrome (RAS)

Question 5

A 24-year old male with end stage cystic fibrosis underwent a bilateral sequential lung transplant with a 50-year old male donor with a 28-pack years smoking history that died of complications due to a stroke. Intra-operatively, the patient had adhesion and bleeding from the chest wall and received 10 units of packed red blood cells. On completion of the surgery the patient returned to the intensive care unit normotensive, afebrile with the following studies and imaging:

- White Blood Cells 48.6 k/ul
- Hemoglobin 8.6 g/dl
- Platelet Count 425 k/ul
- Arterial Blood Gas after arriving to ICU on Pressure Control Ventilation 100% FiO2 Peep 5
  - pH, arterial 7.35
  - pCO₂, arterial 56 mmHg
  - pO₂, arterial 71 mmHg
- Portable Chest Xray done on arrival to the ICU

Which of the following is the best explanation for this patient’s presentation?
A. Transfusion related lung injury (TRALI)
B. Pneumonia
C. Hyperacute rejection (HAR)
D. Primary Graft Dysfunction (PGD)


Question 1

You have established your hospital as a designated site for lung cancer screening. Which of the following statements is true?
A. CMS requires that only positive LDCT screening examinations performed at a designated site should be reported to a national registry.
B. CMS requires data from all LDCT screening examinations performed at a designated site to be reported to a national registry.
C. CMS requires that only information on patients who are ultimately diagnosed with lung cancer needs to be reported to a national registry.
D. CMS recommends that designated sites report LDCT screening examination results to a national registry but that sites may opt out.
E. CMS recommends that all patients with findings from a diagnostic CT scan be submitted to the national registry for screening.
**Question 2**

A hospital wants to initiate a multidisciplinary LDCT program in order to improve outcomes of patients with lung cancer.

Which of the following is true regarding successful LDCT screening programs?

A. Engaged primary care practitioners in the late stages of planning and implementation
B. Involved primary care practitioners early on and conducted educational sessions to emphasize the role of LDCT screening in improving the quality of care
C. Did not educate primary care practitioners regarding the pros and cons, given that this would reduce the rate of screening
D. Involved pulmonologists, rather than primary care practitioners, because they have the widest base of eligible patients
E. Solicited specialist opinion on workflow feedback and concerns regarding the benefits of LDCT

**Question 3**

Mr. Smith is a 54-year old male with a history of liver cirrhosis, most recently admitted to the hospital for decompensation 10 days ago. He has a 30 pack-year smoking history. He is a current smoker and requests a LDCT screening for lung cancer during his transition of care visit.

Which of the following is the most appropriate next step?

A. Offer LDCT as he is a current smoker
B. Defer LDCT till one more year has passed
C. Defer LDCT for ten more years
D. Defer LDCT until the patient has stopped smoking for ten years
E. Do not offer LDCT

**Question 4**

Bruce Wayne is a 58-year old male with a 40 pack-year smoking history. He underwent an LDCT after a shared decision making meeting at his last routine visit with his primary care practitioner. He is now in the office to discuss the results of the scan and is visibly upset that he has a single 4 mm nodule. He is very worried that he now has lung cancer.

What is the most appropriate response in this clinical situation?

A. Referral for nodule biopsy because < 10 % of screens are positive
B. Referral to pulmonology because > 25% of nodules are cancerous
C. Reassurance and follow up, because approximately 95% of screen-detected nodules are benign
D. Refer to oncology because > 40 % of nodules are cancerous
E. Reassure that up to 50% scans can be positive, but refer to pulmonology for follow up scans

**Question 5**

A 70-year old patient with mild COPD presents to his primary care physician’s office for a shared decision discussion regarding whether or not to undergo lung cancer screening.

Which of the following statements is correct?

A. The purpose of shared decision-making is to convince all eligible patients to undergo lung cancer screening
B. The shared-decision making visit is not required by CMS prior to starting lung cancer screening
C. Only a patient’s primary care physician should perform shared decision-making with the patient being considered for lung cancer screening
D. The purpose of the shared decision-making visit is to allow individuals to weigh the risks and benefits of screening based on their personal risk profile
E. CMS allows for a shared decision-making discussion to take place by phone
CC1: Sleep Medicine Clinical Core Curriculum I: Hypersomnia, Narcolepsy, Non-Narcoleptic Hypersomnia

Question 1

A 25-year-old female graduate student with no significant past medical history presents with excessive daytime sleepiness. She reports that her symptoms started in college. She endorses difficulty waking up in time for her 8 am class, and frequently falls asleep during class. She reports that her habitual bedtime is variable, depending on how much studying she needs to accomplish. She endorses trouble falling asleep the night before an exam. This quarter, she has to wake up at 7 am for classes 5 days a week. She sleeps in until as late as noon on the weekends, and sometimes naps in the afternoon. During a recent break from school, she slept from about 4 am until noon and awoke feeling refreshed. Her current Epworth Sleepiness Scale score is 15/24. Her BMI is 22. She does not snore or have witnessed apneas.

What is the best next step in her management?
A. Obtain a home sleep apnea test
B. Refer for psychiatry assessment for suspected depression
C. Refer for cognitive behavioral therapy for insomnia (CBT-I)
D. Obtain two weeks of actigraphy accompanied by sleep diaries
E. Obtain an in-lab polysomnogram followed by multiple sleep latency testing

Question 2

A 19-year-old female college student with no significant past medical history presents for evaluation of excessive daytime sleepiness. Her Epworth Sleepiness Scale score is 16/24. She reports taking no medications. A thyroid stimulating hormone level, complete blood count, and basic metabolic panel were all within normal limits. A urine toxicology test was negative. She does not snore and does not report cataplexy.

Overnight, In-lab Polysomnogram:
Total sleep time: 420 minutes
Sleep efficiency: 90%
Microarousal Index: 10/hour
Sleep onset latency: 10 minutes
REM onset latency: 90 minutes
Apnea hypopnea index: 4 events/hour
Periodic limb movement index: 2/hour
Wake time: 7 am
Multiple Sleep Latency Testing:
Start Time: 9 am

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Mean Sleep Onset Latency: 8 minutes

Actigraphy:
Performed for the two weeks preceding polysomnography:
Average bed time: 3 am
Average wake time during the week: 7 am
Average wake time on the weekends: 12 pm
What is the most likely diagnosis at this time?
A. Idiopathic Hypersomnia
B. Narcolepsy Type II
C. Insufficient Sleep Syndrome
D. Non-24 Hour Sleep Wake Rhythm Disorder
E. Kleine-Levin Syndrome

Question 3
An 18 years old woman with narcolepsy type I (narcolepsy with cataplexy) is seeing you for follow up. She has been on a stable dose of sodium oxybate and armodafinil for the last year. She has had no recent episodes of cataplexy. She is graduating from high school in a few months and is anxious about her ability to perform academically in college. She has been reading about ways to assess her current stimulant regimen, and asks whether she should undergo a maintenance of wakefulness test.

Which of the following is true about the Maintenance of Wakefulness Test (MWT)?
A. MWT is part of the routine assessment of the efficacy of stimulant therapy in narcolepsy
B. MWT consists of four trials performed at two hour intervals
C. MWT is a validated measure of the tendency to fall asleep
D. MWT is recommended during the initial diagnostic evaluation of hypersomnia
E. During the MWT, patients may stand and walk around the room if they wish

Question 4
AA 25-year-old man presents to your clinic for a second opinion. The patient was seen by a local sleep specialist, and after formal evaluation, he was diagnosed with narcolepsy type 1. He complains of approximately 12 months of excessive daytime sleepiness that has worsened with time, and “passing out” when one of his friends tells him a joke. He takes two naps during the day and feels refreshed for several hours after each nap. He wakes up multiple times during the night and does not feel that his sleep is refreshed.

Which of the following features is most commonly associated with cataplexy?
A. Usually caused by negative emotions, such as anger and sadness
B. Loss of consciousness
C. Deep tendon reflexes are preserved
D. Can be made worse by withdrawal of selective serotonin reuptake inhibitors (SSRIs)
E. Episodes become more frequent and intense over time

Question 5
You are evaluating an 18-year-old woman for suspected narcolepsy type 1. She endorses excessive daytime sleepiness, with an Epworth Sleepiness Scale of 16. Her description of cataplexy is somewhat atypical. You decide to proceed with polysomnography (PSG) followed by multiple sleep latency test (MSLT). The following are the results:

Overnight in-laboratory PSG:
Total sleep time: 450 minutes
Sleep efficiency: 75%
Sleep latency: 3 minutes
REM latency: 10 minutes
Apnea Hypopnea Index: 2.5 events/hour
Periodic Limb Movement Index (PLMI): 0.7 events/hour
MSLT:
Given your suspicion for narcolepsy type 1, you contemplate additional testing. Which of the following is needed to establish the diagnosis?

A. No additional testing is required  
B. HLA DQB1*0602 testing  
C. Cerebrospinal fluid (CSF) Hypocretin-1 (orexin) level  
D. Serum Hypocretin-1 (orexin) level  
E. Maintenance of wakefulness testing (MWT)

**Question 6**

You are seeing a 23-year-old woman in clinic for follow-up of narcolepsy type 1, confirmed by multiple sleep latency testing (MSLT). In addition to recommending maintenance of a regular sleep schedule with brief planned naps, you had previously started the patient on modafinil 200 mg twice a day. She notes mild improvement in daytime sleepiness, but continues to experience fragmented sleep at night, as well as frequent episodes of “buckling at the knees” and slurred speech when she laughs or is excited.

What would you recommend in order to effectively treat her ongoing symptoms?

A. Increase modafinil to 400 mg twice a day  
B. Change modafinil to amphetamine  
C. Change modafinil to fluoxetine  
D. Add suvorexant  
E. Add sodium oxybate

**Question 7**

An 18-year-old previously healthy man presents with 11 days of sudden onset profound sleepiness. His parents state that he now sleeps 20 hours daily, and when awake has poor concentration and difficulty participating in conversations. He will over-eat compulsively and then go back to sleep. He had a similar episode 7 months ago that resolved spontaneously. Workup during that episode was normal including toxicology screens, brain MRI, EEG, polysomnography, and laboratory testing for inborn errors of metabolism and Lyme disease.

What is the best treatment for his symptoms?

A. Modafinil  
B. Venlafaxine  
C. Valproic acid  
D. Risperidone  
E. Supportive treatment

**Question 8**

A 23-year-old healthy woman presents with several years of excessive daytime sleepiness and drowsy driving despite sleeping an average of 9.5 hours nightly. She has rare sleep paralysis, and has never experienced cataplexy or hypnopompic or hypnagogic hallucinations. Physical exam is normal and does not include focal neurological abnormalities. She underwent polysomnography (PSG) followed by MSLT. PSG showed high sleep efficiency of 96%, 8.9 hours of sleep, and was otherwise unremarkable with normal REM onset latency, and absence of sleep apnea, respiratory effort related arousals (RERAs), seizures, and abnormal limb or body movements. MSLT showed a mean sleep onset latency of 7 minutes and absence of sleep onset REM periods (SOREMPS).
What additional testing is indicated for this patient?

A. Brain MRI
B. Positron Emission Tomography (PET) scan
C. Lumbar puncture for CSF analysis
D. No additional testing is indicated

**Question 9**

A 27-year-old man has had excessive daytime sleepiness since adolescence. He has sleep inertia with significant difficulty getting out of bed in the mornings despite sleeping for an average of 9 hours nightly and napping for an hour every afternoon. His tendency to doze inadvertently is causing trouble at work and adversely affected his grades while he was in college. He finally presented to a sleep specialty clinic after falling asleep while driving. He underwent polysomnography (PSG) followed by multiple sleep latency testing (MSLT). Based on his history, unremarkable physical examination, and sleep study results, he was given a diagnosis of Idiopathic Hypersomnia (IH).

What is the best initial treatment for excessive daytime sleepiness in Idiopathic Hypersomnia?

A. Modafinil
B. Pitolisant
C. Sodium oxybate
D. Dextroamphetamine
E. Supportive treatment

**A89: Emerging Concepts in Chronic Thromboembolic Pulmonary Hypertension**

**Question 1**

A 66-year old man presents with a one year history of progressive dyspnea on exertion. Two years prior, he had an episode of acute pulmonary embolism diagnosed after knee replacement surgery, for which he was treated with warfarin for six months. His chest x-ray is normal. His pulmonary function testing does not show any abnormalities. You are suspicious of chronic thromboembolic pulmonary disease.

Which of the following diagnostic tests has the highest sensitivity for chronic thromboembolic pulmonary hypertension and can thus be used best to rule out the disease?

A. Echocardiography
B. Ventilation Perfusion Lung Scan
C. CT Pulmonary Angiography
D. Electrocardiogram
E. Chest radiograph

**Question 2**

A 35 month old boy with moderate persistent asthma has a history of loud snoring every night for the past year. He has not had any behavioral changes during wakefulness. Weight has remained constant at the 25th percentile. He has had 2 asthma admissions over the past year. Physical examination is remarkable for an adenoidal facies and 2+ tonsils. Medications include albuterol, inhaled fluticasone (110 mcg) and montelukast. The waiting time for a polysomnogram in his hometown is 2 months.

What is the next best step in the management?

A. Obtain a home pulse oximetry study
B. Refer to an otolaryngologist for adenotonsillectomy
C. Obtain a lateral neck Xray to assess tonsil and adenoid size
D. Obtain an in-laboratory polysomnogram
E. Observe the patient for 6 months to see if symptoms improve
Question 3
Two years prior, a 34-year old male experienced a leg fracture complicated by a DVT. He received three months of anticoagulation with complete resolution of symptoms. Over the last six months he has experienced a gradual decline in exercise capacity. An echocardiogram revealed mild right atrial and right ventricular enlargement; a CT angiogram was read as negative for thromboembolic disease. He was diagnosed with Group I Pulmonary arterial hypertension.

What should be the next step?
A. Initiate pulmonary hypertension targeted medical therapy
B. Perform a right heart catheterization
C. Obtain a lung ventilation-perfusion lung scan
D. Resume anticoagulation
E. Obtain a magnetic resonance angiogram

Question 4
A 55-year old woman with a history of pulmonary embolism two years prior presents for the evaluation of dyspnea on exertion. She has a history of systemic hypertension and dyslipidemia. Her medications include warfarin, atorvastatin and hydrochlorothiazide. Physical examination is unremarkable other than an elevated body mass index of 36. ECG, CXR and pulmonary function tests are normal. A ventilation perfusion lung scan shows large mismatched perfusion defects in both lower lobes. A CT pulmonary angiography shows chronic thromboembolic occlusion of both lower lobe arteries. An echocardiogram shows stage 1 left ventricular diastolic dysfunction, normal estimated pulmonary artery systolic pressure and normal right ventricular size and function. A right heart catheterization shows normal pulmonary artery pressure and vascular resistance.

What should you do next?
A. Refer for pulmonary thromboendarterectomy surgery
B. Refer for balloon pulmonary angioplasty
C. Start riociguat therapy
D. Obtain a cardiopulmonary exercise test
E. Reassure the patient and continue anticoagulation

Question 5
You diagnosed a 68-year old man with chronic thromboembolic pulmonary hypertension (CTEPH) based on ventilation perfusion lung scan, pulmonary angiography and right heart catheterization. The patient tells you he has heard of a new procedure to treat this disease called balloon pulmonary angioplasty (BPA), and asks your opinion.

Before a patient with CTEPH undergoes balloon pulmonary angioplasty (BPA), they should:
A. Be treated with pulmonary hypertension targeted therapies and reassessed to determine if BPA is necessary.
B. Change their current anticoagulant to an alternate agent and reassess if BPA is necessary.
C. Have an evaluation for operability at an experienced pulmonary thromboendarterectomy or CTEPH center.
D. Undergo treatment with catheter directed thrombolytics.
E. BPA is experimental and not recommended at this point

Question 6
You diagnosed a 56-year old woman with chronic thromboembolic pulmonary hypertension (CTEPH) based on a ventilation perfusion lung scan, pulmonary angiography and right heart catheterization. The patient is evaluated at an expert referral CTEPH center with surgical expertise and is deemed to be inoperable. You obtained a second opinion from another CTEPH Center and received the same inoperability opinion. You would like to start medical therapy.

Which therapy is approved for patients with inoperable chronic thromboembolic pulmonary hypertension (CTEPH)?
A. Bosentan
B. Riociguat
C. Sildenafil
D. Iloprost
E. No medical therapy is approved for inoperable CTEPH

A91: Reducing Disparities in Environmental Lung Disease: Emerging Methods in Exposure Mitigation

Question 1
A 54-year old Latino man is a welder with a high school education who recently failed a respirator fit test because of an FEV1/FVC < 65% and an FEV1 of 68% without improvement with inhaled bronchodilator administration. He smoked a half-pack per day for 16 years and quit 20 years ago. He is asymptomatic with unlimited exercise tolerance.

Cessation of work as a welder with maintenance of income would be the ideal outcome for this individual. However, if this situation is not achievable, which of the following would be the most important recommendation for management?
A. Treatment with inhaled corticosteroids
B. Fitting for alternative respiratory protective gear and vaccination for influenza, pneumonia, and pertussis
C. Monthly monitoring with spirometry
D. No specific recommendation, as he is asymptomatic

Question 2
A 5-year old African American boy diagnosed with asthma in early childhood has persistent symptoms of cough and wheeze, despite maintenance therapy with an inhaled corticosteroid. He has increased exercise-related and nocturnal respiratory symptoms and uses his rescue inhaler frequently. He lives in low-income housing in urban Baltimore. His father is an unemployed chronic smoker and admits to smoking in the home. Neighbors in the adjoining residential units are also smokers.

Which of the following is true about the contribution of secondhand smoke to this boy’s symptoms?
A. Indoor smoking results mainly in combustion-generated nitrogen dioxide (NO2) emissions which are provoking this boy’s asthma symptoms.
B. Outdoor pollution in cities such as Baltimore vastly exceeds indoor pollutant levels, and therefore the effects of indoor pollutant levels are negligible.
C. Secondhand smoke predominantly contributes to indoor fine particulate matter (PM2.5) concentrations, which may be responsible for the boy’s increased asthma symptoms.
D. Use of a high-efficiency particulate air (HEPA) filter would not be effective in reducing the adverse effects of secondhand smoke exposure in this boy, since he lives with an active smoker.

Question 3
A 10-year old girl with asthma presents to the pediatric asthma clinic for follow-up. She lives with her mother, father, and grandmother, and two of her siblings in a multiunit housing complex. Her grandmother, the only smoker in the home, quit smoking 6 months ago. They have purchased a high energy particulate air (HEPA) filter for use in the patient’s bedroom. Both the patient and her mother were hopeful that she could stop use of a prescribed inhaled corticosteroid, but attempts at step-down therapy have been unsuccessful. Her mother would like to know which exposures may still be contributing to her daughter’s asthma symptoms.

Which of the following is true regarding relevant exposures?
A. Living in multi-unit housing versus a detached home has no effect on secondhand smoke exposure.
B. Particulate matter concentrations in the home may exceed those outdoors.
C. Sweeping and vacuuming are associated with equivalent amounts of airborne particulate matter.
D. Use of a high-efficiency particulate air (HEPA) filter eliminates exposure to harmful tobacco products in the home of an active smoker.
Question 4

Different approaches exist to characterize workers’ exposure to chemical or biological agents.

Which of the following is true regarding exposure assessment methods?

A. Job title information is easy to collect, but the same job in different contexts may be associated with different exposures.
B. Personal air measurements and biomonitoring always provide the most accurate assessment of exposure.
C. A Job Exposure Matrix provides detailed quantitative information relating jobs to an exposure profile.
D. Expert reviews of individual jobs are more accurate than programmable decision rules in assessing exposure risk.

Question 5

Intervention studies are generally seen as important evidence of causality of exposure response relations and give information about the efficacy of intervention measures.

Why are intervention studies not regularly conducted in the work environment?

A. Intervention studies cannot be implemented easily in the work environment.
B. Observational studies usually give sufficiently strong evidence.
C. Follow-up studies in the work environment are not possible.
D. Intervention studies are only useful in the context of secondary prevention.
E. Intervention studies are not necessary when exposure response relations are available.
Monday, May 21

PCC2: Pediatric Core Curriculum 2: Interstitial Lung Disease

Question 1
Children with progressive neuromuscular weakness undergo a typical evolution of respiratory involvement. Which of the following is a respiratory morbidity commonly associated with patients with neuromuscular disease?
A. Impaired cough
B. Pneumothorax
C. Pulmonary edema
D. Recurrent bronchodilator responsive wheezing
E. Pulmonary embolism

Question 2
A 15-year-old male with Duchenne muscular dystrophy has recently required the use of a wheelchair due to progressive weakness. He has developed daytime somnolence and morning headaches over the past six months. On physical exam, he is not in acute distress, alert and oriented to self, place, and time. He has a respiratory rate of 16/minute, SpO2 of 98% in room air, and awake end-tidal CO2 of 37 mmHg. His lungs are clear to auscultation.

What should be the next step in evaluating for hypoventilation?
A. Spirometry to determine whether the FVC is < 80% predicted.
B. Overnight pulse oximetry to evaluate for hypoxemia
C. There is no need since the awake EtCO2 is normal.
D. Polysomnography with capnography to evaluate for nocturnal hypoventilation
E. Maximal inspiratory pressure (MIP) measurement to determine if the patient has inspiratory muscle weakness and likely hypoventilation

Question 3
A 14-year-old male with spinal muscular atrophy type-2 is here to see you for a pulmonary follow-up visit. He is non-ambulatory and has developed scoliosis and significant contractures of the wrists and elbows bilaterally. You would like to obtain pulmonary function testing and understand the importance of normalizing the data relative using available predicted equations.

Since pulmonary function data are normalized relative to age, race, sex and body size, what would be the best representation of body size for this patient?
A. Height
B. Arm span
C. Ulna length
D. Weight
E. Body mass index
Question 4
This is a picture of a patient with neuromuscular weakness, specifically a patient with spinal muscular atrophy (SMA).

What is the physiologic cause of this abnormality?
A. Pulmonary hypoplasia
B. Weakness of the intercostal muscles
C. Weakness of the diaphragm
D. Abnormal rib development in-utero
E. Scoliosis

Question 5
A 13-year-old male with Duchene Muscular Dystrophy (DMD) has a polysomnogram which reveals moderate sleep efficiency. There were 0 central, 75 obstructive, and 4 mixed apneas recorded. There were 15 hypopneas during the study. AHI was 14, with REM AHI of 57. The lowest saturation recorded was 77%, with 1% of the study time spent with saturation less than 90%. The highest end-tidal CO2 was 50torr, with morning PCO2 of 42mmHg.

What is the most appropriate therapy to best support this patient during sleep?
A. Positional interventions – repeat sleep study in 1 year
B. CPAP 6, further titrated to relieve obstructive apneas
C. Supplemental oxygen to keep saturations >90%
D. BiPAP 8/4, further titrated to relieve obstructive apneas
E. Close monitoring and repeat the study in 1 year

Question 6
A 13-year-old male with Duchenne Muscular Dystrophy (DMD) is scheduled for spinal fusion surgery due to worsening scoliosis. He undergoes appropriate pre-operative testing.

Which of the following measurements has been identified as a predictor of post-operative respiratory complications and need for ventilator assistance following spinal surgery?
A. Peak cough flow 281 L/min
B. Forced vital capacity (FVC) 29% predicted
C. Maximal expiratory pressure (MEP) 65 cmH20
D. SpO2 94%
E. Capillary PCO2 45mmHg

Question 7
AA 2 month-old former term female infant is hospitalized during the winter for a viral illness causing tachypnea and hypoxia. Exam is notable for significant hypotonia with head lag, thin muscle mass, and absent patellar reflexes. Her cough is weak and she seems to be having difficulty tolerating her nasal and oral secretions.

You suspect a neuromuscular disease, particularly spinal muscular atrophy. What is the next step in confirming the diagnosis?
A. Genetic microarray  
B. Urine organic acids, serum amino acids  
C. Creatinine Kinase (CK) levels  
D. Genetic deletion testing  
E. Electromyography (EMG)

B1: Clinical Year in Review 2: COPD, Asthma, Lung Cancer, Pleural Disease

**Question 1**

A 46-year old woman with allergic rhinitis presents to you for a second opinion about the management of her asthma. Aside from seasonal rhinitis symptoms, she has been healthy for most of her life and works as an elementary school teacher. Four years ago, she had a severe upper respiratory viral illness associated with paroxysmal cough as well as occasional wheeze and dyspnea on exertion. She saw her primary care provider after 2 weeks of symptoms and was given a prescription for albuterol. The wheezing and dyspnea improved, but the cough persisted several more weeks until a medium-dose inhaled corticosteroid (ICS) was added for maintenance via a telephone encounter. At follow up six months later, her physician told her that she probably had asthma and that inhaled steroids are the best way to prevent exacerbations. Since that time, she has continued on medium-dose ICS twice daily with rescue albuterol.

On presentation today, she has not had asthma symptoms and her ACT score is 25 of 25 points. She has had one to two colds a year since her asthma diagnosis but has not needed prednisone or urgent medical care. Other than obesity with a BMI of 34 and nasal erythema, her exam today is unremarkable. Spirometry today is also normal, without prior comparisons. She would like to do everything she can to avoid getting sick but is also concerned about the long-term side effects of inhaled steroids.

The best course of action to address her concerns is:

A. Continue her medium-dose ICS and confirm appropriate inhaler technique with a spacer.  
B. Switch from the ICS inhaler to a leukotriene receptor antagonist for better adherence and fewer side effects.  
C. Reduce her ICS dose sequentially until discontinued and order a methacholine challenge study if her spirometry remains normal after 6 weeks of stopping ICS.  
D. Provide assurance that the side effect profile of medium-dose ICS is minimal.

**Question 2**

A 65-year old man with a 30 pack-year history of tobacco use presents to your clinic with a 5.5 cm centrally located right upper lobe mass and enlarged right hilar lymph nodes. You perform bronchoscopic biopsy with endoscopic bronchial ultrasound. Pathology from biopsies of both the primary lesion and ipsilateral hilar lymph nodes shows non-small cell lung cancer. Staging evaluation reveals no distant metastases. The thoracic surgeon at your institution has deemed the tumor unresectable based on its location and the patient’s severe underlying COPD. Genetic testing of the tumor samples is pending.

Which of the following would represent an appropriate personalized treatment algorithm for this patient?

A. If the tumor is epidermal growth factor receptor (EGFR) mutation-negative, treatment with an EGFR tyrosine kinase inhibitor such as erlotinib is recommended as first-line therapy.  
B. If the tumor is EGFR positive, treatment with platinum-based chemotherapy and radiotherapy is recommended as first-line treatment.  
C. If genetic testing does not reveal treatable oncogenic alterations, initial platinum-based chemoradiotherapy, followed by adjuvant immunotherapy with durvalumab, should only be considered for patients with tumor programmed death ligand 1 (PD-L1) expression >50%.  
D. If the tumor is EGFR positive (exon 19 deletion or L858R), osimertinib may be considered as first-line therapy.  
E. Even among non-small cell lung cancer patients with high levels of PD-L1 expression (>50%), immunotherapy with immune checkpoint inhibitors is not an appropriate consideration given unacceptably high rates of severe pneumonitis.
Question 3
A clinical trial published in 2017 compared treatment with tiotropium to placebo for 24 months in patients with mild to moderate COPD.

Compared to placebo, treatment with tiotropium was associated with which of the following observations?
A. Higher study drop-out rate
B. Higher pre-bronchodilator FEV1
C. Greater COPD exacerbation rate
D. Fewer adverse events
E. Worse COPD-specific quality of life measurements

Question 4
A 70-year old man who presents with dyspnea, cough, and weight loss for 1 month is found to have a large pleural effusion. The effusion has positive cytology for adenocarcinoma, likely due to lung cancer. His dyspnea improves with removal of 1L. He now has a chest tube for drainage but, over several days, the lung has not re-expanded. He wishes to go home and take care of himself. His wife is also eager to have him back home, to spend his remaining days in relative comfort. He will consider undergoing chemotherapy as an outpatient but has not decided yet. What should be done about the chest tube for his malignant pleural effusion, which is draining 200 mL per day?

What is the next best step in the management?
A. Perform talc pleurodesis through the chest tube.
B. Refer for VATS to reexpand the lung followed by talc insufflation.
C. Place indwelling pleural catheter.
D. Remove the chest tube and plan for outpatient thoracentesis if needed.
E. Place indwelling pleural catheter and plan for talc instillation to follow.

B10: Lung Cancer Screening: Who's in and Who's Out? A Pro/Con Debate

Question 1
A 60-year old man with a history of COPD presents for a routine visit with his pulmonologist. He has a 40 pack-year history and remains an active smoker. Spirometry from a year ago was notable for an FEV1 that was 46% predicted and an FVC of 80% predicted. He has had one exacerbation in the past year, which resolved after a short course of prednisone and azithromycin. He currently is on tiotropium, fluticasone-salmeterol, and albuterol as needed. He has been using oxygen 2L/min via nasal cannula at rest and with exertion for the past two years.

Which of the following would make this patient ineligible for lung cancer screening?
A. Spirometry results
B. History of a COPD exacerbation
C. Smoking history
D. Use of supplemental oxygen
E. This patient would be eligible for screening

Question 2
A 62-year old woman with a 50 pack-year smoking history presents for routine pulmonary follow-up. At the end of the visit, she mentions she saw a flyer in the waiting room pertaining to lung cancer screening and would like to know if she should have this screening done.

Which of the following is an essential element of lung cancer screening?
A. Pulmonary function tests
B. Chest x-ray
C. Shared decision-making
D. Pulmonary rehabilitation
E. Alpha-1 antitrypsin testing
Question 3
A 65-year old man with a history of severe COPD presents for an annual follow-up visit. Spirometry last year was notable for an FEV1 of 40% predicted. He has not had any exacerbations over the past year and his symptoms are stable on an inhaler regimen of umeclidinium bromate, budesonide-formoterol, and albuterol as needed. He continues to smoke 1 pack per day and has a 75 pack-year smoking history.

Which of the following is true related to the potential benefits and risks of lung cancer screening in this patient?
A. The patient’s history of COPD increases his risk of lung cancer
B. The patient’s history of COPD decreases his risk of a complication during a diagnostic procedure
C. The benefits of lung cancer screening outweigh the risks for this patient
D. There is no role for shared decision-making for lung cancer screening with this patient.

Question 4
A 70-year old woman with a history of asthma presents for annual pulmonary follow-up. She has a 40 pack-year smoking history, having quit smoking 10 years ago. Her asthma is well controlled on inhaled fluticasone and albuterol as needed. You are considering initiating a shared decision-making discussion about lung cancer screening.

Which of the following has been identified as essential to a high-quality lung cancer screening program?
A. Utilization of CT pulmonary angiogram imaging
B. Integrated smoking cessation counseling
C. An established pulmonary rehabilitation program
D. Lung cancer screening decisions managed exclusively by pulmonary specialists

Question 5
A 61-year old woman with a history of COPD presents for an initial evaluation to establish pulmonary care. Her symptoms are well controlled on a regimen of tiotropium and albuterol as needed. She continues to smoke 1/2 a pack of cigarettes daily, with a 35 pack-year history. You are reviewing preventive care measures and engage her in a shared decision-making discussion about lung cancer screening.

Which of the following is true regarding the risks and benefits of annual low-dose CT screening for lung cancer?
A. Annual low-dose CT screening is associated with a 20% reduction in all-cause mortality.
B. Nearly 10% of low-dose CT scans will have a positive finding.
C. The majority of abnormalities detected during lung cancer screening are benign.
D. Annual low-dose CT screening can reduce lung cancer-related mortality but not all-cause mortality.
E. There are no significant risks to performing annual low-dose CT screening.

Question 6
A 72-year old man with a history of chronic systolic heart failure (ejection fraction 30%), poorly controlled diabetes, and severe COPD (FEV1 40%) presents for follow-up after a recent hospitalization. He has recovered well from a recent COPD exacerbation, his 5th hospitalization in the past year. He has received annual low-dose CT screening for the past 3 years.

Which of the following is true regarding lung cancer screening in this patient?
A. The patient’s life expectancy should be considered in the decision to undergo future lung cancer screening.
B. The patient’s comorbidities are an absolute contraindication to lung cancer screening.
C. The benefits of continued lung cancer screening outweigh potential risks.
D. A shared decision-making discussion is not necessary since the patient has already undergone lung cancer screening in the past.
E. The chance of a positive finding on future low-dose CT scans is less than 10%.
B12: Global Care for Sleep Disorders: Towards Universal Access

Question 1
Which of the following parameters is recorded by an actigraph?
A. Total sleep time
B. Rapid eye movement (REM) sleep
C. Movement
D. Respiratory effort
E. Deep sleep

Question 2
A 50-year old man presents with frequent loud snoring, witnessed apneas, tiredness, and unrefreshing sleep. His Epworth Sleepiness Scale Score is 12. He has a history of well-controlled hypertension (on an ACE inhibitor) but no other past medical history. No prior history of smoking. Examination revealed a BMI of 32 kg/m2 and a neck circumference of 19 inches. He had a Mallampati 3 upper airway. The rest of the examination was non-contributory. A Level 3 home sleep study was performed; technically, the signals were adequate. However, no significant sleep apnea was noted.

Which of the following would be the next best step:
A. Reassure him that he does not have significant sleep apnea and advise him to lose weight and exercise more.
B. Prescribe a trial of auto-titrating PAP therapy
C. Refer him for an oral appliance
D. Obtain an in-laboratory polysomnogram

Question 3
Which of the following is a reasonable estimate of the people living with OSA world-wide?
A. 3 million
B. 20 million
C. 30 million
D. 1 billion
E. 5 billion

Question 4
A 68-year old, obese, male patient with a body mass index of 35 kg/m2 and waist circumference 43 inches (110cm) presents to their primary care physician with a history of snoring which has been disturbing to his bed partner.

The primary care physician and a nurse within their practice have received specific training in OSA diagnosis and management, including use and interpretation of a validated home oximeter device and auto-titrating continuous positive airway pressure. They are supported by sleep physicians and staff at an academic sleep center within the same district.

The primary care physician suspects that the patient may be suffering from obstructive sleep apnea (OSA).

Which of the following patient characteristics would favor consideration of the use of an ambulatory diagnostic and management strategy in primary care?
A. On supplemental oxygen for chronic obstructive pulmonary disease
B. A history of hypertension which is well controlled on one antihypertensive medication
C. Frequent and prolonged nocturnal awakenings
D. Excessive daytime sleepiness (Epworth Sleepiness Scale score 14/24)
E. Recent hospital admission for decompensated cardiac failure
CC2: Critical Care Core Curriculum II: Neurology Monitoring, Non-Traumatic Intracranial Hemorrhage, Endocrinologic Emergencies

Question 1
A 58-year-old woman was admitted to the intensive care unit with septic shock from a urinary tract source. She was intubated, mechanically ventilated and treated with intravenous fluids and antibiotics. Her condition stabilized after two days, but twenty-four hours after discontinuation of sedation she remains unresponsive. Her cranial nerve examination is unremarkable and she withdraws all extremities to pain. There is subtle facial twitching. Her WBC count had decreased from 18 to 10 x 103 cells/µL and her serum glucose was maintained below 180 mg/dL on an insulin infusion. A head CT is negative for an acute process.

Which of the following is the most appropriate next step?
A. Lumbar puncture
B. Continuous video electroencephalography
C. Brain MRI
D. CT of the chest/abdomen/pelvis
E. Observation

Question 2
A 52-year-old was brought into the Emergency Department with right sided hemiparesis and global aphasia. A non-contrast head CT showed ischemic changes in greater than 2/3 of the left hemisphere. tPA was not administered because a clear time of onset could not be established. CT angiography demonstrated a proximal left middle cerebral artery occlusion. Three hours after ICU admission, neurologic examination is unchanged except for a newly dilated left pupil with diminished reactivity to light. Lab values, including coagulation tests, are normal.

Which of the following is the most appropriate next step?
A. Repeat CT scan of the brain without contrast
B. Bilateral transcranial Doppler measurements
C. ICP and brain tissue oxygenation monitoring on the left side
D. ICP and brain tissue oxygenation monitoring on the right side
E. Insertion of an extraventricular drain

Question 3
A 26-year-old woman is intubated and admitted to the intensive care following a fall with closed head trauma. Non-contrast head CT demonstrates diffuse effacement of sulci and slit-like ventricles bilaterally. The neurosurgical team placed a combined parenchymal ICP and brain tissue oxygenation monitor (PbtO2). Three hours after admission, the PbtO2 decreases and the ICP monitor displays the following waveform:

What is the most appropriate next step in management?
A. Administer Hypertonic saline
B. Hyperventilate to PaCO2 of 20mmHg
C. Lower the head of the bed
D. Increase the FiO2 1.0
E. Drain 15ml of CSF from the ICP monitor
Question 4

A 55-year-old man with unknown past medical history presents with acute onset somnolence, left-sided weakness and sensory loss. A non-contrast head CT is performed and demonstrates an intracerebral hemorrhage in the right thalamus that compresses the internal capsule.

Which of the following is the most likely etiology for his hemorrhage?

A. Amyloid angiopathy
B. Hypertension
C. Hemorrhagic conversion of infarction
D. Arteriovenous malformation rupture
E. Aneurysm rupture

Question 5

A 48-year-old woman presents with a severe headache and loss of consciousness and is found to have subarachnoid hemorrhage on noncontrast head CT. CT angiography reveals an aneurysm of the left posterior communicating artery, for which she undergoes endovascular coiling. An external ventricular drain is placed for hydrocephalus and set at 10 cm above the external auditory meatus. On post-bleed day 5 she develops right face and arm weakness along with mild aphasia. She is afebrile, heart rate is 74 bpm, and blood pressure is 146/90 mmHg. Intracranial pressure is 14 mmHg. Repeat head CT is unchanged from prior.

Which of the following is the next best step in her management?

A. Start a norepinephrine infusion
B. Administer mannitol 1 g/kg IV
C. Administer nimodipine 60 mg PO
D. Lower the external ventricular drain
E. Administer dexamethasone 10 mg IV

Question 6

A 24-year-old woman with no past medical history presents with headache and left-sided vision loss. On arrival she is afebrile, respiratory rate is 16 breaths per minute, heart rate is 96 bpm, and blood pressure is 136/86 mmHg. Exam confirms a left homonymous hemianopsia with no motor symptoms. Noncontrast head CT reveals a right occipital hemorrhage. CT angiography shows no underlying vascular lesion. She reports that she took aspirin 325 mg for pain control on the advice of her mother. What is the most appropriate next step?

What is the most appropriate next step?

A. Nicardipine infusion
B. Platelet transfusion
C. Conventional cerebral angiogram
D. MRI of the brain
E. Lumbar puncture

Question 7

A 45-year-old man is admitted to the ICU with agitation and vomiting. His girlfriend states he used to take “thyroid pills” but has not seen a doctor or taken medications in years. On exam, he has temperature 100.6°F, heart rate 156 beats per minute and blood pressure 105 / 75 mmHg. He has forward protrusion of both globes with bilateral upper lid retraction and conjunctival irritation, an irregularly irregular heart rhythm and trace pedal edema. An electrocardiogram confirms rapid atrial fibrillation. On laboratory studies, a thyroid stimulating hormone level is undetectably low.

What of the following should be administered next?

A. Methimazole
B. Aspirin
C. Saturated solution of potassium iodide
D. Liothyronine
Question 8

A 29-year-old woman on chronic prednisone and multiple inhalers for severe asthma is admitted to the ICU with altered mental status, nausea and vomiting. Her family reports that she stopped all her medications three days ago after starting a juice cleanse to treat a cold. Her temperature is 102°F, pulse rate 165 beats per minute, respiratory rate 28 breaths per minute, mean arterial pressure (MAP) 50 mmHg despite three liters of normal saline, a norepinephrine infusion of 0.15 mcg/kg/min and antibiotics for presumed pneumonia.

What is the next best step in management?
A. Increase norepinephrine infusion
B. Start vasopressin infusion
C. Bolus with 2 L Lactated Ringers
D. Start dexamethasone
E. Start fludrocortisone

Question 9

A 66-year-old man who has been hospitalized for 3 weeks for allogeneic stem cell transplant is transferred to the ICU following emergent laparotomy for perforated neutropenic enterocolitis. He has received adequate balanced crystalloid resuscitation, is on broad-spectrum antibiotics, and his surgeon reports good source control in the OR. Despite this, he requires moderate dose norepinephrine infusion to maintain a mean arterial pressure of 60 mmHg. A random total plasma cortisol level is 12 mcg/dL. His albumin is 1.9 mg/dL.

What conclusion can be drawn from these data about whether this patient has a “sufficient” adrenal response to his critical illness?
A. This patient is adrenally insufficient based upon a cortisol level of 12 mcg/dL.
B. Given his hypoalbuminemia and duration of illness, his total cortisol level may not accurately reflect his adrenal response.
C. A cosyntropin stimulation test is needed to definitively determine whether he is adrenally insufficient.
D. Given the cortisol level of 12 mcg/dL, an aldosterone level should also be measured to determine whether this patient is adrenally insufficient

CC3: Sleep Medicine Clinical Core Curriculum II: RLS/PMLD, REM Behavior Disorders, Neurologic Disorders, Neuromuscular Disorders

Question 1

A 46-year-old woman presents with difficulty going to sleep because “I just can’t stay in bed at night.” Though she is sleepy when she gets into bed, she feels that she has a need to move or rub her legs repeatedly, often getting up to walk around. She is often unable to fall asleep until 2 or 3 in the morning.

Which of the following patient characteristics most likely contributes to this condition?
A. She is a nonsmoker.
B. She is G3P3.
C. She is of Asian ancestry.
D. She has pre-hypertension.

Question 2

A 35-year-old female has progressive sleep onset insomnia. She describes discomfort in her legs which begins close to bedtime and worsens during the night with resolution in the morning. The symptoms are only temporarily relieved by walking or rubbing her legs and disrupt her sleep. She had similar symptoms during her pregnancies which resolved after delivery. She has a family history of similar symptoms in her mother and sisters as well. Her physical examination is unremarkable with a BMI of 24 and absence of neurologic deficits. Laboratory values are as follows: creatinine 0.9 mg/L, HbA1c 4% and hemoglobin of 11 g/dL.
Which of the following is the next best step in the management of this patient’s symptoms?

A. Referral to neurology for further evaluation.
B. Prescribe dopamine agonist therapy with ropinirole or pramipexole.
C. Prescribe gabapentin enacarbil.
D. Evaluation for and treatment of iron deficiency.

Question 3

A 50-year-old female with a past medical history of depression treated recently with amitriptyline presents for evaluation of insomnia. She goes to bed around 9 PM but is unable to fall asleep as she feels like she has to keep moving her legs. She sometimes gets up and walks around and at other times takes a warm bath to improve her symptoms. She reports that she was diagnosed with restless leg syndrome 5 years ago and prescribed ropinirole. She initially had symptoms only in her legs and close to bedtime but achieved good control with ropinirole 1.5 mg 2 hours before bedtime. A few months ago she noticed that her symptoms began occurring earlier in the evening, so she started taking an extra dose of ropinirole to alleviate her symptoms. Lately she has noticed that her symptoms now begin around 10:00 AM and involve both upper and lower extremities. She has been taking ropinirole 4 mg four times a day for her symptoms without significant relief and looks to you for advice.

As her physician, which of the following statements best conveys your recommendation at this time?

A. Her symptoms are caused by amitriptyline and discontinuation of this medication is expected to resolve them.
B. This is the natural progression of RLS/WED and unfortunately not much can be done medically.
C. Her symptoms are caused by augmentation and she should reduce the dose of ropinirole.
D. Addition of methadone is the next best step.

Question 4

A 65-year-old non-smoking man with a prior history of hypertension, diabetes, hyperlipidemia and obesity presented to neurology clinic after a recent hospitalization for acute ischemic stroke. He presented with symptoms of right hemiparesis and dysarthria for which a CT of the head was negative for acute intracranial pathology including bleeding. Initial ECG revealed normal sinus rhythm. The patient was deemed eligible for thrombolysis with alteplase with complete resolution of his presenting symptoms after 24 hours. The patient was monitored with telemetry for a total of 48 hours during hospitalization with no evidence of arrhythmias, however periodic nocturnal oxyhemoglobin desaturations were noted. Further workup included a CT angiogram of the head and neck, MRI brain and transesophageal echocardiogram which were all normal. He was discharged from the hospital with prescriptions for aspirin, high-dose statin and further modifications to his diabetes regimen.

Given his history of snoring, excessive daytime sleepiness, and periodic oxyhemoglobin desaturations while in the hospital, he was evaluated by a local sleep physician whom performed an overnight polysomnogram which revealed an apnea-hypopnea index of 32/hour and a respiratory disturbance index of 45 events/per hour with a mean oxyhemoglobin saturation of 89% with a nadir of 70% in supine REM sleep. He was successfully titrated to a CPAP pressure of 9cm H2O and has initiated therapy at home. He presents to clinic today for closer evaluation of modifiable risk factors for prevention of future strokes.

Which of the following management strategies would be best address these risk factors?

A. Perform a home sleep test to ensure adequate treatment of OSA.
B. Order an overnight ambulatory oximetry while on CPAP.
C. Order 30-day non-invasive ambulatory electrocardiographic monitoring.
D. Repeat MRI of the brain.

Question 5

A 32-year-old woman with prior history of epilepsy, diabetes and obesity (BMI 37kg/m2) presents for evaluation of difficulties with sleeping. She has noted difficulty with maintaining sleep over the past 2 years, she wakes up two to three times per night for unexplained reasons although she occasionally experiences snort arousals. Following these awakenings, she has difficulty re-initiating sleep, and it usually takes her
approximately 1 hour to fall back asleep. She usually goes to bed at around 11pm and wakes up at 6am to get ready for work. She perceives that she is sleeping a total of 5 hours a night. Her mother has told her that she suffered from febrile seizures as an infant. She also had recurrent generalized tonic-clonic seizures as a child requiring multiple antiepileptic agents, however no organic cause was ever identified. Her seizures were mostly controlled in her late teenage years and during her 20s, but following the birth of her first child three years ago, she has been having up to 5 nocturnal tonic-clonic seizures weekly. Her husband has checked her blood glucose levels following these episodes and noted them to be above 120. Her antiepileptic regimen has since been modified to Levetiracetam, Valproic acid and Lamotrigine resulting in good seizure control. She noted a significant weight gain of 30 lbs over the last 2 years. In that time her husband has noted increased snoring and witnessed apneic events. She underwent a split-night polysomnogram and was noted to have an Apnea-Hypopnea index of 18/hour that was adequately controlled with CPAP 7cm H2O, no epileptiform activity was noted on full electroencephalogram. She has since been started on CPAP therapy at home and has shown good compliance, however she still reports symptoms of insomnia.

Which of the following strategies would reduce the patient’s risk of sudden unexplained death in epilepsy?
A. Refer her for cognitive behavioral therapy for insomnia.
B. Change her lamotrigine to phenytoin in a monitored setting.
C. Change valproic acid to phenytoin in a monitored setting.
D. Add phenytoin to her existing antiepileptic regimen.

Question 6

A 56-year-old man with prior history of hypertension, diabetes and hypothyroidism presents to the sleep clinic. The wife reports that for the last 2 months the patient has had noisy breathing during sleep which is high-pitched and worse with inspiration. He has also recently been evaluated by local neurologist for symptoms of abnormal gait, incoordination, dizziness and recent history of falls. The patient has noted symptoms of urinary frequency and urgency, with a sense of incomplete bladder emptying following micturition. MRI of the brain revealed “hot cross bun” sign in the pons and hyperintensities in the middle cerebellar peduncles. Physical examination was positive for orthostasis, bradykinesia, cogwheel rigidity of the upper extremities, hyperreflexia, gait ataxia and cerebellar dysarthria. A thermoregulatory sweat test was positive for diffuse anhidrosis. The diagnosis of multiple system atrophy was made.

The decision is made to proceed with attended split night polysomnogram. The study revealed evidence of inspiratory stridor, an apnea-hypopnea index of 6 events/hour, a mean end-tidal CO2 of 48mmHg, and evidence of REM sleep without atonia. CPAP was started after 75 minutes of sleep due to severe stridor. CPAP was titrated up to a pressure of 20 cm H2O, however persistent inspiratory stridor remained. A morning arterial blood gas revealed a pH of 7.36 PaO2 of 88mmHg, PaCO2 of 48mmHg and a calculated serum bicarbonate level of 29.

Based on the findings of the polysomnogram, what is the next best step to address the patient’s stridor?
A. Referral to Otolaryngology for tracheostomy evaluation.
B. Repeat titration polysomnogram with Bi-level positive airway pressure with the goal of improving CO2 levels.
C. Repeat titration polysomnogram with Adaptive servoventilation.
D. Referral back to his local neurologist to initiate carbidopa-levodopa for stridor.

Question 7

A 17-year-old Caucasian man with Duchenne muscular dystrophy is referred to pulmonary clinic by his neurologist for assessment of his respiratory muscle function. He notes some mild orthopnea and occasional morning headaches upon awakening. Pulmonary function tests demonstrate a forced vital capacity (FVC) of 55% predicted which is similar to the value obtained 6 and 12 months ago. His maximal inspiratory pressure (MIP) is -62 cm H2O. On ABG, his pCO2 is 42 mmHg. A polysomnogram is performed. The AHI is < 5, but there are oxygen saturations < 88% for > 10 minutes during the night.

Which of the following would indicate the need for non-invasive ventilation (NIV) initiation in this patient?
A. FVC of 55% predicted, stable over 12 months
B. Nocturnal oxygen saturation < 88% for 10 minutes
C. PaCO2 of 42 mmHg
D. MIP of -70 cmH20
E. AHI < 5

**Question 8**

A 62-year-old woman with amyotrophic lateral sclerosis (ALS) and diabetes presents to pulmonary clinic. In the past 4 months, she has developed increasing daytime somnolence, morning headaches, fatigue and irritability. Her husband reports witnessing occasional apneic episodes. The patient denies copious oral secretions. Vitals are normal and physical exam reveals a thin woman in no acute distress with a Mallampati I airway, minimal thin secretions in the oropharynx and decreased strength in the upper and lower extremities with hyperreflexia. Recent pulmonary function tests reveal a forced vital capacity (FVC) of 50% predicted and maximal inspiratory pressure (MIP) of -55 cmH20. A daytime room air arterial blood gas reveals a pH of 7.34, PaCO2 of 46 and PaO2 of 87. A polysomnogram (PSG) is ordered.

During which phase of the sleep cycle is the patient most likely to have sleep disordered breathing?
A. Stage I
B. Stage II
C. Stage III (slow wave sleep)
D. Rapid eye movement sleep (REM)

**Question 9**

A 60-year-old man with ALS is intubated in the ICU with aspiration pneumonia. His neuromuscular doctor documented that both the patient and his family would like to pursue life sustaining interventions. He was previously seen by an outside pulmonologist who initiated him on non-invasive volume assured pressure support (VAPS). His last forced vital capacity (FVC) was 0.95 liters, and he has been compliant with VAPS. Currently, the patient’s wife reports that his sleep quality has been good, and he has not complained of morning headaches or daytime sleepiness. However, in the past 6 months, the patient has developed increased uncontrollable secretions requiring oral suctioning every hour despite the use of drying agents, and the wife notes increasing dysarthria. This is also his 2nd admission for aspiration pneumonia in the last year, and the first time being intubated. The patient and his family wish to have a discussion regarding tracheostomy and home invasive mechanical ventilation.

Which of the following statements is true regarding the use of tracheostomy and invasive ventilation in patients with ALS?
A. Most ALS patients who proceed to tracheostomy have it performed as an elective procedure.
B. Quality of life is consistently improved in patients with tracheostomy placement.
C. Survival is prolonged in patients with FVC < 1 liter if a tracheostomy is placed.
D. Invasive mechanical ventilation has similar costs when compared to non-invasive ventilation.
E. Most patients using invasive mechanical ventilation will become minimally communicative.
Tuesday, May 22

PCC3: Pediatric Clinical Core Curriculum: Pulmonary Physiology

**Question 1**
You are evaluating a 7-month-old female infant in your clinic for recurrent cough, which has not responded to short acting β-agonist treatment. While worsened by upper respiratory infections, her cough also occurs in the absence of a known trigger. Her mother reports that the child occasionally exhibits a cough while drinking from the bottle. Her voice sometimes sounds “wet and gurgly” when she vocalizes. On examination, she has normal vital signs including respiratory rate of 24 breaths/min and oxyhemoglobin saturations are 99%. Her lungs are clear to auscultation with equal breath sounds bilaterally with few transmitted sounds. She has occasional cough during the examination. Chest radiography is normal.

Of the following, the BEST next step for evaluation of this infant’s recurrent cough is

A. Computed tomography of the chest  
B. Contrast esophagram  
C. Laryngoscopy with bronchoscopy  
D. Magnetic resonance imaging of the brain  
E. Multitexture video fluoroscopic swallowing study

**Question 2**
You are asked to evaluate a 16-month-old male for “wheezing since birth”. He has a history of coughing for many months and often chokes on his feeds. A modified barium swallow (videofluoroscopic swallow study) showed aspiration and bronchoscopy showed a type I laryngeal cleft that was surgically repaired. Flexible and rigid bronchoscopy also revealed mild tracheomalacia. He is acyanotic on room air and has a normal voice, but you hear intermittent expiratory wheezes. The mother indicates that her son has had minimal improvement in breathing since the surgical cleft repair. A trial of inhaled corticosteroids and short acting bronchodilators have provided minimal improvement.

Of the following, the MOST likely cause of his ongoing breathing difficulty is

A. Gastroesophageal reflux disease  
B. Vocal cord paralysis  
C. Swallowing dysfunction with aspiration  
D. Tracheo-esophageal fistula  
E. Pulmonary hypertension

**Question 3**
You are seeing a 3-year-old Amish girl in the pulmonary clinic referred by the pediatrician for coughing and intermittent wheezing. She also has difficulty in swallowing with occasional choking on thin liquids. You decide to obtain a multi-texture videofluoroscopic swallow study which came back positive for aspiration of thin liquids. On careful questioning the history is positive for insomnia, occasional vomiting, and slight delay in developmental milestones in the last year. On physical exam, you note mild hypotonia in her extremities and her lung exam is normal except for mild thoracic dextro-scoliosis. There are no dysmorphic facial features.

Which of the following is the next best investigation to arrive at the underlying diagnosis?

A. CT scan of the chest  
B. Referral to Neurology  
C. Flexible bronchoscopy with bronchoalveolar lavage  
D. MRI of the brain  
E. Polysomnogram
Question 4

You are seeing a patient in aerodigestive clinic. He is a 10-month old term boy with recurrent cough, wheeze, and respiratory infections since the age of 4 months. He has required 2 hospitalizations for “bronchiolitis”, and has a daily wet cough which has improved with inhaled corticosteroids and beta agonists but not resolved. He was found to have silent aspiration of thin and nectar thick liquids on 2 modified barium swallows with no improvement in swallow function between the two studies. He has had a normal upper GI fluoroscopy study. The ENT specialist performed a nasal endoscopy which yielded normal results. In clinic he has a wet cough, nasal congestion, no increased work of breathing or tachypnea, and oxygen saturation is 100% in room air with clear lungs on exam. He is developmentally on track for age. His weight had been in the 50th percentile but over the past 2 months has decreased to the 15th, while his height and head circumference have been stable at the 50th percentile.

In collaboration with your colleagues, what is the best next step diagnostically?

A. Admission to the hospital for Nasogastric tube placement, multiple specialty consultations (nutrition, feeding team, pulmonology, gastroenterology) and nutritional rehabilitation.
B. High resolution chest CT scan with inspiratory and expiratory views.
C. Combined triple endoscopy including direct rigid laryngobronchoscopy, flexible bronchoscopy with bronchoalveolar lavage, and upper gastroenterological endoscopy.
D. Pediatric polysomnography.

Question 5

A You have been following a 1 year old patient in pediatric pulmonary clinic for 6 months with persistent cough, gagging, and choking with inability to advance solid foods past puree consistency. She has significant feeding aversion, and her parents have had her on a standard infant formula and a twice daily proton pump inhibitor for 6 months. She has had persistent wheeze that does not improve with beta agonists and inhaled corticosteroids. She has recurrent nasal congestion. Nutritional assessment reveals that while her height and weight have been consistently at the 75th percentile, with 90% of her calories are coming from her infant formula and she is taking upwards of 40 ounces of formula per day. Upper GI fluoroscopy is normal. Modified barium swallow does not show frank aspiration, but her cooperation with the study was limited. Triple endoscopy reveals normal vocal cords, no laryngeal cleft, significant cobblestoning of the trachea, normal lower airways, bronchoalveolar lavage within normal limits, normal ciliary biopsy, and normal appearing upper endoscopy but esophageal biopsies reveal 30 eosinophils per high power field at three esophageal heights.

What is the best next step in treatment for this patient?

A. Add an H2 antagonist to her reflux regimen to try to treat the esophagitis and gagging
B. Surgical nissen placement for GERD.
C. Continued expectant management with upright positioning after meals for treatment of reflux.
D. Add swallowed inhaled corticosteroid such as fluticasone or budesonide.

Question 6

A 16-month old boy with VACTERL presents to you for the first time in pediatric pulmonary clinic. His mother recently moved from out of state and does not have medical records for you to review. His mother reports that post-operatively he had 3 esophageal dilations because of narrowing at the surgical site. He is currently taking whole milk and purees as his primary nutrition source. She brings him to you today because he has developed recurrent wheeze for the past 3 months which has not responded to beta agonists and now recurrent emesis. His appetite has decreased, and he has more choking and gagging with eating and drinking. He has no increased work of breathing and oxygen saturation is 99% in room air. Lung exam is significant for intermittent end expiratory wheeze in the lower lung fields bilaterally without any increased work of breathing and a barky cough.

What is the best next step for evaluation of these symptoms?
A. Initiation of a proton pump inhibitor and routine referral to gastroenterology.
B. Videoflouroscopic swallow study to assess for oropharyngeal dysphagia/aspiration during swallowing
C. Upper gastrointestinal fluoroscopy series to assess for anatomic abnormalities of the esophagus, stomach or duodenum
D. High resolution chest CT scan with inspiratory and expiratory views.
E. Flexible bronchoscopy with bronchalveolar lavage.

C1: Clinical Year in Review 3: Genomics in Lung Disease, ARDS, Cystic Fibrosis, Sleep Disordered Breathing

**Question 1**

A 48-year old man is intubated for ARDS secondary to influenza pneumonia. His ventilator settings include a tidal volume of 6cc/kg of ideal body weight with a resultant plateau pressure of 25 cm H2O, a PEEP of 8 cm H2O, and a FiO2 of 0.6. His PaO2/FiO2 ratio is 95.

What is the best approach to managing PEEP in order to improve this patient's survival?

A. Set PEEP based on the FiO2 using the ARDS network low PEEP ladder
B. Increase the PEEP until the plateau pressure equals 30 cm H2O
C. Perform a recruitment maneuver and set PEEP based on the optimal static compliance
D. No approach to PEEP titration has been shown to improve survival in patients with moderate to severe ARDS
E. Set PEEP based on the FiO2 using the ARDS network high PEEP ladder

**Question 2**

An immunocompromised patient develops severe ARDS and is being evaluated for veno-venous ECMO.

Which of the following characteristics is associated with a higher mortality risk in the setting of ECMO use in this patient?

A. Short duration of immunocompromised status (< 30 days)
B. Underlying hematologic malignancy
C. Elevated pre-ECMO platelet count
D. Younger age
E. Low pre-ECMO driving pressure

**Question 3**

The advent of high-throughput genotyping chips has led to large genome-wide association studies (GWAS) for human traits such as lung function and diseases such as COPD. Multiple GWAS have looked at COPD in large, general populations and cohorts with significant smoking histories. These GWAS have uncovered a number of genetic risk variants for COPD and lung function impairment.

Which of the following statements most accurately describes the state of precision medicine with regard to these GWAS discoveries?

A. The risk variants are applicable across different ethnic and racial groups
B. Risk variants for COPD were only able to predict a small percentage of the observed COPD risk, likely due to limitations in sample size and complex, environmental risk factors such as tobacco smoke exposure
C. GWAS of moderate to severe COPD have identified genetic variants uniquely associated with COPD in the setting of cigarette smoke exposure and have not found variants previously associated with lung function in large general populations
D. The gene pathways represented by the lung function genetic risk scores have not yet implicated genes targeted by currently available FDA-approved drugs
E. These studies have been performed in large cohorts representative of diverse populations
Question 4

In the past several years, there have been increasingly larger genome-wide association studies (GWAS) of pulmonary diseases such as COPD, asthma, IPF, and lung cancer. These GWAS have resulted from the lower costs of higher-throughput DNA genotyping and the availability of data from larger, well-characterized clinical datasets, general population consortia, and the rapid adoption of the electronic medical record (EMR).

Which of the following is important to consider as we generate and interpret GWAS using these data for the development of precision medicine genetic profiles?

A. With a large enough sample size, there is no need to replicate or validate significant findings that are novel and reach genome-wide significance
B. Database resources for mRNA expression in lung tissue provide no supportive evidence for the biologic importance of associated gene variants found in GWAS
C. In asthma and COPD, it is likely that multiple genetic risk factors from multiple biologic pathways interact with environmental exposures to influence disease risk
D. GWAS of a physician’s diagnosis of asthma in large, EMR-based general population cohorts such as the U.K. Biobank and eMERGE network have failed to replicate or confirm asthma risk genes previously found in smaller, well-characterized asthma cohorts
E. An EMR-based diagnosis of COPD or asthma (using ICD codes) is a reliable phenotype for use in genetic studies.

Question 5

A 25-year old woman with cystic fibrosis (CF) presents to outpatient clinic for a 3 month follow-up. She is homozygous for F508del, and her baseline FEV1 is 35% predicted. She has multiple CF complications, including pancreatic insufficiency, chronic sinusitis, and chronic Achromobacter resistant to multiple antibiotics. She did not tolerate Lumacaftor/Ivacaftor due to severe bronchospasm, and her course over the past year has been complicated by 3 pulmonary exacerbations requiring intravenous antibiotics and hospitalization. Chronic medications include alpha-dornase, hypertonic saline, azithromycin, inhaled tobramycin, inhaled colistin, pancreatic enzymes, and fluticasone propionate/salmeterol.

What is the next best step in the management of this patient?

A. Start Ivacaftor 150mg po BID
B. Restart Lumacaftor 400mg/Ivacaftor 250mg concomitantly with prednisone for the first 2 weeks
C. Start high-dose ibuprofen
D. Start Tezacaftor 100mg/Ivacaftor 150mg QAM, Ivacaftor 150mg QPM
E. Stop inhaled tobramycin and start inhaled aztreonam

Question 6

According to the 2017 systemic review conducted to inform recommendations by the U.S. Preventive Services Task Force (USPSTF) on screening for obstructive sleep apnea (OSA) in adults, which of the following statements is true:

A. No screening is recommended for symptomatic adults seeking care at sleep clinics
B. In asymptomatic adults, there is uncertainty about the clinical utility of available screening tools and evidence is limited on the effectiveness of CPAP at reducing mortality
C. The STOP-BANG and Epworth Sleepiness Scale (ESS) should be used to screen all patients seen in primary care offices, regardless of chief complaint
D. A home sleep study should be offered to all patients with sleep complaints

C2: Interstitial Lung Disease From Cradle to Grave

Question 1

Chest imaging of a newborn baby suggests a diagnosis of severe interstitial lung disease in children (chILD). On history, his parents report a possible family history of adult ILD.
Given the severity of the patient’s disease and the family history of ILD, assessment of which of the following is indicated?

A. Metalloproteinase-7 (MMP-7)
B. MUC5B promoter polymorphism
C. SFTPB, SFTPC and ABCA3 mutations
D. Telomere length

Question 2

Shortly after birth, a full-term infant presents with unexplained respiratory distress. She is hypoxemic in room air. A chest radiograph shows non-specific diffuse opacities. You suspect she may have interstitial lung disease in children (chILD).

Which of the following is the next best test to evaluate for the presence of chILD?

A. Bronchoscopy
B. Echocardiogram
C. High-resolution CT scan of the chest (HRCT)
D. Genetic testing
E. Surgical lung biopsy

Question 3

A 62-year old active smoker (> 20 pack-years) develops pleuritic chest pain following air travel. A CT pulmonary angiogram is negative for pulmonary embolism but reveals mild interstitial lung abnormalities (ILA). The patient would like more information about how commonly ILA is seen on chest imaging.

Based on large population studies, how would you counsel the patient regarding the frequency of ILA on chest imaging?

A. 1/10
B. 1/100
C. 1/1000
D. 1/10000
E. 1/100000

Question 4

A 55-year old woman with a longstanding history of interstitial lung disease related to rheumatoid arthritis has been experiencing an increase in her pulmonary symptoms. She has been treated with mycophenolate mofetil for the past 2 years. She now requires supplemental oxygen therapy at all times. High-resolution CT scan of the chest shows progression of fibrosis with a definite usual interstitial pneumonia pattern.

Which of the following is correct in terms of the management of this patient?

A. Therapy with an antifibrotic medication
B. She is ineligible for pulmonary rehabilitation
C. It is appropriate to refer for lung transplant evaluation
D. Evaluation with open lung biopsy

Question 5

A 67-year old male with advanced idiopathic pulmonary fibrosis notices a progressive increase in his shortness of breath over the past 2 months. He uses 3 liters of oxygen at rest and 6 liters while exercising. Last summer, he was able to take short walks with his dog but now prefers to stay inside because he experiences significant dyspnea after walking only a couple of meters. His pulmonary function tests show severe functional impairment and progressive lung function decline over the last 2 years.

Which of the following interventions could help alleviate his dyspnea?

A. Antifibrotic medication
B. Anti-reflux therapy
C. Corticosteroids
D. Low-dose opioids
C10: Novel Strategies to Compliment Pulmonary Rehabilitation in COPD

Question 1
A 76-year old female with severe COPD and frailty is undergoing pulmonary rehabilitation. One month into the rehabilitation program, she does not perceive much benefit and is considering dropping out. She enquires about the use of a walking aid to increase her exercise capacity.

How should you advise this patient regarding the role of walking aids in her pulmonary rehabilitation?

A. All patients with COPD benefit from using walking aids
B. She will likely experience greater improvement in walking distance using a draisine rather than a rollator
C. She will likely experience greater improvement in walking distance using a rollator rather than a draisine
D. The optimal choice of walking aid will depend on her preference in combination with an aided exercise test

Question 2
A 76-year old male with severe COPD and a history of ischemic stroke with residual left lower extremity weakness is enrolled in pulmonary rehab. He has heard about neuromuscular electrical stimulation (NMES) training and is interested in using it to aid in his rehabilitation.

How can you best counsel him regarding the role of NMES in rehabilitation?

A. NMES training requires close supervision and must be performed in a monitored clinical setting
B. NMES training provokes a higher metabolic response than resistance exercise training
C. The training effect from NMES is dependent on generation of sufficient tension in the target muscle
D. NMES training provides a comparable workload to that provided by high-intensity resistance training
E. NMES training can be completed continuously with no rest offered between each period of muscle contraction

Question 3
A 60-year old female with severe COPD is referred to a pulmonary rehabilitation center. Although her insurance covers her pulmonary rehabilitation sessions, the distance from home to the closest center is approximately 60 miles. She says she will be unable to attend rehabilitation sessions.

Which of the following is true regarding her options at this point?

A. Telehealth pulmonary rehabilitation via videoconferencing will likely improve her functional capacity and quality of life
B. Improvements in respiratory outcomes using telehealth pulmonary rehabilitation will not been as significant as those seen with traditional rehabilitation
C. Telehealth pulmonary rehabilitation has not been shown to decrease hospitalizations or emergency department visits for COPD exacerbations
D. Telehealth rehabilitation will be covered by her insurance and is cost-effective

Question 4
An important goal of pulmonary rehabilitation is to reverse the downward spiral of deconditioning induced by a sedentary lifestyle. Increases in objectively measured physical activity in everyday life have been observed in many, but not all, studies of rehabilitation outcomes. However, this observed increase in physical activity may not be durable. The use of pedometers has been suggested as a way to sustain physical activity after pulmonary rehabilitation.

Which of the following is true regarding the use of pedometers in this patient population?

A. Pedometers are too imprecise to provide relevant feedback
B. In recent studies, pedometer feedback has yielded fairly consistent increases in physical activity
C. More complex activity measuring devices will need to be employed before activity monitoring feedback can be practically delivered
D. Studies of pedometer feedback have not been favorable, but larger study groups are needed for definitive evaluation
Question 5

The importance of self-management is outlined in every national and international guideline for the management of COPD. The aim of self-management is to equip individuals with the knowledge and skills to effectively manage their disease and improve their quality of life.

Which of the following statements regarding self-management interventions is true?
A. Self-management training has not been shown to reduce hospitalizations for COPD exacerbations
B. Self-management and pulmonary rehabilitation are distinct interventions
C. There is convincing evidence that telemonitoring and telehealth are important tools for successful self-monitoring
D. Self-management strategies should be integrated into all aspects of delivered care

Question 6

You refer a 69-year old patient with severe COPD and moderate dyspnea to your hospital’s pulmonary rehabilitation center. The pulmonary rehabilitation therapist evaluates the patient and determines that he would be safe to exercise, but is concerned that the patient does not have reliable transportation to attend twice weekly sessions. After completing the initial evaluation, the patient calls you to say that he would prefer to attend once weekly Tai Chi classes at his local YMCA.

How can you best counsel this patient regarding exercise and COPD?
A. Insist that the patient attend pulmonary rehabilitation as it is standard of care for COPD
B. Agree that Tai Chi would be better for this patient
C. Encourage the patient to try pulmonary rehabilitation; if it is too burdensome he can join the Tai Chi classes
D. Advise that Tai Chi has not been shown to improve any COPD outcomes and that he is unlikely to benefit from it

C11: New Paradigms for Resolving Sleep Apnea Heterogeneity: Deep Phenotyping and Genomics

Question 1

A 55-year old female returns to see her sleep physician 1 year after undergoing upper airway surgery in order to treat her obstructive sleep apnea (OSA). She is still experiencing daytime sleepiness and her partner reports that while her snoring has improved since the surgery, it is still present and disrupting their sleep. A repeat sleep study shows the surgery has reduced the apnea-hypopnea index by 10 events/hr (from 35 to 25 events/hr).

Which of the following phenotypical traits is the most probable explanation for this patient’s poor response to upper airway surgery?
A. A high loop gain
B. A low arousal threshold
C. Poor muscle compensation
D. Poor upper airway anatomy
E. Low lung volumes

Question 2

A 45-year old man has been recently diagnosed with obstructive sleep apnea (OSA). His polysomnography report indicates that it is of moderate severity (apnea-hypopnea index = 25 events/hr) with 80% of the respiratory events being obstructive hypopneas and 20% of events central apneas. These events were typically associated with mild desaturations and the nadir saturation experienced during sleep was 90%.

The patient has since undergone a CPAP titration study where it was found that despite being optimally treated with 7cmH2O, there was an appearance of cyclic central events at times. The patient has come back to the sleep clinic for a review one-month following CPAP initiation, reporting difficulty tolerating the pressure from the machine.
Based on physiological principles, which of the following drugs/therapies would be expected to have the greatest immediate impact to reduce the patient’s OSA severity?

A. Desipramine  
B. Eszopiclone  
C. Zolpidem  
D. Supplemental oxygen  
E. Acetazolamide

Question 3

Several studies have examined the impact of CPAP for patients with sleep apnea and hypertension. Which of the following sets of parameters most consistently predicts a favorable blood pressure response to CPAP, for sleep apnea patients?

A. Body mass index and age  
B. Plasma microRNA and uric acid level  
C. Apnea-hypopnea index and oxygen desaturation index  
D. Daytime sleepiness and baseline hypertension  
E. CPAP pressure and humidification settings

Question 4

A 48-year old man with history of gastroesophageal reflux disease and obesity (body mass index of 32 kg/m2) has undergone a sleep study (polysomnogram) for evaluation of sleep disordered breathing after presenting with snoring and unrefreshing sleep. His polysomnogram revealed a total sleep time of 5.5 hours, an apnea hypopnea index (AHI) of 21 events per hour, an arousal index of 27 events per hour, sleep efficiency of 75%, with 18% of sleep time spent in rapid eye movement stage and 10% with an arterial oxygen saturation below 90%.

Which of the following polysomnographic features is associated with increased risk of incident cardiovascular disease or death?

A. Arousal index  
B. Sleep efficiency  
C. Time in bed  
D. Percent rapid eye movement sleep  
E. Percent sleep time with oxygen saturation < 90%

Question 5

A 67-year old man with severe COPD, upper lobe emphysema on chest CT, who continues to smoke, believes his lung disease is primarily due to having been born with “bad genes” and asks about genetic testing. He was tested for alpha one anti-trypsin (AAT) serum levels and underwent AAT genotyping, both of which were normal.

What is the most appropriate advice?

A. Tell him that since his AAT studies were normal, there is no genetic basis for this COPD.  
B. Encourage smoking cessation and inform the patient that measuring serial AAT levels and repeat genotyping will be needed to definitively rule out this genetic disorder.  
C. Tell him that COPD is associated with alterations in many genes, but the most important recommendation is to quit smoking.  
D. Encourage smoking cessation and order DNA sequencing of the COPD gene score risk panel.  
E. Perform gene expression analysis on bronchial airway brushings to assess mortality risk due to his obstructive lung disease.
C12: Choosing Rightly: At the Intersection of Personalized Medicine and De-Adoption of Low-Value Care

Question 1

A 78-year old female presents to the emergency department with shortness of breath, cough, and fevers that have been present for one week. Her chest x-ray shows a right lower lobe opacity. Physical examination reveals the following: temperature 39.1°C, heart rate 110 bpm, blood pressure 101/57 mmHg, respiratory rate 26 breaths/min, oxygen saturation 92% on 6 liters per minute of supplemental oxygen. She has rales in the right lower lobe.

Which of the following is the best method to determine if the patient should be admitted to the ICU?

A. Calculate her CURB-65 score
B. Calculate her APACHE II score
C. Admit to the ICU based on her age
D. Assess her potential to benefit from ICU care
E. Admit to ICU solely if she requires mechanical ventilation

Question 2

A 56-year old woman with a long history of systemic sclerosis presents with worsening shortness of breath and lower extremity edema. She undergoes a non-contrast chest CT that shows clear lungs but enlarged pulmonary arteries. PFTs are normal except for decreased diffusion capacity. An echocardiogram shows normal LV systolic and diastolic function and suggests an elevated mean pulmonary artery pressure (estimated at 60 mm Hg). VQ scan and polysomnography are within normal limits.

Which of the following is the most appropriate next step in the care of this patient?

A. Start treatment with epoprostenol for presumed pulmonary arterial hypertension
B. Obtain a CT pulmonary angiogram to rule out pulmonary embolism
C. Start treatment with sildenafil for presumed pulmonary arterial hypertension
D. Obtain a right heart catheterization to measure pulmonary artery pressures
E. Refer to surgery for consideration of pulmonary thromboendarterectomy

Question 3

A 44-year old man with COPD presents for evaluation of worsening dyspnea on exertion over the past 8 months. He reports that his dyspnea has progressed to the point that he stops for breath after walking 100 yards or after walking a few minutes on level ground (Modified Medical Research Council Score of 3). He has had one outpatient exacerbation in the past year. He quit smoking six months ago, and has been using tiotropium (a long-acting muscarinic antagonist (LAMA)) daily. His only other COPD medication is an as-needed short-acting beta-agonist (SABA), which he requires several times per week. He has no history of asthma. Chest x-ray shows hyper-expansion with no acute changes compared to prior films. On spirometry, his FEV1/FVC is 0.53 and post-bronchodilator FEV1 is 47% predicted.

Which of the following changes to the patient's medication regimen is most appropriate?

A. Continue the patient’s LAMA; add an inhaled corticosteroid (ICS)
B. Stop the patient’s LAMA; start a combination LAMA + long-acting beta-agonist (LABA) inhaler
C. Continue the patient’s LAMA; add a combination LABA + ICS inhaler
D. Stop the patient’s LAMA; start a combination LABA + ICS inhaler
E. Continue the patient’s LAMA; add roflumilast

Question 4

In an effort to reduce costs and conserve quality, a health system wishes to increase prescribing of generic medications rather than trade-name or branded medications. The health system is considering several approaches to modifying its electronic health record (specifically, its physician ordering interface) to achieve this goal.
Which of the following approaches has the strongest evidence base to suggest that the change will achieve its goal?

A. Requiring that physicians justify decisions to order brand medications by selecting from a drop-down menu of reasons
B. Designing a clinical decision pop up that fires when a physician types in a branded medication, notifying the physician that generic medications in the same class produce equal benefits at lower costs
C. Providing physicians with monthly feedback on their rates of prescribing generic medications compared to “top performers”
D. Developing a system in which branded medication orders default to the corresponding generic medications. The physician may sign the generic order or proceed to a new screen to prescribe a branded medication.
E. The existing evidence base does not yet support any of these approaches

Question 5

A 68-year-old ex-smoker is referred to you for lung cancer screening. He has a 50-pack year smoking history but quit smoking 4 years ago. During the shared decision-making visit, you discover that he has GOLD class 4 COPD, is on 2 liters of supplemental oxygen, and had a myocardial infarction 5 years ago followed by stent placement. Using the PLCOm2012 calculator, you estimate that he has a 5% chance of developing lung cancer over the next 6 years.

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

A. He should be referred for a low-dose lung cancer screening CT as he meets all of the eligibility criteria and none of the exclusion criteria
B. He should not be referred for screening because his risk of developing lung cancer is too low to warrant screening
C. He should not be referred for screening because he does not meet the age and/or smoking criteria
D. He should not be referred for screening because he is not a surgical candidate
E. He should be referred for screening because he may be eligible for stereotactic body radiotherapy (SBRT) for a screen-detected cancer

CC4: Critical Care Clinical Core Curriculum II: Infectious Diseases

Question 1

A 31-year-old pregnant woman at 24 weeks’ gestation is evaluated for a 2-day history of fever, neck stiffness, and lethargy. Her pregnancy has been uncomplicated and she has no other significant medical history. On physical examination, temperature is 38.5°C, blood pressure 90/56 mmHg, and heart rate 120 beats per minute. The patient is lethargic, rouses to voice, and is oriented. Physical exam is otherwise normal. White blood cell count is 15,000/μL; her other laboratory studies are normal. Lumbar puncture is performed and demonstrates an opening pressure of 27 cm H2O, leukocyte count 700/μL, with 65% neutrophils and 35% lymphocytes, glucose 35 mg/dL, and protein 1.1 g/L. The Gram stain shows no organisms.

Which of the following is the most appropriate initial antimicrobial regimen?

A. Vancomycin, ceftriaxone, ampicillin, dexamethasone
B. Ceftriaxone, ampicillin, and acyclovir
C. Vancomycin, ceftriaxone, dexamethasone
D. Vancomycin and meropenem
E. Vancomycin and ceftriaxone

Question 2

A 52-year-old man with Type II diabetes mellitus is evaluated for a five-day history of headache and right-sided weakness. There is no antecedent trauma, fever, weight loss or systemic symptoms. Physical examination reveals a temperature of 37.9°C, heart rate 90 beats/minute, blood pressure 145/90 mm Hg. Neurologic exam reveals 3/5 motor strength in right arm, 4/5 motor strength in the right leg, and brisk right-sided deep tendon reflexes. Laboratory studies reveal a white blood cell count of 12,500/μL with left-shift. A computed tomography scan of the brain with contrast shows a 3-cm ring-enhancing lesion in the left occipito-parietal
region with surrounding edema and 2 mm of midline shift. HIV testing is negative.

Which of the following is the most appropriate next step in management?
A. Initiate intravenous ceftriaxone and metronidazole
B. Lumbar puncture
C. Initiate voriconazole
D. Consult a neurosurgeon for aspiration of lesion
E. Whole body Positron Emission Tomography/Computed Tomography scan

Question 3
A 59-year-old woman admitted to the hospital with presumed encephalitis develops weakness on the third hospital day. Notably, she had presented with fever, chills, myalgias, rash, and confusion one week after returning from a camping trip in Colorado. On admission, brain MRI demonstrated diffuse leptomeningeal enhancement. CSF sampling revealed a leukocyte count is 70/µL, (45% neutrophils, 45% lymphocytes), protein 60 mg/dL, glucose 85 mg/dL (serum glucose is 120). She received empiric bacterial coverage and acyclovir, CSF bacterial culture and HSV PCR remain negative.

Exam now reflects a somnolent woman, rousing briefly to voice. A diffuse papular erythematous rash persists. Neurologic examination newly demonstrates 2/5 strength in both arms, 3/5 strength in both legs, diffusely diminished deep tendon reflexes, and right foot drop.

Which of the following CSF diagnostic studies would be most likely to identify the responsible pathogen?
A. West Nile virus IgM antibody assay
B. Borrelia burgdorferi PCR
C. Cytomegalovirus PCR
D. Varicella zoster virus PCR
E. West Nile virus PCR

Question 4
A 41-year-old man returns from West Africa with a fever and diarrhea and presents to your outpatient clinic for evaluation. A blood test for Ebola by PCR is negative.

Which of the following is the most appropriate next steps?
A. Send Ebola IgM and IgG antibodies
B. Repeat the test 72 hours or more after symptom onset
C. Repeat the test one week or more after symptom onset
D. Consider other diseases as more likely

Question 5
On day 14 of illness, your patient is no longer symptomatic and blood tests are negative for Ebola virus RNA. He is being discharged from the Ebola treatment unit.

How long will he need to practice safe sex or abstinence to minimize the risk of sexual transmission of Ebola virus disease?
A. It is not possible for him to transmit the virus sexually
B. 3 months
C. Beginning at 3 months post discharge, Mr. X should undergo monthly testing of semen, if available, until 2 tests are negative separated by one week.

Question 6
PA 37-year-old man presents to the hospital with 5 days of fever, fatigue, headaches, sore throat and facial edema. He says he recently returned from Nigeria 10 days before the onset of his symptoms. Lab tests demonstrate an increased AST out of proportion to ALT and his urinalysis is notable for 2+ proteinuria. A PCR test is positive for Lassa virus RNA.

Which of the following is the next more appropriate step(s)
A. Provide supportive care only as there is no antiviral medications for Lassa fever
B. Provide supportive care and start ribavirin as soon as possible since ribavirin is effective against Lassa virus especially if given within 6 days of onset of symptoms
C. Start oseltamivir since it has efficacy against influenza – another RNA virus.

Question 7

A 77-year-old man is brought to the emergency room with fever, worsening confusion, and a productive cough for 4 days. His vitals are: temp: 38.7°C, heart rate 130 beats per minute, blood pressure 110/70 mm Hg, respiratory rate of 25 breaths per minute, and oxygen saturation of 91% on 2L/min oxygen via nasal cannula. His white blood cell count is 28 x 103/μL. After a chest radiograph demonstrates a right lower lobe opacity with associated effusion, he undergoes an ultrasound-guided thoracentesis with withdrawal of frank pus. The pleural fluid results are below:

- pH 6.9
- LDH 1200 IU/L (serum value 220 IU/L)
- protein 8.6 g/dL (serum value 6.2 g/dL)
- glucose 35 mg/dL
- Gram stain positive for gram negative bacilli

Which of the following is the next best step in management?

A. Treat with ceftriaxone and azithromycin for community acquired pneumonia
B. Treat with vancomycin, cefepime, and metronidazole
C. Treat with vancomycin, cefepime, and flagyl and place a small-bore chest tube for drainage
D. Treat with vancomycin and cefepime and place a large-bore surgical chest tube for drainage
E. Consult thoracic surgery for consideration of VATS decortication

Question 8

A 58-year-old woman with a history of alcohol abuse is admitted to the hospital for one week of fevers and productive cough. After chest radiography reveals a right-sided loculated pleural effusion she undergoes thoracentesis. Gram stain of the fluid demonstrates Gram positive cocci. Antibiotics are initiated and after a chest tube is placed for drainage you decide to instill tissue plasminogen activator (tPA) and DNase for three days.

Which of the following statements is true about the use of intrapleural tPA-DNase?

A. Combined use of tPA and DNAse is shown to have inferior outcomes to either tPA or DNase alone.
B. tPA-DNase acts as a fibrinolytic agent to develop pleural septations and increase the viscosity of pleural fluid
C. The use of tPA-DNase decreases the need for surgical decortication
D. The use of tPA-DNase increases hospital length of stay due to the need for three days of instillation
E. tPA-DNase does not commonly cause intrapleural hemorrhage or hemoptysis

Question 9

A 61-year-old man presents to the emergency department for evaluation of four days of chest pain and fevers. He underwent an uncomplicated coronary artery bypass graft two weeks prior and was discharged home for completion of cardiac rehabilitation. At home, he developed cloudy drainage from his sternotomy incision site which is showing early signs of dehiscence. A chest radiograph demonstrates a widened mediastinum.

What is the next best step in the management of this patient?

A. Discharge home with a 7 day course of clindamycin for treatment of cellulitis
B. Consult interventional radiology for percutaneous drainage of a mediastinal fluid collection
C. Perform vacuum-assisted wound closure over the sternotomy site
D. Place a mediastinal chest tube
E. Initiate vancomycin, zosyn, and fluconazole and consult cardiothoracic surgery for debridement
Question 10
A 47-year-old man with tetraplegia secondary to traumatic spinal cord injury is admitted from his nursing home with hypoxemia. He has been a nursing home resident for 18 months, has a tracheostomy but is not ventilator dependent. He also has a gastrostomy tube for intermittent bolus tube feeds. He has never had a pneumonia but has had two urinary tract infections requiring hospitalization. The most recent hospitalization was for three days about 4 months prior. On presentation, his hemodynamics are stable but he requires 4L of oxygen supplementation via trach mask. His chest radiograph shows a dense right upper lobe opacity.

Which of the following is the most empiric treatment?
A. Azithromycin
B. Vancomycin, levofloxacin, and cefepime
C. Ceftriaxone and azithromycin
D. Vancomycin and azithromycin

Question 11
A 52-year-old woman with no significant medical history is admitted with one week of progressive shortness of breath, cough, and fever. On presentation, temperature is 102.3°F, HR 125 bpm, BP 90/40 mm Hg, RR 36 breaths per minute, O2 saturation 75% on ambient air. She is in respiratory distress with diffuse rhonchi. Chest radiograph reveals diffuse interstitial opacities. The procalcitonin level is low. Viral studies are pending. A trial of high flow nasal cannula is unsuccessful and the patient is intubated and initiated on low tidal volume ventilation, and started on ceftriaxone and levofloxacin.

Which of the following statements is true regarding the use of adjunctive glucocorticoid therapy in this patient?
A. Glucocorticoid therapy should be initiated immediately, prior to evaluation for viral pneumonia, to maximize potential benefit.
B. Evidence of benefit for adjunctive glucocorticoid use is stronger for this patient with severe CAP as compared with patients with non-severe CAP.
C. As this patient has no history of diabetes, she is unlikely to need adjunctive insulin treatment for hyperglycemia with glucocorticoid therapy.
D. The low procalcitonin level means that adjunctive glucocorticoids are not indicated.

Question 12
A 72-year-old man was admitted following an ST-elevation MI. He has had no hospitalizations within the past year. On day 6 of his hospitalization, he develops fever of 101.4°F and non-productive cough. Vital signs otherwise reveal a heart rate of 90 beats per minute, blood pressure 135/78 mmHg, respiratory rate of 18 per minute, and oxygen saturation of 93% on ambient air. Lung exam reveals rhonchi over the right lung base and diffuse expiratory wheezes. A chest radiograph demonstrates hyperinflation and a right lower lobe opacity. White blood cell count is 14 x 103/μL. MRSA screening on admission was negative. The local prevalence of MRSA isolates among S. aureus species is 14% according to the hospital antibiogram.

Which of the following is the most appropriate empiric treatment regimen for this patient’s pneumonia?
A. Aztreonam
B. Cefepime
C. Vancomycin, cefepime
D. Vancomycin, cefepime, and levofloxacin

C83: Pediatric Clinical Chest Rounds

Question 1
An infant has clinical symptoms of central hypoventilation following a negative evaluation for underlying lung disease, neuromuscular weakness, metabolic disorders or anatomic abnormalities of the central nervous system.

Which of the following regarding testing for mutations of the PHOX 2B gene is correct?
A. A positive result will require testing of asymptomatic siblings
B. The type of mutation will predict when the patient can be expected to recover
C. A positive result predicts a one in two risk for each subsequent pregnancy
D. The type of mutation allows targeted screening for other manifestations of CCHS
E. Gene testing is not needed in cases with a high clinical suspicion

**Question 2**

A 6-week old term infant presents with a lower respiratory tract infection and respiratory failure requiring intubation and assisted ventilation. After treatment, the settings on the ventilator are weaned to physiologic for the infant's age and no supplemental oxygen is required to maintain normal oxygen saturation. However, multiple attempts at extubation fail due to poor respiratory effort, apnea, and hypercapnia. A chest radiograph is interpreted as normal while on assisted ventilation.

Which of the following should be performed next?

A. Tracheostomy to facilitate chronic invasive ventilation
B. Lung biopsy
C. Neuromuscular evaluation
D. Phox2B mutation analysis

**Question 3**

A 16-year-old male has been referred to you by his pediatrician for cough. Six months ago, the patient developed a chronic, productive early morning cough that wakes him up at night, nasal congestion and fatigue. He also has had persistent bilateral cervical lymphadenopathy. Despite poor weight gain over the past few years and a BMI of 19.56 kg/m² (29th percentile), there is no history of fever, weight loss, or night sweats. He was adopted from Thailand four years prior.

An initial work up was negative for tuberculosis, HIV, and hepatitis. CBC and complete metabolic panel were unremarkable. A chest CT was performed which revealed diffuse bilateral bronchiectasis with evidence of both hilar and axillary adenopathy. The upper portions of the liver and spleen were noted to be normal.

What would be the next best step in the work up of this patient?

A. Repeat CBC with differential
B. Immunoglobulin panel
C. Sweat test
D. Bronchoscopy with bronchoalveolar lavage
E. Stool elastase

**Question 4**

A 16-year-old male is seen in your office for hospital follow up. A pulmonary consult occurred during his recent hospitalization for intussusception as bronchiectasis was noted on chest CT. He has had one year of bilateral cervical lymphadenopathy, fatigue, poor weight gain, and cough. A sweat chloride was performed and was “quantity insufficient” 6 months ago. A sweat chloride performed while he was in the hospital was 49.5mmol/L right arm, 44mmol/L left arm. He endorses oily, foul-smelling stools and intermittent mild abdominal discomfort. You perform a stool elastase, which reveals a level of 188µg/g stool (normal > 200µg/g stool).

What would be the next most appropriate step in the work up of this patient?

A. Bronchoscopy with bronchoalveolar lavage
B. Amylase/Lipase
C. Repeat sweat test
D. Cystic fibrosis complete mutation analysis
E. CT abdomen with/without contrast

**Question 5**

You are seeing a previously healthy 16-year old girl for chest pain, intermittent fevers, shortness of breath and cough that started approximately one month ago, shortly after playing in soccer tournaments in Southern California and Arizona. A chest CT angiogram was done to assess pulmonary embolism as a cause of her chest pain with a representative image shown below (Figure 1).
On exam she appears tired but acyanotic while breathing RA. Her breath sound intensity was slightly decreased over her left lower lung zone but there were no crackles or wheezes. She had no adenopathy or digital clubbing and her skin was clear.

Her white blood cell count is normal with 7% eosinophils.

Interferon gamma releasing assay (Quantiferon Gold) negative
cANCA and pANCA negative

Coccidiodomycosis: IgM positive; IgG positive; complement fixation 1:128

Figure 1. Representative chest CT scan image.

Which of the following is the most appropriate choice for her initial therapy?

A. Fluconazole
B. Isoniazid
C. Levofloxacin
D. Rituximab

Question 6

You are seeing a previously healthy 15-year old boy with complaints of cough, fever, night sweats and weight loss. On examination the SpO2 while breathing room air was 96% and he had no adventitious pulmonary sounds on auscultation.

Laboratory assessment showed a WBC of 10.9 with a hemoglobin of 11.9, and a platelet count of 353 K. The WBC differential showed 58% neutrophils, 13% lymphocytes, and 27% eosinophils. His CXR is shown:

Which of the following is the next best evaluation to identify the etiology of these abnormalities?

A. Bronchoscopy with bronchoalveolar lavage
B. Complete pulmonary function testing
C. Thoracentesis with pleural fluid analysis
D. Video-fluoroscopic swallow study
Question 7

A 16-month old toddler is admitted with a two-week history of progressive cough and tachypnea with intermittent low-grade fevers. Initial exam was notable for tachycardia and tachypnea, muffled heart sounds, subcostal and supraclavicular retractions and diminished aeration at lung bases bilaterally. His chest radiograph is notable for cardiomegaly and moderate bilateral pleural effusions. He requires pericardiocentesis for a hemodynamically significant pericardial effusion and bilateral chest tube placement.

Pleural fluid
Appearance: Straw colored
pH: 7.61
Protein: 2,710 mg/dL
Lactate dehydrogenase: 125 units/L
Triglyceride level: 30 mg/dL
Serum
Total protein: 6,000 mg/dL
Lactate dehydrogenase: 240 units/L

What is the classification of the pleural fluid?
A. Transudate
B. Exudate
C. Chylothorax
D. Indeterminate

Question 8

A term male infant was transferred to the neonatal intensive care unit after developing tachypnea and cyanosis that progressed to severe respiratory distress within hours following an otherwise uncomplicated vaginal delivery. There were no maternal risk factors during pregnancy and a 20-week fetal ultrasound was notable for polyhydramnios and small bilateral pleural effusions.

The infant required intubation to achieve stable ventilation and oxygenation. On exam, the infant has fine crackles throughout and poor aeration over the right lower lung fields. Initial chest x-ray shows bilateral ground-glass appearance, with moderate sized right-sided and small left-sided pleural effusions.

A right-sided chest tube is placed and the pleural fluid appears milky white. There is a lymphocytic predominance and a triglyceride level of 175 mg/dL.

Which of the following syndromes is associated with the presentation described above?
A. Trisomy 18
B. Williams syndrome
C. Velocardiofacial syndrome
D. Noonan syndrome

C84: Managing Early Acute Hypoxic Respiratory Failure

Question 1

A 75-year old woman with a history of untreated obstructive sleep apnea, diabetes mellitus, and hypertension is admitted to the intensive care unit with acute onset dyspnea and productive cough. She is short of breath and persistently coughs during the exam. Her temperature is 38.5°C, respiratory rate is 29 breaths/min, blood pressure is 104/65 mmHg, heart rate is 110 bpm, and oxygen saturation is 86% on 6LNC. Lung exam is significant for right lower lung crackles with egophony. A chest x-ray shows right lower lobe consolidation with air bronchograms.

ABG: pH=7.42, PaO2=48 mmHg, PCO2=34 mmHg, Sat=86%
What is the next best step in the management?
A. Start heated high-flow nasal cannula oxygen (HHFNC)
B. Start non-invasive positive pressure ventilation
C. Start non-rebreather oxygen mask
D. Continue nasal cannula oxygen
E. Intubation and mechanical ventilation

Question 2
A 67-year old woman with a history of acute myeloid leukemia is undergoing chemotherapy for a planned stem cell transplant. During her hospitalization she develops neutropenic fever and is noted to be tachypneic with a respiratory rate of 32 breaths/min and hypoxic with saturations of 85% on 6L nasal cannula. She is awake and alert and initiated on noninvasive ventilation and transferred to the medical intensive care unit. Exhaled tidal volumes are 10 ml/kg PBW.

Noninvasive ventilation in this patient is associated with which of the following
A. A decreased rate of endotracheal intubation
B. Decreased mortality
C. An increased risk of mortality
D. A decreased length of ICU stay
E. An increased length of ICU stay.

Question 3
A 58-year old female with history of alcoholic abuse and obesity presented to the ED with 3 days of fever, headaches, and cough with yellow sputum. On exam she was hypoxic saturating 95% on 6 L NC. Her in influenza rapid test was positive and she was started on oseltamivir and admitted to the medical floor.

Twenty-four hours later she was found unresponsive, lying flat in bed with evidence of emesis on her pillow. She was emergently intubated and placed on mechanical ventilation at volume control with a set tidal volume of 350 ml (6ml/kg PBW), respiratory rate of 24, PEEP 5 cm H2O, and FiO2 0.4 with O2 sat at 96%. Plateau pressure was 20.

She was given 2L normal saline for a mean arterial pressure of 58 mm Hg and received 2 units pRBC for anemia. Her sputum culture on presentation to the emergency department was positive for Streptococcus pneumoniae and she was started on ceftriaxone for bacterial pneumonia.

After intubation, she continued to worsen requiring FiO2 1.0 and PEEP 12 cm H20. Repeat chest x-ray revealed bilateral lower lobe air space disease, consistent with ARDS.

Which of the following factors contributed to this patient’s ARDS?
A. Aspiration
B. Rapid administration of antibiotics
C. High tidal volume ventilation
D. Oseltamivir therapy

Question 4
A 29-year old man with no past medical history is admitted to the ICU for acute hypoxemic respiratory failure. His chest X-ray shows a right lower lobe airspace opacity and his sputum reveals gram positive cocci in pairs. His airspace filling progresses to involve both lungs. His P/F ratio is 140. He is intubated and subjected to full mechanical ventilatory support for severe ARDS. He receives propofol and fentanyl infusions and is paralyzed with cisatracurium.

What of the following is true for this patient?
A. He will experience diaphragmatic atrophy 48-72 hours after intubation
B. He will experience diaphragmatic atrophy in less than 24 hours after intubation
C. He will not experience diaphragmatic atrophy while on full ventilator support
D. He will not experience diaphragmatic atrophy because of his age.
C92: Disease Modification in COPD: Are We There Yet?

Question 1

COPD is a heterogeneous disease with varying presentations and rates of disease progression.

Which of the following statements about disease progression in COPD is true?

A. Rates of FEV1 decline are faster among individuals with FEV1% predicted ≤ 50%
B. Emphysema has been associated with increased mortality
C. Symptoms always present before lung function decline
D. Initiating inhaled bronchodilators for COPD early in disease reduces mortality
E. The majority of patients with FEV1% predicted ≤ 50% experience ≥ 2 exacerbations annually

Question 2

In the context of lung diseases, such as asthma, “remission” is an ill-defined concept. True spontaneous remission has been documented to occur rarely but therapeutic induction of remission, or even convincing sustained disease modification, remains an elusive therapeutic goal. However, the advent of biological therapies, in particular recombinant monoclonal antibodies has opened the possibility of inducing lasting disease remissions in some other degenerative conditions. Adoption of early detection and early, more aggressive therapy, often with combinations of agents, has seen progress towards true disease modification and remission in arthritis and bowel disease.

Currently, there are no proven therapeutic strategies to induce convincing remissions in asthma. In other inflammatory disorders the main mode of action of biological agents that are effective in inducing remissions is believed to be:

A. Induction of counter-regulatory factors such as soluble ‘decoy’ receptors
B. Induction of 15-lipoxygenase to promote formation of specialized pro-resolving lipid mediators
C. Suppression of inflammation at the level of soluble cytokines, chemokines and alarmins
D. Rebalancing the interleukin-10 (IL-10) to IL-17A/F ratio in mucosal associated invariant T (MAIT) cells.

Question 3

A 38-year old man attends his primary care physician complaining of frequent episodes of cough and sputum production, but without shortness of breath on exertion. He is a smoker of approximately 15 pack-years. Over the past 3 years he gives a history of regular bronchitis also with upper respiratory tract infections (colds) during the winter months. He has no other illnesses and weight has been stable. There is no family history of airways disease.

What is the first action that the primary care physician should do:

A. Prescribe a course of antibiotics
B. Obtain a CT Chest scan
C. Perform spirometry
D. Blood sugar level
E. Perform a blood alpha-1 antitrypsin level

Question 4

A 42-year old woman with an 8 pack-year history of smoking presents with moderate dyspnea for 4 years. She was told she had wheeze in her lungs as a young child and did not perform well in athletic activities. She has tachycardia (98 bpm at rest), weighs 52 kgs with a height of 1.58 M (5 ft 2 inches) and oximetry reads 94% in room air. She has decreased breath sounds with mild thoracic hyperinflation. She walks 210 meters in the 6 minute walk test. On triple bronchodilator/inhaled corticosteroid (ICS) therapy, she has had 2 visits to the ER for “attacks” over the last year. Spirometry shows an FEV1/FVC of 0.52 and an FEV1 of 45% of predicted.

Which of the following tests are potentially helpful for her management?
A. A fat free mass index determination  
B. Lung volumes with diffusion capacity for CO  
C. Cardiopulmonary exercise test (CPET)  
D. Alpha-1 antitrypsin level  
E. Eosinophil count to check if above 2%

**Question 5**

Drugs for disease modification in rheumatoid arthritis have been approved by the FDA based on:

A. Demonstration of improvement in symptoms  
B. Resolution of joint swelling  
C. Slowing of progression based on radiographic evaluation  
D. Improvement based on MRI findings  
E. Induction of remission
Wednesday, May 23

PCC4: Pediatric Clinical Core Curriculum: Pulmonary Physiology

Question 1
An 11-year-old boy undergoes pulmonary function testing. He had a left upper lobectomy performed at 1 week of age for congenital lobar emphysema. Which of these findings is most likely to be decreased?
A. Ratio of residual volume to total lung capacity (RV/TLC)
B. Total lung capacity (TLC)
C. Ratio of forced expiratory volume in 1 second over forced vital capacity (FEV1/FVC ratio)
D. Forced vital capacity (FVC)
E. Specific airway resistance (sRaw)

Question 2
Physiologic measurements are obtained in a cohort of 18 month old infants and a cohort of 6 year old children. Which of the following is most likely to be higher in the infants?
A. Expiratory time constant
B. Chest wall compliance
C. Passively determined FRC
D. Contribution of the rib cage to inspiratory tidal volume
E. Lung compliance

Question 3
Specific airway resistance (sRaw) can be measured using a body plethysmograph in preschool children, who frequently cannot perform the forced expiratory maneuver required for spirometry. Which of the following actions should be performed in order to obtain accurate measurement of sRaw in preschool children?
A. BTPS correction
B. Low inspiratory and expiratory flow rates
C. High inspiratory and expiratory flow rates
D. Perform test at FRC
E. Do not allow parent to be in the box during the test

Question 4
A 29 year old female with history of sickle cell disease (Genotype SS) presents with shortness of breath for one month. She notices that her exercise capacity has been markedly reduced in the last month. She quit smoking 2 months ago. She denies fever, chest pain, and cough. Her chest X-ray is unremarkable for any acute pathology. Her D-Dimer is negative. Of note, she has had 8 vaso-occlusive crises in the last 8 months. What is the next best step in the management?
A. Obtain a Doppler Echocardiogram to evaluate for tricuspid jet velocity during steady state
B. High resolution Chest CT
C. V/Q scan to evaluate for chronic thromboembolic disease.
D. Obtain a Right heart catheterization to look at the filling pressures of the heart
E. Obtain Pulmonary function testing to rule out COPD
Question 5

A 10-year-old boy with a history of asthma presents with 2 days of fevers, rhinorrhea, poor PO intake, and difficulty breathing not relieved by his albuterol inhaler. Current VS include HR 135, RR 24, BP 140/90, O₂ saturation 94% on room air, Temp 38.2. His lung exam reveals diffuse wheezing bilaterally with a prolonged expiratory phase and substernal retractions. Arterial blood gas on room air shows pH=7.31 pCO₂=39 pO₂=72

Which of the following best explains the impact of non-invasive positive pressure ventilation (NIPPV) in this patient?

A. NIPPV is contraindicated because it will aggravate dynamic hyperinflation and auto-PEEP.
B. NIPPV will improve oxygenation by recruiting atelectatic lung and minimizing shunt physiology.
C. NIPPV is contraindicated because it will increase intrathoracic pressure, impede venous return and precipitate cardiac arrest.
D. NIPPV will reduce his work of breathing by relieving the high negative intrathoracic pressure required to overcome intrinsic auto-PEEP.
E. NIPPV is contraindicated because he is in respiratory failure; he should be safely managed with endotracheal intubation, deep sedation and neuromuscular blockade.

Question 6

An 8-year-old, 20 kg, ex-26 week preemie with known chronic lung disease from bronchopulmonary dysplasia (BPD) presents with cough, respiratory distress, and 3 days of fevers and rhinorrhea.

On initial evaluation, her chest xray shows increased interstitial markings, cystic regions, peribronchial cuffing, and mild bronchiectasis, unchanged from previous xrays. On exam, she manifests rapid shallow breathing, coarse breath sounds with scattered wheezing, RR=40, and O₂ saturations of 88% on 100% oxygen via face mask. Her initial arterial blood gas: pH 7.30 pCO₂=70 pO₂=60. Her respiratory viral panel is positive for rhinovirus. The decision is made to intubate for acute on chronic respiratory failure caused by viral infection.

Once intubated, she is placed on VCAC ventilation, PEEP=5, FiO₂=100%, Vt=120 mL, RR=30, i-time=0.5 sec. Subsequent arterial blood gas reveals pH 7.20, pCO₂=82 pO₂=57. Echocardiogram shows estimated right ventricular pressures equal to left ventricular pressures with septal wall flattening.

What is the appropriate ventilator management strategy for her acute-on-chronic respiratory failure, given her known BPD and evidence of pulmonary hypertension?

A. PEEP 18, Vt=120 mL, RR=30, i-time 0.5 sec with inhaled nitric oxide (iNO) at 5 ppm
B. PEEP 10, Vt=200 mL, RR 20, i-time 0.7 sec with iNO at 20 ppm
C. PEEP 5, Vt=120 mL, RR=25, i-time 0.8 sec with iNO at 20 ppm
D. PEEP 12, Vt=180 mL, RR=30, i-time=1 sec with sildenafil 0.5 mg/kg q8h
E. PEEP 7, Vt=220 mL, RR=15, i-time=1.5 sec with bosentan 0.5 tab PO q12h

Question 7

A 16-year-old woman is intubated for status asthmaticus, triggered by viral infection. She receives steroids, nebulized bronchodilators, and IV fluids for volume repletion in the setting of hypotension on positive pressure ventilation. 24 hours after intubation, you are called to bedside for elevated peak pressures on her ventilator. She is sedated and passively ventilated with a volume assist-control mode and square wave flow at 60 LPM. You set a 0.4 second end-inspiratory pause to examine her respiratory mechanics. Panel B shows her waveforms currently, in comparison to Panel A, her respiratory mechanics on admission during the same ventilator diagnostic maneuver after rapid sequence intubation with succinylcholine.

![Panel A and B waveforms](image-url)
Which of the following diagnoses could explain the change in her respiratory physiology on the ventilator?
A. Bronchospasm worsening with time
B. Bronchospasm fully resolving to normal pulmonary physiology
C. Kinked or obstructed tubing
D. Worsening abdominal distention
E. High flow superimposed on bronchospasm

D1: Clinical Year in Review 4: Critical Care, Health Disparities, Nonsococomial Pneumonia

Question 1
According to data on lung cancer incidence and mortality from the National Cancer Institute, which of the following is true?
A. Tobacco smoking rates are higher in large metropolitan areas than in small rural areas.
B. Incidence of lung cancer is higher in urban areas compared to small rural areas, mostly driven by higher smoking rates in inner cities.
C. Incidence of lung cancer is higher in rural areas than in urban areas, but mortality is very similar in both settings.
D. Mortality among patients with lung cancer stages II-IV was significantly higher in rural areas compared to large metro areas.
E. Stage I lung cancer patients in rural areas underwent fewer surgeries and had lower medial survival than those in large urban areas.

Question 2
Your hospital is launching an end-of-life program and would like to engage the ICU team to support family members of deceased patients in the ICU with grief. You're the local champion for quality in your intensive care unit.

What would be your next step in a Quality Improvement project to reduce grief in family members of deceased critically ill patients?
A. Review the existing literature on strategies to improve grief in family members of ICU patients
B. Implement an end-of-life bundle based on current practice from oncology
C. Collect data to determine the prevalence of grief in the family members in your unit
D. Implement a condolence letter for all relatives of deceased patients
E. Identify the risk factors for grief in your population

Question 3
The ICU director in your hospital would like to involve families in bedside rounds. When the interprofessional team was surveyed, several concerns were raised about this new program. You were asked to review the evidence about involving families on rounds in ICUs.

The evidence you gathered supports the following statements:
A. Families usually do not understand discussions during rounds
B. Families provide information that informs the plan of care
C. Families increase the duration of rounds
D. Resident teaching deteriorates when families are on rounds
E. Rounds increase the opportunities for families to ask questions

Question 4
A 67-year old man with a history of diabetes and mild congestive heart failure is admitted to the hospital with a myocardial infarction. Percutaneous angiography and stent placement are successful but the procedure is complicated by acute urinary retention requiring placement of a foley catheter. On the fourth hospital day, the patient has new fever, cough, and increasing shortness of breath on exertion. Review of systems is otherwise negative. On examination, he has a temperature of 101.2°F, respiratory rate of 32 breaths per minute, blood pressure 90/60 mm Hg, oxygen saturation of 86% on ambient air. Exam is otherwise notable for
moderate elevation of the jugular venous pressure, bilateral crackles in the lung bases, and mild swelling of the ankles.

What is the best microbiologic diagnostic testing to send on this patient?
A. Blood and urine cultures
B. Blood and sputum cultures
C. Blood, urine, and sputum cultures
D. Blood, urine, sputum cultures and respiratory virus PCRs
E. Blood and sputum cultures and respiratory virus PCRs

Question 5

A 67-year old female presents with fevers, cough, dyspnea, chest pain, and malaise. Exam is notable for hypotension, tachycardia, and hypoxia. Labwork is notable for leukocytosis and elevated lactate. She is intubated and treated with invasive mechanical ventilation, as well as broad-spectrum antibiotics, crystalloid fluid resuscitation, and norepinephrine infusion. Four hours later, she continues to require vasopressor infusion to maintain mean arterial pressure >65 mm Hg. She has no history of adrenal insufficiency, no prior steroid use, and did not receive etomidate for intubation.

Treatment with hydrocortisone infusion is associated with:
A. Reduced in-hospital mortality.
B. Reduced hospital length of stay.
C. Increased incidence of new bacteremia and fungemia.
D. Reduced duration of mechanical ventilation.
E. Reduced length of hospitalization.

Question 6

A 75-year old male presents with fevers, chills, and dysuria. On exam, he is hypotensive and confused. His extremities are warm. Blood work is notable for leukocytosis and acute renal failure. Urinalysis is notable for bacteria, leukocyte esterase, and nitrites. Broad-spectrum antibiotic, intravenous fluids, and high-dose norepinephrine infusion are started. Your resident would like to start angiotensin II.

What benefit from angiotensin II would this patient be most likely to gain?
A. A reduced 90-day mortality.
B. An improved quality-of-life at 6 months.
C. An improved mean arterial pressure.
D. Fewer serious adverse events.
E. All of the above.

D2: Understanding Chronic Lung Allograft Dysfunction: Update from the Bedside and Bench

Question 1

A 24 year-old female status post bilateral lung transplantation for cystic fibrosis 9 months ago presents with an acute asymptomatic drop in home monitored FEV1 from a baseline of 3.0 liters to 2.7 liters. Physical examination is unremarkable. CXR shows a subtle bilateral upper lobe infiltrate. Laboratory testing confirms FEV1 has fallen to 2.65 with a stable FVC of 3.3 liters. CBC and BAL show an elevated eosinophil count.

What is the next best step in the management?
A. Perform full lung function testing including lung volumes and diffusion
B. Arrange high resolution chest CT
C. Schedule an urgent bronchoscopy with transbronchial lung biopsies
D. Treat empirically for acute rejection with 3gms of methylprednisolone over 3 days
E. Prescribe broad spectrum antibiotics and follow up with repeat spirometry in 3 weeks
Question 2
The same patient, 6 weeks later has progressive loss of lung function despite treatment for biopsy proven acute rejection (Grade A2B1R) with 3gms of methylprednisolone. HRCT chest shows worsening bilateral upper zone infiltrates and her lung function shows an FEV1/FVC ratio of 2.2/ 2.4. The eosinophilia has resolved.

What is the most likely specific diagnosis?
A. Bronchiolitis obliterans syndrome (BOS)
B. Restrictive allograft syndrome (RAS)
C. Restrictive chronic allograft dysfunction (R-CLAD)
D. Recurrent cystic fibrosis
E. Pulmonary drug toxicity

Question 3
Six months later her lung function shows an FEV1/FVC ratio of 1.0 / 1.2 liters with a TLC of 3.0 liters compared to a baseline of 5.0 liters. She is short of breath on minimal exertion and her weight has fallen with her BMI falling from a best of 24 to 18.

What is the preferred management strategy at this stage?
A. Schedule end of life discussion and set reasonable limits of care, including intubation, ECMO or retransplantation
B. Empiric treatment for antibody mediated rejection
C. Change baseline immune suppression
D. Work up for retransplantation
E. Repeat bronchoscopy and biopsy to rule out ongoing acute rejection

Question 4
An overweight (BMI 36) 52-year old male presents with recurrent nocturnal cough 4 years after bilateral lung transplant for idiopathic pulmonary fibrosis. He also reports water brash but no heartburn. There has been a slow decline in his FEV1 over the last 6 months from a baseline of 4.2 liters to the current value of 3.6 liters concurrent with weight gain. Physical examination reveals occasional inspiratory crackles at the bases, which clear partially on deep breathing and coughing. Imaging shows no focal abnormality but expiratory HRCT views show some mosaic changes.

How should one most accurately determine whether gastroesophageal reflux is present?
A. Perform bronchoscopy with BAL and oil red O stains for fat globules
B. Perform upper endoscopy looking for distal esophagitis
C. Perform 24 hour esophageal pH probe monitoring
D. Perform 24 hour esophageal impedance plethysmography
E. Perform exhaled breath analysis for bile acids

Question 5
A 43-year old female status post heart-lung transplant for complex congenital heart disease 12 years ago presents with an asymptomatic fall in home monitored FEV1, 6 weeks after a lower respiratory tract infection where BAL PCR demonstrated Parainfluenza virus. Her FEV1 had fallen from a baseline of 3.0 liters in the first year post transplant to 2.4 liters prior to this infection then to 2.0 liters 6 weeks after the infection. Her chest is quiet on auscultation, imaging shows no focal lesions on HRCT and transbronchial lung biopsy shows minimal rejection (A1B0) with scattered foci of organizing pneumonia and neutrophilic capillaritis in the absence of a positive C4d stain. Repeat BAL PCR is now negative. Single antigen bead testing shows a new donor specific antibody to HLA DQ with an MFI of 2,000, which increases with serial dilution. C1q binding is positive.

What is the most likely diagnosis?
A. Acute LRTI Parainfluenza virus infection superimposed on BOS Grade 1
B. Parainfluenza virus associated acute cellular rejection
C. Antibody mediated rejection
D. Autoantibody mediated rejection
E. BOS Grade 2
**Question 6**

A 42-year old female recipient of a right single lung transplant for necrotizing sarcoidosis at the age of 28 presents with slowly progressive shortness of breath over the last few years. On examination her trachea is deviated towards the right but breath sounds are audible, vesicular with no added sounds. Spirometry reveals her FEV1 has fallen from a baseline of 2.5 liters to 1.5 liters over the last 20 years. Her FVC has fallen from 3.5 to 1.8 liters. Imaging shows herniation of the non-transplanted lung across the midline.

Would you investigate for chronic allograft dysfunction (CLAD) and how?

A. No, the change in FEV1 reflects change in predicted values over time.
B. No, the change in FVC reflects change in predicted values over time.
C. No, as there is no effective treatment.
D. Yes, with lung volumes, diffusion, gas exchange, HRCT, differential V/Q scan, ECHO +/- RH catheter and bronchoscopy.
E. No, as the only treatable condition is acute cellular rejection, so an empiric IV pulse of methylprednisolone is indicated.

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**D3: Women and Lung Disease**

**Question 1**

Which of the following is consistent with the phenotype of perimenopausal asthma?

A. Milder, easily controllable asthma
B. Induced sputum with higher percentage of neutrophils and lower percentage of eosinophils
C. Induced sputum with higher percentage of eosinophils and lower percentage of neutrophils
D. A history of atopy
E. Lower BMI

**Question 2**

Which of the following statements is true?

A. The estrogen paradox in pulmonary arterial hypertension (PAH) refers to findings that female sex is not a significant risk factor for the development of PAH while at the same time conferring a survival advantage to patients with PAH
B. Increased circulating levels of the estrogen metabolite E2 have been associated directly with increased six minute walk distance (6MWD) in men with PAH
C. In a phase II clinical trial in patients with PAH, use of the aromatase inhibitor Anastrazole was associated with an increase in RV function as measured by TAPSE (Tricuspid Annular Plane Systolic Excursion)
D. RV function (as measured by RV ejection fraction or RVEF) improvement in response to PAH therapies appears to be greater in female patients when compared to male patients with idiopathic and heritable PAH.

**Question 3**

A 42-year old woman presents with an 8-month history of worsening shortness of breath on exertion. She endorses a daily, nonproductive cough that has been present for perhaps a year. She denies any past medical history but on review of systems endorses a history of dry eyes, dry mouth, dysphagia with dry solid foods for approximately one year. She has a history of Raynaud’s symptoms since age 15.

Physical exam is notable for clear lungs bilaterally, normal heart exam, and there is no evidence of arthritis or abnormal skin findings.

You perform office spirometry:

- FVC 70%
- FEV1 80%
- FEV1/FVC 0.88
CT imaging was performed and shows:

Which of the following is the most appropriate next step in management?
A. Begin either pirfenidone or nintedanib
B. Obtain CT angiogram to rule out PE
C. Obtain ANA, SCL-70, and Jo-1 antibodies
D. Perform right heart catheterization to measure pulmonary pressures
E. Begin albuterol MDI

Question 4
Which of the following statements regarding gender differences in lung cancer is true?
A. Since 1965, men's and women's smoking prevalence in the United States have fallen at the same rate.
B. Men have greater frequency of epidermal growth factor receptor (EGFR) mutations than women.
C. Men and women with lung cancer have equal DNA repair capacity.
D. Women have higher 5-year survival after non-small cell lung cancer treatment, regardless of disease stage.
E. Estrogen and progesterone appear to have no effect on lung cancer development or outcomes.

Question 5
Which of the following statements is correct?
A. COPD prevalence has been increasing more rapidly in women than in men
B. Women are less susceptible to develop COPD with same risk exposure
C. Exposure to passive smoke and biomass fuel for heating in developed country is not a risk factor for COPD in females
D. Current COPD guidelines are largely based on clinical trials with equal number of male and female participants
E. Women are not likely to develop osteoporosis and high dose inhaled steroids could be associated with less side effects in females

D9: Controversies in Clinical Pediatric Pulmonology

Question 1
Meta-analyses of studies analyzing the clinical response to inhaled salbutamol in infants with bronchiolitis have shown little or no positive benefit in children diagnosed with this condition.
The major reason for this is that:
A. Airway smooth muscle does not develop until after the age of 2 years.
B. Wheeze in bronchiolitis is mostly related to airway edema and mucous plugging, and not to bronchospasm
C. There is no reliable method to administer inhaled salbutamol to young children.
D. β2 sympathomimetic receptors are not present on airway smooth muscle cells in children below the age of 2 years
Question 2
A previously healthy 5-month old term female infant presents with a four-day history of nasal congestion, low grade fever, and cough. On exam she appears non-distressed with a respiratory rate of 42 and SpO2 on room air of 94%. On chest exam there are mild suprasternal retractions with moderate high-pitched polyphonic expiratory wheezing diffusely.

What is the most appropriate course of action?
A. Admit to the hospital for high-flow nasal cannula oxygen
B. Dexamethasone 0.6 mg/kg/day for 2 days
C. Supportive care with nasal hygiene
D. Albuterol 1.25 mg via nebulizer every four hours
E. Albuterol 1.25 mg via nebulizer every four hours and dexamethasone 0.6 mg/kg/day for 2 days

Question 3
10-year old boy with history of Hb SS sickle cell disease (SCD) is being evaluated for episodes of shortness of breath on exertion. His past medical history is significant for two episodes of Acute Chest Syndrome (ACS) before age 6, and 3 episodes of vaso-occlusive crises (VOC) for which he was hospitalized. He has not had any episodes during the past 2 years. His hemoglobin has been stable at 8.5 g/dL. He has no other known medical problems. There are no abnormal findings on physical examination.

His exercise intolerance is most likely due to:
A. Significant restrictive lung disease
B. Severe decrease in his diffusing capacity
C. Chronic thromboembolic disease
D. Exercise-induced bronchospasm
E. Clinically significant pulmonary hypertension

Question 4
Asthma has been recognized as an important risk and/or complicating factor of acute chest syndrome in patients with Sickle Cell Disease. However, its treatment during exacerbations remains controversial in part due to concerns of potential adverse effects.

Which of the following treatments has been associated with serious adverse effects in patients with Sickle Cell Disease (SCD)?
A. Long-term antibiotics
B. Inhaled steroids
C. Hypertonic saline
D. Immunotherapy (allergy shots)
E. Pulse dose oral steroids

Question 5
Colonization of the airways with bacterial and fungal organisms is a constant concern in patients with Cystic Fibrosis (CF) and several infection control measures have been instituted to prevent significant environmental exposures and/or person-to-person transmission.

Which of the following organisms has been associated with hospital outbreaks during construction?
A. Mycobacterium abscessus complex
B. Aspergillus fumigatus
C. Methicillin resistant Staphylococcus aureus (MRSA)
D. Pseudomonas aeruginosa
E. Burkholderia cenocepacia
### CC5: Pulmonary Clinical Core Curriculum I: Lung Disease

#### Question 1
Ms. Smith is a 72-year-old female with diffuse cutaneous scleroderma, manifested by sclerodactyly, Raynaud's phenomenon, interstitial lung disease (ILD) and the presence of anti-topoisomerase (anti-scl70) antibodies. She has progressive dyspnea and evaluation reveals radiographic and physiologic progression of her ILD.

What treatment has been shown in prospective trials to improve lung function in patients with scleroderma-associated interstitial lung disease?

A. Mycophenolate mofetil  
B. Oral cyclophosphamide  
C. Prednisone  
D. Azathioprine  
E. Both A and B

#### Question 2
Which of the following connective tissue diseases is rarely associated with ILD?

A. Polymyositis/dermatomyositis  
B. Systemic lupus erythematosus  
C. Rheumatoid arthritis  
D. Systemic sclerosis

#### Question 3
Why do patients with connective tissue disease-associated-ILD have a better survival than those with idiopathic interstitial pneumonia?

A. Higher incidence of female sex  
B. Higher incidence of non-specific interstitial pneumonia  
C. Lower incidence of smoking  
D. Better survival for usual interstitial pneumonia pattern in all connective tissue diseases

#### Question 4
A 64-year-old man with a history of idiopathic pulmonary fibrosis on baseline supplemental oxygen at 2 liter/minute presents for the evaluation of increased dyspnea. Pulmonary function tests show: FVC 60% of predicted, DLCO 30% of predicted, which are stable from a test performed six months prior. An echocardiogram is remarkable for a pulmonary artery systolic pressure of 68 mmHg and new right ventricle dilation with moderate to severe dysfunction. On right heart catheterization: pulmonary artery pressure 45/21 (30) mmHg, pulmonary capillary wedge pressure 10 mmHg, pulmonary vascular resistance 3.9 WU, cardiac index 2.6 lpm/m^2^. Chest CT (show below) shows no clear worsening of his underlying parenchymal disease and no evidence of pulmonary embolism. Which of the following treatments may improve this patient’s lung function, symptoms of dyspnea and quality of life?

![CT scan of the chest](image_url)
Which of the following therapies are recommended for the initial treatment of this patient’s pulmonary hypertension?

A. Combination therapy with ambrisentan/ tadalafil
B. Inhaled prostacyclin therapy
C. Sildenafil
D. Pirfenidone
E. Supplemental oxygen

Question 5

A 40-year-old woman with recently diagnosed diffuse scleroderma and scleroderma related interstitial lung disease is referred to you for evaluation for worsening dyspnea. Her pulmonary function testing demonstrates FVC 64% predicted, FEV1 74% predicted and DLCO 26% predicted. There are no prior tests for comparison. NT-proBNP level is 900 pg/ml.

When screening this patient which measurement(s) would suggest the presence of pulmonary arterial hypertension?

A. FVC
B. FVC/ DLCO ratio
C. NT-proBNP
D. FVC and NT-proBNP
E. FVC/DLCO and NT-proBNP

Question 6

A 48-year-old woman with limited scleroderma presents for the evaluation of worsening dyspnea on exertion. A recent chest CT shows mild basilar fibrosis. She is referred for right heart catheterization to assess for pulmonary hypertension. The results of that test are: right atrial pressure 5 mmHg, pulmonary arterial pressure 64/30 (41) mmHg, pulmonary capillary wedge pressure 10 mmHg, cardiac output 4 L/min, pulmonary vascular resistance 10 WU. Acute vasoreactivity testing was performed, and she was deemed a non-responder.

Which of the following statements is true regarding the treatment of her pulmonary arterial hypertension?

A. She should be started on a calcium channel blocker.
B. She should be started on pulmonary arterial hypertension-specific therapies.
C. She should be started on cyclophosphamide therapy.
D. All of the above are indicated.
E. None of the above are indicated

Question 7

You diagnose a 38-year-old woman with sporadic lymphangioleiomyomatosis (LAM). She has characteristic findings on HRCT with diffuse lung cysts. She has no renal angiomyolipomas or lymphatic abnormalities. Her VEGF-D level is 923pg/ml. Her FEV1 is 68% of the predicted normal value for her race, age and height. A spirometry performed 12 months ago at a local health fair showed her FEV1% was 73%. She says her dyspnea is a bit more than she would expect when she does her cross-training exercise class.

In addition to recommending that she avoid estrogens, published data support the use of which of the following to stabilize lung function?

A. Supplemental oxygen
B. Sirolimus
C. Bilateral oophorectomy
D. High-dose progesterone

Question 8

A 50-year-old man has long-standing Sjögren's syndrome with sicca (mouth and eyes), Raynaud phenomenon, dysphagia, fatigue and high-titer anti-Sjögren's syndrome-related antigen A (SSA)/Ro and anti-Sjögren's syndrome-related antigen B (SSB)/La antibodies. He is a never-smoker. His primary care provider performed a chest radiograph to evaluate acute symptoms of cough and sputum production. The chest radiograph
was read as abnormal and showing emphysema. A chest HRCT scan was performed to follow-up the chest radiograph. It showed cystic lung disease.

What is the classic distribution of lung cysts seen in patients with this disease?
A. Upper zone predominant
B. Random
C. Peri-bronchovascular and lower zone predominant
D. Peripheral and subpleural

**Question 9**

In your outpatient clinic, you are asked to evaluate a 32-year-old woman who decided to “get healthy” six months ago. At that time, she quit smoking and starting a vigorous daily walking program. Last week, she presented to a local emergency department with the sudden onset of pleuritic chest discomfort and dyspnea. She was found to have a 15% pneumothorax on chest radiograph which was evacuated via needle aspiration. To further evaluate her lungs, she was instructed to get a chest CT scan and see you in clinic. The CT done yesterday shows cystic lung disease, with cysts predominantly found in the medial aspects of the basilar zones bilaterally. On exam, you notice several, small grayish-white papules on the cheeks and neck.

Besides a second pneumothorax, which of the following is this patient at risk for?
A. Leukemia
B. Melanoma
C. Renal cancers
D. Splenic rupture

**CC6: Pulmonary Clinical Core Curriculum II: Lung Disease**

**Question 1**

A 62-year old man presents to your office complaining of slowly progressive dyspnea on exertion. He now has difficulty lying flat at night due to dyspnea and has been sleeping propped up on several pillows. He also complains of occasional swallowing difficulty. His exam is consistent with subtle proximal muscle weakness and he has notable fasciculations in his tongue. He has normal reflexes.

You suspect that he may have amyotrophic lateral sclerosis. His pulmonary function tests demonstrate a decreased FVC with normal FEV1/FVC ratio and a reduced TLC.

Which of the following is true of a diffusing capacity maneuver in a patient with neuromuscular weakness?
A. The diffusing capacity for carbon monoxide (DLCO) is always reduced
B. The transfer coefficient (often referred to as the DLCO/VA or KCO) is generally supranormal in patients with pure neuromuscular weakness
C. While the FVC is usually low in neuromuscular disease, the alveolar volume maneuver (VA) is generally normal
D. The diffusing capacity adjusted for hemoglobin will always be unchanged in this scenario
E. A normal DLCO rules out neuromuscular disease

**Question 2**

A 21-year old man with Duchenne muscular dystrophy has the following pulmonary function test results:

- FVC: 31% of predicted
- FEV1: 34% of predicted
- FEV1/FVC: 79%
- TLC: 38% of predicted
- DLCO: 67% of predicted
- Maximum Inspiratory Pressure: -28 cm H2O
- Maximum Expiratory Pressure: 40 cm H2O
- Peak cough flow: 220 L/minute
He uses average volume assured pressure support ventilation at night via a full face mask, with good compliance.

Based on these pulmonary function tests, which of the following is most likely to benefit this patient in addition to his nocturnal ventilation?

A. Twice daily bronchodilators
B. Sip ventilation during the day regardless of symptoms
C. A feeding tube
D. Cough clearance maneuvers
E. Vest percussive chest physiotherapy

Question 3

You are called from an outside hospital for transfer to your institution of a 54-year-old woman with newly diagnosed Guillain-Barré syndrome. The referring physician described a woman who appears comfortable on room air but is drooling, has severe facial muscle weakness, and is unable to move her legs. Her upper extremities have 3/5 strength.

They have measured her Vital Capacity at 1.8 liters (25 mL/kg) and her Maximum Inspiratory Pressure is 50 cm H2O; these have been stable since admission to their hospital 24 hours ago. They have not obtained an ABG.

Which of the following clinical features most strongly supports a need for monitoring this patient in an intensive care unit, rather than a stepdown unit or other general medical ward?

A. Lower extremity paralysis
B. Most recent vital capacity measurement
C. Most recent maximum inspiratory pressure measurement
D. Her age
E. The presence of severe bulbar muscle weakness

Question 4

A 63-year-old man with idiopathic pulmonary fibrosis (IPF), being treated with pirfenidone, sees you in clinic. He has never been treated with other medications for IPF. His pulmonary function tests have declined with a current FVC of 55% of predicted. He has also developed a new requirement for supplemental oxygen at 2LPM on exertion only. He remains active and exercises regularly. His other medical problems include obesity with a body mass index of 36.3kg/m2, and well-controlled hypertension. He has a history of early stage prostate cancer, resected at age 57, without evidence of recurrence. You discuss referral for lung transplant evaluation with him.

Which of the following is true regarding referral for lung transplant in this patient?

A. The patient’s current weight is a relative contraindication to transplantation
B. Disease severity does not warrant transplant referral at the current time
C. Current antifibrotic therapy is associated with complications after transplant
D. Transplant referral should be considered only after medical therapies (other antifibrotics) have been exhausted
E. A history of prostate cancer is a contraindication to transplantation

Question 5

A 45-year-old woman with chronic hypersensitivity pneumonitis who is active on the waiting list for lung transplantation has been admitted with progressive hypoxemic respiratory failure due to progression of her interstitial lung disease. She was treated with high-dose corticosteroids upon admission without improvement in respiratory status and is now being supported with veno-venous extracorporeal membrane oxygenation (ECMO). She is awake and able to both ambulate and participate in physical therapy with no other significant organ dysfunction. She is 60 inches tall with a BMI of 24 kg/m2 and remains actively listed for transplant.

Which of the following is most likely to prolong this patient’s waiting time for lung transplantation?
A. Diagnosis of fibrotic ILD (as compared to COPD)
B. Height
C. Female sex
D. Need for ECMO support
E. Recent corticosteroid therapy

Question 6

A 60-year old woman with a history of interstitial lung disease secondary to systemic sclerosis underwent successful bilateral lung transplant 11 months ago. She was CMV seropositive at the time of transplant. In addition to standard 3-drug immunosuppression, she has been treated with azithromycin, trimethoprim-sulfamethoxazole, valganciclovir and voriconazole since transplant. Her extra-pulmonary manifestations of scleroderma have been well controlled. She has not experienced any episodes of acute rejection, infection or other significant complications.

In this patient, which of the following has been shown to significantly increase the risk of complications after lung transplant?

A. Underlying scleroderma
B. Bilateral rather than unilateral lung transplant
C. Prolonged azithromycin exposure
D. Prolonged valganciclovir exposure
E. Prolonged voriconazole exposure

Question 7

A 66-year old male coal miner presents with slowly progressive shortness of breath and cough. The patient has a 40 pack-year history of smoking. Chest computed tomography (CT) demonstrates multiple small (<1 cm) nodular opacities in the bilateral upper lung zones (Figure 1). There is mild enlargement of several mediastinal lymph nodes. Given the presence of multiple pulmonary nodules, the patient undergoes positron emission tomography with 18F-fluorodeoxyglucose (FDG) and concurrent computed tomography (PET-CT). This examination demonstrates markedly increased FDG-avidity in several of the upper lobe pulmonary nodules. An open lung biopsy is performed and representative histology is shown in Figure 2.

Which of the following is the most likely diagnosis?

A. Silicosis
B. Coal worker’s pneumoconiosis
C. Progressive massive fibrosis
D. Berylliosis
E. Metastatic lung carcinoma
Question 8
A 42-year-old male custodian is referred to the pulmonary clinic with 3 months of cough, wheezing and shortness of breath. He has a 10 pack-year history of cigarette smoking, but quit at the onset of these symptoms. He has been treated with inhaled albuterol as a rescue therapy followed by a trial of an inhaled corticosteroid with improvement in symptoms. One day prior to the onset of the wheezing and shortness of breath, the patient remembers spilling a large container of bleach cleaning solution in the custodial closet. Vital signs are normal. Physical examination demonstrates mild wheezes in bilateral lung fields. Spirometry is performed and shows mild airflow obstruction without a bronchodilator response. Laboratory results include a normal eosinophil count and a serum IgE level of 64 kU/L (normal <214 kU/L).

What is the most appropriate next diagnostic test?
A. High resolution CT of the chest without contrast
B. Direct laryngoscopy
C. Sputum eosinophil count
D. Methacholine challenge test
E. Specific bronchoprovocation testing with the cleaning agent of interest

Question 9
The state legislature is debating a bill to increase the renewable portfolio standard (RPS) and to address vehicle emissions. As part of this debate, you are asked to give expert testimony on the potential health effects of air pollution due to the burning of fossil fuels. During the hearing, the lawmakers make a series of claims about the relationship between air pollution and health.

Which of the following statements about the effects of air pollution on human health is most accurate?
A. Surface temperature has no effect on ground-level ozone formation.
B. Impaired mucus clearance exacerbates the effects of air pollution among patients with chronic obstructive pulmonary disease (COPD).
C. Short-term exposure to air pollutants has not been associated with increased rates of myocardial infarction.
D. Reduction in air pollution during the 1996 Atlanta Summer Olympic Games was negligible and therefore had a limited effect on asthma outcomes.
E. Asthma exacerbations occur independently of ground-level ozone concentrations.

Question 10
A 52-year-old man with coronary artery disease and atrial fibrillation controlled with amiodarone presents with one week of worsening dyspnea and cough. A high-resolution chest CT is obtained and lung toxicity from amiodarone is strongly suspected.

Which of the following HRCT patterns, if found, would most favor a diagnosis other than amiodarone pulmonary toxicity?
A. Interstitial pneumonitis
B. Organizing pneumonia
C. Diffuse parenchymal cysts
D. Pulmonary nodules or masses
E. Pleural effusions

Question 11
A 64-year-old man, former smoker, with stage IV non-small cell lung cancer is initiated on nivolumab, a programmed death-1 inhibitor, after disease progression on standard platinum-based chemotherapy. Nine weeks later he presents with new-onset dry cough and dyspnea on exertion. He does not report fever or chest pain. Oxygen saturation is 97% on room air. Chest computed tomography demonstrates scattered ground glass opacities. Bronchoscopy with bronchoalveolar lavage is performed showing a lymphocyte-predominant cell differential with negative microbiologic studies for viruses, bacteria, fungi including Pneumocystis.

Which of the following is an indication for initiating corticosteroids for this patient’s illness?
A. Time of symptom onset less than 3 months after starting nivolumab  
B. Ground glass opacities present on chest CT  
C. Lymphocyte-predominance on BAL  
D. Negative microbiological evaluation on BAL  
E. Symptoms of dyspnea on exertion and cough

**Question 12**

A 68-year-old woman with hypertension, hyperlipidemia, and rheumatoid arthritis presents to clinic for evaluation of several weeks of dyspnea on exertion, cough, and fevers. She is a lifelong non-smoker with no known prior history of pulmonary disease. Medications include aspirin, atorvastatin, lisinopril, and methotrexate. Exam is notable for coarse crackles and oxygen saturation of 94% on room air. Leukocyte count is 9,000 cells/mm³ and chest CT demonstrates lower lung zone-predominant ground glass opacities and centrilobular nodules. Pulmonary function tests demonstrate a restrictive pattern with a reduced DLCO. Blood and sputum cultures are negative for infection.

Which of the following features would favor a diagnosis of methotrexate lung toxicity in this patient over interstitial lung disease from underlying rheumatoid arthritis?

A. Peripheral blood eosinophilia and granulomas with eosinophils on histopathology  
B. Peripheral blood lymphocytosis and usual interstitial pneumonia on histopathology  
C. Nonspecific interstitial pneumonia on histopathology  
D. Diffuse alveolar damage on histopathology  
E. Organizing pneumonia on histopathology
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