# QUESTION BANK

# HFSA Virtual AHFTC Board Certification Review 2022

# ACKNOWLEDGEMENTS

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**HFSA Virtual Board Certification Review 2022**
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# PRACTICE

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# QUESTIONS

## QUESTION 1

A 38-year old presents to the emergency room with four days of shortness of breath, chest discomfort, and nausea. Upon arrival, heart rate is 115 beats per minute with blood pressure of 95/65 mm Hg. On physical examination, jugular venous pressure is 15 cm H2O, lungs are clear to auscultation, cardiac exam reveals tachycardia and S3. Extremities are cool. Bedside echocardiography reveals severely reduced left ventricular systolic function. 12-lead electrocardiography reveals sinus tachycardia with deep T wave inversions throughout the precordium and poor R-wave progression. On telemetry, frequent episodes of non-sustained ventricular tachycardia are seen.

Which is the best next diagnostic test?

A. Cardiac magnetic resonance imaging
B. Right heart catheterization with endomyocardial biopsy
C. Pharmacologic myocardial perfusion stress imaging
D. Coronary angiogram

## QUESTION 2

A 72-year old presents for evaluation of persistent fatigue and lower extremity edema. The patient was recently hospitalized for heart failure requiring intravenous diuretics. Yet, he still continues to notice swelling in his legs and feels dizzy when standing up.

His past medical history is notable for lumbar spinal stenosis, carpal tunnel syndrome, and aortic stenosis status-post transcatheter aortic valve replacement a few months earlier. In the office, his blood pressure seated is 100/60 mm Hg with heart rate of 65 bpm. With standing, blood pressure drops to 80/60 mm Hg with heart rate of 90 bpm.

Transthoracic echocardiography shows preserved left ventricular ejection fraction with interventricular septum of 15 mm and posterior wall thickness of 14 mm. The aortic valve is well-seated and gradients across the valve are normal.

Laboratory testing reveals a sodium of 128 mmol/L, potassium of 3.7 mmol/L, creatinine of 2.2 mg/dL. Albumin is 3.2 g/dL. Kappa free light chains are 1.8 mg/dL with lambda free light chains of 1.2 mg/dL.

What is the best diagnostic test?

A. Cardiac magnetic resonance imaging
B. Endomyocardial biopsy
C. Technetium pyrophosphate scintigraphy
D. Cardiac positron emission tomography

## QUESTION 3

A 63-year old with hypertension, insulin-dependent diabetes mellitus, and hyperlipidemia is admitted for heart failure. The patient has been admitted three times in the last six months for heart failure. Home medications include aspirin 81 mg daily, atorvastatin 80 mg daily, long-acting insulin 30 units nightly, furosemide 20 mg daily, losartan 50 mg daily.

On examination, blood pressure is 150/80 mm Hg, heart rate is 80 beats per minute. Body mass index is 38 kg/m$^2$. Jugular venous pressure is 15 mm H2O. There are decreased breath sounds at the base bilaterally. Abdomen is distended with a positive fluid wave. Cardiac examination is regular rate and rhythm. There is a III/VI holosystolic murmur at the left lower sternal border that increases with inspiration. Lower extremities have 2+ pitting edema bilaterally.

Echocardiography has repeatedly demonstrated left ventricular ejection fraction of 65% without significant valvular disease.

Which of the following approaches is most likely to decrease rehospitalization for heart failure?

A. Therapy adjustment guided by pulmonary artery pressure monitoring
B. Transition furosemide to torsemide
C. Addition of metoprolol succinate
D. Medication changes guided by daily weights

## QUESTION 4

A 76-year old is brought to the hospital by emergency medical personnel after multiple shocks from his implantable cardioverter-defibrillator and syncope. His medical history is notable for diabetes, hypertension, and coronary artery disease status-post coronary artery bypass grafting eight years ago and resulting ischemic cardiomyopathy with left ventricular ejection fraction of 20%. He is a current, every-day smoker.

On arrival, his heart rate is 110 bpm, blood pressure 80/60 mm Hg, oxygen saturation 99% on room air. His jugular venous pressure is elevated, lungs are clear and cardiac exam reveals tachycardia without murmurs and an S3. His legs are cool but without edema.

He is started on an amiodarone infusion and taken emergently to the cardiac catheterization lab where coronary angiogram reveals a patent left internal mammary artery graft to the left anterior descending artery with known occlusion of his saphenous vein grafts without revascularizable native vessels. Right heart catheterization reveals right atrial pressure of 15 mm Hg, pulmonary artery pressure of 63/35 mm Hg with pulmonary capillary wedge pressure of 30 mm Hg. Cardiac index by Fick is 1.4. An intra-aortic balloon pump is placed in the right femoral artery and the patient is admitted to the coronary care unit.

Initial laboratory results are notable for a sodium of 121 mmol/L, creatinine of 2.5 mg/dL, alanine

transaminase of 345 U/L and aspartate transaminase of 512 U/L with arterial lactate of 5 mmol/L.

Which of the following is the best next step to manage this patient?

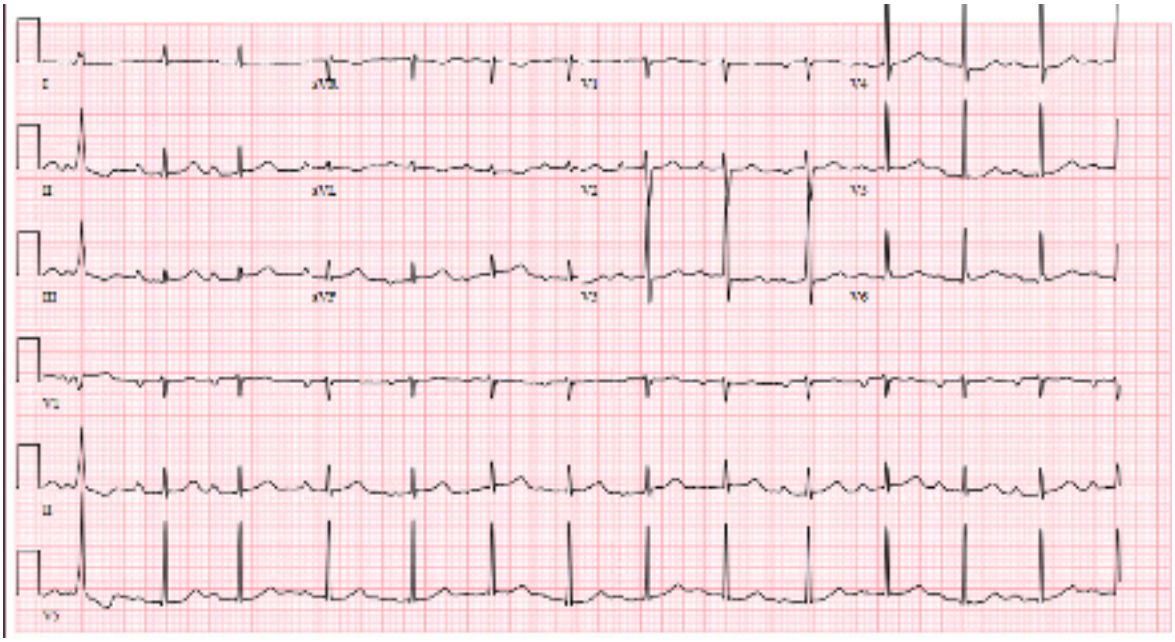
A. Implantation of a durable left ventricular assist device
B. Administration of continuous inotropes
C. Placement of an extracorporeal membrane oxygenation device
D. Enrollment in a palliative care program

## QUESTION 5

A 65-year old with ischemic cardiomyopathy presents for a follow-up outpatient visit for progressive shortness of breath. The patient has been hospitalized twice in the last year for decompensated heart failure despite adherence with diet and medications. Due to symptomatic hypotension the dose of sacubitril-valsartan was decreased from 97-103 mg twice daily to 24-26 twice daily.

After discharge, a cardiopulmonary exercise test revealed a peak exercise VO2 of 12 ml/min/kg with an RER of 1.2.

12-lead electrocardiogram is shown below. Echocardiogram shows a left ventricular end-diastolic dimension of 8.0 cm and a left ventricular ejection fraction of 15-20% with severe mitral regurgitation. Current medications include sacubitril-valsartan 24-26 mg BID, metoprolol succinate 25 mg daily, spironolactone 12.5 mg daily and torsemide 80 mg daily.



Which is the best next step in management?

A. Refer for transcatheter mitral valve repair
B. Refer for implantation of a pulmonary artery pressure sensor
C. Refer for evaluation for heart transplantation
D. Refer for cardiac resynchronization therapy.

## QUESTION 6

A 48-year old with longstanding but well-controlled human immunodeficiency virus infection presents with progressive dyspnea on exertion.

Vitals signs are notable for blood pressure of 100/70 mm Hg, heart rate of 90 beats per minute, respiratory rate 12 breaths per minute with oxygen saturation of 91% on room air. Physical examination reveals jugular venous distension with prominent v waves. Lungs are clear to auscultation. Cardiac examination reveals a right ventricular heave, regular rate and rhythm with prominent P2 and a systolic ejection murmur at the left lower sternal border. Extremities are without peripheral edema and warm.

Transthoracic echocardiogram reveals left ventricular ejection fraction of 65%, moderately reduced right ventricular systolic function with moderately dilated right ventricle. There is moderate tricuspid regurgitation. Estimated right ventricular systolic pressure is 45 mm Hg.

Subsequently, a right heart catheterization is performed and reveals the following: right atrial pressure of 14 mm Hg, pulmonary artery pressure of 84/42 mm Hg with mean PA of 56 mm Hg, pulmonary capillary wedge pressure is 13 mm Hg. Cardiac output is 4 L/min with pulmonary vascular resistance of 11 Wood units.

Inhaled nitric oxide (NO) is administered at escalating doses from 20 PPM to 80 PPM.

Which of the following hemodynamic responses to NO suggest the patient would benefit from treatment with calcium channel blockers?

A. Mean pulmonary artery pressure of 50 mm Hg with cardiac output of 3L/min
B. Pulmonary capillary wedge pressure of 15 mm Hg with cardiac output of 4L/min
C. Mean pulmonary artery pressure of 35 mm Hg with cardiac output of 4L/min
D. Pulmonary vascular resistance of 8 Wood units with cardiac output of 5L/min

## QUESTION 7

A 72-year-old woman presents to the outpatient clinic with severe, gradually worsening exertional dyspnea. Symptoms include shortness of breath with one flight of stairs and light household chores. Upon examination, BMI is 23 kg/m$^2$, heart rate is 65 bpm, jugular venous pressure is 8 cm of water, and there is no peripheral edema. Electrocardiogram shows sinus rhythm and normal intervals. A pharmacologic nuclear stress test is negative for infarction and inducible ischemia; coronary artery calcium score was 0.

Echocardiogram reveals a left ventricular ejection fraction of 65%, LV wall thickness of 1.0 cm throughout, mitral calcification with mild mitral regurgitation and moderate mitral stenosis with a diastolic mitral valve gradient of 6 mm Hg (at a heart rate of 65 bpm) and mitral valve area of 1.5 $cm^2$. Right ventricular size and function are normal, with tricuspid regurgitation jet velocity of 2.0 m/s.

What is the next best diagnostic test?

A. Coronary angiography
B. Right heart catheterization with shunt series
C. Exercise positron emission tomography stress test
D. Exercise echocardiography

## QUESTION 8

A 43-year-old woman with dilated non-ischemic cardiomyopathy (LV 8.0 cm) with LV ejection fraction 10% presents with anorexia and mild elevation of liver chemistry tests (2X upper limit of normal). Examination reveals an estimated jugular venous pressure of 15 cm of water. Echo reveals stable LV size and systolic function, mild-to-moderate aortic regurgitation and new RV dilation and severe RV contractile dysfunction. She diureses poorly over the first 24 hours in response to intravenous milrinone and high dose IV diuretics. Creatinine rises to 1.4 mg/dl from 1.0 mg/dl. Lactate is 5 mmol/L. Blood pressure is 80/60/70 mm Hg. CT angiography reveals a sub-segmental pulmonary embolism in the left lower lung. She is referred for right heart catheterization which reveals:

| | |
|---|---|
| RA | 22 mm Hg |
| PA | 63/36/49 mm Hg |
| PCW | 34 mm Hg |
| CO | 3.0 L/min |
| CI | 1.2 L/min/$m^2$ |
| PVR | 5.0 Wood units |
| PA sat | 26% |

Which of the following would be the best next step to manage this patient?

A. Intra-aortic balloon pump insertion
B. Ventriculo-arterial ECMO
C. Addition of sodium nitroprusside
D. Addition of intravenous norepinephrine

## QUESTION 9

A 38-year-old previously healthy woman has been suffering from slowly progressive exertional dyspnea over the last 3 years. Her medication history is notable for remote use of dieting pills. Pulmonary function tests reveal a 20% drop in DLCO. Echocardiogram reveals normal left ventricular size and function, a dilated

pulmonary artery, right ventricular and right atrial dilation, and moderate tricuspid regurgitation with an estimated RV systolic pressure of 70 mm Hg. She undergoes a right heart catheterization which reveals:

| | |
|---|---|
| RA | 8 mm Hg |
| PA | 76/40 (mean 53 mm Hg) |
| PCW | 11 mm Hg |
| CO | 5.0 L/min |
| PVR | 8.4 Wood units |

Which of the following would constitute a positive vasodilator challenge that would prompt initiation of long-term calcium channel blocker therapy?

A. After 5 minutes of 40 p.p.m. inhaled NO, mPAP falls to 25 mm Hg and CO remains at 5.0 L/min
B. After 5 minutes of 40 p.p.m. inhaled NO, mPAP falls to 41 mm Hg and CO rises 10% to 5.6 L/min
C. After 5 minutes of 40 p.p.m. inhaled NO, mPAP falls to 25 mm Hg and CO decreases to 3.5 L/min
D. Vasodilator challenge is not indicated.

## QUESTION 10

A 66-year-old woman with a history of mild COPD and heart failure with reduced ejection fraction (EF 25%) experiences reduced exertional tolerance. She was recently hospitalized and treated for both a COPD flare and a heart failure exacerbation. In clinic, vital signs are BP 105/72 mm Hg, heart rate 90 bpm, SpO2 96% on room air, BMI 31 kg/m$^2$. She is referred for a cardiopulmonary exercise test. Results are as follows:

Exercise time: 11 minutes and 9 seconds (stops due to dyspnea)
Peak workload: 125 Watts (4.3 METS)
Absolute peak VO2: 1371 ml/min
Peak heart rate: 144 bpm
Normalized peak VO2: 14.1 ml/Kg/min (57% predicted)
Maximum ventilatory volume: 150 L/min
Peak ventilation: 99 L/min
Respiratory exchange ratio: 1.39
VE/VCO2: 32
SpO2 at peak activity: 92%

What is the primary cause of his exertional limitation?

A. Cardiac limitation
B. Deconditioning
C. Cannot interpret due to obesity
D. Pulmonary limitation

## QUESTION 11

A 55-year-old patient is referred to your clinic for evaluation of left ventricular hypertrophy. The patient's past medical history includes hypertension, diabetes mellitus, and obesity. Symptoms include progressive exertional dyspnea and chest pressure over the prior 3 years especially with strenuous activities. Nuclear stress test and coronary angiography were consistent with the absence of obstructive coronary artery disease. Vital signs in clinic are: HR 90 bpm, BP 102/70 mm Hg, BMI 31 kg/m$^2$. On examination, jugular venous pressure is normal and there is no peripheral edema. Medications include: aspirin 81 mg/day, pravastatin 40 mg/day, metformin 1000 mg/day, lisinopril 10 mg/day. Echocardiogram shows left ventricular hypertrophy with interventricular septum measuring 21 mm and LV posterior wall 18 mm. The patient is noted to have a resting LVOT gradient of 8 mm Hg that increases to 82 mm Hg with the Valsalva maneuver ,along with concomitant systolic anterior motion of the mitral valve.

What is the best first step in management?

A. Initiate verapamil ER 180 mg/day
B. Refer for septal myectomy
C. Refer for alcohol septal ablation
D. Discontinue lisinopril 20 mg/day
E. Initiate disopyramide 600 mg/day

## QUESTION 12

A 66 year old man presents to clinic for his monthly follow-up. He notes feeling short of breath when walking to the bathroom and has difficulty dressing himself. He also complains of dyspnea and a persistent, nonproductive cough, even at rest. He says these symptoms have worsened over the past few days, though he denies any chest pain or neurological deficits. He states that he has been adherent to medications and has had no dietary indiscretions. The patient has a past medical history of HF with reduced ejection fraction (LVEF: 30%, NYHA III, Stage C), hyperlipidemia, type 2 diabetes, mood disorder, depression, renal insufficiency, central sleep apnea, and hypertension. Today his physical exam indicates a blood pressure of 145/95 mm Hg, heart rate of 58 beats per minute, and bilateral 2+ pitting edema in his legs. Pre-visit laboratories shows a B-type natriuretic peptide (BNP) level of 1021 pg/dL (baseline of 600 pg/dL), HbA1c of 7.0%, potassium (K) of 4.3 meq/L, and a digoxin level of 0.9 ng/ml. His home medications include atorvastatin 40 mg once daily, carvedilol 25 mg twice daily, valsartan 160 mg twice daily, aspirin 81 mg once daily, glyburide 5 mg twice daily, hawthorn 450 mg twice daily, pregabalin 75 mg twice daily, sertraline 200 mg once daily, aripiprazole 10 mg at bedtime, nitroglycerin as needed, digoxin 125 mcg once daily, spironolactone 25 mg twice daily, and furosemide 20 mg once daily.

Which of the following medications could be exacerbating this patient's heart failure?

A. Hawthorn
B. Aripiprazole
C. Sertraline
D. Pregabalin

## QUESTION 13

The patient is a 66-year-old woman with trastuzumab-induced cardiomyopathy (LVEF: 40%, NYHA class III, Stage C). Her past medical history is significant for atrial fibrillation, type 2 diabetes, hypertension, and depression. Her medications include lisinopril 20 mg daily, furosemide 40 mg twice daily, carvedilol 12.5 mg twice daily, spironolactone 25 mg daily, and digoxin 0.125 mg daily, apixaban 5 mg twice daily, and metformin 1000 mg twice daily. She has been stable on these medications for the past month. Her most recent laboratory results include sodium (Na) 140 mEq/L, potassium (K) 4.0 mEq/L, chloride (Cl) 105 mEq/L, bicarbonate 26 mEq/L, blood urea nitrogen (BUN) 12 mg/dL, serum creatinine (SCr) 0.8 mg/dL, creatinine clearance (CrCL) 55 ml/min, glucose 70 mg/dL, calcium 9.0 mg/dL, phosphorus 2.8 mg/dL, magnesium 2.0 mEq/L, and digoxin 0.9 ng/mL. Her vital signs today include blood pressure 112/70 mm Hg and heart rate 68 beats/minute. Her lung examination is clear. She will be admitted for elective hip surgery.

Which of the following should be done prior to surgery?

A. Stop the apixaban two days prior to surgery.
B. Stop the metformin seven days prior to surgery.
C. Stop carvedilol three days prior to surgery.
D. Stop atorvastatin two days prior to surgery

## QUESTION 14

Which patient is the best candidate for initiation of ivabradine?

A. 55-year-old man with NYHA functional class HF II (LVEF of 60%) and heart rate 75 beats/minute taking lisinopril 20 mg daily and carvedilol 25 mg twice daily for hypertension.
B. 45-year-old man with NYHA functional class II HF (LVEF of 35%) and heart rate 62 beats/minute taking lisinopril 40 mg daily, metoprolol succinate 150 mg daily, and spironolactone 25 mg daily.
C. 65-year-old woman with NYHA functional class III HF (LVEF of 20%) with heart rate 78 beats/minute taking lisinopril 20 mg daily, eplerenone 50 mg daily, and carvedilol 25 mg twice daily.
D. 85-year-old man with NYHA function class IV HF (LVEF of 10%) with heart rate 110 beats/minute receiving outpatient dobutamine therapy and awaiting transplantation.

## QUESTION 15

A 55 year old man who is status post orthotropic heart transplant returns to clinic for initial follow-up following transplantation. His immunosuppressant regimen consists of tacrolimus 5 mg twice daily (tacrolimus trough: 8 ng/ml), mycophenolate mofetil 1000 mg twice daily , and prednisone 20 mg daily

postoperatively. The patient demonstrates both resting and intention tremors. The tremors began to impact the patient's activities of daily living, requiring supervision for eating and minimal assistance for grooming, bathing, and dressing.

Which of the following interventions would be the most appropriate to treat his tremors?

A. Switch tacrolimus to cyclosporine modified.
B. Reduce the dose of MMF to 750 mg twice daily
C. Add oxycodone 10 mg every 4-6 hours as needed.
D. Add primidone 50 mg daily and titrate to 250 mg/day

## QUESTION 16

The patient is a 35 year old man status post LVAD implantation one year ago who presents to the emergency room with hematuria and epistaxis. He has currently been on warfarin 5mg daily with a goal INR of 2-2.5. His INR in the ED is 10. A urine toxicology drug screen shows the following:

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| Amphet/Metamphetaine: | Positive |
| Barbiturates, Urine: | Negative |
| Benzodiazepines, Urine: | Negative |
| Cannabinoids, Urine: | Positive |
| Cocaine, Urine: | Positive |
| Cotinine: | >20 ng/ml |
| Opiates, urine: | Positive |
| Phencyclidine, urine: | Negative |

Which of the following could be responsible for his elevated INR?

A. Methamphetamine
B. Cannabis
C. Cocaine
D. Opiates

## QUESTION 17

A 68 year old man presents to clinic for evaluation for advanced therapies. He has a past medical history significant for ischemic cardiomyopathy, ventricular tachycardia, and chronic left ventricular thrombus. He denies alcohol, cannabis, and illicit substances and does not take any OTC or herbal medications. He is currently receiving the following medications: amiodarone 200 mg daily, mexiletine 150 mg twice daily, apixaban 5 mg twice daily, omeprazole 20 mg daily, bisoprolol 5 mg daily, sacubitril/valsartan 49-51 mg daily, pregabalin 75 mg daily, rosuvastatin 10 mg daily, spironolactone 25 mg daily. When evaluating the patient's labs, his urine toxicology screen returns positive for methamphetamine. When asked again about use, he admittedly denies methamphetamine use.

Which of the following medications is responsible for this patient's positive methamphetamine test?

A. Omeprazole
B. Pregabalin
C. Mexiletine
D. Spironolactone

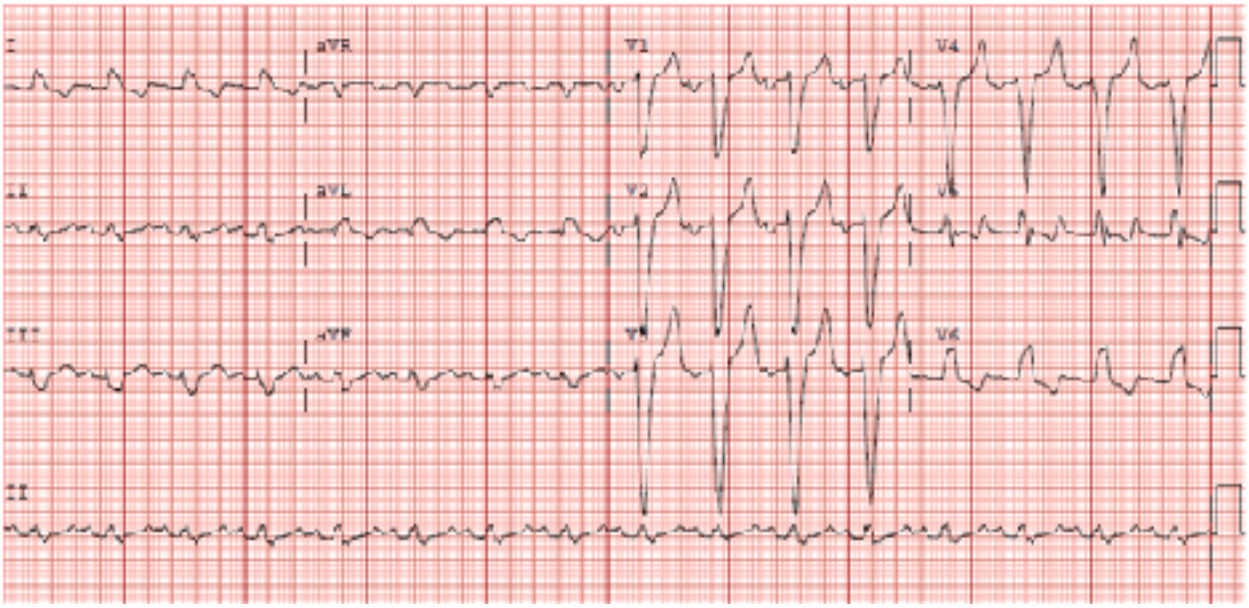
## QUESTION 18

A 68-year-old female with a nonischemic cardiomyopathy (NICM), mild chronic kidney disease (CKD), hypertension, diabetes and obesity is referred to you for evaluation for ventricular assist device (VAD) candidacy.

The patient reports longstanding history of mild dyspnea on exertion (DOE) for the preceding 10 years. In the past year, she noted some worsening of her DOE in addition to some mild orthopnea and fatigue. She was seen by her primary care physician and eventually underwent an echocardiogram (TTE) that revealed a left ventricular end-diastolic dimension (LVEDD) of 6.3 cm, an ejection fraction (EF) of 20% and moderate to severe mitral regurgitation (MR). Coronary angiogram showed no significant coronary artery disease. She was referred to a cardiologist and was initiated on appropriate guideline directed medical therapy (GDMT) with Carvedilol 25 mg twice daily, Entresto 97-103 mg twice daily, and spironolactone 25 mg daily. She was also initiated on maintenance diuretic with Bumex 1 mg daily. With this regimen, her symptoms improved but did not resolve. Repeat TTE after 6 months of GDM T demonstrated an EF of 30% with moderate MR and she was referred to your clinic.

In clinic today, her blood pressure is 103/74 mmHg, heart rate is 68 bpm, oxygen saturation is 98% on room air. Her physical exam is only notable for a JVP of 7 cm of H20 with positive HJR, regular rate and rhythm with III/VI holosystolic murmur radiating to the apex. The remainder of her physical exam is unrevealing. Her blood work is notable for a sodium of 136 mmol/L, potassium of 4.7 mmol/L, creatinine of 1.5 mg/dL. CBC, LFTs and TSH are unremarkable.

EKG shows:




Which is the best next step?

A. Initiate evaluation for ventricular assist device
B. Initiate evaluation for heart transplantation
C. Referral to electrophysiology for a cardiac resynchronization therapy (CRT) with a biventricular defibrillator
D. Referral to interventional cardiology for evaluation for MitraClip

## QUESTION 19

A 52-year-old male presents to his primary care doctor for recurrent episodes of syncope at the gym. TTE reveals a preserved EF, small LV cavity (LVEDD 4 cm) , severe concentric LVH with posterior and septal wall thickness of 1.8 cm and an apical aneurysm. TTE was also notable for mild MR without evidence of systolic anterior motion of his mitral valve. He is referred to his local cardiologist who diagnoses him with hypertrophic cardiomyopathy (HCM) and referred him for ICD implantation given prior unexplained syncopal events and presence of apical aneurysm. He is initiated on metoprolol and continues to do well for several years.

Five years after diagnosis, he developed symptoms of palpitations and dyspnea and presents to his cardiologist after 5 days of symptoms. His JVP is elevated at 15 mmHg and he has 1+ LE edema. EKG reveals atrial fibrillation.

Which of the following anticoagulation strategies should be pursued?
A. Direct-acting oral anticoagulant
B. Left atrial appendage occlusion device
C. Full dose aspirin
D. No anticoagulation is needed

## QUESTION 20

A 73-year-old male with stage D ischemic cardiomyopathy, diabetes, hypertension and hyperlipidemia presents to VAD clinic for a routine visit 12 months after DT HVAD implantation. He reports he is doing well with excellent functional status. He reports poor dietary habits over the prior 6 months and has noted a 15 pound weight gain. He otherwise reports excellent adherence with his medications and has been following with his local doctors routinely.

He reports recently noticing a gradual decrease in his HVAD flows from 5-6 L/min previously to low 3s and has had 1 low flow alarm.

In the office today, examination is notable for a loud VAD hum, JVP estimated at 7 cm of water, no lower extremity edema and driveline appears clean, dry and intact. His blood pressure measured by Doppler is 118/87 mmHg. His HVAD waveforms are notable for a mean flows of ~ 2.8 L/min, peak flows of 6.5L/min and nadir flows of 1 L/min.

His blood work is unrevealing. TTE shows intermittent aortic valve opening, mild RV dysfunction, trivial tricuspid/mitral regurgitation and midline septum.

Which of the following is the best next step to manage this patient?
A. Decrease VAD speed
B. Increase VAD speed
C. Increase bumex dose
D. Initiate losartan

## QUESTION 21

A 32-year-old female status post orthotopic heart transplantation 6 years prior for non-ischemic cardiomyopathy presents to your office for preconception counseling. Her post transplant course has been unremarkable with no episodes of rejection or infection. Donor specific antibodies are negative. She has developed mild hypotension since transplant which is successfully managed with Norvasc 2.5 mg daily. She is maintained on tacrolimus and mycophenolate for immunosuppression. Her regimen otherwise consists of aspirin 81 mg, pravastatin 40 mg, vitamin C and E.

You plan to refer her to maternal-fetal medicine for more detailed preconception counseling and to

establish a multidisciplinary team for her management before conception, during pregnancy and in the postpartum period.

She asks you about management of her immunosuppressive regimen from preconception to postpartum.

Which of the following statement is correct in counseling her?

A. Transition from tacrolimus to sirolimus
B. Discontinuation of mycophenolate mofetil before planned conception.
C. Discontinuation of tacrolimus from first trimester until completion of breast feeding
D. Discontinuation of tacrolimus during the first trimester only.

## QUESTION 22

A 59-year-old man with chronic heart failure and reduced ejection fraction, non-obstructive CAD, systemic hypertension, T2DM and chronic kidney disease stage 3, was hospitalized for an episode of acute decompensated heart failure. He recovered well and you are seeing him for follow up in the office after discharge. His heart failure is compensated now and current medications include: ASA 81 mg/d, LosartanHCT 100/25 mg/d, Rosuvastatin 20 mg/d, Torsemide 20 mg/d, rosiglitazone 4 mg/d and metformin 2,000 mg/d.

Physical exam revealed an oriented patient with BP 117/80 mmHg and HR 80bpm, BMI 33 kg/m2, elevated JVP at 12cmH20, basilar crackles, S3 gallop and 2+ pitting edema of lower extremities. Echocardiogram showed LVEF of 40%, grade 2 diastolic dysfunction, 1+MR and systolic, and estimated PAP of 45 mmHg. CXR showed pulmonary vascular congestion.

Laboratory data: A1C: 8.7%; eGFR: 52 ml/min/1.73m2; fasting glucose 176 mg/dl; Hb 11.4g/dl; LDL cholesterol 78 mg/dl; triglycerides 256 mg/dl.

Which is the best glucose lowering regimen to reduce this patient's A1c to approximately 7.5%?

A. Add dapagliflozin 10 mg/d to rosiglitazone and metformin
B. Add empagliflozin 10 mg/d to metformin 2,000 mg/d and discontinue rosiglitazone
C. Discontinue rosiglitazone and initiate insulin
D. Lower rosuvastatin dose and initiate insulin
E. Discontinue Metformin and start saxagliptin

## QUESTION 23

A 72-year-old patient with end-stage heart failure NYHA class IV with non-ischemic cardiomyopathy was referred to you for evaluation for LVAD therapy. His ECG showed sequential AV pacing at 70/min with BP of 100/54 mmHg. His blood tests showed creatinine 1.3mg/dl, sodium 132 mEq/l and pro-BNP 5,455 pg/ml. Echo doppler revealed markedly dilated LV cavity with LVEF 15%, moderate MR and moderate TR with mildly enlarged RV chamber. Doppler showed an increased E/E' ratio of 20. Right heart catheterization showed RA

pressure of 22mmHg, PCWP of 24mmHg, mean PA of 60/30/45mmHg, cardiac index of 1.8 l/min/m2, and PVR of 4.4 Wood Units.

Which of the following hemodynamic and echocardiographic measures are most predictive of RV failure after LVAD implantation?

A. Severe pulmonary hypertension with enlarged right ventricle
B. RV stroke work index of 558mmHg-ml/m2 and moderate TR
C. Moderate MR and moderate TR with high E/E' ratio of 20
D. RA pressure / PCWP ratio of 0.9 and PAPi (pulmonary artery pulsatility index) of 1.36
E. TAPSE (tricuspid annular plane systolic excursion) of 1.6cm and high E/E' ratio of 20

## QUESTION 24

A 72-year-old woman was referred for evaluation of progressive dyspnea and fatigue. You are the third cardiologist she sees because the etiology of her symptoms remains elusive. She has a long-standing hypertension, dyslipidemia, T2DM, carpal tunnel syndrome. She can no longer climb a flight of stairs without stopping frequently due to gradual worsening of dyspnea on exertion over the last 6 months. She denies orthopnea, leg edema, chest pain or dizziness. Her current medications include Losartan 50 mg/d, amlodipine 5mg/d, atorvastatin 20 mg/d, and metformin 500 mg twice a day.

A previous echocardiogram showed normal LV and RV systolic function, mildly biatrial enlargement, and no valvular pathology. Systolic RV pressure was 34 mmHg and E/E' ratio was 8.5 by doppler. Pulmonary function tests were normal. CXR was unremarkable. Coronary arteries were normal angiographically with LVEDP of 12mmHg.

Physical exam revealed an oriented patient with BP 127/80 mmHg and HR 78bpm, BMI 28 kg/m2, no neck vein distension, clear lungs, regular rate with no cardiac gallop and no leg edema.

Laboratory data: A1C 6.7%; NT-pro-BNP 390 pg/ml; creatinine 0.9 mg/dl; potassium 4.3 mEq/L, sodium 139 mEq/L.

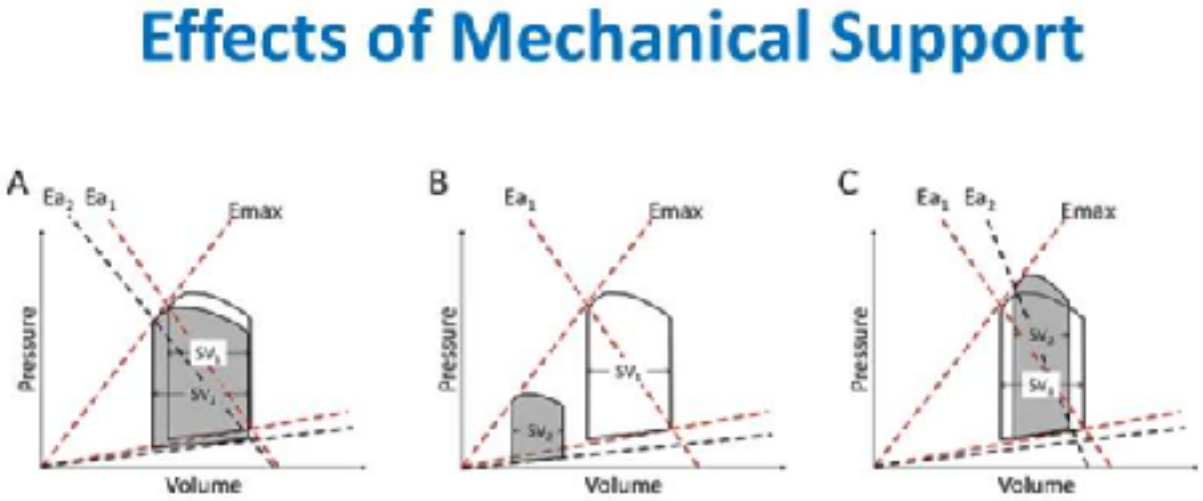
Which of the following tests is most likely to uncover the diagnosis of the patient's exertional dyspnea?

A. Chest computed tomography with PE protocol.
B. Right heart catheterization with 300 ml normal saline volume challenge.
C. High resolution chest computed tomography.
D. Right ventricular endomyocardial biopsy.
E. Right heart catheterization with exercise hemodynamics.

## QUESTION 25

A 45-year-old man with known non-ischemic dilated cardiomyopathy LVEF of 20%, moderate mitral valve

insufficiency, secondary severe pulmonary hypertension, and chronic kidney disease, presented with acute viral respiratory infection. He required mechanical ventilation. Due to refractory hypoxia and complicated cardiogenic shock with hypotension on 3 pressors, you are deciding on a temporary mechanical circulatory support device.




Which set of three devices produces the hemodynamic changes in the pressure-volume loops (gray loops) illustrated in the figure from left to right?

A. IABP, Impella, ECMO
B. Impella, ECMO, IABP
C. IABP, ECMO, Impella
D. ECMO, Impella, IABP
E. ECMO, IABP, Impella

## QUESTION 26

A 51-year-woman underwent heart transplantation for non-ischemic dilated cardiomyopathy 4 years ago. She has had no rejection. Her maintenance immunosuppressive drugs include tacrolimus 3 mg twice a day, mycophenolate mofetil (MMF) 1,000 mg twice a day and prednisone 5 mg po daily. She also takes medications for hypertension and hyperlipidemia. She feels fine and stays well hydrated, but her laboratory data shows serum creatinine 2.3mg/dl, which is a new finding. Fasting glucose level is 106 mg/dL with HbA1c 5.6%. Tacrolimus level is 5.1mg/dl. Annual cardiac catheterization shows normal RHC hemodynamics and only mild intimal hyperplasia in the proximal LAD artery by intracoronary ultrasound, not appreciated angiographically.

Which of the following immunosuppressive drug changes would be most appropriate to decrease risk of further renal dysfunction?

A. Add sirolimus and discontinue tacrolimus.
B. Increase prednisone and increase MMF doses
C. Add sirolimus, discontinue MMF, and decrease tacrolimus dose.
D. Increase tacrolimus dose to suppress cardiac allograft vasculopathy.
E. Continue current regimen and recheck metabolic panel in 3 months.

## QUESTION 27

A 48-year-old woman presented with worsening dyspnea on exertion for the last 4 months. She also reported mild intermittent leg edema, but denied chest pain, palpitations or syncope. Physical exam revealed elevated JVP of 9cmH2O at 45 degrees and 1+ bilateral lower leg edema. The rest of physical exam was normal. The patient stated that she takes Warfarin since an abnormal V/Q scan a year ago. NT-pro-BNP was 2690 ng/L. She was able to cover 240 meters during a 6-minute walk test.

Echocardiogram showed mild RV enlargement, mildly reduced RV systolic function, RVSP of 40 mmHg, and trace tricuspid valve regurgitation. V/Q scan was unchanged. Chest CT angiography showed multiple bilateral segmental and more distal vessel filling defects. RHC revealed RAP 9mmHg, severe pulmonary hypertension 62/28/45mmHg, cardiac index of 2.0 L/min/m2 and PCWP of 12mmHg.

Which therapeutic approach would be the most effective for this patient?

A. Change Warfarin to rivaroxaban
B. Recommend a catheter directed thrombolytic therapy
C. Initiate a soluble guanylate cyclase (sGC) stimulator
D. Initiate dual therapy with sildenafil and treprostinil
E. Refer for surgical pulmonary thromboendarterectomy

## QUESTION 28

A 24-year-old male presents to the emergency room with abdominal pain. He was in his usual state of health until 1 month prior when he developed a viral syndrome. His fever resolved, but he has continued to have malaise and fatigue. In the last week his malaise has gotten worse, but he has also developed abdominal pain and nausea. On physical exam he has a blood pressure of 95/60 and a pulse of 104. Jugular venous distention is present. On cardiac auscultation he has a regular rate and rhythm, a 2/6 systolic murmur and an s3. The lung fields are clear, and peripheral pulses fluctuate. His extremities are warm, and he has 1+ edema.

Which statement is true regarding the patient's physical exam?

A. Lack of pulmonary rales excludes the diagnosis of heart failure.
B. Presence of lower extremity edema is a sensitive and specific sign of heart failure.
C. Presence of jugular venous distention and an S3 indicate that a heart failure diagnosis is likely
D. Fluctuations of the pulse suggest cardiac tamponade.

## QUESTION 29

A 48-year-old male with cardiomyopathy due to a pathogenic mutation in the Titin gene is 3 weeks post LVAD implantation and being prepared for discharge. He had an ejection fraction of 22% prior to LVAD but has no history of ventricular arrhythmia. Repeat echo post discharge demonstrates an ejection fraction of 25%. ECG shows atrial fibrillation with a QRS of 132 msec, and a left bundle branch block pattern.

Which is the best advice for this patient regarding the need for a CIED?

A. The patients should be referred for a CRT device prior to discharge
B. The patient should be discharged with a Life Vest a
C. The patient should be referred for single lead ICD 90 daysafter discharge.
D. ICD at this time is not recommended

## QUESTION 30

A 63-year-old male 4 with a history of diabetes and coronary disease who has undergone implantation of a LVAD 6 months ago, presents in the office with shortness of breath and fatigue with minimal exertion. He is on aspirin 325mg daily and coumadin 3 mg daily. On exam BP is 124/90 mmHg (MAP 101mmHg) measured by an automated cuff. HR is 84 bpm. There is a slightly elevation of JVP and 1+ edema. LVAD Speed is 6000 RPM with a flow of 3.2 L/min and Power of 4.5 Watts. No LVAD alarms are noted. Labs are notable for a Hct of 36%, a creatinine of 1.1 mg/dl and potassium of 4.1.mmol/l

Which of the following should be recommended next?

A. Refer for hemodynamic "Ramp" study
B. Initiation of Lisinopril 5mg daily.
C. Initiation of Hydralazine 50mg TID.
D. Refer for Cardiac Rehabilitation.

## QUESTION 31

Which is correct regarding specialized palliative care in patients with advanced HF?

A. Discussions about code status and goals of care should preferably be initiated by specialized palliative care providers.
B. Specialized palliative care consultation improves cost of care and survival in patients with advanced heart failure.
C. Specialized palliative care consultation improves quality of life measures, in inpatient and outpatient settings.
D. Specialized palliative care consultation is reserved for patients who have elected to defer life prolonging therapies.

## QUESTION 32

A 56-year-old female with history of rheumatoid arthritis presents for evaluation of new onset heart failure. She has a long history of arthritis which is well controlled on daily ibuprofen and hydroxychloroquine as well as intermittent steroids. She has NYHA class III symptoms for the last 2 months. Her ECG is notable for a 1st degree AV block and right bundle branch block. A still image of her echocardiogram is shown in figure 1.

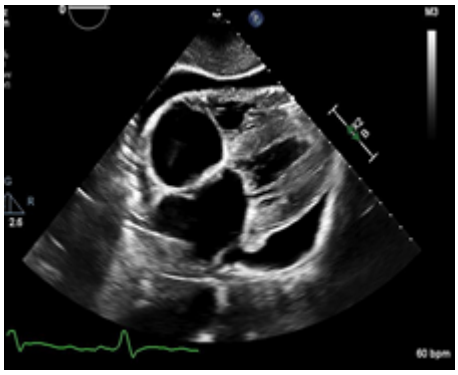


*Figure 1. Parasternal short axis image of patient.*

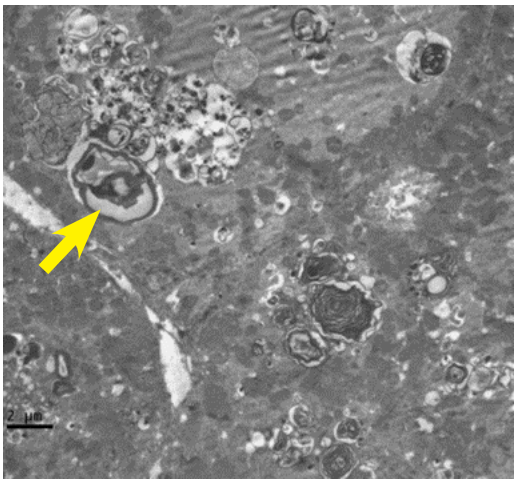


*Figure 2. Electron microscopy of a patient with chloroquine toxicity. Note curvilinear bodies (yellow arrow).*

Which evaluation is most likely to identify the diagnosis?

A. Genetic Testing
B. Cardiac MRI
C. Echocardiography
D. Endomyocardial biopsy
E. No further testing is required

## QUESTION 33

A 49-year-old female 6 days post-transplant develops mental status changes. She had an uneventful transplant and was extubated on post-operative day 2. On the morning of day five she started complaining of headache and was noted to have visual hallucinations. At the time she was afebrile with a blood pressure of 147/96. AM Labs were notable for a Tacrolimus trough level of 14 ng/ml, a WBC of 5.3 $10^9$/L and a creatinine of 4mg/dl. That evening she had a grand mal seizure and needed urgent intubation. The next best diagnostic test for this patient is:

A. Spinal tap.
B. Magnetic Resonance Imaging of the brain.
C. Serum quantitative cytomegalovirus polymerase chain reaction test.
D. Non contrast CT scan of the brain.

## QUESTION 34

The CHAMPION trial investigating the use of the CardioMEMS implantable pulmonary artery pressure monitoring device demonstrated which of the following:

A. Reduction of HF hospitalizations in all NYHA Class III patients.
B. Reduction of HF hospitalizations in only NYHA Class III patients with LVEF <40%
C. Reduction of HF hospitalizations and mortality in NYHA Class III patients.
D. Reduction of HF hospitalizations and mortality in NYHA Class III patients with LVEF <40%.

## QUESTION 35

A 37-year-old African American female presents with shortness of breath with minimal exertion. She also notes weight gain and persistent swelling in her legs after having twins 2 months prior. She complains of bilateral chest discomfort while breastfeeding. She has a history of hypertension preceding pregnancy, gestational diabetes, and obesity. Medications include labetalol and a multivitamin. BP is 132/85, HR 109 bpm, BMI is 33, JVP 13 cm H2O, chest tender to palpation, abdomen obese, and 1+ bilateral edema on exam. ECG demonstrates sinus tachycardia. Echocardiogram reveals severely reduced biventricular function with LVEF 18% and large apical thrombus. Which of the following treatments should be prescribed next?

A. Sacubitril-valsartan
B. Hydralazine/Isosorbide dinitrate
C. Bromocriptine
D. Ivabradine
E. Apixaban

## QUESTION 36

A 45-year-old gentleman with a history of non-ischemic cardiomyopathy presents to the office after recent admission for treatment of acute decompensated heart failure. This was his 3rd hospitalization for HF in the past year. He was previously on guideline directed medical therapy with carvedilol, lisinopril, and spironolactone but these were discontinued while admitted due to hypotension and he was discharged on metoprolol succinate. Despite increasing doses of bumetanide, the patient continues to complain of dyspnea with minimal exertion, orthopnea, PND, and weight gain. Exam is notable for BP 92/78, HR 89, elevated JVP, and warm extremities. ECG demonstrates atrial fibrillation, QRS 94 ms, LVH, and abnormal R wave progression. TTE performed during recent hospitalization revealed severe LV dilatation (LVEDD 6.6 cm), LVEF 23%, normal RV size and function, and severe mitral regurgitation.

Which of the following treatments is not indicated for this patient:

A. Remote pulmonary artery pressure monitoring
B. CRT-D
C. Transcatheter mitral valve repair
D. Left ventricular assist device
E. Cardiac transplantation

## QUESTION 37

Which of the following valve prostheses should be replaced at the time of left ventricular assist device implantation?

A. Bioprosthetic mitral valve
B. Bioprosthetic aortic valve
C. Mechanical mitral valve
D. Mechanical aortic valve

## QUESTION 38

A 56 year-old man with recently diagnosed ischemic cardiomyopathy presents for routine clinic visit. History is notable for anterior myocardial infarction status post PCI, diabetes mellitus type 2 (Hgb A1c 7%), hypertension, and chronic kidney disease. Recent echocardiography showed ejection fraction of 25%. He has dyspnea walking 1 block. Medications include metoprolol succinate 100 mg daily, losartan 100 mg daily, furosemide 80 mg twice daily, aspirin 81 mg daily, atorvastatin 80 mg daily, and insulin. Physical exam is notable for heart rate 66 bpm, blood pressure 110/65 mmHg, regular rhythm, 2/6 holosystolic murmur at the apex, and no signs of excess volume. Labs show creatinine 2.8 mg/dl, which is stable.

Which of the following is the next best action?

A. Start ivabradine
B. Start eplerenone
C. Refer for cardiac rehabilitation
D. Refer for nutritional assessment

## QUESTION 39

A 70 year-old female with history of hypertension, obesity (BMI 38 Kg/m$^2$), and prior smoking (20 pack-years) presents for evaluation of dyspnea. For the last year, she has noted dyspnea and associated chest tightness ascending one flight of stairs or walking up inclines. Her heart rate is 82 bpm and blood pressure 131/83 mmHg. She takes amlodipine 10 mg daily. JVP is difficult to assess due to body habitus. The heart rate is regular with distant S1 and S2 and no murmurs or gallops. The lung fields are without crackles or wheezes. There is trace pedal edema. ECG demonstrates sinus rhythm. Chest x-ray is unremarkable. Echocardiogram shows normal left ventricular wall thickness, chamber size, and ejection fraction of 55-60%. She undergoes cardiac catheterization, showing no coronary artery disease.

Which of the following additional findings would be most consistent a diagnosis of heart failure with preserved ejection fraction?

A. E/e' ratio of 10
B. NT-proBNP of 120 pg/ml
C. Left atrial volume index of 30 ml/m2
D. Pulmonary capillary wedge pressure of 16 mmHg

## QUESTION 40

A 36-year-old man is referred for evaluation and management of hypertrophic cardiomyopathy. Echocardiogram showed asymmetric septal hypertrophy (interventricular septum 2.2 cm, posterior wall 1.2 cm) with ejection fraction of 65-70%, systolic anterior motion of the anterior mitral leaflet, left ventricular outflow gradient of 55 mmHg at rest, 1+MR, and no apical aneurysm. Holter monitor shows no atrial fibrillation, 1% premature ventricular contraction burden, and occasional ventricular couplets. He takes metoprolol XL 100 mg daily. He walks 2 miles per day and swims occasionally with his children without exertional dyspnea or lightheadedness. He has not had syncope. A paternal uncle had diabetes and died of a heart attack at age 49. Otherwise, there is no known family history of cardiac disease.

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

A. Refer for an implantable cardioverter defibrillator
B. Refer for genetic testing
C. Recommend septal myectomy
D. Recommend reduction in physical activity

## QUESTION 41

A 62 year-old man with ischemic cardiomyopathy and diabetes mellitus status post HeartMate 3 LVAS implantation as bridge-to-transplant 1 year ago, listed status 4 for transplant as blood group O, presents with 2 weeks of malaise, fevers, and dull upper abdominal discomfort. He had a driveline exit site infection 2 months ago with methicillin-resistant staph aureus that resolved with oral doxycycline and ciprofloxacin. Abdominal exam is notable for slight erythema and thin yellow drainage at the driveline exit site without induration. VAD hum is normal. Device interrogation reveals normal power and flow at 5000 rpm. Labs are notable for leukocyte count of 14, hemoglobin 11.5 mg/dl, platelets 256, creatinine 1.3 mg/dL. Blood cultures are pending. He is started on intravenous vancomycin and cefepime. CT scan shows a 3.3 x 2.1 x 6.5 cm fluid collection adjacent to the pump pocket with soft-tissue stranding near the internal driveline.

What is the next best treatment?

A. Surgical washout and debridement
B. Targeted intravenous antibiotics for 6 weeks
C. Switch to oral antibiotics if blood cultures are negative
D. Status upgrade for urgent transplant

## QUESTION 42

A 29-year-old female with no significant past medical history presents with gradually progressive dyspnea on exertion over the past two years. She has dyspnea climbing a flight of stairs or walking up inclines, but she can perform activities of daily living without difficulty. Her paternal aunt died at a young age, and post-mortem genetic testing showed a pathologic variant in the BMPR2 gene. Vitals are within normal limits. A complete metabolic panel, complete blood count, and B-type natriuretic peptide level are normal. ECG shows sinus rhythm 68 bpm. She walks 450 meters in 6 minutes. She undergoes right heart catheterization, with results shown below:

Baseline: RA 5 mmHg, PA 65/35 (45) mmHg, PCW 12 mmHg, CO 5.1 L/min, PVR 6.5 Wood units
Nitric oxide (20 ppm for 5 minutes): RA 4 mmHg, PA 50/24 (33) mmHg, PCW 10 mmHg, CO 5.3 L/min, PVR 4.3 Wood units

What would be the next most appropriate treatment?

A. Ambrisentan plus tadalafil
B. Intravenous treprostinil
C. Nifedipine
D. Verapamil

## QUESTION 43

A 61-year-old female with a 5-year history of chronic ischemic cardiomyopathy and left ventricular ejection fraction of 25% is hospitalized for decompensated heart failure. She has generally done well on daily oral diuretic dose of furosemide 80 mg, but was recently discharged from her first heart failure hospitalization 3 weeks prior. Due to increase in serum creatinine from 1.0 to 1.5 mg/dL after in hospital diuresis and achievement of euvolemia, she was discharged on a decreased maintenance oral diuretic dose of furosemide 40 mg daily. She now presents with dyspnea on exertion, mild leg edema, abdominal bloating and an 8 lb. weight gain. During her current admission, she undergoes diuresis with intravenous loop diuretics and optimization of heart failure guideline recommended medical therapy.

Which of the following is the most effective guideline recommended intervention to reduce this patient's risk of 30-day readmission?

A. Transition from oral furosemide to oral torsemide
B. Referral for nephrology evaluation.
C. Reassessment of renal function and electrolytes within 1-2 weeks
D. Follow up appointment with provider within 1-2 weeks

## QUESTION 44

A 25-year-old female former college athlete presents for evaluation of her second episode of chest pain and palpitations occurring within two months of each other. The pain occurs both at rest and with exertion, and

there is some relief with leaning forward. She has a history of childhood asthma, and about three months ago she had an upper respiratory tract infection. She is not taking any medications. The patient has never had syncope or limitations to exercise. A 54-year-old uncle experienced sudden cardiac arrest while mowing the lawn several years ago. The patient's evaluation is notable for an electrocardiogram demonstrating normal sinus rhythm, normal voltage, and T wave inversions in leads V1 and V2. Serum cardiac troponin I is 0.98 ng/dL. Transthoracic echocardiography shows low normal biventricular systolic function without pericardial effusion. Cardiac magnetic resonance imaging demonstrates biventricular late gadolinium enhancement with a right ventricular ejection fraction of 45% and left ventricular ejection fraction of 48%.

What is the highest yield next diagnostic test for this patient?

A. Cardiac fluorodeoxyglucose-positron emission tomography
B. Right Ventricular endomyocardial biopsy
C. Coronary angiography
D. Genetic counseling and testing

## QUESTION 45

A 33-year old female is admitted with a 4-week onset of progressive dyspnea on exertion and leg edema. Her medical history is otherwise notable for several years of a nonproductive cough, a history of obesity and gestational diabetes. On examination, blood pressure is 144/80 mm Hg, heart rate is 65 beats per minute. Jugular venous pressure is 15 mm $H_2O$. Cardiac examination shows regular rate and rhythm without murmurs. Lower extremities have 1+ edema bilaterally.
Electrocardiogram demonstrates sinus rhythm with low-normal voltage and a right bundle branch block. On transthoracic echocardiography she has normal right and left ventricular systolic function, with mitral E to A velocity reversal. The patient is effectively diuresed during hospitalization, with symptomatic improvement, and initiated on an oral diuretic. During her hospitalization she is noted to have several episodes of second degree Mobitz type II heart block.

Which of the following additional tests should be considered in this patient with new heart failure and preserved ejection fraction?

A. Cardiac fluorodeoxyglucose-positron emission tomography imaging
B. Technetium pyrophosphate scan
C. Coronary angiography
D. Genetic counseling and testing

## QUESTION 46

A 70-year old man is admitted with recurrent decompensated heart failure. Medical history is notable for diabetes, stage 2 chronic kidney disease, hypertension, and coronary artery disease status-post coronary artery bypass grafting 6 years ago. The patient now has ischemic cardiomyopathy with left ventricular ejection fraction of 20%. He is a current, every-day smoker. During admission, he has progressive pulmonary

edema and cardiogenic shock necessitating inotropic support. His course is complicated by a stroke resulting in dysarthria with intact fine and gross motor function. During his four-week long hospitalization, he is diuresed and stabilized on milrinone at 0.375 mcg/kg/min, furosemide 80 mg oral twice a day and spironolactone 25 mg oral daily. A right heart catheterization is repeated on current milrinone infusion, revealing blood pressure 94/60 mmHg, right atrial pressure of 8 mm Hg, pulmonary artery pressure of 49/19 mm Hg with pulmonary capillary wedge pressure of 19 mm Hg. Cardiac index is 2.1 L/min/$m^2$. At this time, he demonstrates New York Heart Association class III symptoms, is able to walk without significant dyspnea, but reports fatigue requiring a rest after ambulating 50 feet. Your team discusses next best management steps with the patient and his family. This includes a discussion of risks, benefits and candidacy for durable ventricular assist device and heart transplantation. You express that he has favorable hemodynamics for a durable left ventricular assist device however has contraindications to heart transplantation given ongoing tobacco use. The patient is able to demonstrate understanding of the recommendation for durable left ventricular assist device but is adamant about discharge home today.

Which of the following is the best next step to manage this patient?

A. Discuss continued hospitalization for implantation of durable left ventricular assist device with the patient's healthcare power of attorney
B. Offer later admission for additional evaluation of advanced heart failure therapies
C. Set up a session with the patient to discuss goals of care
D. Refer for neurocognitive evaluation

## QUESTION 47

A 43-year old female patient is status post heart transplantation 2 months ago. Her post-transplant course was notable for treatment of fungal infection with fluconazole for 6 weeks but was otherwise without incident. Her immunosuppression regimen consists of prednisone, tacrolimus and mycophenolate mofetil. Her tacrolimus trough level was maintained in the 10-12 ng/mL range. She now presents with dyspnea, edema and is found to have a newly depressed ejection fraction of 45%. The patient reports a gastrointestinal illness characterized by 2 days of profuse diarrhea which self-resolved last week. She was able to take her immunosuppressive medications during the episode of diarrhea. Her current tacrolimus trough is 4.4 ng/mL.

Which of the following is most likely to decrease tacrolimus levels?

A. Discontinuation of fluconazole
B. Recent diarrheal illness
C. Nonadherence to immunosuppressive medications
D. Inappropriate timing of tacrolimus level assessment

## QUESTION 48

A 56-year-old woman presents to clinic for follow up. She has a five-year history of nonischemic dilated HFrEF (25-30%), diabetes mellitus, hypertension, hyperlipidemia and paroxysmal atrial fibrillation. She has been on stable doses of metoprolol succinate 150mg daily, sacubitril/valsartan 97/103mg twice daily, spironolactone 25mg daily, furosemide 40mg daily, apixaban 5mg twice daily, metformin 1000mg twice daily, and atorvastatin 80mg nightly. She is reporting NYHA class III symptoms. She underwent CRT-D placement and has regular device interrogations with no reported therapies delivered since implant. Last hospitalization for decompensated heart failure was 9 months ago. On examination her blood pressure is 100/68 mm Hg, heart rate is 60 bpm, respiratory rate is 12/min, and oxygen saturation rate is 99% while breathing ambient air. She appears comfortable; there is no appreciable JVD. Her cardiovascular examination is notable for normal rate, regular rhythm, with no murmurs or gallops heard. Lungs are clear, abdomen is soft, and distal extremities are warm to touch.

Which one of the following is the best next step in the management of this patient?

A. Initiate Dapagliflozin
B. Decrease Sacubitril/ Valsartan
C. Refer for CPET
D. Decrease metoprolol succinate to 100mg daily

## QUESTION 49

A 65-year-old man presents to clinic for follow up. He has a seven-year history of nonischemic dilated HFrEF (25-30%) who presents earlier than planned with abdominal discomfort. The patient reports dull and diffuse stomachaches, nausea and vomiting that started 3 days prior. Patient's other medical history includes CRT-D placement for primary prevention, DM, HTN, HLD, obesity. He has been on stable doses of carvedilol 25mg twice daily, sacubitril/valsartan 49/51mg twice daily, spironolactone 25mg daily, furosemide 60mg daily, and rosuvastatin 40 nightly. About a week ago empagliflozin 10mg daily was added to medical therapy. On exam he appears uncomfortable. His blood pressure is 114/56 mm Hg, heart rate is 72 bpm, respiratory rate is 16/min, and oxygen saturation rate is 99% while breathing ambient air. Orthostatic vital signs were normal. There is no appreciable JVD. His cardiovascular examination reveals normal rate, regular rhythm, with no murmurs or gallops heard. Lungs are clear, abdomen is soft, and distal extremities are warm to touch. Patient is sent to emergency department for further evaluation. Laboratory studies reveal sodium of 147 mEq/L, potassium of 4.4 mEq/L, glucose 106 mg/dL, urea nitrogen is 24 mg/dL and creatinine of 1.38 mg/dL. His electrocardiogram and chest X-ray are normal and transthoracic echocardiogram is unchanged from prior.

Which one of the following is the best next step in the management of this patient?

A. Admit patient for evaluation of advanced HF therapies
B. Obtain urine and serum ketones
C. Obtain Gi consult
D. Admit for IV fluids

## QUESTION 50

A 60-year-old man with ischemic cardiomyopathy and left ventricular ejection fraction of 20-25% 9 years post SC-ICD, history of VT ,paroxysmal atrial fibrillation/flutter status/post ablation, hypertension, hyperlipidemia, CKD stage 3b, presents to emergency room with fatigue, sweats, and dyspnea. Patient reports adherence to recommended fluid and salt restrictions and medication regimen which includes: apixaban 5 mg twice daily, amiodarone 200 mg daily, carvedilol 12.5mg twice daily, lisinopril 20mg daily, and spironolactone 25mg daily. On examination his temperature is 100.1oF, blood pressure is 117/55 mm Hg, heart rate is 114 bpm, respiratory rate is 14/min, and oxygen saturation rate is 98% while breathing ambient air. BMI is 38kg/$m^2$. He is diaphoretic, anxious appearing; there is no appreciable JVD. His cardiovascular, pulmonary and abdominal examinations are unremarkable. Distal extremities are warm to touch. Laboratory studies reveal sodium of 138 mEq/L, potassium of 4.0 mEq/L, glucose 106 mg/dL, creatinine of 1.63 mg/dL white blood cell count of 10.38 K/uL, and hemoglobin of 10.0 g/dL, ferritin 300 µg/L. Transthoracic echocardiogram shows a stable left ventricular ejection fraction of 20-25%. However right ventricular dilation of 5.3cm with reduced function on visual assessment is now present. TAPSE is measured at 0.8cm, lateral s' is 0.08m/sec. IVC is normal in size with blunted respirophasic diameter changes. Right heart catheterization shows RA 12/12/8, RV 74/7/15, PA 76/32/47, PCWP 17/16/14, Fick CO 10.3L/min, CI 5L/min/m2, without shunt present, SVR 640 dynes, PVR 3.2 Wood's units.

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

A. Obtain transferrin saturation
B. Refer for infectious disease evaluation
C. Refer patient to bariatric clinic
D. Obtain Thyroid Function Tests

## QUESTION 51

A 47-year-old man with history of HTN, prior tobacco use and non-small cell lung cancer presented to ER with 2-day history of worsening fatigue, malaise, dizziness, shortness of breath and palpitations. In the emergency room patient's vital signs are: heart rate of 120 beats per minute, blood pressure is 90/47 mmHg, and respiratory rate is 28/min. On examination he is ill appearing, sitting up, crackles are noted to mid lung fields bilaterally. Cardiovascular exam reveals regular rhythm, 120 beats per minute, and an S3 gallop. Chest Xray is notable for vascular congestion changes. During the patient's examination salvos of non-sustained ventricular tachycardia are noted on telemetry.

Initiation of which medication can be associated with patient's current presentation?

A. Carboplatin
B. Pamextrexed
C. Pembrolizumab
D. Gemcitabine

## QUESTION 52

A 45-year-old man presents with concerns about personal risk for heart failure because his father was diagnosed with HF after a myocardial infarction at age 72. His medical history includes HTN and DM. Medications include metformin 1000mg twice daily, lisinopril 10mg daily, and amlodipine 10mg daily. Patient smokes ½ pack per day for 15 years, but does not drink alcohol or use recreational drugs. He reports he does not exercise on a regular basis but walks 5 blocks and climbs 3 flights of stairs with no activity related limitations. On examination patient vital signs are: blood pressure is 148/78 mmHg, heart rate is 92 beats per minute, respiratory rate is 16/min, oxygen saturation 98% on room air, BMI 39 Kg/m$^2$. Patient appears comfortable, lungs are clear. Cardiovascular exam is normal. Pulses are palpable in all extremities.

In addition to intensifying anti-hypertensive regimen, counseling on tobacco cessation and lifestyle modifications, which is the next appropriate step in management of this patient?

A. Obtain BNP or NT-proBNP
B. Refer for weight loss surgery
C. Obtain an exercise treadmill test
D. Refer for a computerized tomography of the chest

## QUESTION 53

A 66-year-old man with history of nonischemic HFrEF (EF 10-15%) s/p implantation of a dual chamber ICD is admitted to the hospital with acutely decompensated HF. Patient's other medical history includes osteoarthritis. Social history includes remote tobacco use and occasional ETOH intake. In emergency room patient's vital signs are: heart rate of 104 beats per minute, blood pressure is 92/58 mmHg, and respiratory rate is 24/min. On examination he is ill appearing, sitting up, crackles are noted to mid lung fields bilaterally. Cardiovascular exam reveals a heart rate of 114 beats per minute, regular rhythm with S3 gallop present. Abdominal exam is notable for palpable liver. Extremities are cool on exam. Chest Xray is notable for vascular congestion. ECG shows sinus tachycardia, RBBB, QRS 146msec. Labs are significant for sodium of 132 mmol/L, potassium of 4.5mmol/L, chloride 93mmol/L, BUN 9mg/dL, creatinine 0.8 mg/dL, albumin 2.8 g/dL, INR 2.2 sec, hemoglobin 11.6 g/dL, platelets of 89. Patient is given furosemide 80mg IVP with minimal urine output, he becomes more lethargic and is started on dobutamine 5mcg/kg/min. His blood pressure improves to 110/64, HR is 128 bpm, and mental status improves as dose urine output and respiratory status.

What is the next best step in patient management?

A. Referral for evaluation for OHT
B. Referral for upgrade to CRT
C. Referral for palliative care
D. Referral for durable LVAD implantation

## QUESTION 54

Which of the following drugs decreases tacrolimus levels?

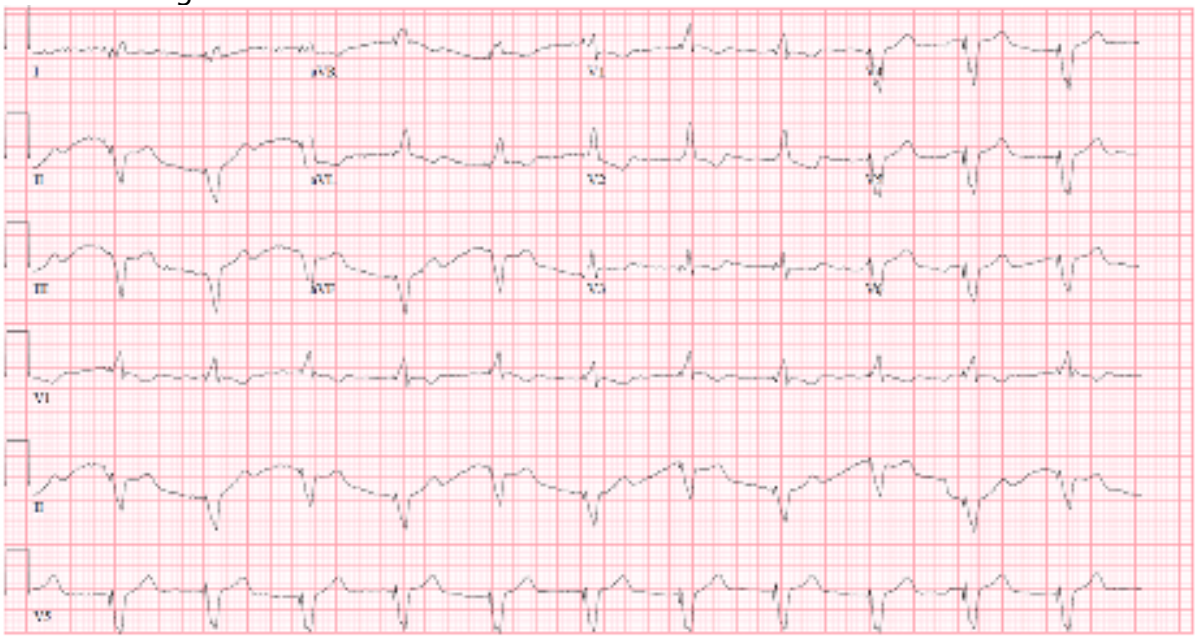
A. Ketoconazole
B. Gemfibrozil
C. Diltiazem
D. Fluoxetine

## QUESTION 55

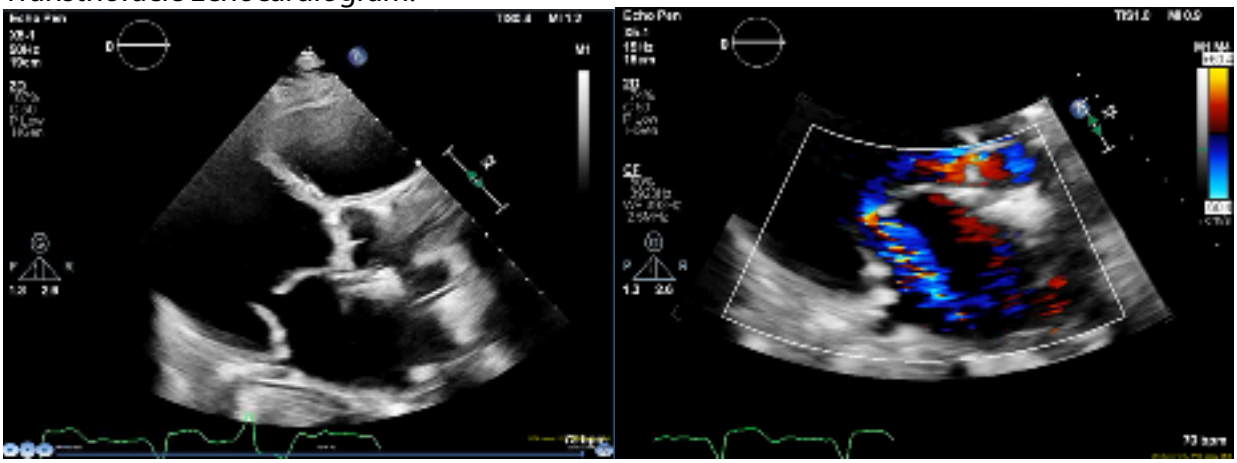
A 79-year-old man with permanent atrial fibrillation, hypertension, nonischemic cardiomyopathy with a left ventricular ejection fraction partially recovered to 40%, and biventricular implantable cardioverter-defibrillator presents to your office with progressive shortness of breath over the last month while ascending one flight of stairs. Prior to this he was well compensated. Medications include carvedilol 6.25 mg twice daily, sacubitril/valsartan 49/51 mg twice daily, spironolactone 25 mg daily, dapagliflozin 10 mg daily, torsemide 20 to 40 mg daily, and apixaban 5 mg twice daily. Recent coronary angiography demonstrates no obstructive coronary disease. He has been unable to tolerate further uptitration of medical therapy because of hypotension.

On examination, his blood pressure is 95/74 mmHg, heart rate is 72 beats per minute, respiratory rate is 16 respirations per minute, and oxygen saturation is 98% on room air. He exhibits no jugular venous distension. Cardiac examination demonstrates a laterally displaced point of maximum impulse, and a III/VI holosystolic murmur best heard at the apex. There is no peripheral edema. The remainder of the examination is benign.

Electrocardiogram:



Transthoracic Echocardiogram:



Transesophageal echocardiography demonstrates a mitral regurgitant orifice area of 72 $mm^2$, a regurgitant volume of 65 ml, and a vena contracta jet width of 9 mm.

Which of the following should be considered as a next step?
A. Pursue echocardiographic dyssynchrony study
B. Implantation of a pulmonary artery pressure monitor
C. Implantation of a left atrial appendage occlusion device
D. Initiate transcatheter edge-to-edge repair (TEER) of the mitral valve evaluation
E. Initiate ventricular assist device evaluation

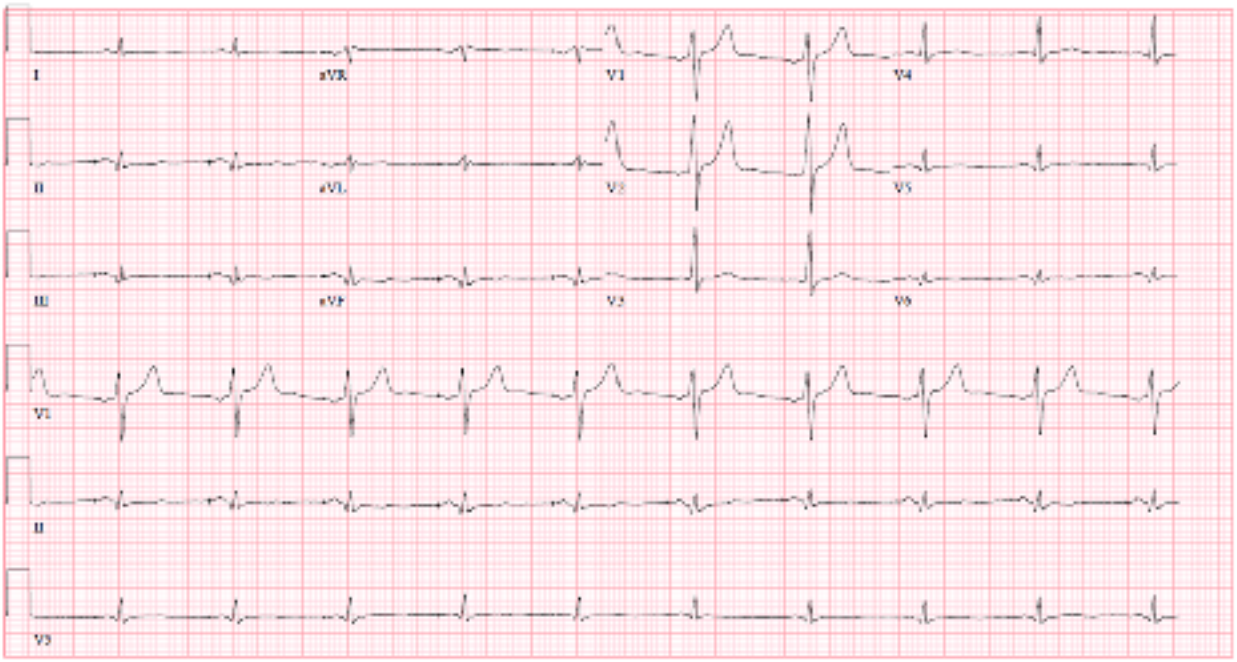
## QUESTION 56

A 50-year-old man comes to your office for routine follow-up. He has a history of dyslipidemia, coronary artery disease, and is status-post primary percutaneous intervention for left circumflex territory myocardial infarction two years ago. One year ago, he experienced stent thrombosis while taking high doses of non-steroidal anti-inflammatory drugs for gout. This resulted in a large circumflex territory infarct, was complicated by ventricular fibrillation arrest, and resulted in placement of a dual chamber implantable cardioverter-defibrillator. He feels generally well, though while walking into his home with a bag of groceries he gets short of breath.

Medications include atorvastatin 80 mg daily, aspirin 81 mg daily, metoprolol succinate 200 mg daily, sacubutril/valsartan 97/103 mg twice daily, spironolactone 25 mg daily, and ticagrelor 90 mg twice daily.

On examination, blood pressure is 112/72 mmHg, pulse is 60 beats per minute. Examination demonstrates no jugular venous distension, a PMI that is minimally displaced laterally from the mid-clavicular line, a soft I/VI systolic murmur, and no peripheral edema. The remainder of the examination is normal.

Electrocardiogram:




Echocardiography demonstrates a moderately dilated left ventricle, with an LV internal diameter in systole at 5.9 cm. Ejection fraction is calculated at 32%, with an akinetic inferior and inferolateral wall. There is trivial mitral regurgitation.
Laboratory studies reveal a GFR of 65 ml/min, and a BNP of 65 pg/ml.

Which of the following is the best next step?
A. Start dapagliflozin 10 mg daily.
B. Refer for upgrade of ICD to dual chamber ICD.
C. Start ivabradine 5 mg twice daily.
D. Add lisinopril 10 mg daily.

## QUESTION 57

A 34-year-old man comes to clinic for cascade genetic screening, as his father had been recently diagnosed with a Lamin A/C gene mutation. The patient is asymptomatic and feels generally well. He is active, jogging 30 minutes five times a week. He takes no medications. His blood pressure is 114/68 mmHg, heart rate is 78 beats per minute. Examination is unremarkable.

Electrocardiography demonstrates normal sinus rhythm, and first-degree AV block, with a PR interval of 280 ms. Echocardiography demonstrates a mildly dilated left ventricle, and an ejection fraction of 42%. The patient's genetic testing indicates the same mutation in LMNA (the gene encoding Lamin A/C) as his father.

Which of the following represents the best next step?
A. Initiate metoprolol succinate 25 mg daily.
B. Continue surveillance of left ventricular ejection fraction with repeat echocardiography in one year.
C. Institute home blood pressure monitoring.
D. Recommend placement of implantable cardioverter-defibrillator.

## QUESTION 58

A 22-year-old college rower presents for three-month follow-up after an episode of severe viral myocarditis. On initial presentation he had had left sided pleuritic chest pain, headache, and malaise. His troponin I peaked at 22.4 ng/ml, echocardiogram demonstrated LV ejection fraction of 55%, and in hospital monitoring disclosed a 5-beat run of nonsustained ventricular tachycardia. Electrocardiogram demonstrated diffuse ST-elevations. He was initiated on metoprolol tartrate 25 mg twice daily, which you discontinued two months ago. He remains on colchicine 0.6 mg twice daily. Ibuprofen was started initially and is now tapered off.

He currently feels well and is eager to return to training.

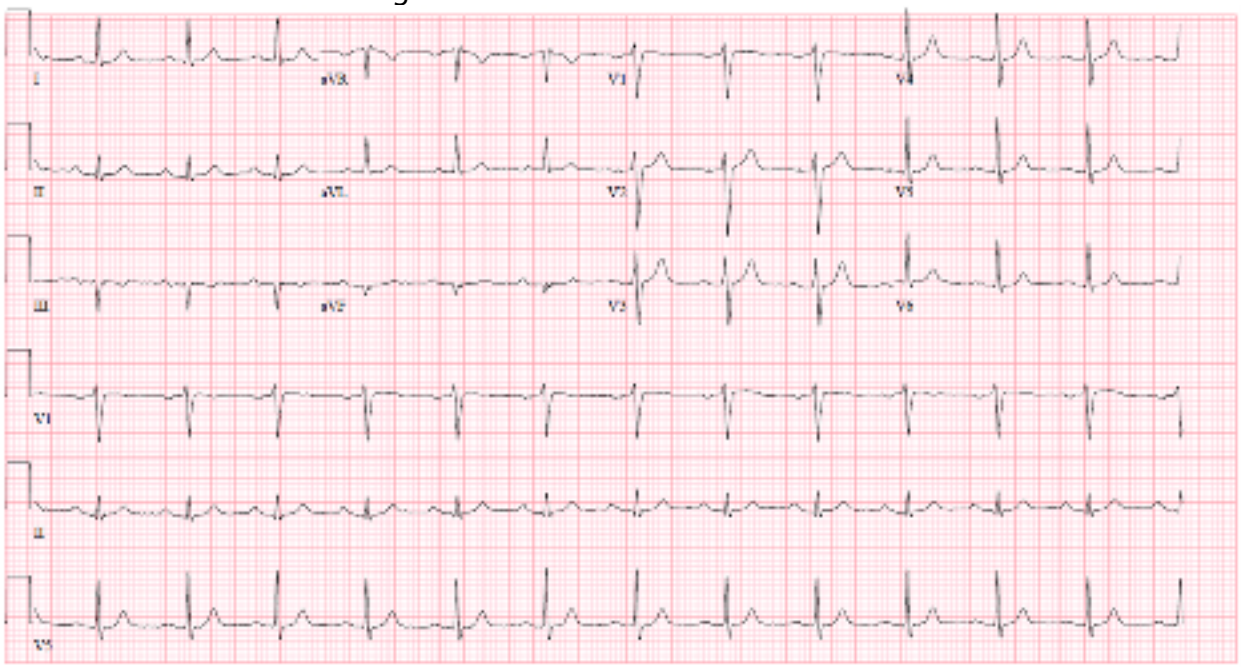
What should you advise?
A. He may return to play without restrictions.
B. Restart metoprolol tartrate 25 mg twice daily.
C. Obtain a stress electrocardiogram.
D. Perform cardiac magnetic resonance imaging.
E. Advise permanent restriction from training and competition.

## QUESTION 59

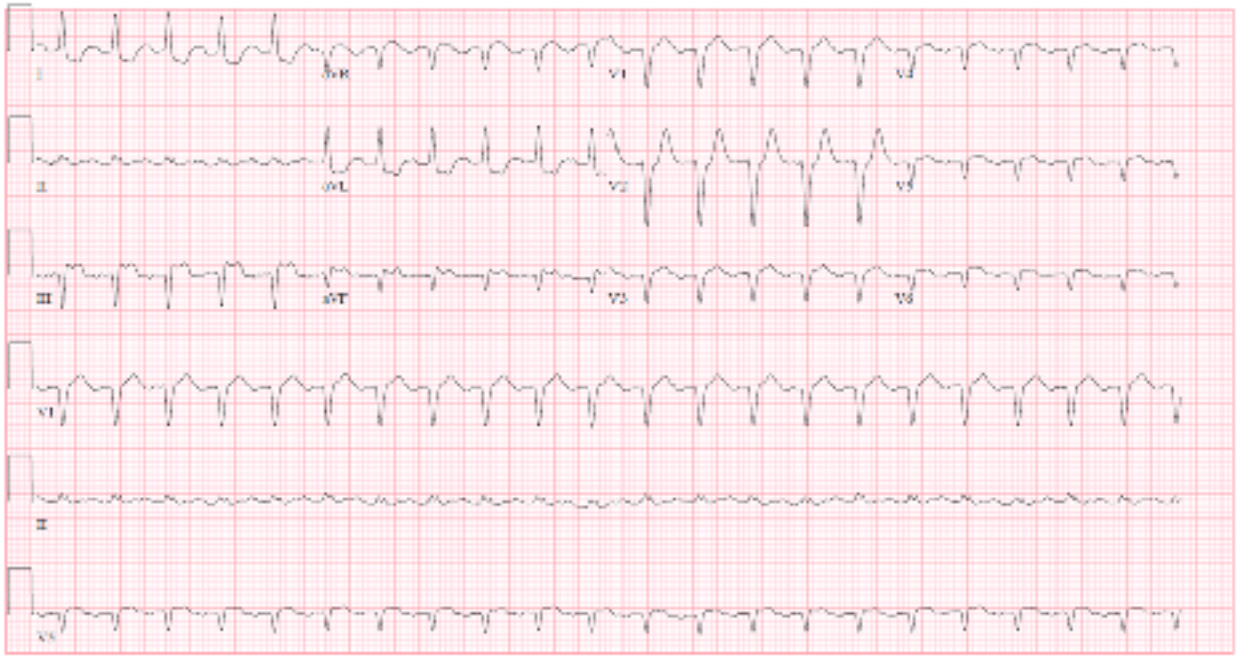
A 78-year-old man with a past medical history of coronary artery disease status-post three vessel CABG, COPD, and high-grade metastatic prostate cancer presents to hospital with progressive dyspnea on exertion and orthopnea. His prostate cancer had been treated with androgen deprivation therapy and enzalutamide until six weeks prior to admission, when he was started on pembrolizumab, an immune checkpoint inhibitor. Echocardiography the day of admission demonstrates a deterioration in left ventricular ejection fraction from 65% to 25% in the span of three months.

Laboratory evaluation reveals high-sensitivity troponin I rising to a peak of 44,802 pg/ml.

Baseline ECG from 2 months ago:



ECG on presentation:



Which of the following should be considered as a next step in this patient's management plan?
A. Empiric treatment with prednisone 10 mg daily.
B. Technetium-99 m pyrophosphate scintigraphy
C. Endomyocardial biopsy
D. Empiric treatment with nonsteroidal anti-inflammatory drugs

## QUESTION 60

A 68-year-old man with a history of ACC/AHA Stage D nonischemic cardiomyopathy, heart failure with reduced ejection fraction (HFrEF), type II diabetes mellitus, hypertension, obesity (body mass index 34 kg/m$^2$) presents to the emergency room with acute decompensated heart failure. He reports exertional dyspnea, orthopnea, and 9 lb. weight gain over the past 2 weeks. He has had two hospitalizations for acute decompensated heart failure for the past year. His medications include torsemide 100 mg twice daily, metoprolol succinate 12.5 mg daily, dapagliflozin 10 mg daily, sacubitril-valsartan 24-26 mg twice daily, spironolactone 12.5 mg daily, and metformin 1000 mg twice daily. Physical examination reveals blood pressure (BP) 90/65 mm Hg, heart rate (HR) 115 bpm, jugular venous pressure 12 cm H2O, hepatojugular reflux, +S3 gallop, bilateral basal crackles, and 2+ lower extremity edema. Laboratories reveal sodium 132 mEq/L, blood urea nitrogen (BUN) 50, creatinine (Cr) 2.2 mg/dL, and potassium 5.2 mEq/L. Bedside echocardiography reveals left ventricular end-diastolic dimension 7.5 cm, ejection fraction 15%, moderate-severe centrally directed mitral regurgitation, and estimated right ventricular systolic pressure 40 mm Hg plus 15 mmHg right atrial pressure.

He is admitted to the cardiac intensive care unit and initiated on continuous infusions of furosemide titrated to 20 mg/hour and milrinone titrated to 0.375 mcg/kg/minute. Over the next 24 hours, his urine output is 2.5 liters but his BP decreases to 80/60. Furthermore, titration of milrinone results in frequent salvos of non-sustained ventricular tachycardia on continuous telemetry monitoring. Repeat labs demonstrate sodium 130 mEq/L, potassium 5.5 mEq/L, BUN 64, Cr 2.5 mg/dL. The patient has advanced directives, indicating he would want "reasonable measures to preserve a good quality of life."

Which of the following is the most appropriate next step in his management?
A. Refer for percutaneous mitral valve repair.
B. Refer for right heart catheterization.
C. Refer for implantable pulmonary artery pressure sensor.
D. Refer for initiation of continuous renal replacement therapy.
E. Refer for palliative care consultation.

## QUESTION 61

A 66-year-old woman with morbid obesity, hypertension, type II diabetes mellitus, hyperlipidemia, and obstructive sleep apnea on continuous positive airway pressure therapy, presents with progressive exertional dyspnea, orthopnea, and lower extremity edema over the last 6 days. The patient has been

admitted three times in the last twelve months for acute decompensated heart failure. Home medications include aspirin 81 mg daily, atorvastatin 40 mg daily, long-acting insulin 30 units nightly, bumetanide 4 mg twice daily, carvedilol 25 mg twice daily, and losartan 50 mg daily.

On examination, blood pressure is 150/90 mm Hg, heart rate is 90 beats per minute. Body mass index is 42 kg/m$^2$. Jugular venous pressure is 14 cm H2O. There are decreased breath sounds at the base bilaterally. Cardiac examination is regular rate and rhythm without murmurs or rubs. S4 is present. Lower extremities have 2+ pitting edema bilaterally. Labs on admission showed sodium 133, potassium 4.5, blood urea nitrogen 38, serum creatinine 1.9 mg/dl. Echocardiography reveals left ventricular ejection fraction of 65% without significant valvular disease, consistent with findings from the prior study 3 months ago.

Which of the following interventions is most likely to decrease rehospitalization for heart failure?

A. Transition from bumetanide 4 mg twice daily to torsemide 40 mg twice daily.
B. Transition from carvedilol 25 mg twice daily to metoprolol succinate 100 mg daily.
C. Addition of spironolactone 25 mg daily.
D. Transition from losartan 50 mg daily to sacubitril-valsartan 49-51 mg twice daily.
E. Addition of sildenafil 20 mg three times daily.

## QUESTION 62

A 22-year-old man with newly diagnosed hypertrophic cardiomyopathy is referred by his general cardiologist to your clinic for further evaluation. He is able to walk several blocks and climb 2 flights of stairs without limitation. He is disappointed that he can no longer participate in his competitive recreational soccer league. Upon further questioning, he denies any chest pain, orthopnea, paroxysmal nocturnal dyspnea, palpitations, lightheadedness or syncope. He does not recall any family members who have had sudden cardiac death, although his maternal grandfather died of unknown causes at age 70. On examination, his vital signs are blood pressure 120/80, heart rate 80, respiratory rate 22, and pulse oximetry of 94% on room air. His exam is notable for III/VI systolic murmur at the apex but otherwise unremarkable.

He had an echocardiogram performed 2 weeks ago which showed left ventricular systolic function of 55% without apical aneurysm, left ventricular end diastolic dimension of 55 mm, interventricular septum of 27 mm, and posterior wall of 19 mm. His resting left ventricular outflow tract gradient is 30 mmHg and increases to 50 mmHg with provocation. A cardiac MRI shows left ventricular ejection fraction of 42%, right ventricular ejection fraction of 50%. You order an ambulatory event monitor which shows some occasional premature ventricular contractions but no episodes of nonsustained or sustained ventricular tachycardia.

Which of the following is the best next step in managing this patient?

A. Start verapamil 120 mg daily.
B. Refer for alcohol septal ablation.
C. Start metoprolol succinate 25 mg daily.
D. Refer for implantable cardioverter defibrillator placement.
E. Refer for septal myectomy.

## QUESTION 63

A 65-year-old woman status post heart transplantation presents for her tenth annual office visit with progressive shortness of breath. The patient denies any hospitalizations for acute decompensated heart failure and states adherence to a low salt, low fluid diet and her immunosuppression medications. Her routine annual testing is reviewed.

12-lead electrocardiogram reveals normal sinus rhythm at 75 beats per minute with a nonspecific intraventricular conduction delay of 120 milliseconds. Transthoracic echocardiogram shows a left ventricular end-diastolic dimension of 6.0 cm and a left ventricular ejection fraction of 45% with anterior and apical wall motion abnormalities and mild to moderate mitral regurgitation. She is referred for coronary angiography, which reveals 70% proximal stenosis in the left anterior descending artery, 80% stenosis in the mid left circumflex artery, and 70% stenosis in the proximal right coronary artery with diffuse and distal disease noted in all three vessels. Current medications include atorvastatin 80 mg daily, tacrolimus 2 mg twice daily (recent trough level, 6.8; target level 5-8) and mycophenolate 500 mg twice daily. She has had no history of rejection episodes and was weaned off corticosteroids at the end of the first year after heart transplantation.

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

A. Switch tacrolimus to everolimus.
B. Refer for multivessel percutaneous coronary intervention.
C. Begin evaluation for cardiac re-transplantation.
D. Refer for coronary artery bypass grafting.
E. Switch atorvastatin to rosuvastatin.

## QUESTION 64

A 51-year-old woman is being evaluated for gradually progressive dyspnea on exertion over the past year. She has no known prior history of hypertension, cardiac or lung disease. She was never a smoker. She is not on any medications. She now has to stop after walking half a block and can no longer climb stairs in her home without resting periodically.

On examination, her blood pressure is 120/80 mm Hg, heart rate is 90 bpm, respiratory rate is 16 breaths/min, and oxygen saturation is 93% RA. Her jugular venous pressure is 8 cm H2O. Cardiac examination is unremarkable except for a widely split $S_2$ and loud $P_2$. Lungs are clear to auscultation. There is 1+ bilateral

pedal edema.

An echocardiogram shows normal left ventricular ejection fraction, but a moderately dilated right ventricle (RV) with moderate RV systolic dysfunction. The interventricular septum is D-shaped. The pulmonary artery systolic pressure is estimated at 50 mm Hg. She is referred for a ventilation/perfusion (V/Q) scan, which demonstrates bilateral segmental mismatched perfusion defects. The patient is started on therapeutic anticoagulation and then referred for definitive surgical management.

Which one of the following peri-procedure hemodynamic parameters is associated with the highest risk of perioperative mortality?

A. Central venous pressure (CVP)/Pulmonary capillary wedge pressure (PCWP) ratio of 0.6.
B. Mean pulmonary artery pressure of 50 mm Hg.
C. Pulmonary vascular resistance of 1200 dyn·s·cm–5
D. Pulmonary artery pulsatility index of 1.9.
E. Right ventricular stroke work index of 400.

## QUESTION 65

A 29-year-old woman with a history of NYHA class 2 chronic systolic heart failure, diabetes mellitus, and hypertension is seen in heart failure clinic. 9 months ago, she was diagnosed in the office with asymptomatic rapid atrial fibrillation. She did not tolerate dose escalation of beta-blockers and was referred for pulmonary veins isolation one month after diagnosis. She was lost to follow up due to social stressors. On returning, she explains that she is now 5 months pregnant. Current medications include sacubitril-valsartan, bisoprolol, spironolactone, amiodarone, warfarin and furosemide. Interrogation of her ICD shows no breakthrough atrial fibrillation since her PVI. Her renal function, liver function, INR, and thyroid function tests are normal.

Which pharmacologic agents should be discontinued at this time?

A. bisoprolol, furosemide and warfarin
B. spironolactone and warfarin
C. spironolactone and amiodarone
D. warfarin alone
E. amiodarone and warfarin

## QUESTION 66

The patient is a 45-year-old man with a PMH of HTN, obesity, diabetes mellitus and paroxysmal atrial fibrillation presenting for evaluation of slowly progressive severe dyspnea on exertion for 1 year. He has had many presentations with rapid atrial fibrillation and to date has been treated with multiple anti-arrhythmics and 4 atrial fibrillation ablations. At this time, recent ambulatory event monitors suggest no breakthrough atrial fibrillation within the last year. Surface echo showed a preserved EF, normal wall thickness, biatrial enlargement, and grade II diastolic dysfunction without significant resting valvular abnormalities. A

myocardial spect is normal. A cardiac CTA and cardiac pulmonary vein CT are normal. A VQ scan is unremarkable. Moderate dose diuretics are offered without relief and cause orthostasis. He underwent an exercise left and right heart catherization which included a transseptal puncture to measure left atrial pressure directly. The values from this study are below:

| | HR | BP | RAP | mPAP | LAP | LVEDP | V wave |
|---|---|---|---|---|---|---|---|
| Rest | 73 | 120/80 | 2 | 18 | 9 | 7 | 16 |
| Moderate exercise (30 w) | 86 | 140/70 | 19 | 50 | 35 | 15 | 44 |
| Peak exercise (70 w) | 110 | 160/55 | 21 | 60 | 38 | 17 | 48 |

What is the most likely etiology of this patient's symptoms?

A. Dynamic mitral valve stenosis
B. Pulmonary vein stenosis
C. Stiff left atrial syndrome
D. idiopathic heart failure with preserved ejection fraction
E. cardiac amyloidosis
F. idiopathic exercise induced pulmonary hypertension

## QUESTION 67

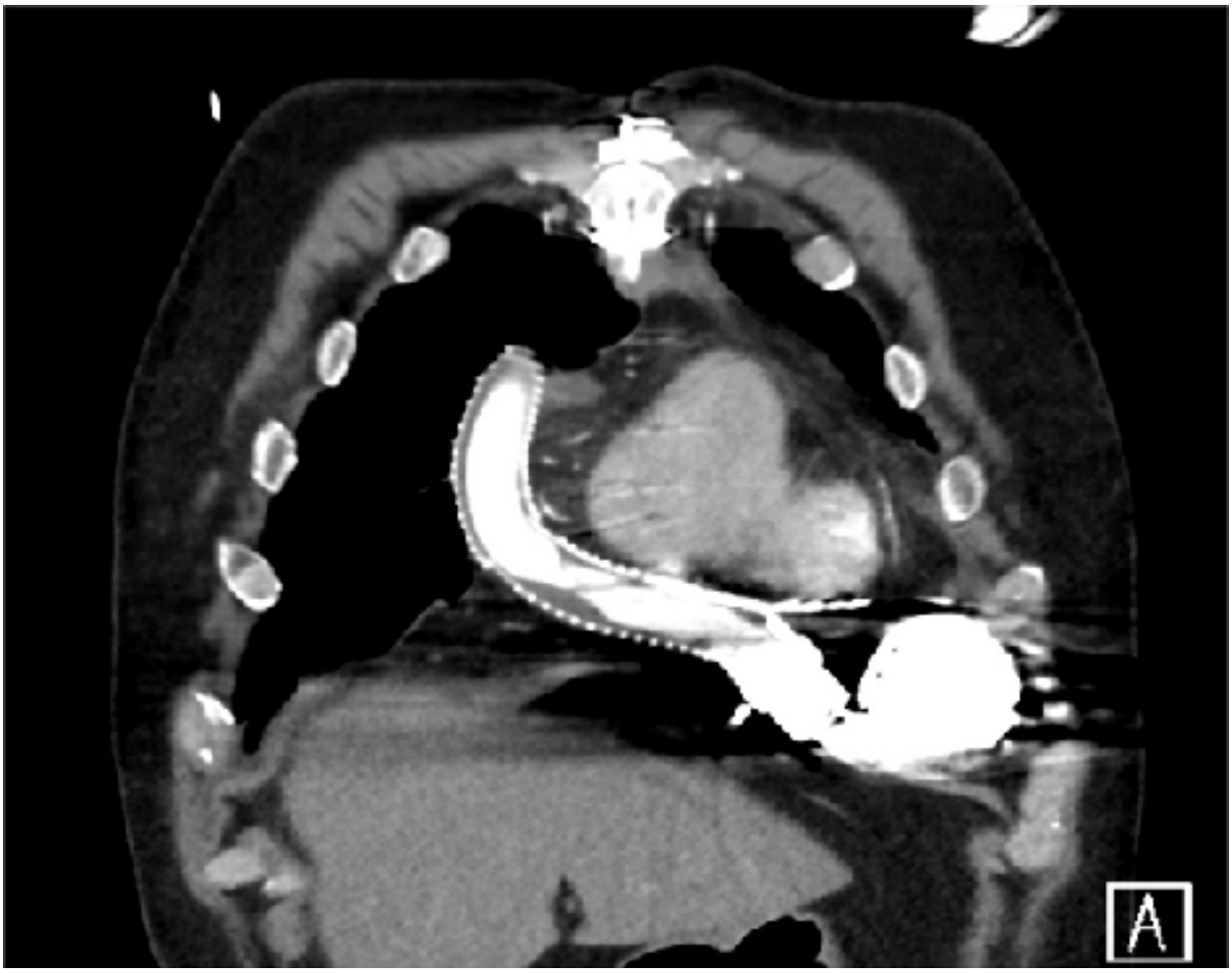
A 20-year-old woman presents to your clinic. She states her father was recently diagnosed with hypertrophic cardiomyopathy at age 50 after presenting with aborted sudden cardiac death. She is interested in knowing whether she and her future children will develop the disorder and is willing to get tested as needed. She and her father are somewhat estranged and she does not like him. However she thinks she will be able to get him to get whatever testing is needed for the sake of his future grandchildren. Her surface echocardiogram is normal. Her father undergoes genetic testing which does not identify a known sarcomeric gene mutation associated with HCM

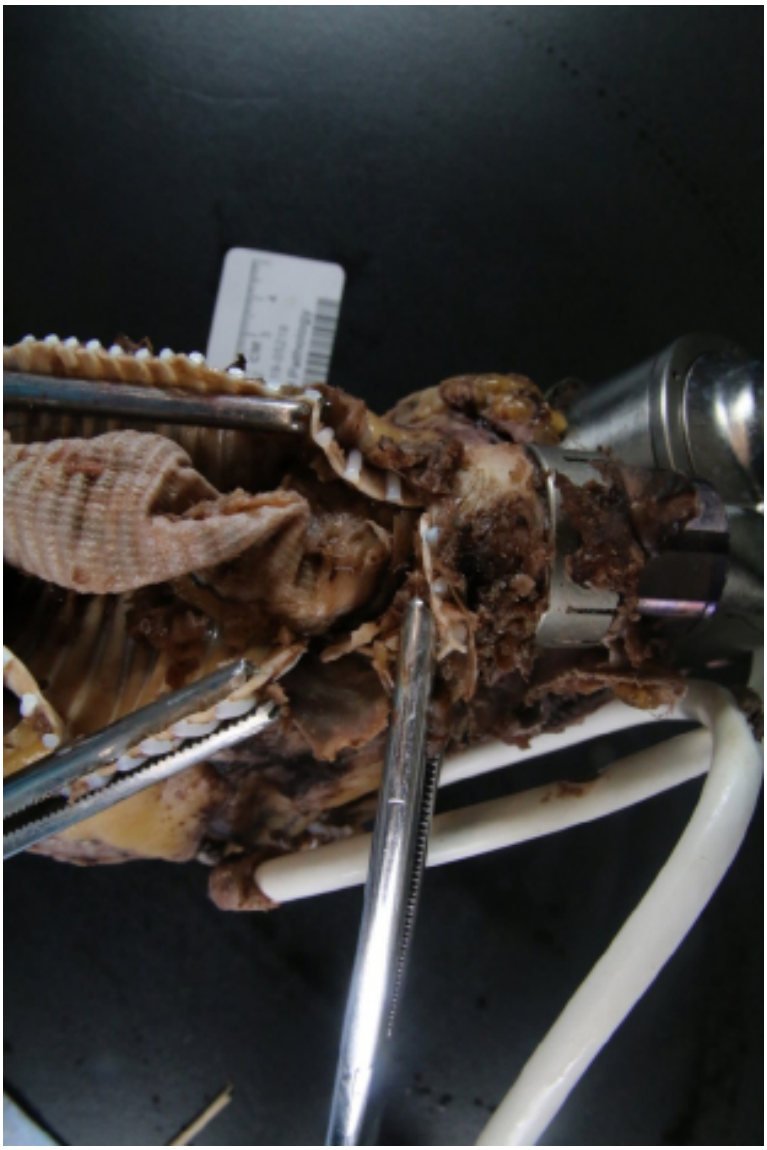
What should you recommend to her?

A. Her echocardiogram is normal and she is unlikely to develop the disease in her lifetime
B. She should undergo genetic testing
C. She should ask her father to repeat genetic testing
D. She should undergo clinical evaluation with ECG and echocardiograms every 5 years starting now
E. She should undergo clinical evaluation with ECG and echocardiograms every 5 years starting at age 40.

## QUESTION 68

A 60 year old patient received a continuous flow LVAD 4 years ago for Stage D non-ischemic cardiomyopathy. He is currently admitted for volume overload and worsening heart failure. . In the last 24 months he has had 4 admissions for decompensated heart failure. The left ventricle has progressively dilated during this time despite repeated increases in LVAD speed. TEEs have shown only trace aortic regurgitation. During this admission the patients is notd to have new low flow alarms. LDH is approximately 370 u/L (about 1.5 his baseline value) and RHC shows a wedge pressure of 30 mmHg and cardiac index of 1.7 L/min/m2, which did not improve increasing of LVAD pump speed by 600 RPM. A CTA is performed and shown below. The patient ultimately undergoes a LVAD pump exchange and an image of his explanted device is below





What is the etiology of the patient's pump dysfunction?

A. Worsening aortic insufficiency
B. Extra-luminal outflow graft compression
C. Intraluminal thrombosis of the outflow graft
D. Outflow graft twisting
E. Progression of underlying

## QUESTION 69

A 51-year-old recipient of OHT 5 years ago, a history of depression, ethanol use and non-adherence to immunosuppressive medications for 6 months is admitted to the hospital in severe cardiogenic shock with an LVEF of <20%. Endomyocardial biopsy shows 3R ACR and AMR 2. He is placed on intravenous heparin and cannulated on VA-ECMO. The patient was then treated with IV solumedrol and IV rabbit anti-thymocyte globulin and IV rituximab. The LVEF improved to 35-40% and he was transitioned to high dose oral MMF, tacrolimus, and low dose prednisone. VA-ECMO is uneventfully removed on hospital day 5. On day 12, the patient complains of diffuse joint discomfort and pruritus. He is now febrile and has a new urticarial rash. Mild new neutropenia and thrombocytopenia are noted on laboratory examination. The patient's spleen is mildly enlarged.

Which is the most likely etiology of these findings?

A. A re-exacerbation of acute rejection
B. A complication of intravenous immunosuppressive agents
C. Heparin induced thrombocytopenia
D. An opportunistic infection
E. A complication of oral immunosuppressive agents

## QUESTION 70

A 69-year-old woman with past medical history significant for sinus node dysfunction s/p dual-chamber PPM implantation, hypertension, chronic kidney disease stage III, breast cancer sp mastectomy and chemotherapy (trastuzumab and doxorubicin (cumulative dose 100mg/m2), 15 years ago) and recent diagnosis of non-small cell lung cancer (diagnosed ten months prior to presentation and treated with pembrolizumab) presents with worsening dyspnea, weight gain and lower extremity edema. Physical exam is notable for HR 60bpm, BP 110/82mmHg, normal rate, regular, JVP ~16cm H2O, bibasilar crackles, and 2+ LE edema. Serologic work up reveals SCr 1.4mg/dL, NT-proBNP 2768 pg/mL, hsTnT 32ng/L, CK 50 U/L and TSH 6.3 mU/L. PPM interrogation reveals sinus rhythm with ventricular pacing, and 22% atrial and 34% ventricular pacing. TTE shows mild biventricular dilation, LVEF 40% (from 55% one year prior), mild RV dysfunction, no significant valvular dysfunction, and no pericardial effusion. Coronary angiogram is without obstructive epicardial coronary artery disease.

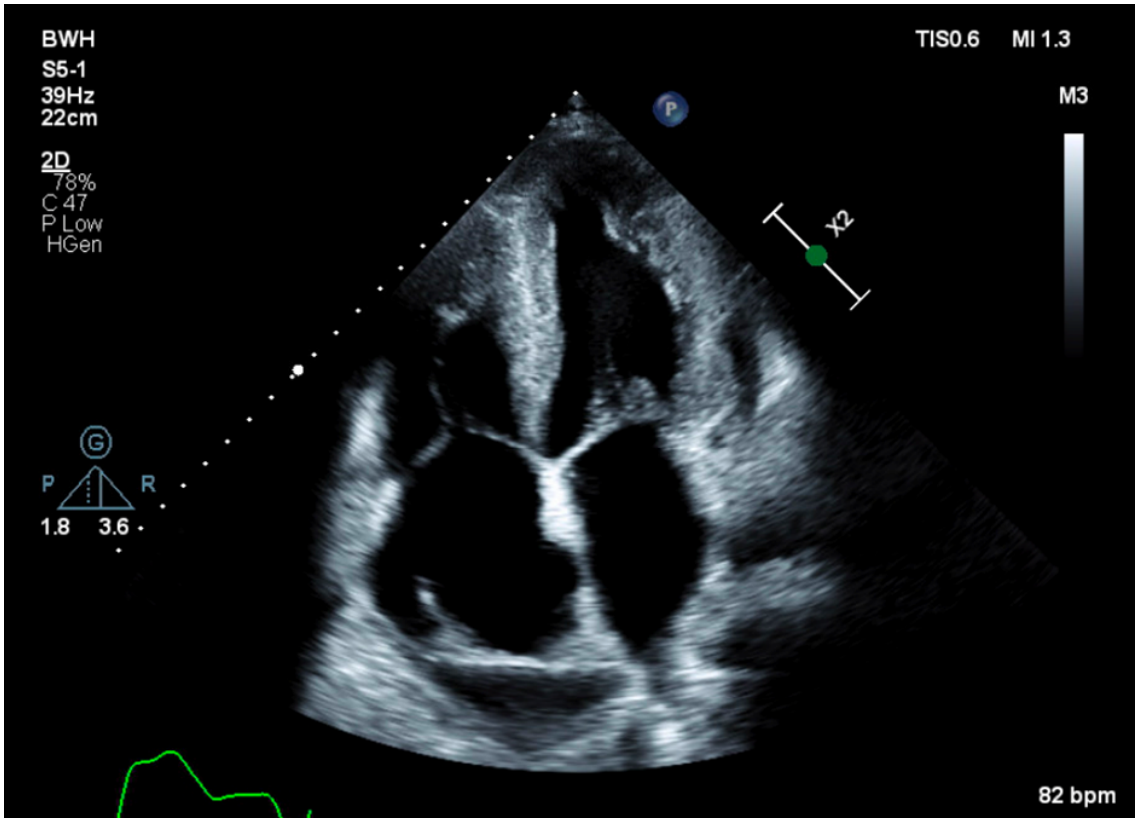
What is the most likely reason for this patient's decompensation?

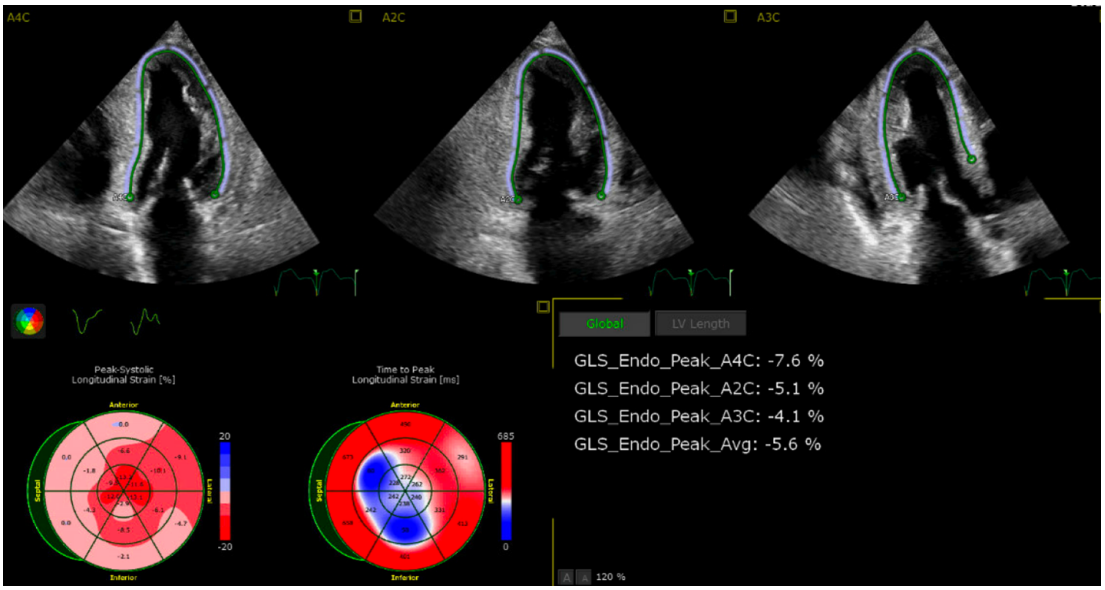
A. Pembrolizumab
B. Doxorubicin
C. RA pacing
D. RV pacing

## QUESTION 71

A 62-year-old African American gentleman with a history of long-standing hypertension, diabetes,

neuropathy, and chronic kidney disease stage III presents with worsening dyspnea and lower extremity edema. His physical exam is notable for HR 118 with an irregularly irregular rhythm, BP 110/85, macroglossia, normal S1S2 with II/VI systolic murmur at the LLSB and apex, JVP 14cm H2O with V waves, diminished breath sounds at the bases with dullness to percussion, and 2+ lower extremity edema. Laboratory studies are notable for SCr 1.7mg/dl, NT-proBNP 8826pg/ml, kappa free light chains of 16.6mg/L, lambda free light chains of 116.6mg/L and K/L of 0.14. ECG demonstrates atrial fibrillation with rapid ventricular response and NSSTWA. Representative images of his TTE are shown below. Grade 2 uptake is visualized on 99mTc-labeled pyrophosphate (PYP) scan.



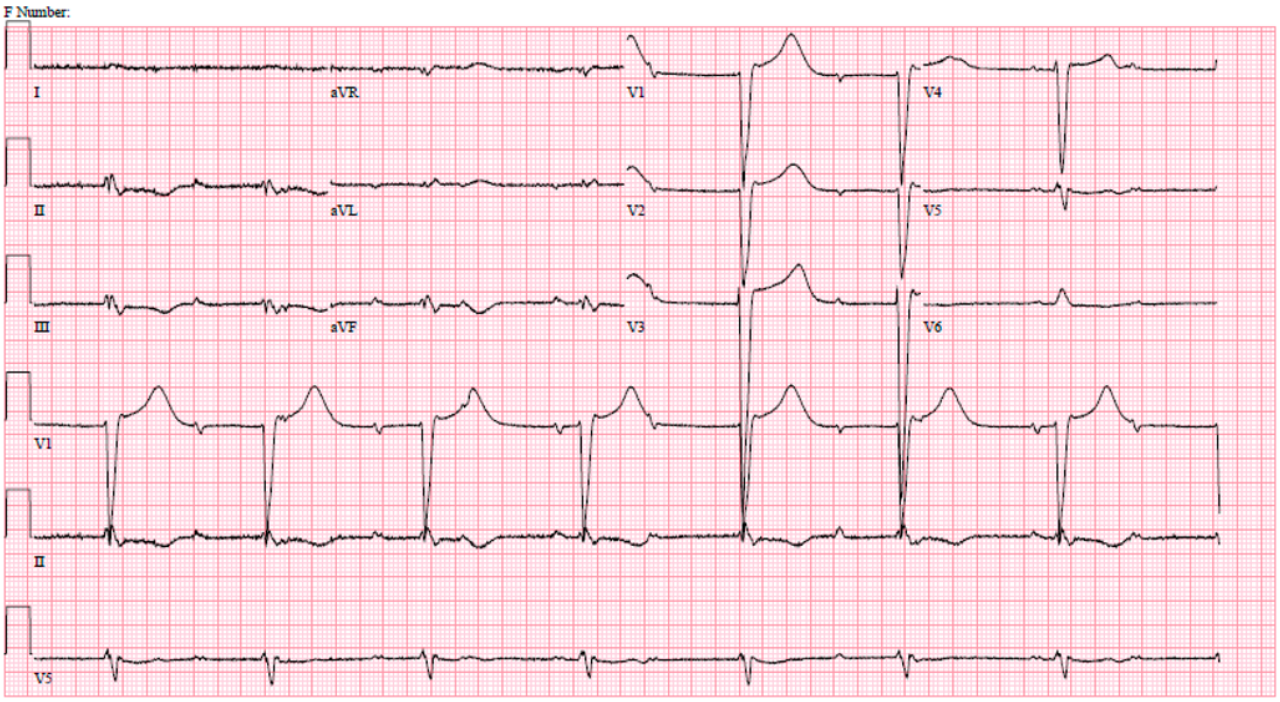



What is the next best step in patient management?

A. Start tafamidis
B. Arrange for TEE/DCCV
C. Perform endomyocardial biopsy
D. Obtain TTR genotyping

## QUESTION 72

A 51-year-old man with a history of hypertension presents with presyncope and shortness of breath. Initial vital signs are notable for HR of 39bpm and BP 170/84mmHg. Physical exam is notable for bradycardia with a regular rhythm, II/VI systolic murmur at the LLSB, JVP ~9cm H2O with cannon A waves, crackles at the bases on pulmonary auscultation and trace LEE. High-sensitivity troponin T is 47ng/ml, TSH 8 mU/L and Lyme serologies are negative. ECG and representative images of his TTE and cardiac PET are shown below. CCTA does not show any significant obstructive epicardial coronary artery disease.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25mm/s 10mm/mV 150Hz 9.0.9 12SL 243 CID: 0
EID:278 EDT: 20:51 05-APR-2022 ORDER: 983559923 ACCOUNT: 3436195038
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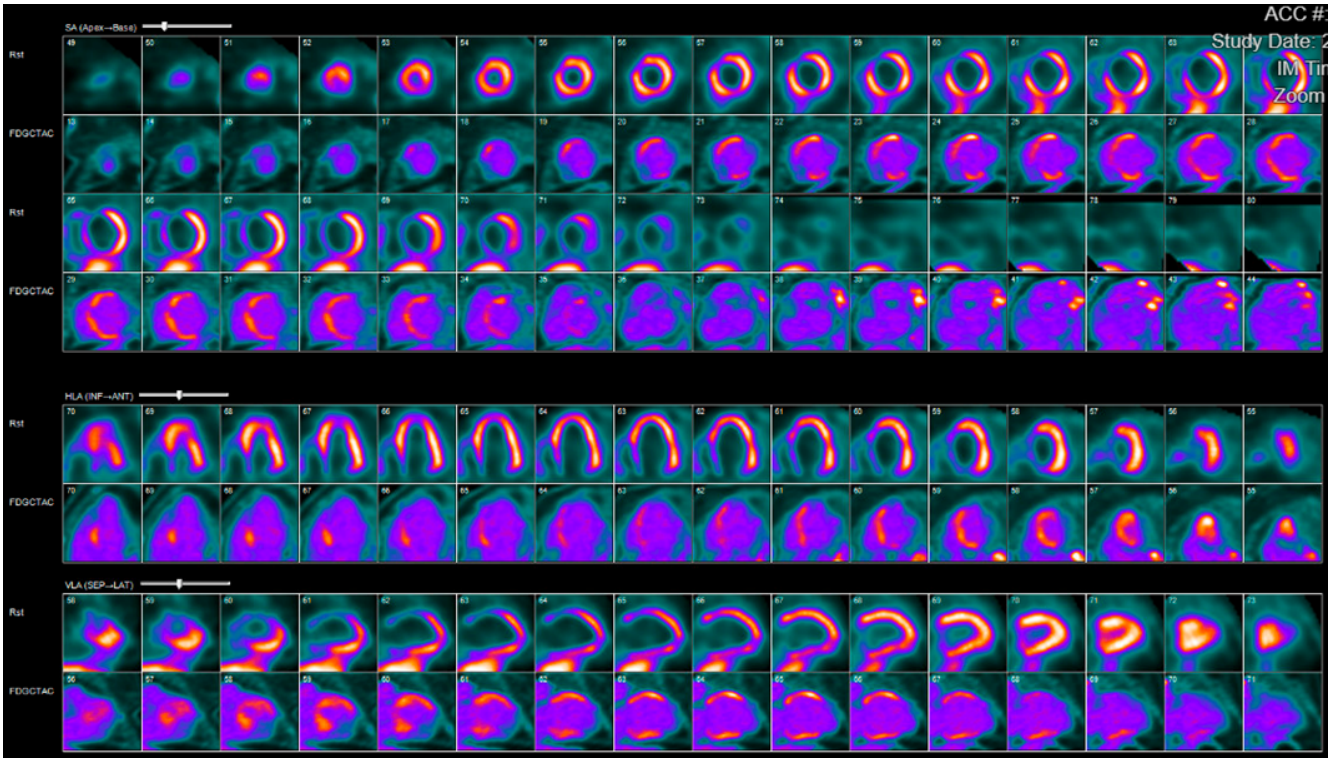


What is the next best step in management of this patient?

A. Refer the patient for implantation of a PPM
B. Refer the patient for implantation ICD
C. Start corticosteroids
D. Start methotrexate

## QUESTION 73

A 76-year-old patient with iCM s/p HM3 LVAD (implanted three years ago), pAF, HTN, HLD and DM presents with worsening dyspnea and LE edema despite escalation of diuretics and antihypertensives during serial clinic visits. The physical exam is notable for HR 88bpm with an irregularly irregular rhythm, BP 92/D mmHg, +LVAD hum, JVP ~15cm H2O, crackles at the bases, mild hepatomegaly, no palpable peripheral pulse and 2+ LE edema. LVAD interrogation demonstrates flows of 5.3L/min at 5800rpm, PI 2.6, power 5.0W and frequent PI events. Serologic work up is notable for SCr 1.4mg/dl (bl ~1.1mg/dl), hemoglobin 12.1mg/dl, LDH 312 U/L, total bilirubin 1.8mg/dl, plasma free hemoglobin 10mg/dl, and INR 2.4.
Representative clips of the patient's TTE are shown below.

Which echocardiographic finding is most specific for the patient's condition?

A. Vena contracta width of 2mm
B. Persistent LV cavity dilation with LVAD speed increases
C. Peak systolic-to-diastolic velocity ratio of 3 of the LVAD outflow cannula
D. Diastolic acceleration of the LVAD outflow cannula of 30cm/sec$^2$

## QUESTION 74

A 43-year-old patient with end-stage dilated cardiomyopathy s/p OHT nine months ago (CMV D+/R+, EBV D+/R-, toxoplasma D-/R-) presents with worsening fatigue and dyspnea. Vitals are notable for HR 110bpm, RR 18, BP 118/68mmHg, SpO2 98%. The patient's exam is notable for subconjunctival pallor, mild sublingual jaundice, II/VI systolic murmur at the LUSB, clear lungs, benign abdomen, and no LEE. Laboratory studies show SCr 1.6mg/dL (baseline 1.1mg/dL), WBC 3.1k/uL, Hgb 7.1g/dL (baseline 11mg/dL), platelets 120 k/uL, reticulocyte count 10.2%, total bilirubin 4.9mg/dL, direct bilirubin 1.2mg/dL, LDH 789U/L, INR 1.2. EBV VL 106IU/mL. Peripheral smear demonstrates rare burr cells and schistocytes. Home medications include tacrolimus, mycophenolate mofetil, prednisone, valganciclovir, sulfamethoxazole/trimethoprim, nystatin, aspirin, and pravastatin.

What is the most likely cause of this patient's anemia?

A. Tacrolimus
B. Mycophenolate mofetil
C. Ebstein-Barr virus infection
D. CMV infection

## QUESTION 75

A 42-year-old woman just relocated to your area and is referred to your clinic for evaluation of pulmonary hypertension (PH). She has a history of morbid obesity (BMI 40 kg/m2), type II diabetes mellitus, hypertension, former tobacco use, and remote deep vein thrombosis (DVT) for which she completed a six-month course of anticoagulation. Upon further questioning, she is only able to walk less than half a block due to shortness of breath.

Her prior evaluation for HIV, hepatitis serologies, and connective tissue disease was negative. A helical computed tomography (CT) scan with contrast revealed no acute pulmonary emboli (PE). She had been prescribed sildenafil 20 mg three times daily, but she reported feeling worse and self-discontinued the medication approximately three months prior to this office visit. VS: BP 110/70 HR 90 RR 20 SpO2 90% RA. Her examination is notable for jugular venous pressure at 15 cm $H_2O$, increased $P_2$, and parasternal heave with III/VI systolic murmur at the left lower sternal border and II/VI diastolic murmur in the left upper chest. A right-sided $S_3$ is present. Clear lung fields are noted on auscultation. Her liver is mildly enlarged. She has 1+ bilateral lower extremity edema.

Her echocardiogram is remarkable for normal left ventricular ejection fraction, dilated right ventricle (RV) with moderately decreased RV function, diastolic shift of the RV septum, RV systolic pressure 64 mm Hg, moderate to severe tricuspid regurgitation, mitral E/e' of 8, and mild pulmonary insufficiency.

Which of the following is the best next step in determining the etiology of her dyspnea?

A. Right heart catheterization with exercise
B. Polysomnogram
C. Ventilation/perfusion scan
D. High-resolution chest computed tomography
E. Pulmonary function tests

## QUESTION 76

A 65-year-old woman who underwent heart transplant approximately one year ago for ACC/AHA Stage D ischemic cardiomyopathy presents to the emergency room with sudden onset of shortness of breath. She has a past medical history of a seizure disorder, gout, hypertension, and hyperlipidemia. Her post-transplant course was complicated by an Aspergillus pneumonia three months prior, for which she remains on antifungal therapy. Her outpatient medication list includes tacrolimus, mycophenolate mofetil, trimethoprim-sulfamethoxazole, allopurinol, levetiracetam, pravastatin, voriconazole, and aspirin. On exam, she is ill appearing but afebrile. Her blood pressure is 90/78 mm Hg, heart rate is 120 bpm, oxygen saturation is 96% on 2 L nasal cannula. Examination shows jugular venous distention to 14 cmH2O, hepatojugular reflux, bilateral pulmonary crackles, and an S3 gallop, with 2+ pitting edema bilaterally. Urgent transthoracic echocardiography demonstrates reduced severely reduced left ventricular systolic function with LV ejection fraction at 20%. Her labs are notable for N-terminal-pro-BNP 575 pg/ml and an undetectable FK506 (tacrolimus) level.

She is given 1 gram of intravenous methylprednisolone, 80 mg of intravenous furosemide, and taken for urgent endomyocardial biopsy, which ultimately reveals International Society of Heart and Lung Transplantation (ISHLT) grade 3R rejection. The patient insists she was taking medications as directed until she ran out of medical insurance two weeks ago.

Which of the following medications is most likely to have been discontinued resulting in this patient's presentation?

A. Phenytoin
B. Voriconazole
C. Allopurinol
D. Trimethoprim/Sulfamethoxazole

## QUESTION 77

A 37-year-old woman with ACC/AHA Stage D nonischemic cardiomyopathy status post single chamber

implantable cardioverter defibrillator in 2020, morbid obesity (BMI 40), obstructive sleep apnea and longstanding persistent atrial fibrillation presents to the emergency room with sudden-onset exertional dyspnea, orthopnea, and paroxysmal nocturnal dyspnea. She has had multiple ER visits and hospitalizations for acute decompensated heart failure in the last 12 months and has had difficulty affording her medications. Upon initial examination, she is found to be "cold and wet." She is started on intravenous norepinephrine and intravenous bumetanide infusion and subsequently admitted to the cardiovascular intensive care unit for further evaluation and management. On exam, vital signs are: BP 90/60 HR 120 RR 30 SpO2 90% 4 liters/minute nasal cannula. She is confused, restless, and has poor urine output. Her initial labs show a serum creatinine 1.9 mg/dL (baseline unknown), lactic acid 3.0 mmol/L, with elevated liver function tests. Norepinephrine is increased to 20 mcg/min, and epinephrine 5 mcg/min is started.

She is taken for a right heart catheterization: right atrial pressure 40 mmHg, pulmonary artery pressure 62/48 mmHg (mean 52 mmHg), pulmonary capillary wedge pressure 45 mmHg, Fick cardiac output/cardiac index 2.1/1.0, and pulmonary artery pulsatility index 0.3. A cardiogenic shock team is activated in the catheterization laboratory and the decision is made to escalate to veno-arterial extracorporeal membrane oxygenation (VA ECMO). During induction, she develops a cardiac arrest due to pulseless electrical activity requiring 5 minutes of cardiopulmonary resuscitation and emergent intubation prior to cannulation. She is neurologically intact, noted to have minimal pulsatility of her left ventricle (LV), a severely dilated LV of 89 mm, and a dilated and moderately dysfunctional right ventricle (RV). A percutaneous microxial left ventricular assist device, Impella CP, is inserted for LV venting.

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

A. Evaluation for heart transplant
B. Evaluation for durable left ventricular assist device with temporary right ventricular assist device support
C. Wean VA ECMO and leave Impella CP in place
D. Refer to hospice and withdraw care

## QUESTION 78

A 51-year-old woman with a past medical history of estrogen receptor/progesterone receptor/human epidermal growth factor receptor 2–negative metastatic breast cancer status post mastectomy on pembrolizumab presents urgently to the emergency department with sudden onset of fatigue, dyspnea, and orthopnea for 48 hours since returning from a weeklong camping excursion to Connecticut with family and friends.

She has no known prior personal or family history of cardiac disease. Vital Signs: BP 100/70 HR 120 RR 28 SpO2 90% 6 L/min NC. Her examination is significant for a jugular venous pressure > 15 cm $H_2O$, hepatojugular reflux, tachycardic, irregularly irregular rhythm, bilateral crackles extending halfway up to the apex, an erythematous maculopapular rash on the tibial surface of her left leg, and otherwise cool

extremities bilaterally.

An electrocardiogram (ECG) reveals atrial fibrillation (AF) with ventricular rate 115 bpm and a new right bundle branch block. Limited bedside echocardiography reveals normal left ventricular (LV) cavity size, mild concentric LV hypertrophy, and estimated ejection fraction 35%. She is administered an intravenous furosemide challenge but becomes progressively hypotensive and bradycardic, prompting an emergent right heart catheterization which reveals: RA 14; PA 40/24 PCWP 36 and Fick cardiac index 1.4 L/min/$m^2$.

Which one of the following is the most appropriate next diagnostic test?

A. Cardiac Magnetic Resonance Imaging
B. FDG-Positron Emission Tomography
C. Endomyocardial Biopsy
D. Lyme serology

## QUESTION 79

A 68-year-old man presents to your heart failure clinic endorsing progressive exertional dyspnea over the past year. He is unable to walk one block or ascend a flight of stairs without dyspnea. His past medical history includes hypertension, chronic obstructive pulmonary disease, paroxysmal atrial fibrillation status post electrical cardioversion six months earlier, and type II diabetes mellitus. He has a remote history of tobacco and alcohol use but has been abstinent from both substances for 10 years.

Which one of the following factors is the most predictive of a diagnosis of heart failure with preserved ejection fraction?

A. Systemic hypertension
B. Body mass index
C. Atrial fibrillation
D. Mitral E/e' ratio
E. Pulmonary hypertension

## QUESTION 80

A 65-year-old man with longstanding nonischemic cardiomyopathy, heart failure with reduced ejection fraction (HFrEF) with LVEF 35% status post primary prevention implantable cardioverter defibrillator two years ago, paroxysmal atrial fibrillation, obstructive sleep apnea, and stage IIIa chronic kidney disease presents following recent hospitalization for acute decompensated heart failure. He underwent aggressive intravenous diuresis and was discharged home on metoprolol 25 mg twice daily, sacubitril/valsartan 97/103 twice daily, spironolactone 25 mg daily, apixaban 5 mg twice daily, and torsemide 20 mg daily.

In clinic today, he reports exertional dyspnea walking into clinic from the parking garage but denies orthopnea, bendopnea, and paroxysmal nocturnal dyspnea. Vital signs are: BP 101/64 mmHg HR 62 beats

per min RR 12 breaths per min SpO2 98% on room air. Weight is 253 lbs. Exam shows regular rate and rhythm, normal S1, S2, jugular venous pressure 8 cmH2O, no hepatojugular reflux, and trace ankle edema bilaterally. His labs are notable for NTproBNP 200 pg/ml, sodium 133 mEq/L, potassium 4.5 mEq/L, blood urea nitrogen 35 mg/dL, and serum creatinine 1.7 mg/dL (estimated GFR 46).

Which of the following is the best next step to reduce this patient's risk of hospitalization for heart failure (HF) and cardiovascular mortality?

A. Refer for transcatheter edge to edge mitral valve repair
B. Add empagliflozin 10 mg daily
C. Add ivabradine 5 mg twice daily
D. Refer for implantable pulmonary artery sensor

# ANSWER KEY

| | | | | | |
|---|---|---|---|---|---|
| 1. | B | 28. | C | 55. | D |
| 2. | C | 29. | D | 56. | A |
| 3. | A | 30. | B | 57. | D |
| 4. | D | 31. | C | 58. | C |
| 5. | C | 32. | D | 59. | C |
| 6. | C | 33. | B | 60. | B |
| 7. | D | 34. | A | 61. | C |
| 8. | B | 35. | B | 62. | D |
| 9. | A | 36. | B | 63. | C |
| 10. | A | 37. | D | 64. | C |
| 11. | D | 38. | C | 65. | C |
| 12. | D | 39. | D | 66. | C |
| 13. | A | 40. | B | 67. | D |
| 14. | C | 41. | A | 68. | D |
| 15. | D | 42. | C | 69. | B |
| 16. | B | 43. | D | 70. | D |
| 17. | C | 44. | D | 71. | C |
| 18. | C | 45. | A | 72. | B |
| 19. | A | 46. | C | 73. | C |
| 20. | D | 47. | A | 74. | A |
| 21. | B | 48. | A | 75. | C |
| 22. | B | 49. | B | 76. | B |
| 23. | D | 50. | D | 77. | B |
| 24. | E | 51. | C | 78. | C |
| 25. | A | 52. | A | 79. | C |
| 26. | A | 53. | C | 80. | B |
| 27. | E | 54. | B | | |

# RATIONALE

## QUESTION 1

Heart Failure with Reduced Ejection Fraction

**Testing Point:** The Board-certified AHFTC specialist should recognize the indications for an endomyocardial biopsy in the setting of acute onset heart failure with hemodynamic consequences.

**Answer:** B. Right heart catheterization with endomyocardial biopsy.

According to the American College of Cardiology/American Heart Association guidelines, it is a Class I recommendation to perform endomyocardial biopsy for patients with new-onset failure of <2 weeks' duration associated with a normal-sized or dilated left ventricle and hemodynamic compromise.

Cardiac magnetic resonance imaging may not provide the needed hemodynamic information in a patient nearing cardiogenic shock, but the option may be selected by the less knowledgeable AHFTC physician because of CMR may demonstrate LGE patterns consistent with myocarditis.

Distractors C and D are not the best answer because the patient's age and presentation are not consistent with ACS. However, the less knowledgeable AHFTC physician may select these options because of the symptoms of chest discomfort.

**References:**

1. Cooper LT et al. The Role of Endomyocardial Biopsy in the Management of Cardiovascular Disease: A Scientific Statement from the American Heart Association, the American College of Cardiology

## QUESTION 2

Special Etiologies

**Testing Point:** The Board-certified AHFTC specialist should recognize the presentation of ATTR amyloidosis and non-invasive methods for diagnosis.

**Answer:** C. Technetium pyrophosphate scintigraphy.

Technetium pyrophosphate scintigraphy, also known as a PYP scan, is a non-invasive test that can be used to diagnose ATTR amyloidosis. The patient's clinical presentation (unexplained increased LV wall thickness together with persistent heart failure symptoms and orthostatic hypotension) including associated conditions such as carpal tunnel syndrome, lumbar spinal stenosis, and aortic stenosis are all associated with ATTR amyloidosis. Given that the patient has a normal serum k/λ free light chain ratio, AL amyloidosis is much less likely. The use of PYP scans has dramatically reduced the need for endomyocardial biopsies which

are more invasive as a first diagnostic test.

Cardiac positron emission tomography is more helpful for evaluating for cardiac sarcoidosis or myocarditis, which is not consistent with this patient's history. While cardiac magnetic resonance imaging will provide additional information, the PYP scan would be a better test given the high pretest probability for ATTR.

**References:**

1. Dorbala S, Ando Y, Bokhari S, et al. ASNC/AHA/ASE/EANM/HFSA/ISA/SCMR/SNMMI expert consensus recommendations for multimodality imaging in cardiac amyloidosis: Part 1 of 2-evidence base and standardized methods of imaging. J Nucl Cardiol 2019;26:2065-123.
2. Dorbala S, Ando Y, Bokhari S, et al. ASNC/AHA/ASE/EANM/HFSA/ISA/SCMR/SNMMI Expert Consensus Recommendations for Multimodality Imaging in Cardiac Amyloidosis: Part 2 of 2-Diagnostic Criteria and Appropriate Utilization. J Card Fail 2019;25:854-65.
3. Gillmore JD, Maurer MS, Falk RH, et al. Nonbiopsy Diagnosis of Cardiac Transthyretin Amyloidosis. Circulation 2016;133:2404-12.

## QUESTION 3

Heart Failure with Preserved Ejection Fraction

**Testing Point:** The Board-certified AHFTC specialist should know that implantable pulmonary artery pressure sensor monitoring has been demonstrated to reduce heart failure hospitalizations in patients with NYHA Class II or III regardless of ejection fraction.

**Answer:** A. Therapy adjustment guided by pulmonary artery pressure monitoring.

In the CHAMPION trial (the CardioMEMS Heart Sensor Allows Monitoring of Pressor to Improve Outcomes in NYHA Class III Heart Failure Patients), implantable pulmonary artery pressure sensor monitoring was shown to reduce the rate of heart failure hospitalizations. The Post-Approval Study also showed that this effect was consistent across ejection fraction groups.

While decongestion is key for symptom relief and the patient may benefit from increased diuretic dose, it has not been demonstrated to decrease hospitalization. Beta-blockers have not been shown to reduce heart failure hospitalizations or survival in patients with preserved ejection fraction. Carvedilol may be reasonable to improve blood pressure control as this patient's blood pressure is elevated. Spironolactone may be considered to decrease hospitalizations.

**References:**

1. Givertz MM, Stevenson LW, Costanzo MR, et al., on behalf of the CHAMPION Trial Investigators. Pulmonary Artery Pressure-Guided Management of Patients With Heart Failure and Reduced Ejection Fraction. J Am

Coll Cardiol 2017;70:1875-86.
2. Abraham WT, Stevenson LW, Bourge RC, Lindenfeld JA, Bauman JG, Adamson PB, on behalf of the CHAMPION Trial Study Group. Sustained efficacy of pulmonary artery pressure to guide adjustment of chronic heart failure therapy: complete follow-up results from the CHAMPION randomised trial. Lancet 2016;387:453-61.
3. Shavelle DM, Desai AS, Abraham WT et al. Lower Rates of Heart Failure and All-Cause Hospitalizations During Pulmonary Artery Pressure-Guided Therapy for Ambulatory Heart Failure: One-Year Outcomes from the CardioMEMS Post-Approval Study. Circ Heart Fail. 2020; 13(8):e006863.

## QUESTION 4

Mechanical Circulatory Support

**Testing Point:** The Board-certified AHFTC specialist should recognize when advanced therapies including LVAD or heart transplantation are futile in patients with end-stage heart failure.

**Answer:** D. Enrollment in a palliative care program

This patient has end-stage heart failure secondary to ischemic cardiomyopathy. He has presented with ventricular tachycardia storm with multiple shocks from his ICD with poor hemodynamics and no coronary lesion to be revascularized.

Additionally, he has evidence of multi-organ failure which preclude him from candidacy for durable left ventricular assist device. Given the lack of an advanced therapy option, escalation of support to extracorporeal membrane oxygenation (ECMO) would be unlikely to meaningfully change his outcome given his advanced age and comorbidities.

Administration of continuous inotropes in some studies has been associated with improvement in quality of life but overall data is lacking. Additionally, inotropes are pro-arrhythmogenic and likely to worsen his ventricular tachycardia.

Therefore, the best option would be to discuss goals of care and engage palliative care.

**References:**

1. Mehra et al. The 2016 International Society for Heart Lung Transplantation Listing criteria for heart transplantation: A 10-year update. JHLT. 2016;35(1):P1-23.
2. Yancy et al. 2013 ACCF/AHA Guideline for the management of Heart Failure. Circulation 2013; 128; e240-e327.

## QUESTION 5

Heart Transplantation

**Testing Point:** The Board-certified AHFTC specialist should recognize when to refer a patient with end-stage heart failure for evaluation for heart transplantation.

**Answer:** C. Refer for evaluation for heart transplantation.

The patient has advanced heart failure as indicated by repeated hospitalizations for heart failure, intolerance to neurohormonal blockade due to hypotension with persistent symptoms. His cardiopulmonary exercise test (CPET) demonstrates severely impaired functional capacity in the setting of maximal volitional effort.

Although this patient may benefit from an implantable pulmonary artery pressure sensor given repeated hospitalizations, he has evidence of progressive disease and should be evaluated for heart transplantation in a timely manner. With respect to transcatheter mitral valve repair, the patient has severe functional mitral regurgitation, but given a severely dilated left ventricle with an LVEDD of 8.0 cm, is unlikely to benefit. Lastly, the patient does not have a sufficient prolongation of the QRS to warrant consideration of cardiac resynchronization therapy.

**References:**

1. Yancy et al. 2013 ACCF/AHA Guideline for the management of Heart Failure. Circulation 2013; 128; e240-e327.

## QUESTION 6

Pulmonary Hypertension

**Testing Point:** The Board-certified AHFTC specialist should know what constitutes a positive response to vasodilator testing for pulmonary arterial hypertension.

**Answer:** C. Mean pulmonary artery pressure of 35 mm Hg with cardiac output of 4L/min.

A positive response to vasodilators is defined as a decrease in mean pulmonary arterial pressure by at least 10 mm Hg to a value less than 40 mm Hg while maintaining cardiac output. In patients with a positive response, treatment with calcium channel blockers should be considered as a first-line agent.

**References:**

1. McLaughlin VV, Archer SL, Badesch DB, et al. ACCF/AHA 2009 expert consensus document on pulmonary hypertension: a report of the American College of Cardiology Foundation Task Force on Expert Consensus Documents and the American Heart Association: developed in collaboration with the American College of Chest Physicians, American Thoracic Society, Inc.and the Pulmonary Hypertension Association. Circulation. 2009;119:2250–94.

## QUESTION 7

Valvular Heart Disease

**Testing Point:** The Board-certified AHFTC specialist should be able appropriately evaluate valvular heart disease in heart failure patients.

**Answer:** D. Exercise echocardiography.

According to the ACC/AHA Guidelines, provocative exercise testing is warranted to evaluate any valvular disease where there is a significant discrepancy between symptoms and valve disease severity based on resting assessment. In this case, although the echocardiogram report concludes moderate mitral stenosis, the gradient and mitral valve area are actually borderline for severe mitral stenosis. The patient has demographics and echo features consistent with rheumatic heart disease too. Exercise echo would reveal pathologic elevation in mitral valve gradients and estimated RVSP.

Normal electrocardiogram and lack of paroxysmal symptoms argues against arrhythmic causes. Normal right heart features on echocardiogram argues against pulmonary hypertension. Reassuring ischemic evaluation argues against coronary artery disease. PET stress testing would not add much beyond a reassuring pharmacologic nuclear stress test.

**References:**

1. Nishimura R et al. 2014 ACC/AHA Guideline for the Management of Patients with Valvular Heart Disease. Circulation. 2014;129:2440-92.

## QUESTION 8

Mechanical circulatory support

**Testing point:** The Board-certified AHFTC specialist should recognize which is the best treatment for cardiogenic shock according to clinical situation.

**Answer:** B. ventriculo-arterial ECMO.

This patient has severe cardiogenic shock despite inotropic support and requires aggressive measures to save her life. VA-ECMO could also be employed with concomitant LV venting strategies, but that can be done as the next step.

The other choices here would remain insufficient to salvage perfusion. The patient's LV cardiac power output, or (MAP x CO) / 451 = 0.46, which is too low to be remedied with nitroprusside alone. Norepinephrine may boost blood pressure but at the expense of imposing more afterload. alone. Due to

very low cardiac power as well as the aortic regurgitation, intra-aortic balloon placement, or even Impella alone, would both be poor choices for managing his shock.

**References:**

1. Van Diepen S, et al. Contemporary Management of Cardiogenic Shock: A Scientific Statement From the AHA. Circulation. 2017;136:e232-268.

## QUESTION 9

Pulmonary Hypertension

**Testing point:** The Board-certified AHFTC specialist should recognize the indications for vasodilator therapy in pulmonary arterial hypertension patients.

**Answer:** A. After 5 minutes of 40 p.p.m. inhaled NO, mPAP falls to 25 mm Hg and CO remains at 5.0 L/min.

This patient has anorexigen-associated pulmonary arterial hypertension. The original study revealing pulmonary vasoreactivity as a predictor of calcium-channel blocker responsiveness was done in idiopathic PAH subjects.[1] This defined pulmonary vasoreactivity as a drop in mPAP by at least 10 mm Hg from an initial value above 40 mm Hg, with either unchanged or increased cardiac output. Choice A fits the definition of vasoreactivity. In Choice B, the mPAP does not fall enough, even though the cardiac output increases. In Choice C, mPAP falls, but this may be because flow (cardiac output) falls significant as well.

Note that a follow-up study showed that vasoreactivity should only be tested in those with idiopathic, heritable, or anorexigen-associated PAH benefit, because those are the only three groups that, if vasoreactive, respond to first-line CCB treatment.[2] That same follow-up study showed that other sub-groups of PH, including connective tissue disorder-associated Group I PAH, and other World Health Organization (WHO) Groups of PH (Groups 2-5), do not benefit from CCB therapy, even if vasoreactive.

**References:**

1. Sitbon O, et al. Long-Term Response to CCB in Idiopathic Pulmonary Arterial Hypertension. Circulation. 2005;111:3105-3111.
2. Montani D, et al. Long-term response to CCB in non-idiopathic pulmonary arterial hypertension. European Heart Journal. 2010;31:1898-1907.
3. Galie N, et al. Risk stratification and medical therapy of pulmonary arterial hypertension. European Respiratory Journal. 2019;53:1801889.

## QUESTION 10

Heart failure with reduced ejection fraction

**Testing point:** The Board-certified AHFTC specialist should be able identify the cause of exertional limitation on cardiopulmonary exercise test.

**Answer:** A. Cardiac limitation.

The patient has a moderate reduction in exertional capacity, having only reached 57% of her predicted peak VO2. VE/VCO2 is elevated as well but this does not always help distinguish between cardiac and pulmonary limitation, as abnormalities in either can result in ventilatory inefficiency. The patient's estimated oxygen pulse, however, is < 10 ml/min (1371/144) which argues for reduced exercise stroke volume as the culprit for the patient's limitation.

Although the patient's $SpO_2$ drops to 92% at peak activity, the patient retains a breathing reserve of 0.34 (1 - 99/150) arguing against a pulmonary limitation. The patient's BMI is indeed elevated but not enough to warrant discounting the peak $VO_2$. The patient reaches an adequate respiratory exchange ratio, which indicates achievement of maximal volitional effort

**References:**

1. Malhotra R, et al. Cardiopulmonary Exercise Testing in Heart Failure. JACC Heart Failure. 2016;4:607-16.

## QUESTION 11

Special Etiologies

**Testing point:** The Board-certified AHFTC specialist should be able to identify which is the first therapy action to be undertaken in a patient with hypertrophic cardiomyopathy

**Answer:** D. Discontinue lisinopril 10 mg/day.

This patient is referred for what was thought to be left ventricular hypertrophy and has been managed as hypertension in the setting of other cardiovascular risk factors. However, the patient does not have very high afterload, and LVH is out of proportion to blood pressure. Instead, hypertrophic cardiomyopathy is present, as evidenced by the concentric hypertrophy, somewhat asymmetric LVH, LVOT gradient that is especially pronounced with Valsalva, and systolic anterior motion of the mitral valve. Given the tendency to exhibit inducible obstruction of the LVOT, afterload reducing agents should first be withdrawn, and the patient should be counselled about the importance of hydration. These efforts help reduce the tendency to precipitate LVOT obstruction and exertional symptoms.

After removal of offending issues, the next step in the management of obstructive HCM would be to initiate verapamil ER. Disopyramide should only be considered if beta blockers or calcium channel blockers are first tried and maximized. Even though the provoked gradient is reasonably high, interventional and surgical

therapies are considered after failure of medical therapies.

**References:**

1. Ommen SR, et al. 2020 AHA/ACC Guideline for the Diagnosis and Treatment of Patients with Hypertrophic Cardiomyopathy. Circulation. 142:e558-631.

## QUESTION 12

**Testing Point:** The Board-certified AHFTC specialist should recognize potential medications which could exacerbate or worsen heart failure.

**Answer:** D. Pregabalin

Pregabalin and gabapentin use in patients with heart failure has been associated with increased development of both peripheral edema and pulmonary edema, which is resistant to loop diuretics. The mechanism of the edema associated with pregabalin and gabapentin (in doses exceeding 1600mg/daily) is believed to be similar to that of a dihydropyridine calcium channel blocker, which affects the microvascular system, leading to lower-extremity edema, making Answer D correct. Answer A is incorrect as it has been evaluated in the SPICE trial and found to be safe in patients with HFrEF. While atypical antipsychotics do have cardiovascular side effects such as QT prolongation, hyperlipidemia, worsening glucose control, and weight gain, this class of drugs have not been shown to directly exacerbate symptoms of heart failure, making Answer B incorrect. Based on the SADHART trial, sertraline was found to be safe in patients with significant heart failure making these Answer C incorrect.

**References:**

1. Page RL, Cheng D, Dow T, et al. Drugs that may cause or exacerbate heart failure: a scientific statement from the American Heart Association; on behalf of the American Heart Association Clinical Pharmacology and Heart Failure and Transplantation Committees of the Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, Council on Cardiovascular and Stroke Nursing, and Council on Quality of Care and Outcomes Research. Circulation 2016;134:e261.
2. O'Connor CM, Jiang W, Kuchibhatta M, et al. Safety and efficacy of sertraline for depression in patients with heart failure: results of the SADHART-CHF (Sertraline Against Depression and Heart Disease in Chronic Heart Failure) trial. J Am Coll Cardiol 2010; 56 (9): 692-9.
3. Holubarsch CJ, Colucci WS, Meinertz T, et al. The efficacy and safety of Crataegus extract WS 1442 in patients with heart failure: the SPICE trial. Eur J Heart Failure. 2008; 10 (12): 1255-63.

## QUESTION 13

**Testing Point:** The Board-certified AHFTC specialist should have an understanding of perioperative management of a patient with HFrEF in order to reduce the risk of adverse events.

**Answer:** A. Stop the apixaban two days prior to surgery.

Major adverse cardiac events (MACE) are some of the most common complications occurring in the perioperative period, particularly for patients with heart failure. The use of the direct acting oral anticoagulants (DOACs) are now recommended over warfarin by the AHA/ACC Atrial Fibrillation management guidelines for therapeutic anticoagulation. As more than 35% of patients with heart failure have concomitant atrial fibrillation, management of anticoagulation in the perioperative setting is important to minimize MACE in this population. When considering discontinuation of a DOAC, one must balance thromboembolic risk, renal function, and bleeding risk of the surgery. This patient has a CHADs-Vasc score of 4, which is moderate-high risk of stroke. Her renal function is adequate as her CrCl > 30 ml/min, but the surgery is considered a high bleed-risk. With this in mind, apixaban should be stopped 48 hours prior to surgery based on the PAUSE study, making Answer A the best choice. If this surgery was low risk for bleeding, stopping the apixaban one day prior would be acceptable. Due to the risk of lactic acidosis, stopping metformin will be important; however, stopping seven days prior to surgery is excessive and could lead to hyperglycemia prior to surgery. The American Diabetes Association recommends withholding metformin the day of surgery, particularly in patients with a CrCL > 30 ml/min, making Answer B incorrect. A sudden stop in carvedilol can result in rebound hypertension, tachycardia and potential worsening of heart failure. The 2014 ACC/AHA perioperative practice guidelines for non-cardiac surgery recommend beta blockers and statins be continued in patients who are on these medications chronically (1B recommendations for both). Thus, Answers C and D are not the best choice.

**References:**

1. American Diabetes Association. 15. Diabetes Care in the Hospital: Standards of Medical Care in Diabetes-2019. Diabetes Care 2019;42(Suppl. 1):S173–S181
2. Douketis JD, Spyropoulous AC, Duncan J, et al. Perioperative management of patients with atrial fibrillation receiving a direct oral anticoagulant. JAMA Internal Medicine 2019; 179(11): 1469-1478.
3. Doherty JU, Gluckman TJ, Hucker WJ, et al. 2017 ACC Expert Consensus Decision Pathway for Periprocedural Management of Anticoagulation in Patients With Nonvalvular Atrial Fibrillation: A Report of the American College of Cardiology Clinical Expert Consensus Document Task Force. J Am Coll Cardiol. 2017;69(7):871-898.
4. Fleisher LA, Kleischmann KE, Auerback AD, et al. 2014 ACC/AHA guideline on perioperative cardiovascular evaluation and management of patients undergoing noncardiac surgery: executive summary: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. Circulation. 2014; 130(24): 2215-45.

## QUESTION 14

**Testing Point:** The Board-certified AHFTC specialist should have an understanding of appropriate populations to initiate GDMT in patients with HFrEF

**Answer:** C. 65-year-old woman with NYHA functional class III HF (LVEF of 20%) with heart rate 78 beats/minute taking lisinopril 20 mg daily, eplerenone 50 mg daily, and carvedilol 25 mg twice daily.

According to data from the SHIFT trial, the ideal candidate for ivabradine would be a patient with HFrEF with NYHA functional classes II and III and a heart rate greater than 70 beats/minute who is taking maximally tolerated dose of a β-blocker and is in normal sinus rhythm. Answer C is correct because this patient matches each of these criteria. Answer A is incorrect because this patient has HFpEF. Additionally, the EDIFY trial found heart rate reduction with ivabradine did not improve outcomes and do not support the use of ivabradine in HFpEF. Answer B is incorrect because the patient has a heart rate of 62 beats/minute on maximal doses of metoprolol succinate. Answer D is incorrect because the patient is NYHA class IV, and the patient's heart rate of 110 beats/minute was most likely caused by dobutamine.

**References:**

1. Yancy CW, Jessup M, Bozkurt B, et al. 2017 ACC/AHA/HFSA focused update of the 2013 ACCF/AHA guideline for the management of heart failure. A report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Failure Society of America. Circulation 2017;136:e137-61.
2. Komajda M, Isnard R, Cohen-Solal A, et al. Effect of ivabradine in patients with heart failure with preserved ejection fraction: the EDIFY randomized placebo-controlled trial. Eur J Heart Fail. 2017 ;19(11):1495-1503.
3. Swedberg K, Komajda M, Bohm M, et al. Ivabradine and outcomes in chronic heart failure (SHIFT): a randomised placebo-controlled study. Lancet 2010; 376 (9744): 875-85.

## QUESTION 15

**Testing Point:** The Board-certified AHFTC specialist should be able to identify and manage common adverse medication side effects in heart transplant recipients.

**Answer:** D. Add primidone 50 mg daily and titrate to 250 mg/day

Tacrolimus and cyclosporine, both calcineurin inhibitors, can cause significant neurological side effects. Tremor is the most common tacrolimus-associated side effect and can also be seen with cyclosporine. In the case of this patient, primidone has been shown to be safe and affective at reducing tremor by 60% 1 to 7 hours after ingestion, with stable serum primidone levels but no detectable phenobarbital levels at doses of 250 mg/day. Thus, no drug-drug interactions would be expected with the patients CNI regimen. The addition of primidone to this patients regimen could reduce his tremors and improve his quality of life, making Answer D correct. While switching to cyclosporine modified could be considered, the patient is a fresh transplant and is currently at his goal tacrolimus trough, making Answer A as not the best option. Reducing the dose of MMF will not address his tremors and again, he is newly transplanted making Answer B as incorrect. Adding an opiate is not recommended for the management of tremors and can lead to hazardous side effects, thus Answer C is not the best choice.

**References:**

1. Koller WC, Royse VL. Efficacy of primidone in essential tremor. Neurology 1986; 36(1): 121-124.

2. Bechstein WO. Neurotoxicity of calcineurin inhibitors: impact and clinical management. Transpl Int. 200; 13(5): 313-326.

## QUESTION 16

**Testing Point:** : In the management of patients with LVADs, the Board-certified AHFTC specialist should have an understanding of potential drug-drug interactions with anticoagulation.

**Answer:** B. Cannabis

Management of INRs in patients with LVAD can be a conundrum, especially as it relates to drug-drug interactions. Substances of abuse can cause potential pharmacokinetic interactions. With the pervasive use of cannabis in today's society, providers need to be aware of potential drug-drug interactions. Warfarin is metabolized via the CYP450 hepatic enzyme complex, however each stereoisomer is metabolized differently. The S-isomer is predominantly metabolized by CYP2C9 and R-warfarin by way of CYP3A4 with lesser involvement of CYP1A1, CYP1A2, CYP2C8, CYP2C9, CYP2C18 and CYP2C19. Resultantly, factors that impact the CYP2C9 enzyme (genetic polymorphisms, other medications, etc.) alter warfarin activity. THC has the potential to inhibit CYP 3A4/4, CYP2C9, CYP2C19, and CYP2D6, whereas CBD also has the potential to inhibit CYP3A4/5, CYP2C19, CP2D6, and CYP1A2. This interaction between cannabinoids and warfarin leading to an elevated INR is well documented in the literature, making Answer B correct. Both methamphetamine and opiates are metabolized through the CYP2D6, thus should not impact warfarin metabolism, making Answers A and D incorrect. Approximately 30-50% of cocaine is metabolized by hepatic esterases and plasma pseudocholinesterase thus should not effect warfarin metabolism, making Answer C incorrect.

**References:**

1. Page RL2, Allen LA, Kloner RA, et al. Medical Marijuana, Recreational Cannabis, and Cardiovascular Health: A Scientific Statement from the American Heart Association. Circulation 2020; 142(10): e131-e152.
2. Hollbrook AM, Pereira JA, Labiris R, et al. Systematic overview of warfarin and its drug and food interactions. Arch Intern Med. 2005;165:1095-1106.

## QUESTION 17

**Testing Point:** As part of the advanced therapies evaluation, the Board-certified AHFTC specialist should recognize potential medications which could cause false positive urine drug screens.

**Answer:** C. Mexiletine

Illicit drug use is widely accepted as an absolute contraindication to many advanced medical therapies such as transplantation. Clinical practice guidelines list active or recent illicit drug use as a class III recommendation for heart transplantation and durable mechanical circulatory support. Patients with

advanced heart failure commonly have ventricular arrhythmias treated with antiarrhythmics, such as mexiletine. Mexiletine is structurally similar to amphetamine and methamphetamine which explains the false-positive amphetamine immunoassay result, making Answer C correct. Proton pump inhibitors have been documented to cause a false positive urine drug screen for TCH, making Answer C incorrect. Neither pregabalin or spironolactone have been associated with false positive urine drug screens, making Answers B and D as incorrect.

**References:**

1. Bui QM, LA Allen, Monte AA, et al. Amphetamine-positive urine drug screens in the setting of mexiletine use: A case series. J Heart Lung Transplant. 2016;35(8):1045-8.
2. Snozke CL, Kaleta EJ, Jannetto PJ, et al. False-positive amphetamine results on several drug screening platforms due to mexiletine. Clin Biochem. 2018; 58: 125-127.

## QUESTION 18

Heart Failure with Reduced Ejection Fraction

**Testing Point:** The Board-certified AHFTC specialist should recognize the indications and appropriate timing of cardiac resynchronization therapy (CRT).

**Answer:** C. Referral to electrophysiology for a cardiac resynchronization therapy (CRT) with a biventricular defibrillator

Referral to electrophysiology for a cardiac resynchronization therapy (CRT) with a biventricular defibrillator. The patient has a recently diagnosed stage C NICM on GDMT for > 6 months with persistent LV dysfunction, mod-severe MR, NYHA II and a LBBB. The American College of Cardiology/American Heart Association guidelines have a Class I indication for cardiac resynchronization therapy for patients with an EF ≤ 35%, LBBB with QRS of ≥ 150ms, NYHA class II-IV symptoms on GDMT (Level of evidence of a for NYHA class III/IV, level of evidence B for NYHA class II). The guidelines also provide class I (level of evidence of A) indication for primary prevention ICD therapy in patients with ischemic or nonischemic cardiomyopathy, EF ≤ 35%, NYHA class II-III, on GDMT with reasonable expectation of 1-year survival and who are ≥ 40 days after a myocardial infarction). CRT–D implantation in this group of patients improves mortality, morbidity and quality of life. Further, data has shown higher rates of CRT response in women and in patients with NICM.

Distractors A and B are not the best answer because there is no evidence provided to suggest that the patient has refractory end-stage heart failure. She is tolerating excellent GDMT with relatively minimal symptoms, exhibits no markers of stage D HF and has several potential interventions available to improve her clinical status with proceeding to advanced therapies. Further, if she was exhibiting end-stage heart failure and required evaluation for VAD therapy, she does not exhibit any contraindications to heart transplantation and so this would need to be considered in the evaluation process.

Distractor D is not the best answer because 1) CRT has been shown to improve functional mitral regurgitation, LV geometry and LVEF and 2) the MR is only moderate. This patient would not qualify for the inclusion and exclusion criteria for the COA PT trial showing efficacy of MitraClip for treating functional MR. Namely, the patient has not undergone full guideline directed therapy for their LV dysfunction as they have not yet received the CRT. In fact, implantation of CRT or CRT–D within the last 30 days is an exclusion criteria given the potential for improvement in LVEF, LV geometry and severity of MR. The less knowledgeable AHFTC physician may select this option if they do not recognize that the patient has a LBBB or that CRT therapy needs to be addressed first given potential improvement in EF and severity of MR.

**References:**

1. Yancy et al. 2013 ACCF/AHA Guideline for the management of Heart Failure. Circulation 2013; 128; e240-e327
2. Stone et al. Transcatheter Mitral-Valve Repair in Patients with Heart Failure. NEJM 2018; 379 (24): 2307-2318
3. Zusterzeel et al. Cardiac resynchronization therapy in women: US Food and Drug Administration meta-analysis of patient-level data. JAMA Int Medicine. 2014 Aug;174(8):1340-8.
4. Cardiac resynchronization therapy in women: US Food and Drug Administration meta-analysis of patient-level data. JAMA Int Medicine. 2014 Aug;174(8):1340-8.
5. van Bommel et al. Characteristics of heart failure patients associated with good and poor response to cardiac resynchronization therapy: a PROSPECT (Predictors of Response to CRT) sub-analysis. Eur Heart J. 2009 Oct;30(20):2470-7

## QUESTION 19

Special Etiologies

**Testing Point:** The Board-certified AHFTC specialist should know that patients with hypertrophic cardiomyopathy (HCM) and atrial fibrillation should be anticoagulated regardless of their CHA2DS2-VASc score.

**Answer:** A. Direct acting oral anticoagulant.

Data from a meta-analysis of more than 7000 patients with HCM and AF demonstrated a higher prevalence an incidence of thromboembolism (27% and 3.75/100 patients, respectively). This risk was found to be independent of the CHA2DS2-VASc score. As such, the 2020 AHA/ACC guidelines for the diagnosis and treatment of patients with HCM gives anticoagulation with either a direct acting oral anticoagulant (DOAC) or Coumadin a class I, level of evidence B–NR recommendation. DOACs are considered first-line agents given ease of use and possible improvement in outcomes and warfarin is considered a second line agent.

Distractor B, left atrial appendage occlusion device has not been well studied in HCM and should not be the

first approach for this patient.

Distractors C and D are incorrect as full dose aspirin will not provide enough anticoagulation to prevent thromboembolic embolism and anticoagulation is needed.

**References:**

1. Ommen et al. 2020 AHA/ACC Guideline for the Diagnosis and Treatment of Patients With Hypertrophic Cardiomyopathy: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. Circulation. 2020 Dec 22;142(25):e533-e557
2. Guttmann OP, Rahman MS, O'Mahony C, et al. Atrial fibrillation and thromboembolism in patients with hypertrophic cardiomyopathy: systematic review. Heart. 2014;100:465–72

## QUESTION 20

Mechanical Circulatory Support

**Testing Point:** The Board-certified AHFTC specialist should recognize hypertension as a possible cause of low flow alarms in VAD supported patients.

**Answer:** D. Initiate losartan.

The patient is presenting to the VAD clinic with progressive decline in VAD flows in the setting of significantly elevated blood pressure. His recent weight gain and dietary indiscretion are likely contributors to worsening of his blood pressure control. Elevated blood pressure increases afterload of the pump i.e. the head pressure seen by the pump as demonstrated by the HQ curves published for individual devices. Notably, the detrimental effect of hypertension on flow rates is more prominent during diastole and so peak flow rates can remain stable or see mild decreases while the effect on mean and trough flows is far more significant. Further, uncontrolled hypertension is a known risk factor for stroke one of the most debilitating adverse events seen after VAD implantation. As such, initiation of antihypertensives is the first step in managing this patient.

Distractor A is not the correct answer for several reasons. While decreasing VAD speed may decrease blood pressure, echocardiogram shows optimized VAD speed and he otherwise feels well. Decreasing VAD speed can further decrease flows and potentially increase symptoms. The correct approach to addressing his hypertension is the initiation of antihypertensives.

Distractor B. The less knowledgeable AHFTC physician may select this option if they do not recognize that hypertension rather than inappropriate VAD speed is driving the low flows. Increasing VAD speed in this clinical scenario may increase flows slightly, however speed increases can also worsen the already poorly controlled blood pressure. This, in turn, results in only slight changes in flows at the expense of even further

increasing the patient's risk for stroke and other detrimental effects of hypertension.

Distractor C. Increasing diuretic dosage is not the correct answer as volume overload and RV dysfunction are not the primary drivers of the patient's low flows. RV dysfunction which is exacerbated by fluid overload is a cause of underfilling of the LV and VAD and is on the differential of low flow alarms in a VAD supported patient. However, there is no evidence of RV failure contributing to the clinical scenario presented here. The patient JVP demonstrates euvolemia and the echocardiogram reveals only mild RV dysfunction, trivial TR and midline septum all arguing against RV dysfunction/failure causing low flow alarms. Further, in HVAD patients with RV failure causing low VAD flows, peak, mean and trough flows would all be depressed leading to low flows and low pulsatility. This patient is demonstrating low flows and high pulsatility consistent with a hypertensive waveform.

**References:**

1. Rich et al. HVAD Flow Waveform Morphologies: Theoretical Foundation and Implications for Clinical Practice. ASAIO J. 2017 Sep; 63(5): 526–535.
2. Teuteberg JJ, Slaughter MS, Rogers JG, et al. ; ADVANCE Trial Investigators: The HVAD left ventricular assist device: Risk factors for neurological events and risk mitigation strategies. JACC Heart Fail 20153: 818–828

## QUESTION 21

Heart Transplantation

**Testing Point:**. The Board-certified AHFTC specialist should know how immunosuppression must be adjusted when a heart transplant female recipient plans a pregnancy.

**Answer:** B. Discontinuation of mycophenolate mofetil before planned conception.

She will need to discontinue mycophenolate mofetil 6-12 weeks before planned conception due to severe teratogenic effects.

Mycophenolate mofetil is categorized as category D by the FDA and has known teratogenicity causing structural malformations including anophthalmia, adenopathy, hydrocephaly and fetal death. As such it is recommended that mycophenolate be discontinued 6 to 12 weeks prior to conception or at minimum immediately after unplanned pregnancy is discovered. This is detailed further in a 2018 ISH LT consensus report. This is also given a class I evidence level C indication in the 2010 ISHLT guidelines.

This was further highlighted in the recent publication in JH LT highlighting 157 pregnancies in 91 female heart transplant patients in North America. This study demonstrated that of pregnancies exposed to MPA, 63% had miscarriages and 23% of live births suffered birth defects (compared to miscarriage percentage of 17% and a birth defect percentage of 6% in patients not exposed to mycophenolate). The birth defects

in this study including duodenal atresia, laryngomalacia and facial defect. Of note, live births were noted in 69% of pregnancies, there were no neonatal deaths, a rejection rate of 9.1% was reported, and other pregnancy complications including preeclampsia were seen in 23% of patients and infections in 14% of patients. Other series have reported higher rejection rates up to 19%.

In general, it is recommended to delay planned pregnancies until after the first year after heart transplantation. Pregnancy is typically avoided in heart transplant patients with LV dysfunction, significant CAD, significant and/or uncontrolled comorbidities and who have donor specific antibodies. Ideally the transplant recipient should have no rejections in the prior year, "stable graft function", no recent infections and stable immunosuppressive dosing.

Distractor A is incorrect as the ISHLT consensus report also recommends discontinuation of proliferation signal inhibitors (PSI) 6 to 12 weeks prior to conception. Data on PSI during pregnancy is very limited however animal studies show teratogenicity including fetal demise, structural malformations and growth retardation. It is classified by the FDA as category C.

Distractors C and D are incorrect as tacrolimus has not been shown to be teratogenic and Tacrolimus use during pregnancy is considered "acceptable" as described by the 2018 ISHLT consensus statement. There is a concern for kidney dysfunction and hyperkalemia in the newborn who was exposed to tacrolimus during late pregnancy. Breast-feeding is also considered acceptable with tacrolimus. Tacrolimus is categorized by the FDA as category C.

**References:**

1. Punnoose et al. Pregnancy outcomes in heart transplant recipients. JHLT. 2020 May;39(5):473-480.
2. DeFilippis et al. Pregnancy after Heart Transplantation. Journal of Cardiac Failure. J Card Fail. 2021 Feb;27(2):176-184
3. Cochrane et al. Report from the 2018 consensus conference on immunomodulating agents in thoracic transplantation: Access, formulations, generics, therapeutic drug monitoring, and special populations. JHLT. 2020 Oct;39(10):1050-1069
4. Costanzo et al. The International Society of Heart and Lung Transplantation Guidelines for the care of heart transplant recipients. J Heart Lung Transplant. 2010 Aug;29(8):914-56.

## QUESTION 22

Acute Heart Failure New Onset.

**Testing Point:** Exam Takers should know that SGLT2i reduce HF re-admission rate and decrease CV mortality in diabetic patients.

**Answer:** B. Add empagliflozin 10 mg/d to metformin 2,000 mg/d and discontinue rosiglitazone

This patient recovered from HF exacerbation hospitalization and currently compensated during follow-up. He takes metformin and rosiglitazone for glycemic control. Current guidelines recommend SGLT2 inhibitors for diabetes control in heart failure patients and in patients with high cardiovascular risk. SGLT2 inhibitors have proven cardiovascular efficacy for patients in whom HF predominates regardless of baseline A1C. The primary mechanisms of SGLT2 inhibitors which account for their renal and cardiovascular protection in clinical trials are natriuresis and glucosuria. Rosiglitazone is contraindicated in HF patients. Empagliflozin is an SGLT2i that is indicated to improve glycemic control and reduce the risk of cardiovascular death in adults with T2DM and established cardiovascular disease.

Regarding choice D, lowering rosuvastatin dose may be selected since this statin may increase glucose level modestly but there is more benefit from statin in this case. Insulin regimen may be more effective to lower glucose level than oral hypoglycemic agents but has no proven cardioprotective effect by itself. In choice A, adding dapagliflozin to rosiglitazone and metformin seems to be an effective regimen to lower HbA1c. However, rosiglitazone (TZD) should be discontinued in HF patients as it causes fluid retention and an increase in HF hospitalizations in these patients by increasing plasma volume related to a decreased renal excretion of sodium with TZDs.

DPP-4 inhibitors have neutral or even harmful effects on cardiovascular patients. Saxagliptin or Alogliptin may increase the risk of HF decompensation in patients who already have heart or kidney disease with FDA warning posted not to use these medications in HF patients.

SGLT2i reduce HF re-admission rate and decrease CV mortality in diabetic patients. Rosiglitazone (Avandia) is contraindicated in cardiac patients due to the fact that TZDs increase cardiovascular morbidity and mortality.

**References:**

1. Zinman B at al. EMPA-REG -NEJM 2015;373:2117-28.
2. DECLARE Study. Circulation. 2019;139:2528–2536.
3. McMurray JJV et al. N Engl J Med. 2019 Sep 19.

## QUESTION 23

Mechanical Circulatory Support Devices

**Testing Point:** The AHFTC specialist should know which values are best at predicting RV function after LV implant Question Line: Which of the following combinations of hemodynamic measures or echo doppler findings are the most predictive of RV failure after LVAD implantation?Distractors: valve regurgitations, severe pulmonary HTN, high LV filling pressure.

**Answer:** D. RA pressure / PCWP ratio of 0.9 and PAPi (pulmonary artery pulsatility index) of 1.36
RV failure after LVAD placement is not an isolated problem, and therefore, assessment of RV function is very

important prior to LVAD implant. Post-LVAD RV failure decreases success of LVAD therapy and increases morbidity and mortality in these patients. A high CVP/wedge ratio is a known predictor of poor RV function. TAPSE may not be accurately measured and 1.6cm is consistent with mild RV dysfunction. High E/E' ratio correlates with high LV filling pressures. Insufficient mitral or tricuspid valve is not a contraindication to LVAD and does not provide information about degree of RV dysfunction. A moderate degree mitral regurgitation and tricuspid regurgitation are common in advanced heart failure and may easily change with volume status changes.

In clinical trials and registries, PAPi, RVSWI and CVP/PCWP ratio were also superior to other variables in predicting RV failure and need for RV assist device support following LVAD implantation.

PAPi is the pulmonary artery pulsatility index and is calculated: (systolic PA – diastolic PA) / RAP. Based on clinical trials, PAPi >2 seems to be protective for RV when considering LVAD. A calculated PAPi for this case is 1.36. There is also a different cut off value for RV Impella placement in cardiogenic shock and is <0.9. Low PAPi score is associated with elevated RAP, elevated mean PAP, elevated wedge and lower cardiac index. PAPi is prognostic of CV death and HF readmissions at 6 months.

CVP/PCWP ratio is a good marker of RV dysfunction. CVP>20mmHg and CVP/PCWP ratio >0.6 correlate with poor R function.

RVSWI is an RV stroke work index and is calculated: [(cardiac index/HR) x (mean PAP-mean RAP)]. Cut off of 600 is a predictor of risk of prolonged RV support post LVAD.

Know well RV function prior to final decision re: LVAD implantation. Know how to calculate RVSWI and PAPi.

**References:**

1. Kochav et al. Journal of Cardiac Failure 2018;24:453-449.
2. Schenk et al. Journal of Thoracic CV Surgery 2006;131:447-54
3. Drazner et al. Circulation Heart Failure. March 2013.

## QUESTION 24

Acute on Chronic Heart Failure

**Testing Point:** The AHFTC should know that resting hemodynamics are inadequate to diagnose exertional symptoms and exercise hemodynamics are required in this setting.

**Answer:** E. Right heart catheterization with exercise hemodynamics.

Measurement of hemodynamics during exercise can be very helpful to determine the cause of exertional

dyspnea in patients with unremarkable resting cardiac work-up and pulmonary work-up. Normal PCWP and normal NT-pro-BNP at rest do not preclude potential the presence of a cardiac problem. Increase in PCWP above 25 mmHg with exercise suggests cardiac etiology of dyspnea on exertion, consistent most likely with undiagnosed HFpEF. Provocative RHC can be very useful in these patients. Abnormal rise in pulmonary systolic pressure above 45 mmHg with abnormal rise in PCWP above 25 mmHg during exercise is highly diagnostic to identify HFpEF with very high sensitivity 96% and specificity 95%.

Normal saline bolus (300ml) during RHC may not be sufficient and exercise hemodynamics are more reliable to diagnose HFpEF in cath lab. Normal RV and only mildly elevated RVSP with no TR speak against significant pulmonary emboli. Chest HRCT would be unlikely positive for pulmonary parenchymal abnormalities with normal PFTs and normal CXR. Heart biopsy is not indicated without suggestion of inflammatory or infiltrative process in myocardium.

Exercise hemodynamics have been shown to enhance the diagnosis of unexplained exertional dyspnea.

Distractors: carpal tunnel syndrome, mild biatrial enlargement.

**References:**

1. Borlaug BA et al. Exercise hemodynamics enhance diagnosis of early heart failure with preserved ejection fraction. Circ HF 2010;3:588-95.

## QUESTION 25

Mechanical Circulatory Support Devices.

**Testing Point:** The AHFTC must understand the hemodynamic changes produced by different mechanical circulatory support devices

**Answer:** A. IABP, Impella, ECMO

IABP – positioned in descending aorta deflates in systole and inflates in diastole. There is augmentation of BP in diastole and peak coronary flow increases. During systole, balloon pump reduces afterload and systolic BP and increases SV, but it only modestly enhances C.O. Balloon pump is more about increasing MAP and coronary flow than unloading LV.

Impella – percutaneous LVAD – for short term use. Impella pumps blood continuously out of LV into the ascending aorta, increasing forward flow. It is a catheter mounted trans-valvular non-pulsatile axial blood flow device. As one increases flow on Impella, PV loop shifts progressively leftward, indicating greater LV unloading and greater uncoupling between increasing aortic pressure and decreasing peak LV pressure. PV loop area correlates with myocardial work and decreases with Impella support. This direct unloading also

results in decreased LAP, wedge pressure and PAP. Impella also decreases native heart work and wall stress and improves systemic perfusion.

V-A ECMO – provides full cardio- and pulmonary support for the patients whose heart and lungs can not provide adequate physiological support. ECMO withdraws deoxygenated blood from RA / venous system and returns oxygenated blood to the arterial system using a centrifugal, non-pulsatile pump. ECMO utilizes a blood gas exchange unit, a membrane oxygenator, to help to normalize acid-base abnormalities. As one increases ECMO blood flow, native SV decreases and PV loop gets narrower; systemic BP increases and the work of the heart is done by ECMO device.

The primary hemodynamic effect of ECMO is increased afterload and blood pressure but the device does not unload LV. LV has to overcome increasing afterload caused by ECMO. Since LVEDP and wedge pressure increase with ECMO, sometimes LV may need to be unloaded. The shape of the PV loop is narrower and the loop moves up and later to the right along the EDPVR curve. ECMO increases central aortic pressure and improves coronary perfusion. ECMO is not designed to unload LV unlike Impella.

The different mechanical circulatory support devices produce specific changes in the PV loops.

**References:**

1. 2015 SCAI/ACC/HFSA/STS Clinical Expert Consensus Statement on the Use of Percutaneous Mechanical Circulatory Support Devices in Cardiovascular Care. JACC 2015 vol. 65. NO. 19.
2. D. Burkhoff et al. Hemodynamics of Mechanical Circulatory Support. JACC 2015 vol. 66, NO. 23:2663-74

## QUESTION 26

Heart Transplant

**Testing Point:** know side effects of anti-rejection medications and know co-morbidity of heart transplant like renal dysfunction and CAV

**Answer:** A. Add sirolimus and discontinue tacrolimus.

This patient has more significant renal dysfunction than cardiac allograft vasculopathy (CAV). Discontinuation of tacrolimus (calcineurin inhibitor) and initiation of sirolimus (mTOR inhibitor) seems to be the most optimal therapeutic move here. Worsening renal function occurs commonly in heart transplant patients. Calcineurin inhibitors (CNIs) contribute to renal dysfunction. A transition of CNI to mTOR inhibitor can slow down progression of renal dysfunction. Sirolimus may be a substitute for CNIs to reduce CNI-induced nephrotoxicity. Lowering tacrolimus dose is not indicated with the trough level already at the lower limit of therapeutic range.
CAV is a form of chronic allograft rejection and is a major limitation of post-transplant survival. Statin

therapy and mTOR inhibitors like sirolimus demonstrated efficacy in slowing progression of CAV. Substitution of calcineurin inhibitors (CNIs) with sirolimus may improve renal function and may also slow down CAV progression. Sirolimus is not indicated for use in early posttransplant course due to increased rejection risk. In this case, the patient is 4 years after heart transplant.

Increasing prednisone or tacrolimus dose would not help with worsening renal function.
A significant rise in serum creatinine requires prompt intervention and it is not recommended to wait 3 months.

Calcineurin Inhibitors (CNIs): Cyclosporine, Tacrolimus.
Proliferation Signal Inhibitors (PSIs) / mechanistic target of rapamycine (mTOR) inhibitors: Sirolimus, Everolimus.

Distractors: CAV, different medications

**References:**
1. Gullestad L et al. Transplantation 2010;89:864-72.
2. Gustafsson F et al. JHLT 2007.
3. Costanzo MR et al. The International Society of Heart and Lung Transplantation Guidelines for the care of heart transplant recipients. JHLT 2010;29:914-56.
4. Zuckermann A at al. JHLT 2008; 27:141-9.

## QUESTION 27

Pulmonary Hypertension

**Testing Point:** Exam Takers should know that the optimal treatment for chronic, multi-segments CTEPH is pulmonary thromboembolectomy.

**Answer:** E. Refer for surgical pulmonary thromboendarterectomy

This patient has chronic thromboembolic disease (CTEPH), which is potentially treatable by surgical pulmonary thromboendarterectomy. This patient had symptomatic chronic PE proven by V/Q scan a year earlier and has not responded to Warfarin therapy.

Riociguat is a soluble guanylate cyclase (sGC) stimulator approved for CTEPH treatment, but it would not be as effective as surgical removal of pulmonary thrombi. Riociguat would be a second choice if surgery is not possible or not effective. Riociguat (Adempas) is a soluble guanylate cyclase (sGC) stimulator approved for treatment of inoperable CTEPH or persistent / recurrent CTEPH after surgery (PAH WHO Group 4) and PAH WHO Group 1.

Catheter directed thrombolytic therapy would be another option if the patient did not meet criteria for surgical thrombectomy, but it may not be effective with chronic thrombi.

Switching to rivaroxaban probably would not change the outcome. There is no proven effectiveness of NOACs over warfarin in treatment of CTEPH.

Sildenafil (phosphodiesterase inhibitor) and treprostinil (prostacyclin mimetic) have not been shown to be effective in CTEPH.

Pulmonary thromboembolectomy is a potentially curative approach for WHO IV PAH.

**References:**

1. Piazza G at al. NEJM 2011:364:351-60
2. Banks DA et al. Semin Cardiothorac Vasc Anesth 2014;18:319-30.

## QUESTION 28

Acute Heart Failure/Physical Exam

**Testing Point:** The Board-certified AHFTC specialist should know the most sensitive and specific physical exam findings for the diagnosis of heart failure.

**Answer:** C. Presence of jugular venous distention and an S3 indicate that a heart failure diagnosis is likely

*Presence of elevated jugular venous pressure and an S3 are of diagnostic utility and prognostic.* An elevated jugular venous pressure is both sensitive and specific for heart failure. Furthermore, a landmark paper study in the New England Journal demonstrated that patients with an elevated JVP have significantly worse prognosis up to five years later. Though peripheral edema is specific for heart failure, it lacks sensitivity. Pulsus alternans is associated with low cardiac output, whereas pulsus paradoxus is associated with cardiac tamponade.

**References:**

1. JACC Heart Fail. 2018 Jul;6(7):543-551. doi: 10.1016/j.jchf.2018.04.005. Epub 2018 Jun 6. PMID: 29885957.
2. N Engl J Med 2001; 345:574-81.
3. *JAMA*. 1989;261(6):884-888. doi:10.1001/jama.1989.03420060100040

## QUESTION 29

Management of Patients after LVAD

**Testing Point:** The Board-certified AHFTC specialist should know the indications for ICD placement in LVAD patients.

**Answer:** D. ICD at this time is not recommended

*Given his lack of arrhythmia pre/post operatively no ICD is indicated.*
The risk of death from cardiac arrhythmia is extremely low post LVAD in patients without history of arrhythmia. This is particularly true in patients with non-ischemic cardiomyopathy, in whom risk of arrhythmic death is low and the benefit of ICD implant is uncertain, as shown in the DANISH ICD trial. No evidence thus far, suggests benefit of CRT in LVAD patients. Given the lack of any sustained or non-sustained ventricular arrhythmia an implantable ICD nor loop recorder are indicated.

**References:**
1. JACC. Clinical electrophysiology, 4(9), 1166–1175. doi.org/10.1016/j.jacep.2018.05.006
2. Curr Heart Fail Rep. 2019 Dec;16(6):229-239. doi: 10.1007/s11897-019-00449-8.
3. Heart Rhythm. 2017 Dec;14(12):1812-1819. doi: 10.1016/j.hrthm.2017.07.027. Epub 2017 Jul 27.
4. N Engl J Med 2016; 375:1221-1230. DOI: 10.1056/NEJMoa1608029

## QUESTION 30

Management of Patients After LVAD

**Testing Point:** The Board-certified AHFTC specialist should realize the importance of optimal blood pressure management in LVAD patients.

**Answer:** B. Initiation of Lisinopril 5mg daily.

Hypertension is a common cause of heart failure post LVAD. The Ideal MAP in LVAD patients is between 65-80. This patient clearly has an elevated MAP which is decreasing his cardiac flow and is likely contributing to his heart failure symptoms. Increasing evidence specifically suggests benefit of ACE inhibitors in LVAD patients. Though further prospective study is required to confirm the benefit of ACE inhibitors, in this case they would be preferable to hydralazine given their once daily dosing and the history of diabetes and coronary disease. Though cardiac rehabilitation is beneficial in LVAD pateints, optimizing blood pressure should be done prior if possible. Furthermore, a RAMP study would be indicated if symptoms persisted despite bp control.

**References:**
1. J Am Coll Cardiol. 2019 Apr 16;73(14):1769-1778. doi: 10.1016/j.jacc.2019.01.051. PMID: 30975293.
2. ASAIO J. 2020 Aug;66(8):881-885. doi: 10.1097/MAT.0000000000001104. PMID: 32740347.
3. Yousefzai R, Brambatti M, Tran HA, Pedersen R, Braun OÖ, Baykaner T, Ghashghaei R, Sulemanjee ASAIO J. 2020 Apr;66(4):409-414. doi: 10.1097/MAT.0000000000001022. PMID: 31192845.

## QUESTION 31

Palliative Care/Hospice in End Stage Heart Failure

**Testing Point:** The AHFTC specialist should know the effects and indications for specialized palliative care in advanced HF patients.

**Answer:** C. Specialized palliative care consultation improves quality of life measures, in inpatient and outpatient settings.

*Palliative care interventions have shown benefit in quality of life in inpatient and outpatient settings*

Palliative care is both a medical specialty (aka Specialized Palliative Care) as well as an overall approach to care that improves quality of life and relieves suffering for patients with serious illness, regardless of prognosis. It differs from hospice in that patients are not required to be at end of life and patients may be on life prolonging therapy. Primary palliative care refers to the routine administration of palliative interventions, such as basic pain management and routine end of life discussions. Primary palliative care can be administered by various care providers such as internists, cardiologists, and advanced heart failure physicians. Conversely, specialized palliative care providers (those with extra dedicated training in palliation) may be of assistance with complex medical decision making at the end of life, such as contentious transitions to hospice or patients with familial conflicts. Randomized Interventions in palliative care have shown improvement in quality of life, psychosocial domains and costs but have not been proven to prolong survival in randomized studies. Palliative care consultation is appropriate for advanced heart failure patients even if they are interested in life prolonging care, such as AICDs and stents.

**References:**

1. J Am Coll Cardiol. 2017 Jul 18;70(3):331-341. doi: 10.1016/j.jacc.2017.05.030.
2. J Am Coll Cardiol. 2017 Oct 10;70(15):1919-1930. doi: 10.1016/j.jacc.2017.08.036.
3. Circulation. 2009 Dec 22;120(25):2597-606. doi: 10.1161/CIRCULATIONAHA.109.869123.
4. Gelfman, L.P., Kavalieratos, D., Teuteberg, W.G. et al. Primary palliative care for heart failure: what is it? How do we implement it?. Heart Fail Rev 22, 611–620 (2017). https://doi.org/10.1007/s10741-017-9604-9

## QUESTION 32

New Onset Heart Failure/Drug Toxicity

**Testing Point:** The Board-certified AHFTC specialist should be able to recognize chloroquine toxicity

**Answer:** D. Endomyocardial biopsy

*Cardiac Biopsy*. Chloroquine toxicity is a rare but severe side effect of sustained use of the drug. It presents

with heart block and its most severe form heart failure. Diagnosis requires cardiac biopsy and should be performed. Electron microscopy of biopsy tissue is notable for numerous autophagic vacuoles and curvilinear bodies (see Figure 2). Though an MRI could be useful is not specific for the diagnosis of chloroquine toxicity. As the patient may benefit from chloroquine, a biopsy is indicted to rule out disease. Genetic testing, though could be useful, would not be diagnostic in this scenario.

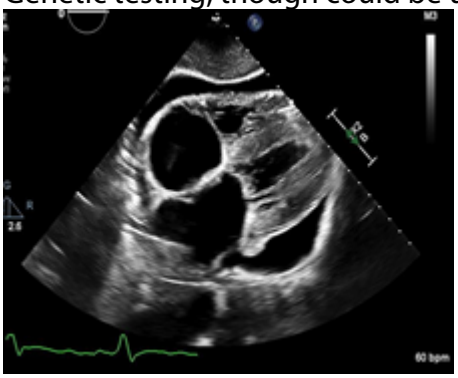

*Figure 1. Parasternal short axis image of patient.*

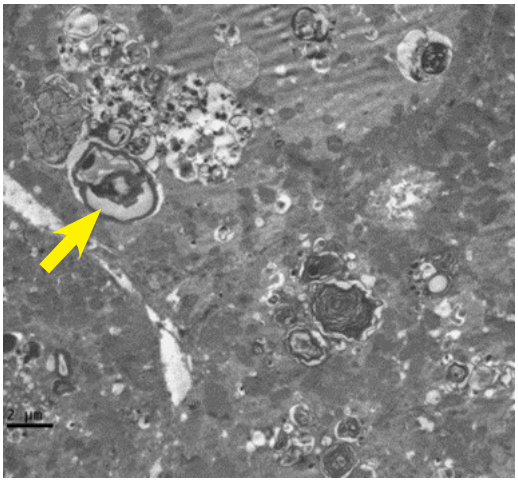

*Figure 2. Electron microscopy of a patient with chloroquine toxicity. Note curvilinear bodies (yellow arrow).*

**References:**

1. N Engl J Med 1987; 316:191-193 https://doi: 10.1056/NEJM198701223160405
2. Drug Saf 41, 919–931 (2018). https://doi.org/10.1007/s40264-018-0689-4
3. ESC Heart Fail. 2018 Jun;5(3):372-375. doi: 10.1002/ehf2.12276.

## QUESTION 33

Early Complications Heart Transplant

**Testing Point:** The Board-certified AHFTC specialist should recognize PRES as a cause of mental status changes in the immediate peri-transplant period.

**Answer:** B. Magnetic Resonance Imaging of the brain.

*MRI of the Brain.* Given the patients symptoms and elevated blood pressure, tacrolimus and creatinine she most likely has posterior reversible encephalopathy syndrome (PRES). PRES is characterized by a headache, seizures, altered mental status and visual loss. MRI is notable for white matter vasogenic edema affecting the posterior occipital and parietal lobes of the brain predominantly. Though infection is possible, the clinical scenario and laboratory findings are more consistent with PRES. Though CMV is in the differential, it is less likely given the constellation of clinical findings. Leaving PRES untreated may lead to seizures and possibly death. Though non contrast CT would identify intracranial hemorrhage, it would not identify PRES.

**References:**

1. Lancet Neurol. 2015 Sep;14(9):914-925. doi: 10.1016/S1474-4422(15)00111-8.
2. Curr Opin Neurol. 2019 Feb;32(1):25-35. doi: 10.1097/WCO.0000000000000640

## QUESTION 34

**Answer:** A. Reduction of HF hospitalizations in all NYHA Class III patients.

The goal of the question is to review the main findings of the CHAMPION trial, which concluded that management of NYHA Class III HF, regardless of LVEF, based on home transmission of pulmonary artery pressure with an implanted pressure sensor (CardioMEMS) has significant long term benefit in lowering hospital admission rates for HF.

**References:**

Abraham WT, Adamson PB, Bourge RC, Aaron MF, Costanzo MR, Stevenson LW, Strickland W, Neelagaru S, Raval N, Krueger S, Weiner S, Shavelle D, Jeffries B, Yadav JS; CHAMPION Trial Study Group. Wireless pulmonary artery haemodynamic monitoring in chronic heart failure: a randomised controlled trial. Lancet. 2011 Feb 19;377(9766):658-66. doi: 10.1016/S0140-6736(11)60101-3. Erratum in: Lancet. 2012 Feb 4;379(9814):412. PMID: 21315441.

## QUESTION 35

**Answer:** B. Hydralazine/Isosorbide dinitrate
The goal of the question is to understand appropriate treatment of suspected peripartum cardiomyopathy in a patient who is breastfeeding.

**References:**
1. Vera Regitz-Zagrosek, Jolien W Roos-Hesselink, Johann Bauersachs, Carina Blomström-Lundqvist, Renata Cífková, Michele De Bonis, Bernard Iung, Mark Richard Johnson, Ulrich Kintscher, Peter Kranke, Irene Marthe Lang, Joao Morais, Petronella G Pieper, Patrizia Presbitero, Susanna Price, Giuseppe M C Rosano, Ute Seeland, Tommaso Simoncini, Lorna Swan, Carole A Warnes, ESC Scientific Document Group, 2018 ESC Guidelines for the management of cardiovascular diseases during pregnancy: The Task Force for the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology (ESC), European Heart Journal, Volume 39, Issue 34, 07 September 2018, Pages 3165–3241.

## QUESTION 36

**Answer:** B. CRT-D

The goal of the question is to recognize a patient with NYHA Class III HF who demonstrates signs and symptoms of advanced disease and refer them for appropriate device and other advanced therapies, recognizing that there is no stand-alone indication for CRT in severe mitral regurgitation.

**References:**
1. Epstein AE, Dimarco JP, Ellenbogen KA, Estes NA 3rd, Freedman RA, Gettes LS, Gillinov AM, Gregoratos G, Hammill SC, Hayes DL, Hlatky MA, Newby LK, Page RL, Schoenfeld MH, Silka MJ, Stevenson LW, Sweeney MO; American College of Cardiology; American Heart Association Task Force on Practice Guidelines; American Association for Thoracic Surgery; Society of Thoracic Surgeons. ACC/AHA/HRS 2008 Guidelines for device-based therapy of cardiac rhythm abnormalities. Heart Rhythm. 2008 Jun;5(6):e1-62. doi: 10.1016/j.hrthm.2008.04.014. Epub 2008 May 21. Erratum in: Heart Rhythm. 2009 Jan;6(1):e2. PMID: 18534360.

## QUESTION 37

**Answer:** D. Mechanical aortic valve

The goal of the question is to highlight important surgical considerations regarding valvular pathology prior to/during LVAD implantation. Based on the guidelines, it is a Class I recommendation to replace a pre-existing aortic mechanical valve with a bioprosthetic valve (or oversewing the valve) at the time of LVAD implantation. With adequate anticoagulation, pre-existing mechanical mitral valves in LVAD patients have not been shown to result in clinically significant thromboembolic events. Therefore, routine replacement of a mechanical mitral valve with a bioprosthesis is not recommended, but may be considered.

**References:**
1. Feldman D, Pamboukian SV, Teuteberg JJ, Birks E, Lietz K, Moore SA, Morgan JA, Arabia F, Bauman ME, Buchholz HW, Deng M, Dickstein ML, El-Banayosy A, Elliot T, Goldstein DJ, Grady KL, Jones K, Hryniewicz K, John R, Kaan A, Kusne S, Loebe M, Massicotte MP, Moazami N, Mohacsi P, Mooney M, Nelson T, Pagani F, Perry W, Potapov EV, Eduardo Rame J, Russell SD, Sorensen EN, Sun B, Strueber M, Mangi AA, Petty MG, Rogers J; International Society for Heart and Lung Transplantation. The 2013 International Society for Heart and Lung Transplantation Guidelines for mechanical circulatory support: executive summary. J Heart Lung Transplant. 2013 Feb;32(2):157-87. doi: 10.1016/j.healun.2012.09.013. PMID: 23352391.

## QUESTION 38

Heart failure with reduced ejection fraction

**Testing Point:** The Board-certified AHFTC specialist should prescribe cardiac rehabilitation to eligible patients to improve functional status, improve quality of life, and reduce mortality

**Answer:** C. Refer to cardiac rehabilitation

Distractors: The minimally-qualified AHFTC specialist might select other answers:

Ivabradine is recommended to reduce heart failure hospitalizations in patients with HFrEF, but it is indicated for patients with resting heart rate above 70 bpm despite a beta-blocker or those unable to tolerate a beta-blocker. Mineralocorticoid antagonists are recommended to reduce morbidity and mortality in patients with LVEF<35%, but patients with significant renal dysfunction (creatinine >2.5 mg/dl for men or >2.0 for women) or hyperkalemia were excluded from trials due to potential for harm. The serum potassium level is not provided. A nutritional assessment may be useful for education and management of heart failure, diabetes, weight loss, and malnutrition. This patient's diabetes is controlled, and no history of obesity or dietary non-adherence is mentions.

Clinical trials and meta-analyses have shown that exercise training such as cardiac rehabilitation is associated reduction in all-cause mortality and heart failure hospitalizations.

**References:**
1. Yancy CW, Jessup M, Bozkurt B et al. 2013 ACCF/AHA guideline for the management of heart failure: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol 2013; 62:e147-239.
2. O'Connor CM, Whellan DJ, Lee KL et al. Efficacy and Safety of Exercise Training in Patients With Chronic Heart Failure: HF-ACTION Randomized Controlled Trial. JAMA 2009;301:1439-1450.
3. Piepoli MF, Davos C, Francis DP, Coats AJ. Exercise training meta-analysis of trials in patients with chronic heart failure (ExTraMATCH). BMJ 2004;328:189.

## QUESTION 39

Heart failure with preserved ejection fraction

**Testing Point:** The Board-certified AHFTC specialist should be able to diagnose heart failure with preserved ejection fraction

**Answer:** D. Pulmonary capillary wedge pressure of 16 mmHg

Distractors: The minimally-qualified AHFTC specialist might select another answer…

The E/e' ratio from Doppler echocardiography can provide a non-invasive estimation of left ventricular diastolic function that correlates with invasive measurement of left ventricular filling pressures. There is consensus that E/e' ratio ≥15 is most supportive, with values 10-14 having less strong association. Elevated B-type natriuretic peptide levels are useful in the diagnosis of HFpEF, but normal values do not exclude heart failure. Levels may be normal in obese patients with heart failure. NT-proBNP level >220 pg/dl is supportive of a diagnosis. The NT-proBNP value in this patient is not supportive, so other evidence is needed. Left atrial enlargement, consistent with increased left ventricular filling pressures, can be useful in the diagnosis of HFpEF. There is consensus that left atrial volume index > 34 ml/m2 supports a diagnosis of HFpEF. The left atrial volume index of this patient is within normal limits.

A diagnosis of HFpEF requires clinical signs/symptoms of heart failure, evidence of normal/preserved ejection fraction, and echocardiographic or hemodynamic evidence of elevated left ventricular filling pressures. This patient has dyspnea and preserved LVEF and the risk factors of age and obesity. A pulmonary capillary wedge pressure of ≥ 15 mmHg at rest is abnormal and definitive for a diagnosis of HFpEF in this scenario.

**References:**

1. Yancy CW, Jessup M, Bozkurt B et al. 2013 ACCF/AHA guideline for the management of heart failure: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol 2013; 62:e147-239.
2. Redfield MM. Heart Failure with Preserved Ejection Fraction. N Engl J Med 2016; 375:1868-1877.
3. Pieske B, Tschöpe C, de Boer RA et al. How to diagnose heart failure with preserved ejection fraction: the HFA–PEFF diagnostic algorithm: a consensus recommendation from the Heart Failure Association (HFA) of the European Society of Cardiology (ESC). European Journal of Heart Failure 2020; 22:391-412

## QUESTION 40

Specific etiologies of heart failure (hypertrophic cardiomyopathy)

**Testing Point:** The Board-certified AHFTC specialist should know when it is appropriate to refer patients

with hypertrophic cardiomyopathy for genetic testing.

**Answer:** B. Refer for genetic testing

Distractors: The minimally-qualified AHFTC specialist might select another answer…

Primary prevention defibrillators are appropriate for some patients with hypertrophic cardiomyopathy, but this patient does not meet major (extreme hypertrophy, family history of SCD likely attributable to HCM, left ventricular dysfunction, or apical aneurysm) or minor criteria (episodes of non-sustained ventricular tachycardia, unexplained syncope, or extensive scar burden) per 2020 AHA/ACC guidelines. The death of a close family member < 50 years warrants investigation of the cause, but there is a plausible alternative here. This patient has good functional status (NYHA 1), so septal reduction therapy is not recommended, even with a resting gradient >50 mmHg. Strenuous activity and competitive sports should be avoided, but moderate recreational activity, as in this patient without limiting symptoms, is beneficial.

Offering genetic testing to patients with hypertrophic cardiomyopathy (and other inherited heart disease) is recommended (class I) to elucidate the genetic basis, evaluate for phenocopies, and facilitate screening at-risk family members. This patient has multiple family members at risk who should be screened traditionally or via cascade testing if a pathologic variant is found.

**References:**

1. Ommen SR, Mital S, Burke MA et al. 2020 AHA/ACC Guideline for the Diagnosis and Treatment of Patients With Hypertrophic Cardiomyopathy: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. J Am Coll Cardiol 2020; 76:e159-e240.

## QUESTION 41

Mechanical circulatory support (durable left ventricular assist devices)

**Testing Point:** The Board-certified AHFTC specialist should know how to treat pump pocket infection.

**Answer:** A. Surgical washout and debridement

Distractors: The minimally-qualified AHFTC specialist might select another answer…

While extended intravenous antibiotics will be necessary, antibiotics alone are ineffective without adequate source control. Even if blood cultures return negative, oral antibiotics are unlikely to be effective for a pump-pocket infection with abscess. The patient is not a candidate for transplant while he has an untreated pump pocket infection and possibly bacteremia (cultures pending). In addition, he is likely to have a prolonged wait as group O and needs treatment first. Pump removal at the time of transplant, following

treatment, is the long-term cure.

The patient has a pump pocket infection, as evidenced by fever, tenderness, leukocytosis, and a fluid collection with stranding adjacent to the pump. He was previously treated with oral antibiotics for superficial MRSA driveline infection, but some bacteria like MRSA form a biofilm that resists antimicrobial treatment, resulting in a driveline or pump pocket abscess. Adequate source control is essential with debridement, washout, and possibly a wound-vac.

**References:**

1. Kirklin JK, Pagani FD, Goldstein DJ et al. American Association for Thoracic Surgery/International Society for Heart and Lung Transplantation guidelines on selected topics in mechanical circulatory support. J Heart Lung Transplant 2020; 39:187-219.

## QUESTION 42

Pulmonary hypertension

**Testing Point:** The Board-certified AHFTC specialist should be able to treat group 1 pulmonary hypertension.

**Answer:** C. Nifedipine

Distractors: The minimally-qualified AHFTC specialist might select another answer…

Combination therapy is appropriate for patients without vasoreactivity or those with vasoreactivity who do not improve on a calcium channel blocker. This patient had a positive vasoreactivity study, and is treatment naïve. Parenteral or inhaled prostacyclins are not recommended as initial therapy for WHO functional class II symptoms. Verapamil has significant negative inotropy and should generally be avoided in pulmonary hypertension.

This patient has group 1 pulmonary hypertension, WHO functional class II symptoms, and a positive vasoreactivity test (PA mean pressure decreases by >10 mmHg to < 40 mmHg). Calcium channel blockers such as nifedipine, amlodipine, and diltiazem are recommended treatment options.

**References:**

1. Klinger JR, Elliott CG, Levine DJ et al. Therapy for Pulmonary Arterial Hypertension in Adults: Update of the CHEST Guideline and Expert Panel Report. CHEST 2019; 155:565-586.
2. Galiè N, Humbert M, Vachiery JL et al. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension: The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS): Endorsed by:

Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT). European Heart Journal 2016; 37:67-119.
3. McLaughlin VV, Archer SL, Badesch DB et al. ACCF/AHA 2009 expert consensus document on pulmonary hypertension a report of the American College of Cardiology Foundation Task Force on Expert Consensus Documents and the American Heart Association developed in collaboration with the American College of Chest Physicians; American Thoracic Society, Inc.; and the Pulmonary Hypertension Association. J Am Coll Cardiol 2009; 53:1573-619.

## QUESTION 43

Heart Failure with Reduced Ejection Fraction

**Testing Point:** The Board-certified AHFTC specialist should recognize optimal management strategies at time of hospital discharge to prevent rehospitalization.

**Answer:** D. Follow up appointment with provider within 1-2 weeks of discharge.

According to the 2013 American College of Cardiology/American Heart Association guidelines, it is a Class II recommendation for a post-discharge follow-up appointment within 14 days of hospital discharge. Given the complexity of the heart failure medication regimen, and changes that occur once a patient is home in regards to lifestyle and diet, the first post-discharge visit affords an important opportunity to perform medication reconciliation, patient education and treatment optimization. Scheduling an appointment prior to patient discharge may improve rates of appointment completion. Close follow-up with a heart failure specific disease management program when available is recommended.

Transitioning from oral furosemide to oral torsemide may be considered in patients on high (>80 mg/day) doses of furosemide, with demonstrated lack of response or decreased bioavailability. However, in this patient, the dose of furosemide was recently reduced, and there is no existing evidence that a switch to torsemide at this juncture would reduce readmission.

Distractor B may be playing a role in this patient's care, but more so in the longer term. This answer may be chosen given the patient's prior worsening renal function, however is less likely to impact the patient's short-term risk of readmission. In fact, based on existing diuretic clinical trial data, worsening renal function may be expected with effective diuresis, without impact on longer term outcomes.

Close monitoring of renal function and serum potassium is encouraged given the nuances of diuretic and other heart failure guideline directed medical therapies, however usually prompted by a specific recent change in medication or dosing. This intervention should be considered as part of the more comprehensive post-discharge follow up appointment.

**References:**
1. Hollenberg SM et al. 2019 ACC Expert Consensus Decision Pathway on Risk Assessment, Management, and Clinical Trajectory of Patients Hospitalized With Heart Failure. A Report of the American College of Cardiology Solution Set Oversight Committee. JACC 2019(74);15:1966-2011.
2. Yancy CW et al. 2013 ACCF/AHA guideline for the management of heart failure: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2013;62:e147–239.
3. Felker GM, Lee KL, Bull DA, et al. Diuretic strategies in patients with acute decompensated heart failure. N Engl J Med. 2011;364:797–805.

## QUESTION 44

Special Etiologies

**Testing Point:** The Board-certified AHFTC specialist should recognize the indications for genetic counseling and testing.

**Answer:** D. Genetic counseling and testing.

Arrhythmogenic inherited cardiomyopathies (ACM) should be recognized by the AHFTC as a cause for otherwise unexplained cardiomyopathy. This patient has a myopericarditis like syndrome, with chest pain, elevated troponin and abnormal findings on cardiac magnetic resonance imaging. With the patient's mild biventricular cardiomyopathy, one key component of her history that should not be ignored is the family history of sudden cardiac death in an uncle. A 3-generational family history should be performed in any patient presenting with cardiomyopathy. Additionally, she has T wave inversions in V1 and V2, RV systolic dysfunction and a history of competitive athletics the latter of which is a known risk factor for ACM progression. Arrhythmogenic inherited cardiomyopathies are increasingly recognized as having an inflammatory or myocarditis like presentation particularly when due to gene mutations in the desmosomal genes such as DSP. Appropriate referral for genetic counseling and testing can help identify inherited cardiomyopathies, and thus inform arrhythmic as well as family member risk.

Answer choices A-C may all be considered as part of the diagnostic work up in such a patient however there are a few caveats to consider. Cardiac fluorodeoxyglucose-positron emission tomography imaging can identify cardiac inflammation, for example in the diagnosis of cardiac sarcoidosis. In this patient, it is unlikely to yield further information. Endomyocardial biopsy would be indicated as the next step if the patient presented with hemodynamic compromise and/or documented ventricular arrhythmia or conduction disease specifically to diagnose more fulminant forms of myocarditis such as giant cell myocarditis. The history of an upper respiratory tract infection 3 months prior is a distractor for the risk of myocarditis. Lastly, coronary angiography is likely to be of low yield in this young patient with atypical chest pain features for acute coronary syndrome. It is reasonable however to rule out coronary artery disease, but this could be

considered via coronary computed tomography instead given her low risk profile and adequately controlled heart rate.

**References:**

1. Hershberger RE, Givertz MM, Ho CY, Judge DP, Kantor PF, McBride KL, Morales A, Taylor MRG, Vatta M, Ware SM. Genetic Evaluation of Cardiomyopathy-A Heart Failure Society of America Practice Guideline. J Card Fail. 2018 May;24(5):281-302. doi: 10.1016/j.cardfail.2018.03.004. Epub 2018 Mar 19. PMID: 29567486.
2. Marcus FI, McKenna WJ, Sherrill D, Basso C, Bauce B, Bluemke DA, Calkins H, Corrado D, Cox MG, Daubert JP, Fontaine G, Gear K, Hauer R, Nava A, Picard MH, Protonotarios N, Saffitz JE, Sanborn DM, Steinberg JS, Tandri H, Thiene G, Towbin JA, Tsatsopoulou A, Wichter T, Zareba W. Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia: proposed modification of the task force criteria. Circulation. 2010 Apr 6;121(13):1533-41. doi: 10.1161/CIRCULATIONAHA.108.840827. Epub 2010 Feb 19. PMID: 20172911; PMCID: PMC2860804.
3. Cooper LT et al. The Role of Endomyocardial Biopsy in the Management of Cardiovascular Disease: A Scientific Statement from the American Heart Association, the American College of Cardiology, and the European Society of Cardiology. Circulation. 2007;116:2216-2233.

## QUESTION 45

Heart Failure with Preserved Ejection Fraction

**Testing Point:** The Board-certified AHFTC specialist should recognize the importance of identifying the underlying etiology in patients with heart failure with preserved ejection fraction.

**Answer:** A. Cardiac fluorodeoxyglucose-positron emission tomography imaging (FDG-PET).

This patient presents with new heart failure with preserved ejection fraction. Risk factors for heart failure with preserved ejection fraction in the setting of cardiometabolic syndrome include her history of diabetes and obesity, and she is also noted to be hypertensive during this admission. However, there are several additional key historical points to consider and prompt further evaluation for specific underlying etiologies for heart failure in the setting of preserved ejection fraction. Key history includes conduction disease and low voltage on ECG which should prompt consideration of infiltrative cardiomyopathies. Given her young age and chronic cough, sarcoidosis is the most likely possibility. Cardiac sarcoidosis specifically can present with ventricular arrhythmias, conduction disease and/or heart failure either with systolic ventricular dysfunction or as a restrictive phenotype with preserved ejection fraction. Diagnosis of cardiac sarcoidosis is made through a combination of histopathologic findings and advanced cardiac imaging including cardiac FDG-PET and magnetic resonance (CMR) imaging, both which would be reasonable in this case. Diagnosis carries treatment implications, namely in the form of immunosuppression.

Though the low voltage and conduction disease should also raise suspicion for amyloidosis, the patient's

younger age and other presenting features such a chronic cough make sarcoidosis more likely that transthyretin amyloidosis for which technetium pyrophosphate scan may be useful. Genetic counseling would be indicated if family history and/or further cardiac imaging reveal concern for an inherited infiltrative cardiomyopathy.

In regards to an ischemic evaluation given risk factors of hypertension, diabetes and obesity, FDG-PET imaging can include nuclear stress imaging to evaluation for potential reversible ischemia as a cause of her heart failure as well.

**References:**

1. Birnie DH, Sauer WH, Bogun F, Cooper JM, Culver DA, Duvernoy CS, Judson MA, Kron J, Mehta D, Cosedis Nielsen J, Patel AR, Ohe T, Raatikainen P, Soejima K. HRS expert consensus statement on the diagnosis and management of arrhythmias associated with cardiac sarcoidosis. Heart Rhythm. 2014 Jul;11(7):1305-23. doi: 10.1016/j.hrthm.2014.03.043. Epub 2014 May 9. PMID: 24819193.

## QUESTION 46

Mechanical Circulatory Support

**Testing Point:** The Board-certified AHFTC specialist should recognize the role of shared decision making in patients with end-stage heart failure.

**Answer:** C. Set up a session with the patient Discuss to discuss goals of care with the patient

This patient has end-stage heart failure secondary to ischemic cardiomyopathy. He is demonstrating inotropic dependence despite treatment for decompensated heart failure. Additionally, he has CKD and recent stroke. Goals of care should be discussed with end-stage heart failure patients, including in the evaluation for advanced therapy candidacy. The patient's desire to be discharged home after this prolonged complex hospitalization should not be used in isolation to make inferences about the patient's longer and short-term goals of care. The role of chronic home inotropes should be discussed, including a bridge to LVAD versus palliative strategy and prognosis. The patient has capacity to make independent medical decisions and therefore answer choice A (contact the patient's healthcare power of attorney) should only be done with the patient's agreement. Prior to outpatient scheduling of admission for LVAD implantation (choice B), the patient's goals of care and hesitancy to proceed with LVAD at this time should be further explored. His history of stroke and renal dysfunction should be presented as risks for post LVAD complications such as recurrent stroke or need for dialysis, to allow shared decision making with patient. Distractor D, neurocognitive testing, may be employed during advanced heart failure therapy evaluation in select patients. However, a patient's desire to be discharged home should not be misinterpreted as need for neurocognitive evaluation, and should instead prompt better understanding of patient's understanding of his disease and discussion of goals of care.

**References:**
1. Mehra et al. The 2016 International Society for Heart Lung Transplantation Listing criteria for heart transplantation: A 10-year update. JHLT. 2016;35(1):P1-23.
2. Yancy et al. 2013 ACCF/AHA Guideline for the management of Heart Failure. Circulation 2013; 128; e240-e327.

## QUESTION 47

Heart Transplantation

**Testing Point:** The Board-certified AHFTC specialist should recognize drug-drug interactions when managing immunosuppression regimens post-transplantation.

**Answer:** A. Discontinuation of fluconazole.

Tacrolimus is a calcineurin inhibitor (CNI) and part of the backbone of the immunosuppression regimen post transplantation. Several drug-drug interactions may occur and should be considered when aiming for optimal drug level maintenance to minimize risk of infection. The azole antifungal agents inhibit metabolism of CNI through their interactions with CYP3A4 and P-glycoprotein, both of which are important in CNI metabolism. When the patient was taken off fluconazole two weeks prior to presentation, this likely resulted in reduced tacrolimus levels due to less inhibition of CYP3A4 and P-glycoprotein by the azole.

P-glycoprotein is a membrane associated protein. In the intestine, it causes intestinal efflux of drug from the enterocyte back into the gut, thereby reducing CNI levels. However, in the setting of severe diarrhea, intestinal epithelial cells may actually be destroyed, thereby abrogating P-glycoproteins and increasing CNI levels through increased absorption. Therefore, answer choice B is incorrect, as tacrolimus levels would have been expected to have increased during the patient's recent severe diarrheal illness.

Though nonadherence to immunosuppression medications (answer choice C) can result in decreased tacrolimus levels, the discontinuation of the azole is more likely contributing. Drug adherence and tolerance should be routinely assessed post-transplant. Inappropriate timing of tacrolimus levels would result in a higher measured tacrolimus level if drawn soon after a dose is given, or prior to the actual trough.

**References:**
1. Lindenfeld J, Miller GG, Shakar SF, Zolty R, Lowes BD, Wolfel EE, Mestroni L, Page RL 2nd, Kobashigawa J. Drug therapy in the heart transplant recipient: part I: cardiac rejection and immunosuppressive drugs. Circulation. 2004 Dec 14;110(24):3734-40. doi: 10.1161/01.CIR.0000149745.83186.89. PMID: 15596559.

## QUESTION 48

HFrEF Stage C

**Testing Point:** SGLT2 inhibitors reduce MACE

**Answer:** A. Initiate dapagliflozin

Consider an SGLT2 inhibitor when your patient has established ASCVD, HF, CKD or is at high risk for ASCVD.

Distractor 1
Decrease Sacubitril/Valsartan
Will be selected due to perception of marginal blood pressure
Distractor 2
Refer for CPET
Will be selected due to duration of heart failure and lack of knowledge of high-risk features (I NEED HELP)
Distractor 3
Decrease metoprolol succinate to 100mg daily
Will be selected due to borderline resting heart rate

**References:**

1. Yancy, Clyde W., et al. "2017 ACC expert consensus decision pathway for optimization of heart failure treatment: answers to 10 pivotal issues about heart failure with reduced ejection fraction: a report of the American College of Cardiology Task Force on Expert Consensus Decision Pathways." Journal of the American College of Cardiology 71.2 (2018): 201-230.

## QUESTION 49

HFrEF Stage C

**Testing point:** SGLT2 inhibitors can precipitate euglycemic DKA

**Answer:** B. Obtain urine and serum ketones

Euglycemic DKA in a DM patient on SGLT2 inhibitors

**References:**

1. Rosenstock, Julio, and Ele Ferrannini. "Euglycemic diabetic ketoacidosis: a predictable, detectable, and preventable safety concern with SGLT2 inhibitors." Diabetes care 38.9 (2015): 1638-1642.

Distractor 1 - Admit patient for evaluation of advanced HF therapies
Will be selected due to lack of knowledge of cardiogenic shock signs
Distractor 2 - Obtain GI consult
Will be selected due to lack of knowledge of SGLT2 inhibitor adverse effects

Distractor 3 Admit for IV fluids
Will be selected due to mild AKI

## QUESTION 50

Pulmonary Hypertencsion

**Testing point:** Recognize amiodarone-induced thyrotoxicity

**Answer:** D. Obtain thyroid function tests

Distractor 1 - Obtain transferrin saturation
Will be selected as cause of high output heart failure, less useful as ferritin not indicative of iron deficiency
Distractor 2 - Refer for infectious disease evaluation
Will be selected as cause of high output elevated CO/CI and low SVR
Distractor 3 - Refer patient to bariatric clinic
Will be selected as cause of high output/elevated CO/CI and low SVR

Rationale: Recognize amiodarone induced thyrotoxicity leading to pulmonary hypertension/high output heart failure

**References:**

1. von Haehling, Stephan, et al. "Iron deficiency in heart failure: an overview." JACC: Heart Failure 7.1 (2019): 36-46
2. Marvisi, Maurizio, et al. "Pulmonary hypertension is frequent in hyperthyroidism and normalizes after therapy." European journal of internal medicine 17.4 (2006): 267-271.

## QUESTION 51

Special etiologies of HF

**Testing point:** Recognize myocarditis associated with check point inhibitors

**Answer:** C. Pembrolizumab

Distractor 1 – Carboplatin
Will be selected due to lack of knowledge that this drug is unlikely to have cardiac toxicity.
Distractor 2 - Pamextrexed
Will be selected due to lack of knowledge that antifolate anti-neoplastic agents are unlikely associated with cardiac effects
Distractor 3- Gemcitabine

Will be selected due to lack of knowledge that nucleoside analogs are unlikely to have cardiotoxic effects

Myocarditis associated with check point inhibitors. Choices A, B, and D are not typically associated with myocarditis.

**Refrences**

1. Mahmood, Syed S., et al. "Myocarditis in patients treated with immune checkpoint inhibitors." Journal of the American College of Cardiology 71.16 (2018): 1755-1764.

## QUESTION 52

HFpEF – Stage A

**Testing point:** Heart Failure Prevention

**Answer:** A. Obtain BNP or NT-proBNP

Distractor 1- Refer for weight loss surgery
Will be selected due to lack of knowledge that bariatric surgery should be considered for adults with a BMI >40kg/m$^2$ or BMI >35 kg/m$^2$ with obesity-related comorbid conditions who are motivated to lose weight and who have not responded to behavioral treatment (with or without pharmacotherapy) with sufficient weight loss to achieve targeted health out- come goals
Distractor 2 - Obtain an exercise treadmill test
Will be selected due to lack of knowledge of recommendations regarding ETT in asymptomatic patients
Distractor 3 - Refer for a computerized tomography of the chest
Will be selected due to patients smoking history

Utilizing natriuretic peptide biomarker-based screening for those at risk of developing HF, followed by team-based care including a cardiovascular specialist optimizing guideline-directed medical therapy (GDMT), to prevent the development of left ventricular dysfunction (systolic or diastolic) or new-onset HF.

**References:**

1. Jensen, Michael D., et al. "2013 AHA/ACC/TOS guideline for the management of overweight and obesity in adults: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines and The Obesity Society." Journal of the American college of cardiology63.25 Part B (2014): 2985-3023
2. Lauer, Michael, et al. "Exercise testing in asymptomatic adults: a statement for professionals from the American Heart Association Council on Clinical Cardiology, Subcommittee on Exercise, Cardiac Rehabilitation, and Prevention." Circulation112.5 (2005): 771-776.
https://www.uspreventiveservicestaskforce.org/uspstf/recommendation/lung-cancer-screening

## QUESTION 53

MCS

**Testing point:** The AHFTC Physician should know when to refer patients with advanced HF to palliative care.

**Answer:** C. Referral for palliative care

Distractor 1 Referral for evaluation for OHT
Will be selected due to lack of knowledge of contraindications to transplantation – evidence of liver cirrhosis
Distractor 2 Referral for upgrade to CRT
Will be selected due to lack of knowledge of CRT guidelines
Distractor 3 Referral for durable LVAD implantation
Will be selected due to lack of knowledge of MCS contraindications – evidence of liver cirrhosis suggestive of alcohol use disorder.

## QUESTION 54

Heart Transplant

**Answer:** B. Gemfibrozil

Distractor 1. Ketoconazole increases Tacrolimus Levels
Distractor 2. Diltiazem increases Tacrolimus Levels
Distractor 3. Fluoxetine may increase Tacrolimus levels

Recent initiation of gemfibrozil can lower tacrolimus levels

**References:**

1. Page, Robert L., Geraldine G. Miller, and JoAnn Lindenfeld. "Drug therapy in the heart transplant recipient: part IV: drug–drug interactions." Circulation 111.2 (2005): 230-239.

## QUESTION 55

**Testing Point:** The Board-certified AHFTC specialist should know that transcatheter edge to edge repair (TEER) of the mitral valve is an indicated procedure for severe secondary mitral regurgitation.

**Answer:** D. Initiate transcatheter edge-to-edge repair (TEER) of the mitral valve evaluation

This patient has severe secondary mitral regurgitation. The transthoracic echocardiogram clips demonstrate a dilated left ventricle, and tenting of the mitral valve, indicating restricted leaflet motion due to tethering of the valve leaflets. This is considered secondary (or functional) mitral regurgitation. The COAPT trial using

the MitraClip device to effect transcatheter edge-to-edge repair (TEER) of the mitral valve demonstrated that patients with moderate-to-severe (3+) or severe (4+) mitral regurgitation had reduced time to heart failure hospitalization or mortality compared to patients on optimal medical therapy alone. Based on this study, the 2020 ACC/AHA guideline for the Management of Valvular Heart Disease recommends that "patients with chronic severe secondary MR related to LV systolic dysfunction (LVEF < 50%) who have persistent symptoms (NYHA class II, III, or IV) while on optimal GDMT for HF (Stage D), TEER is reasonable in patients with…LVEF between 20% and 50%, LVESD ≤ 70 mm, and pulmonary artery systolic pressure ≤ 70 mmHg."

While the QRS is paced and wide, attempts to improve cardiac synchrony after CRT-P implantation using echocardiographic dyssynchrony studies have demonstrated only mixed results. The patient has remained euvolemic, and thus would not benefit from implantation of a pulmonary artery pressure monitor. There is no evidence for clinical bleeding to suggest that left atrial appendage occlusion would represent an improvement over the current anticoagulation strategy. Finally, the patient's age and the presence of an alternative treatment make VAD evaluation less attractive.

**References:**

1. Otto CM, et al. 2020 ACC/AHA Guideline for the Management of Patients with Valvular Heart Disease. Circulation. 2021; 143:e72-e227.
2. Stone GW, et al. Transcatheter Mitral-Valve Repair in Patients with Heart Failure. NEJM. 2018;379:2307-18.

## QUESTION 56

**Testing Point:** The Board-certified AHFTC specialist should know that dapagliflozin is indicated for patients with heart failure with reduced ejection fraction, regardless of whether the patient has diabetes or not.

**Answer:** A. Start dapagliflozin 10 mg daily.

This patient has a reduced ejection fraction and continues to have at least NYHA class II symptoms. Therefore, dapagliflozin 10 mg daily is indicated, based on the results of the DAPA-HF trial. The key enrollment criteria were LVEF ≤ 40%, NYHA Class II-IV symptoms, which this patient meets. The QRS complex is narrow on the electrocardiogram, obviating any utility of resynchronization therapy. The heart rate is 60, and atrial paced, making ivabradine less helpful, as this agent is indicated when the heart rate remains ≥70 beat per minute when a patient is on maximally tolerated doses of beta-blockers or have a contraindication to beta-blocker use. There is no role for addition of ACE-inhibitor to sacubutril-valsartan, and in fact this combination is contraindicated because of the risk of angioedema.

**References:**

1. McMurray, JJV et al. Dapagliflozin in Patients with Heart Failure and Reduced Ejection Fraction. NEJM 2019; 381:1995-2008.

2. Maddox, TM et al. 2021 Update to the 2017 ACC Expert Consensus Decision Pathway for Optimization of Heart Failure Treatment: Answers to 10 Pivotal Issues About Heart Failure with Reduced Ejection Fraction: A Report of the American College of Cardiology Solution Set Oversight Committee. JACC 2021; 77(6):772-810.

## QUESTION 57

**Testing Point:** The Board-certified AHFTC specialist should know that genetic cardiomyopathy associated with Lamin A/C mutations portend a high risk of cardiac sudden death, and ICD can be considered at ejection fractions up to 45% in patients with other risk factors.

**Answer:** D. Recommend placement of implantable cardioverter-defibrillator.
Lamin A/C mutations portend a very high risk of cardiac sudden death and represent an important cause of dilated cardiomyopathy with prominent conduction system disease. The disease typically manifests with cardiac dysfunction or skeletal myopathy. Inheritance is autosomal dominant. Clinical features include heart block, LV dysfunction, and arrhythmia. Arrhythmic risk is high. The 2017 AHA/ACC/HRS guidelines recommend that patients with Lamin A/C mutation who have 2 of the 4 following features: NSVT, LVEF < 45%, nonmissense mutation, and male sex) consider placement of an ICD. Since this patient meets two of the four criteria, ICD placement should be considered.

Continued surveillance of LV dysfunction, home blood pressure monitoring, and initiation of metoprolol succinate are all reasonable additional steps. However, these steps do not offer sufficient protection against cardiac sudden death, a critical component of the natural history of this disease.

**References:**

1. Al-Khatib SM. 2017 AHA/ACC/HRS Guideline for Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death. Circulation. 2018;138:e272-e391.
2. Hershberger RE. Genetic Evaluation of Cardiomyopathy—A Heart Failure Society of America Practice Guideline. Journal of Cardiac Failure. 2018 May;24(5):281-302.

## QUESTION 58

**Testing Point:** The Board-certified AHFTC specialist should know that athletes with suspected myocarditis should be restricted from training and competition for 3-6 months.

**Answer:** C. Obtain a stress electrocardiogram.

Myocarditis portends a risk of residual scar and arrhythmia. According to AHA/ACC Eligibility and Disqualification Recommendations for Competitive Athletes with Cardiovascular Abnormalities, after having myocarditis, athletes should be restricted from competition for 3 months. At 3 months, athletes should have a resting echocardiogram, a 24-hour Holter monitor, and stress electrocardiogram to evaluate for LV

dysfunction or arrhythmia, whether provoked or unprovoked. Concerning findings merit further follow-up. A cardiac MRI may be reasonable in selected circumstances where there is concern for residual scar.

**References:**

1. Maron BJ., et al. Eligibility and Disqualification Recommendations for Competitive Athletes with Cardiovascular Abnormalities: Task Force 3: Hypertrophic Cardiomyopathy, Arrhythmogenic Right Ventricular Cardiomyopathy and Other Cardiomyopathies, and Myocarditis. Circulation. 2015 Dec 1;132(22):e273-80.

## QUESTION 59

**Testing Point:** The Board-certified AHFTC specialist should know that immune checkpoint inhibitors are important causes of cardiomyopathy in cancer patients.

Answer. C. Endomyocardial biopsy

The clinical scenario is that of new onset acute decompensated heart failure, temporally associated with the start of immune checkpoint inhibitor therapy. This class of agents—including ipilimumab, nivolumab, pembrolizumab, cemiplimab, avelumab, atezolizumab, and durvalumab—works by disinhibiting the T cell response. This can result in a variety of toxicities, including myocarditis. Myocarditis related to immune checkpoint inhibitor therapy exists on a spectrum, ranging from minor elevations in troponin to fulminant disease. When myocarditis related to immune checkpoint inhibitors is suspected, endomyocardial biopsy can be helpful in establishing the diagnosis. The decision to continue or discontinue immune checkpoint inhibitors is often high-stakes, thus definitive diagnosis with tissue is often preferred.

Treatments of choice include high-dose steroids or other immune modulating agents such as mycophenolate. NSAIDs are not indicated, nor are low-dose steroids such as prednisone 10 mg daily. The electrocardiographic changes are striking, including new left bundle branch block, and diffuse pseudoinfarction pattern, which can be seen in myocarditis. Left heart catheterization would likely be a part of the diagnostic strategy. However, the presentation is not consistent with transthyretin amyloidosis, for which technetium-99 m pyrophosphate scintigraphy (technetium PYP scan) may be helpful.

**References:**

1. Moslehi J, et al. Immune checkpoint inhibitor-associated myocarditis: manifestations and mechanisms. JCI. 2021;131(5):e145186.
2. Hu JR, et al. Cardiovascular toxicities associated with immune checkpoint inhibitors. Cardiovasc Res. 2019;115:854-68.

## QUESTION 60

Heart Failure with Reduced Ejection Fraction (HFrEF)

**Testing Point:** The Board-certified AHFTC specialist should recognize when to refer a patient for right heart catheterization.

**Answer:** B. Refer for right heart catheterization.

The patient has signs and symptoms of advanced heart failure with several high-risk features. These include but are not limited to recurrent HF admissions over the prior 12 months, inability to tolerate neurohormonal antagonists, hyponatremia, escalation of high-dose diuretics, and worsening renal function. Given that the patient has been refractory to initial challenge with high dose inotropes and intravenous diuretics, the next best step would be to pursue a right heart catheterization to characterize intravascular hemodynamics and facilitate pulmonary artery catheter guided therapy. This can also facilitate an evaluation for advanced therapies, including durable left ventricular assist devices and cardiac transplantation.

Percutaneous mitral valve repair was shown to reduce morbidity and mortality in patients with disproportionately severe mitral regurgitation, including those with significantly less dilated ventricles on optimal medical therapy (as per the COAPT [Cardiovascular Outcomes Assessment of the MitraClip Percutaneous Therapy for Heart Failure Patients with Functional Mitral Regurgitation] Trial) and should not be used in patients with stage D HF requiring inotropic therapy in the acute setting. Pulmonary artery pressure monitoring has been demonstrated to reduce heart failure hospitalizations in patients with NYHA Class II or III regardless of ejection fraction (and this patient is NYHA Class IV). Initiation of continuous renal replacement therapy is not yet appropriate as the patient may benefit from oral or intravenous thiazide challenge, milrinone titration or transition to other inotropes/vasodilators pending the results of the right heart catheterization. Finally, while palliative care engagement and consultation may be reasonable, it may be premature to refer to hospice without consideration of further diagnostic testing given the patient's stated wishes and advanced directives.

**References:**

1. Yancy CW, Januzzi JL Jr, Allen LA, et al. 2017 ACC expert consensus decision pathway for optimization of heart failure treatment: answers to 10 pivotal issues about heart failure with reduced ejection fraction: a report of the American College of Cardiology Task Force on Expert Consensus Decision Pathways. J Am Coll Cardiol 2018;71:201-30.
2. Hollenberg SM, Warner Stevenson L, Ahmad T, et al. 2019 ACC expert consensus decision pathway on risk assessment, management, and clinical trajectory of patients hospitalized with heart failure: a report of the American College of Cardiology Solution Set Oversight Committee. J Am Coll Cardiol 2019;74:1966-2011.
3. Fang JC, Ewald GA, Allen LA, et al.; Heart Failure Society of America Guidelines Committee. Advanced (stage D) heart failure: a statement from the Heart Failure Society of America Guidelines Committee. J Card Fail 2015;21:519-34.

## QUESTION 61

Heart Failure with Preserved Ejection Fraction (HFpEF)

**Testing Point:** The Board-certified AHFTC specialist should know which intervention(s) prevent hospitalizations for heart failure with preserved ejection fraction (HFpEF).

**Answer:** C. Addition of spironolactone 25 mg daily.

Multiple large randomized controlled trials over the previous two decades have failed to show mortality benefit in HFpEF with a specific pharmacologic agent. Although the TOPCAT (Treatment of Preserved Cardiac Function Heart Failure With an Aldosterone Antagonist) trial did not reach its primary composite endpoint (CV death, congestive HF hospitalization, resuscitated cardiac arrest), spironolactone did reduce HF hospitalizations. Therefore, therapy with spironolactone is reasonable to consider for patients with similar characteristics as those enrolled in this trial (EF ≥45%, elevated BNP or HF admission within 1 year, serum creatinine <2.5 mg/dL, potassium <5 mEq/L). A recent analysis combining the PARAGON-HF (Prospective Comparison of ARNI With ARB Global Outcomes in HF With Preserved Ejection Fraction) and PARADIGM-HF (Prospective Comparison of ARNI With ACEI to Determine Impact on Global Mortality and Morbidity in Heart Failure) trials suggested a potential benefit of sacubitril-valsartan in patients with HF and LVEF >40% (and possibly as high as 57%), although statistical significance for the primary endpoint was not achieved. Thus, sacubitril-valsartan is not currently considered standard therapy for such patients.

Beta-blockers have not been shown to reduce heart failure hospitalizations or survival in patients with heart failure with preserved ejection fraction. It may be reasonable to titrate carvedilol up to 50 mg twice daily to improve blood pressure control as this patient's blood pressure is elevated; however, transition to metoprolol succinate 100 mg daily is unlikely to provide additional benefit or prevent rehospitalization. Also, while diuretics are the cornerstone of de-congestion management in these HFpEF patients, it is not reasonable to transition from bumetanide 4 mg twice daily to torsemide 40 mg twice daily, as the latter is a less potent diuretic. Lastly, phosphodiesterase-5 inhibitors (e.g., sildenafil) have not shown benefit in these HFpEF patients, as shown in the RELAX trial.

**References:**

1. Redfield MM. Heart failure with preserved ejection fraction. N Engl J Med 2016;375:1868-77.
2. Pitt B, Pfeffer MA, Assmann SF, et al., on behalf of the TOPCAT Investigators. Spironolactone for Heart Failure with Preserved Ejection Fraction. N Engl J Med 2014;370:1383-92.
3. Solomon SD, McMurray JJV, Anand IS, et al., on behalf of the PARAGON-HF Investigators and Committees. Angiotensin–Neprilysin Inhibition in Heart Failure with Preserved Ejection Fraction. N Engl J Med 2019;381:1609-20.

## QUESTION 62

Special Etiologies of Heart Failure

**Testing Point:** The Board-certified AHFTC specialist should recognize indications for implantable cardioverter defibrillator placement in adult patients with hypertrophic cardiomyopathy.

**Answer:** D. Refer for implantable cardioverter defibrillator (ICD) placement.

Based on the 2020 ACC/AHA Guidelines for the Diagnosis and Treatment of Patients with Hypertrophic Cardiomyopathy, an ICD is recommended (Class I) in HCM patients with sudden cardiac death, ventricular fibrillation, or sustained ventricular tachycardia and is reasonable (Class IIa) in patients with at least one of the following: massive left ventricular hypertrophy ( 30 mm on cardiac imaging); family history of sudden cardiac death; unexplained syncope; apical aneurysm; LVEF 50%. In this case, the patient meets the latter criteria by both transthoracic echocardiography and cardiac magnetic resonance imaging and ICD is recommended using shared decision making.

Non-vasodilating beta blockers and/or non-dihydropyridine calcium channel blockers may be reasonable if the patient has symptoms related to left ventricular outflow tract obstruction (LVOTO) or symptoms of exertional angina or dyspnea (if nonobstructive). This patient is asymptomatic and thus medical therapy is not indicated at this time. Septal reduction therapy, including septal myectomy or alcohol septal ablation therapy, may be considered if a patient has severe LVOTO symptoms and has failed a trial of medical therapy.

**References:**

1. Ommen SR et al. 2020 ACC/AHA Guideline for the Diagnosis and Treatment of Patients with Hypertrophic Cardiomyopathy. Circulation. 2020;142:e558-e631.

## QUESTION 63

Heart Transplantation

**Testing Point:** The Board-certified AHFTC specialist should recognize the indications for repeat heart transplantation.

**Answer:** C. Begin evaluation for cardiac re-transplantation.

The patient has evidence of severe cardiac allograft vasculopathy (consistent with International Society for Heart and Lung Transplantation Grade 3 CAV since 2 or more primary vessels have 70% stenosis). There are insufficient clinical data to suggest that pharmacologic therapy can alter the natural history of CAV once it develops or has become this severe. Percutaneous coronary intervention may be considered, especially

when severe stenosis is seen with proximal primary vessels, but no improvement in long-term outcomes has been noted. Similarly, the use of surgical revascularization is limited by the diffuse and distal process of coronary arterial remodeling. Cardiac re-transplantation is the only definitive management of advanced CAV; however, this approach may be limited by patient candidacy and scarcity of donor organs.

**References:**

1. Yancy et al. 2013 ACCF/AHA Guideline for the management of Heart Failure. Circulation 2013; 128; e240-e327.
2. Mehra MR, Crespo-Leiro MG, Dipchand A, et al. International Society for Heart and Lung Transplantation working formulation of a standardized nomenclature for cardiac allograft vasculopathy-2010. J Heart Lung Transplant 2010;29:717-27.
3. Mehra MR. The scourge and enigmatic journey of cardiac allograft vasculopathy. J Heart Lung Transplant 2017;36:1291-3.

## QUESTION 64

Pulmonary Hypertension

**Testing Point:** The Board-certified AHFTC specialist should know the risk factors for mortality with respect to definitive surgical management of WHO Group 4 chronic thromboembolic pulmonary hypertension.

**Answer:** C. Pulmonary vascular resistance of 1200 dyn·s·cm$^{-5}$.

Based on the clinical findings of severe pulmonary hypertension with a V/Q scan demonstrating bilateral pulmonary emboli, the patient has chronic thromboembolic pulmonary hypertension. Pulmonary thromboendarterectomy (PTE) offers the best chance at symptomatic improvement and long-term survival in eligible patients. At experienced centers, 1-year reported survival post-PTE is > 90%, 5-year survival is > 80% and 10-year survival is > 70%. Even at experienced centers, however, perioperative mortality is increased with markedly elevated preoperative pulmonary vascular resistance (PVR) >1000 dyn·s·cm$^{-5}$, corresponding to > 12.5 Wood Units. At the UCSD center, 4.1% of patients with a pre-operative PVR >1000 dyn·s·cm$^{-5}$ died, whereas only 1.6% who had a PVR <1000 dyn·s·cm$^{-5}$ died. Data from the international CTEPH registry showed in-hospital mortality approximately three times greater in those with PVR >1200 dyn·s·cm$^{-5}$ at diagnosis compared with PVR 400–800 dyn·s·cm$^{-5}$. Although the other answer choices represent markers of increased postsurgical mortality risk, the markedly elevated PVR shown here has been demonstrated to confer the highest risk.

**References:**

1. McLaughlin VV, Archer SL, Badesch DB, et al. ACCF/AHA 2009 expert consensus document on pulmonary hypertension: a report of the American College of Cardiology Foundation Task Force on Expert Consensus Documents and the American Heart Association: developed in collaboration with the American College of

Chest Physicians, American Thoracic Society, Inc.and the Pulmonary Hypertension Association. Circulation. 2009;119:2250–94.
2. Mahmud E, Madani MM, Kim NH, et al. Chronic thromboembolic pulmonary hypertension: evolving therapeutic approaches for operable and inoperable disease. J Am Coll Cardiol 2018;71:2468-86.
3. Jenkins D, Madani M, Fadel E, et al. Pulmonary thromboendarterectomy in the management of chronic thromboembolic pulmonary hypertension. Eur Resp Rev. Mar 2017; 26(143): 160111.

## QUESTION 65

Systolic heart failure during pregnancy

**Testing point:** The board certified AHFTC specialist should recognize medications that are contraindicated when pregnancy occurs in the setting of heart failure.

**Answer:** C. amiodarone and spironolactone.

Amiodarone is US FDA category D agent and is strongly associated with fetal hypothyroidism, goiter, intrauterine growth retardation, and neurological developmental delay. According to the 2018 ESC guidelines, amiodarone should only be used when other therapies have failed. This patient underwent a pulmonary vein isolation and currently has no further ongoing rapid atrial fibrillation. It would be reasonable to stop amiodarone entirely at this time, with or without replacement with an alternative anti-arrhythmic.
Spironolactone is a US FDA category D agent. In addition to mineralocorticoid antagonism, it also exhibits antiandrogenic effects. It has been shown to significantly impair fetal CNS and urogenital formation in animal models. Whilst human data is lacking, current ESC guidelines as well as expert consensus on this topic, recommend against the use of aldosterone inhibition in pregnancy.

The other agents are generally considered safe during pregnancy (FDA category B). Of note, whilst some guidelines establish that low molecular weight heparin could be considered in lieu of warfarin in the first 6-12 weeks of pregnancy, the use of warfarin in the 2nd and 3rd trimesters is generally preferred, with the caveat that at week 36 to facility safe delivery low molecular weight heparin is preferable.

**References:**

1. Vera Regitz-Zagrosek, Jolien W Roos-Hesselink, Johann Bauersachs, ESC Scientific Document Group et al. 2018 ESC Guidelines for the management of cardiovascular diseases during pregnancy: The Task Force for the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology (ESC), European Heart Journal, Volume 39, Issue 34, 07 September 2018, Pages 3165–3241

## QUESTION 66

Heart failure with preserved ejection fraction

**Testing point:** The Board-certified AHFTC specialist should recognize etiologies of dyspnea on exertion that may mimic heart failure with preserved ejection fraction but have different prognoses and treatment strategies.

**Answer:** C. stiff left atrial syndrome.

The patient presents with worsening dyspnea on exertion. He carries a history of pAF and multiple ablations. His AF has been reasonably controlled. His extensive cardiovascular workup excludes many common etiologies of dyspnea including coronary disease, valvular heart disease, and PE. His echo is not suggestive of amyloidosis and a CTA pulmonary vein does not find pulmonary stenosis. He does not respond to diuretics because at rest he is not generally volume overloaded and his hemodynamic abnormality is largely with exertion.

The exercise study shows normal resting hemodynamics. His blood pressure and heart rate response are relatively unremarkable. With exercise, there is a minimal elevation in his LVEDP but a dramatic elevation in his left atrial pressure. In the absence of mitral valve disease, this likely represents an atrial compliance abnormality. In addition, a low left ventricular transmural pressure, defined as the difference between the left ventricular end diastolic and right atrial pressure, suggests right ventricle pressure overload during exercise contributed to the small increases in LV filling pressures seen. His right sided filling pressures dramatically increase as a result of the rise in his left atrial pressures. Large V waves are generated as well, again a sign of left atrial non-compliance. Whilst the patient does have pulmonary hypertension during exercise, this is secondary to stiff left atrial syndrome and not idiopathic in nature.
Stiff left atrial syndrome has been well described in the literature as a complication of repeated ablation that results in extensive left atrial scarring. There is no specific treatment for it, though transplant may be definitive and atrial septostomy could be considered. It is however important to diagnose and differentiate this disorder from similarly presenting forms of exertional dyspnea such as pulmonary vein stenosis, HFpEF, cardiac amyloidosis, and idiopathic exercise induced pulmonary hypertension.

**References:**

1. Gibson, Douglas N., et al. “Stiff left atrial syndrome after catheter ablation for atrial fibrillation: clinical characterization, prevalence, and predictors.” Heart Rhythm 8.9 (2011): 1364-1371.
2. Mehta S, Charbonneau F, Fitchett DH, Marpole DG, Patton R, Sniderman AD. The clinical consequences of a stiff left atrium. Am Heart J. 1991 Oct;122(4 Pt 1):1184-91. doi: 10.1016/0002-8703(91)90498-7. PMID: 1927878.

## QUESTION 67

Heart failure special etiologies

**Testing point:** The Board-certified AHFTC specialist should know how to guide genetic testing for HCM.

**Answer:** D: she should undergo serial clinical evaluation for HCM with ecg and echocardiograms intermittently at this time

The patient reports a first degree family member with confirmed HCM. The family member is genotype negative. For patients with clinically confirmed HCM who have undergone genetic testing and were found to have no pathogenic variants, cascade genetic testing of their family members is not useful. Similarly, if a genetic variant of unknown significance is identified, cascade genetic testing of a family is also not useful. The presence of a normal echocardiogram at this time does not mean she is unlikely to develop clinical HCM in the future. Repeating her father's genetic testing is unlikely to yield a different result. Screening for HCM clinically when genetic testing is not available or is unlikely to be useful should occur at the time of diagnosis of HCM in a first degree family member. There is no indication to wait till age 45 and the patient would be exposed to risk by not entering a 2-5 year surveillance program immediately.

**References:**

1. Ommen, Steve R., et al. "2020 AHA/ACC guideline for the diagnosis and treatment of patients with hypertrophic cardiomyopathy: a report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines." Journal of the American College of Cardiology 76.25 (2020): e159-e240.

## QUESTION 68

MCS

**Testing point:** The Board-certified AHFTC specialist should recognize etiologies of persistently poor left ventricular unloading in patients with continuous flow LVADs.

**Answer:** D. outflow graft twisting.

The patient presents with worsening heart failure despite the presence of a durable LVAD. His clinical picture is highlighted by evidence of slowly progressive poor LV unloading as demonstrated by his increasingly dilated LV, high wedge pressure and resultant HF hospitalizations and eventual development of low flow alarms. Whilst other processes may present similarly (progressive AI, chronic pump thromboses, etc), the question stem gives enough information to exclude them and most likely an outflow graft complication is to blame. He does not have AI on TEE and his LDH is not in range to raise concern for an acute pump thrombosis. The CTA image provided shows a clear high grade out flow graft twist and the exploration of the explanted device confirms that as well.

The question stem deliberately leaves out the model of the device which has to be a heartmate 3. Outflow graft twisting is phenomena specific to the heart mate 3 due to the presence of a swivel joint initially designed to allow surgeons to address minor outflow graft kinks during initial device implants. However,

post implant, that joint appeared to over time allow cardiac motion to convey kinetic forces to the outflow graft that allowed for preferential twisting in one direction. Whilst the heart mate 2 uses the same swivel joint, the pump body and connectors are intra-abdominal and are insulated from cardiac motion. This issue has not been described with the HVAD. The treatment of choice is an outflow graft revision or heart transplantation. In the Momentum 3 trial, 8 out of 515 patient that received the HM3 developed outflow graft twisting within 2 years of implant. Circa 2018 a clamp was developed to prevent this issue and currently should be widespread use for new device implants. Nonetheless, a large number of patients are receiving care who remain at risk for this and physicians should be able to quickly recognize it and address it.

**References:**

1. Grüger, Thomas, et al. "Late post-pump blood flow obstruction in a novel left ventricular assist device: The unusual case of a twisted outflow graft." The Journal of thoracic and cardiovascular surgery 155.1 (2017): e33-e35.
2. Mehra, Mandeep R., et al. "A tale of the twist in the outflow graft: an analysis from the MOMENTUM 3 trial." The Journal of Heart and Lung Transplantation 37.11 (2018): 1281-1284.

## QUESTION 69

Heart Transplant

**Testing point:** The Board-certified AHFTC specialist should recognize the complications of immunosuppressive agents commonly used in heart transplant care.

**Answer:** B. a complication of either intravenous immunosuppressive agents, in this case likely thymoglobulin or rituximab.

The patient presents with high grade ACR/AMR complicated by cardiogenic shock. He is treated aggressively for both. Whilst there is some practice variation on the management of these cases, it is reasonable to offer both thymoglobulin and rituximab for these forms of high grade rejection. The patient improved with therapy but developed a classic presentation of acute serum sickness. Serum sickness presents with fevers, polyarthralgias, dermatoses (often urticarial in nature), cytopenias and multiorgan failure. The onset is 1-2 weeks after first receipt of the offending agent and is often fatal if not recognized and treated. Treatment commonly involves high dose corticosteroids and/or plasmapheresis.

Both thymoglobulin and rituximab are known agents that cause acute serum sickness. Thymoglobulin has the stronger association (some estimates of up to 16% of patients) but it is well described with rituximab as well. The presence of jaw pain is more specific to thymoglobulin. The timing of his presentation does not exclude either drug as the culprit. Acute rejection or acute HIT do not explain the findings. Whilst the patient is at risk for opportunistic reactions, no common opportunistic infection would result in the constellation

of symptoms described, particularly when his urticarial rash is considered. Acute serum sickness should be recognized and timely treatment as delays in doing so are often associated with high mortality.

**References:**

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## QUESTION 70

HFrEF

**Testing point:** The Board-certified AHFTC physician should know that excess RV pacing may lead LV dilatation and Failure

**Answer:** D. RV pacing

Right ventricular pacing is known to cause electrical and mechanical dyssynchrony. In MOST (Mode Selection Trial) ventricular pacing >40% in patients with sinus node dysfunction and normal QRS duration was associated with greater risk of HF hospitalization. The DAVID (Dual Chamber and VVI Implantable Defibrillator) trial also showed a greater risk of death and HF hospitalization in patients with LV systolic dysfunction and >40% ventricular pacing. More recent studies have suggested that an even lower threshold (as low as 20%) of RV pacing may be associated with LV systolic dysfunction.

A. - Immune checkpoint inhibitors (ICI) act on inhibitory signaling molecules (CTLA-4, PD1, PDL1) on T-cells and potentiate the T-cell response to cancer cells. ICIs have been associated with arrhythmias, cardiomyopathy, vasculitis, pericarditis and myocarditis. ICI-induced myocarditis typically occurs within the first few months following ICI initiation with a reported incidence of 0.04-1.1%. It is thus less likely to emerge in a patient who has tolerated ICI therapy for almost one year and who does not have significant elevations in cardiac biomarkers. ICI-induced myocarditis may occur concomitantly with myositis and myasthenia gravis (both absent in this patient). Prompt recognition of ICI-induced myocarditis and initiation of high-dose steroids (or other immunosuppressive therapies in steroid-refractory cases) are of paramount importance as ICI-induced myocarditis has been associated with mortality of up to 50%.

B. - Anthracycline-associated cardiotoxicity occurs in a dose-dependent fashion and has been traditionally

classified as acute, subacute or chronic although a more recent large prospective cohort study has challenged this concept, showing that incidence was highest during the first year after completion of anthracycline therapy. Risk factors associated with the development of anthracycline-related cardiotoxicity include cumulative anthracycline dose (typically doxorubicin ≥250mg/m2 or epirubicin ≥600mg/m2), chest radiation, presence of conventional cardiac risk factors, and age at initial cancer diagnosis. With a cumulative exposure of doxorubicin 100mg/m2, even with concomitant trastuzumab therapy, the risk of cardiac dysfunction attributed to anthracyclines is relatively low.

C. - RA pacing does not lead to ventricular mechanical dyssynchrony and has not been associated with development of LV dilation or heart failure.

**References:**

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## QUESTION 71

HFpEF

**Testing point:** The Board-certified AHFTC physician should be able to distinguish between ATTR and AL amyloid cardiomyopathy.

**Answer:** C. Perform endomyocardial biopsy

Although the positive PYP scan suggests a diagnosis of transthyretin amyloid cardiomyopathy (ATTR-CM), false positive studies can be seen in patients with immunoglobulin light chain amyloid cardiomyopathy (AL-CM). Furthermore, the presence of only mild concentric LVH with severely decreased GLS, markedly elevated NT-proBNP and elevated free lambda light chains (K/L ratio of 0.03) as well as macroglossia should raise suspicion for AL-CM. While chronic kidney disease can lead to elevations in free light chains, these are usually only modest and frequently associated with a kappa rather than lambda free light chain predominance. To distinguish between ATTR-CM (with MGUS) and AL-CM a tissue biopsy should be pursued.

A. - The TTR stabilizer tafamidis would be indicated for treatment of ATTR-CM. The ATTR-ACT trial showed a reduction in all-cause mortality and cardiovascular-related hospitalizations among patients with ATTR-CM (confirmed by histopathology/immunohistochemical analysis or scintigraphy with at least one prior heart failure hospitalization or clinical evidence of heart failure) randomized to tafamidis compared with placebo (win ratio 1.70; 95% CI, 1.26-2.29). However, in this case a diagnosis of AL-CM should be excluded first.

B. - Patients with amyloid cardiomyopathy often poorly tolerate atrial fibrillation due to their restrictive physiology. While this patient would benefit from diuresis and restoration of sinus rhythm, it is imperative to establish a correct diagnosis promptly so that appropriate and disease-specific therapy can be initiated without delay.

D. - In patients with ATTR-CM genetic counseling and TTR genotyping should be performed for prognostication and family screening purposes. In patients of African descent, the prevalence of the Val122Ile mutation has been estimated to be 3-3.5%, and its presence has been associated with more rapid disease progression. Nevertheless, presence of a pathogenic TTR mutation would not exclude a concomitant diagnosis of AL-CM and biopsy should therefore be pursued for diagnostic purposes (to diagnose AL-CM and exclude concomitant ATTR-CM).

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## QUESTION 72

HF – special etiologies

**Testing point:** The Board-certified AHFTC physician should know the indications for ICD implantation in patients with cardiac sarcoidosis.

**Answer:** B. Refer the patient for implantation ICD

The presentation with third-degree heart block at young age and typical imaging findings on cardiac PET showing classic perfusion-metabolism mismatch in the basal inferior septum, basal to mid anterior septum and the anterior wall in the absence of CAD is consistent with a diagnosis of cardiac sarcoidosis (CS) even in the absence of a tissue diagnosis. Other etiologies for his clinical presentation have been excluded. Current U.S. society guidelines suggest that implantation of a primary prevention ICD can be useful in patients with CS who have a requirement for permanent pacing and/or syncope and/or evidence of extensive myocardial scar by CMR or PET, even in the presence of preserved left ventricular systolic function (class IIa recommendation). Other indications for ICD implantation in patients with CS include secondary prevention of sudden cardiac death and LVEF ≤35% (class I recommendation) as well as inducible sustained ventricular arrhythmias on electrophysiology study with LVEF >35% (class IIa recommendation).

A. - Presence of advanced atrioventricular block in patients with CS has been associated with a substantial risk of ventricular arrhythmias and sudden death. Current guidelines have therefore issued a modest recommendation for a primary prevention ICD in patients with CS who have an indication for permanent pacing (class IIa recommendation).

C. - While AV conduction abnormalities may be reversible and immunosuppression can be useful in patients with CS with Mobitz II or third-degree heart block, reversibility is unpredictable and device implantation is therefore recommended. To decrease the risk of device infection, immunosuppression should be started once the wound has healed following device implantation.

D. - Similarly to C, steroid-sparing immunosuppression with methotrexate should be delayed to facilitate wound healing and minimize risk of infection following ICD implantation.

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## QUESTION 73

MCS

**Testing point:** The Board-certified AHFTC physician should be familiar with echocardiographic parameters to non-invasively estimate the severity of aortic insufficiency in patients supported on LVAD.

**Answer:** C. Peak systolic-to-diastolic velocity ratio of 3 of the LVAD outflow cannula

Traditional echocardiographic measures may underestimate the severity of aortic insufficiency (AI) as they do not take the pancyclic nature of AI in LVAD patients into account. Continuous regurgitant flow back into the LV cavity augments LV preload. At the same time aortic afterload increases during systole due to enhanced flow through the LVAD or across the aortic valve (if opening). During diastole afterload decreases, thereby diminishing the diastolic transaortic pressure gradient. As a result diastolic flow accelerates through the LVAD outflow cannula and the systolic-to-diastolic velocity ratio decreases as AI severity worsens. A peak systolic-to-diastolic velocity ratio of <5.0 has been associated with clinically important AI and is strongly

correlated with left-sided filling pressures and regurgitant fraction measured by Doppler echocardiography/ right heart catherization.

A. - Although the vena contracta is a well validated measure of AI in non-LVAD patients it may underestimate the severity of AI in LVAD patients in whom AI is pancyclic, occurring during both systole and diastole, and may be eccentric. ASE guidelines cite a vena contracta of ≥3mm as suggestive of clinically important AI (≥moderate) in LVAD patients.

B. - A lack of a change in LV dimensions during an echocardiographic ramp study can be seen with clinically significant AI but also occurs in other circumstances, e.g. pump thrombosis or LVAD graft obstruction, and is therefore not specific for AI.

D. - Diastolic flow accelerates as AI severity worsens. Diastolic flow acceleration of the outflow cannula of >49cm/sec$^2$ has been associated with clinically important AI and elevations in PCWP.

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## QUESTION 74

Heart transplant

**Testing point:** The Board-certified AHFTC physician should recognize thrombotic microangiopathy as a rare but potentially fatal side effect associated with tacrolimus.

**Answer:** A. Tacrolimus

The presence of schistocytes on the peripheral smear, anemia, thrombocytopenia, unconjugated hyperbilirubinemia, elevated LDH and reticulocyte count with acute renal injury point to thrombotic microangiopathy (TMA) as the unifying diagnosis for this patient's presentation. TMA is a rare but potentially life-threatening side effect associated with tacrolimus with an estimated incidence of <5% among solid organ transplant recipients. The pathophysiology remains poorly understood, and its emergence has

been attributed to a toxic, dose-dependent rather than an immune-mediated effect. Treatment consists of supportive care and withdrawal of the calcineurin inhibitor, or dose reduction if discontinuation is not feasible. Plasma exchange may be appropriate if there is diagnostic uncertainty about the possibility of thrombotic thrombocytopenic purpura (defined by ADAMTS-13 activity <10%). However, therapeutic efficacy of plasma exchange has not been established in the treatment of tacrolimus-induced TMA and the American Society for Apheresis guidelines therefore do not recommend routine plasma exchange given the lack of robust evidence and possible complications associated with its use. Case reports have suggested possible benefit with eculizumab, a terminal complement inhibitor, in patients with normal ADAMTS-13 activity not responding to supportive care.

B. - Mycophenolate mofetil is associated with myelosuppression and colitis, which can lead to gastrointestinal blood loss, but not with TMA.

C. - This patient is at increased risk of post-transplant lymphoproliferative disorder (PTLD) given the EBV mismatch. PTLD typically presents in a bimodal fashion early within the first year or late 5-15 years post-transplant. PTLD can manifest with myelosuppression if bone marrow invasion is present, gastrointestinal bleeding from mucosal erosions if gastrointestinal involvement is present, autoimmune hemolytic anemia or spontaneous tumor lysis syndrome but is not generally associated with TMA.

D. - CMV infection can be associated with pancytopenia related to myelosuppression and hemolytic anemia. This patient's elevated reticulocyte count in the setting of hemolysis argues against myelosuppression and schistocytes on the peripheral smear point to TMA.

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## QUESTION 75

Pulmonary Hypertension

**Testing point:** The Board-certified AHFTC specialist should be able to recognize the appropriate diagnostic evaluation for patients with chronic thromboembolic pulmonary hypertension.

**Answer:** C. Ventilation/perfusion scan

The patient most likely has a diagnosis of chronic thromboembolic pulmonary hypertension (CTEPH), a disease with high morbidity and mortality without appropriate recognition and treatment. Although the incidence of CTEPH following an acute PE is low (< 10%), a history of PE is present in up to 75% of patients and DVT in up to 56% of patients presenting with CTEPH. Importantly, a normal CT chest does not exclude PE and has a sensitivity of 51% as compared with >95% for ventilation/perfusion (V/Q) scan. A normal V/Q scan effectively excludes CTEPH.

Pulmonary endarterectomy (PEA) is the only definitive curative therapy for CTEPH. If mismatched defects are found on V/Q scan, RHC and pulmonary angiogram should be performed, and the patient should be referred to a center specializing in management of CTEPH, particularly PEA.The periprocedural mortality is < 5% in experienced centers. PEA also results in near-normalization of hemodynamics and improvement in symptoms in most CTEPH patients.

Although the patient is a former smoker and pulmonary function tests and high-resolution CT chest may be useful in evaluating parenchymal and airway disease, her presentation is most suspicious for CTEPH given the aforementioned history and exam. A right heart catheterization (RHC) will be useful in determining hemodynamics and guiding further therapy after confirming the diagnosis of CTEPH; however, exercise RHC is most useful in making the diagnosis of heart failure with preserved ejection fraction with pulmonary venous hypertension. While this is a common cause of PH, it is unlikely given the absence of dilated left atrium or elevated left ventricular filling pressure (E/e' < 10). A polysomnogram would be recommended given her morbid obesity and resting hypoxia, and associated symptoms, but would not be the best next step to determine the etiology of her dyspnea.

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## QUESTION 76

Cardiac Transplantation

**Testing point:** The Board-certified AHFTC specialist should be able to recognize important drug-drug interactions in managing post transplant immunosuppression.

**Answer:** B. Voriconazole

The calcineurin inhibitors (CNIs), specifically tacrolimus, serve as integral components of the standard immunosuppressive medication regimen following heart transplantation. Both tacrolimus and cyclosporine are metabolized by the cytochrome P450 (CYP) system, especially the 3A4 family; therefore, medications that either inhibit or induce CYP3A4 must be used with caution in patients on calcineurin inhibitors. CYP3A4 inhibitors include erythromycin, clarithromycin, diltiazem, verapamil, and azole antifungals (including voriconazole); CYP3A inducers include phenobarbital, phenytoin, and rifampin. Initiation or cessation of these medications without CNI adjustment and/or close monitoring of levels can lead to either subtherapeutic CNI levels or toxicity.

In this patient, cessation of voriconazole (a CYP3A4 inhibitor) without concomitant monitoring or adjustment in tacrolimus led to subtherapeutic tacrolimus levels and subsequent severe acute cellular rejection (ISHLT Grade 3R). Notably, allopurinol and trimethoprim/sulfamethoxazole do not impact the metabolism of tacrolimus. Allopurinol should not be prescribed with azathioprine because buildup of a metabolite due to xanthine oxidase inhibition can cause severe anemia. Whereas antiseizure medications such as phenytoin can have drug-drug interactions with tacrolimus, phenytoin is a CYP3A inducer and thus discontinuation of the drug would have led to supratherapeutic CNI levels or CNI toxicity.

**References:**

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## QUESTION 77

Temporary Mechanical Circulatory Support

**Testing point:** The Board-certified AHFTC specialist should be able to recognize the appropriate exit strategies for patients with temporary mechanical circulatory support in cardiogenic shock.

**Answer:** B. Evaluation for durable left ventricular assist device with temporary right ventricular assist device support

This is a critically ill patient with heart-failure related cardiogenic shock now status post cardiac arrest requiring VA ECMO with Impella CP for LV venting. Based on the revised and updated classification scheme proposed by the Society for Cardiovascular Angiography and Intervention, she would be classified as SCAI D or E with the cardiac arrest modifier. The patient requires an exit strategy following escalation of temporary mechanical circulatory support. Consultation of the advanced heart failure team will be helpful in evaluating her candidacy for advanced therapies. As an INTERMACS 1 profile, she may be a high-risk candidate for durable left ventricular assist device with temporary right ventricular assist device support pending multidisciplinary evaluation.

Her BMI alone is likely an absolute contraindication for cardiac transplantation at most programs. In addition, her difficulty affording medications may serve as a potential barrier to transplantation. While it may be reasonable to wean VA ECMO as a bridge to decision or bridge to recovery, it is not prudent to pursue this strategy immediately following resuscitation from cardiac arrest. Finally, palliative care specialists will be critical to shared decision-making with the patient's caregivers and family and elucidating the patient's goals, wishes, and values. However, a referral to hospice may be premature given the patient's age and if the patient is deemed a suitable candidate for durable LVAD with temporary RVAD support.

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## QUESTION 78

Special Etiologies of Heart Failure

**Testing point:** The Board-certified AHFTC specialist should be able to recognize that the use of immune checkpoint inhibitors can be associated with myocarditis.

**Answer:** C. Endomyocardial Biopsy

This patient is presenting in cardiogenic shock. Therefore, the most appropriate next step is to perform an endomyocardial biopsy, the diagnostic "gold standard" in myocarditis, to evaluate for diffuse immune infiltration of T cells and macrophages. Immune checkpoint inhibitors (ICIs) such as pembrolizumab are monoclonal antibodies designed to activate the immune system against malignant cells and are often used

to treat triple-negative breast cancer. The use of ICIs, however, has been associated with myocarditis, which can be fatal (approximately 30-50%) despite its low incidence (0.5-1%). Symptoms are often nonspecific, including dyspnea and weakness. The majority of patients present with ECG abnormalities, including atrial or ventricular fibrillation; an echocardiogram will often demonstrate regional wall motion abnormalities, thickened septum, or an enlarged left ventricular cavity size with reduced systolic function.

According to the 2021 Heart Failure Association/Heart Failure Society of America/Japanese Heart Failure Society (HFA/HFSA/JHFS) Position Statement on endomyocardial biopsy, the indications for performing endomyocardial biopsy include: 1) suspected fulminant myocarditis or acute myocarditis with acute heart failure, LV dysfunction, and/or rhythm disorders; and 2) suspected ICI-mediated cardiotoxicity, including acute heart failure with or without hemodynamic instability early after drug initiation. In patients with confirmed ICI-mediated myocarditis, ICI treatment should be discontinued and high-dose immunosuppression should be instituted in addition to standard heart failure and cardiogenic shock care.

This patient's hemodynamic instability likely precludes a cardiac magnetic resonance imaging (MRI) or position emission tomography (PET). The latter can be used to detect active inflammation, particularly when cardiac sarcoidosis is suspected, but its use in other forms of myocarditis is reserved for situations in which a cardiac MRI is contraindicated. Lyme carditis, which typically manifests several weeks after initial exposure, is not likely to be the etiology of her current symptoms.

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## QUESTION 79

Heart Failure with Preserved Ejection Fraction (HFpEF)

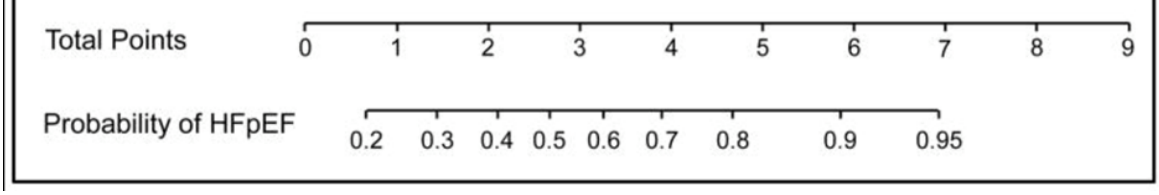
**Testing point:** The Board-certified AHFTC specialist should be able to recognize which factors in the H2FPEF scoring system is the strongest predictor of the diagnosis of heart failure with preserved ejection fraction (HFpEF).

**Answer:** C. Atrial fibrillation

The etiology of dyspnea can be difficult in patients with suspected heart failure with preserved ejection fraction (HFpEF) in the absence of overt intravascular volume overload. The H2FPEF score is a well-validated instrument that can be useful. Clinical variables associated with HFpEF as the cause of shortness of breath include: *h*eavy (BMI >30 kg/m2), *h*ypertension (requiring two or more medications for control), atrial *f*ibrillation, pulmonary *h*ypertension, *e*lderly (age >60 years), and elevated *f*illing pressures by echocardiography (E/e' >9). The score is calculated based on these clinical parameters, with an increasing score coinciding with increased likelihood of HFpEF as the etiology of dyspnea. All parameters are assigned 1 point based on clinical risk, except for elevated BMI (2 points) and the presence of paroxysmal or persistent AF (3 points).

This patient's H2FPEF score was 9, which is associated with a 97% probability of his symptoms being caused by HFpEF. Of all the parameters, the history of AF carries the most weight. His BMI was >30 kg/m2 and importantly, LVH is not a characteristic noted to contribute significantly to this model. See figure below.

| | Clinical Variable | Values | Points |
|---|---|---|---|
| $H_2$ | **H**eavy | Body mass index > 30 kg/m$^2$ | 2 |
| | **H**ypertensive | 2 or more antihypertensive medicines | 1 |
| F | Atrial **F**ibrillation | Paroxysmal or Persistent | 3 |
| P | **P**ulmonary Hypertension | Doppler Echocardiographic estimated Pulmonary Artery Systolic Pressure > 35 mmHg | 1 |
| E | **E**lder | Age > 60 years | 1 |
| F | **F**illing Pressure | Doppler Echocardiographic E/e' > 9 | 1 |
| $H_2$FPEF score | | | Sum (0-9) |



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## QUESTION 80

Heart Failure with Reduced Ejection Fraction (HFrEF)

**Testing point:** The Board-certified AHFTC specialist should be able to recognize the clinical indications and appropriate use of SGLT2 inhibitors in Stage C heart failure with reduced ejection fraction patients.

**Answer:** B. Add empagliflozin 10 mg daily

The EMPEROR-Reduced (EMPagliflozin outcomE tRial in Patients With chrOnic heaRt Failure With Reduced Ejection Fraction) trial demonstrated a 25% reduction in composite of cardiovascular death or HF hospitalization and a 30% reduction in HF hospitalization for individuals with NYHA II-IV HF and LVEF < 40% when empagliflozin was added to contemporary guideline-directed medical therapy for HF. SGLT2 inhibitors have received a Class I recommendation for the treatment of patients with symptomatic Stage C HF with reduced ejection fraction (HFrEF) and consequently, serve as one of the four major classes of guideline-directed medical therapy for HF.

Transcatheter edge-to-edge mitral valve repair carries a Class 2a recommendation for individuals with severe stage D secondary mitral regurgitation who have persistent symptoms on optimal guideline-directed medical therapy for HF. The recent TEE showed mild to moderate regurgitation and so referral for TEER would be inappropriate.

The Systolic Heart Failure Treatment With the IF Inhibitor Ivabradine Trial (SHIFT) clinical trial randomized 6558 patients with stable (≥4 weeks) but symptomatic chronic HF with reduced ejection fraction, hospitalized with HF decompensation in the previous 12 months, New York Heart Association class II–IV, left ventricular ejection fraction (LVEF) ≤35%, and sinus rhythm with HR >70 beats per minute to ivabradine versus placebo. SHIFT demonstrated a 18% reduction in the composite primary endpoint of cardiovascular death or HF hospitalization. However, ivabradine has not been shown to improve cardiovascular mortality and the patient appears to be rate controlled on his current regimen of beta blockade.

In selected adult patients with NYHA class III HF and history of a HF hospitalization in the past year or elevated natriuretic peptide levels, on maximally tolerated stable doses of GDMT with optimal device therapy, wireless monitoring of pulmonary artery pressure by an implanted hemodynamic monitor may reduce the risk of subsequent HF hospitalizations. However, no cardiovascular mortality benefit has been demonstrated using this diagnostic strategy.

**References:**

1. Heidenreich P, Bozkurt B, Aguilar D, et al. 2022 AHA/ACC/HFSA Guideline for the Management of Heart Failure. *J Am Coll Cardiol*. 2022 May, 79 (17) e263–e421. https://doi.org/10.1016/j.jacc.2021.12.012
1. Packer M, Anker SD, Butler J, et al. Cardiovascular and Renal Outcomes with Empagliflozin in Heart Failure. *N Engl J Med*. 2020 Oct 8;383(15):1413-1424. doi: 10.1056/NEJMoa2022190.

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