Case Presentation (Michael H. Kim, MD)
A 77-year-old woman was brought to a community hospital emergency room after an episode of syncope. She had been seated at the table and had passed out for ~2 minutes. She had a history of hypertension, paroxysmal atrial fibrillation, and an abdominal aortic aneurysm repair 10 years earlier. She had no complaints immediately before the syncopal episode. When she awoke, she complained of mild chest and back discomfort that resolved quickly. Over the past several months, she had fatigue, malaise, and abdominal pain, and she had lost 10 lbs of weight. At the hospital, her systolic blood pressure was noted to be 47 mm Hg, and she was transferred to the University of Michigan Medical Center.

Clinical Discussion (Eric M. Isselbacher, MD)
Although there are numerous causes of syncope, a careful history can often help narrow the differential diagnosis considerably. In this patient, syncope appears to have occurred without warning, and because the patient remained unconscious for 2 minutes, vasovagal syncope is not likely. Her chest and back pain immediately raises the suspicion of either acute myocardial infarction or aortic dissection. She has a history of both hypertension and an abdominal aortic aneurysm, so she is at risk for both coronary artery disease and aortic dissection. Her hypotension is consistent with either true cardiogenic shock or “pseudohypotension,” a falsely low blood pressure measurement that may occur with aortic dissection. This finding is due to acute compromise of arterial flow to the subclavian artery. If she was awake and responding to questions when this blood pressure was measured, such pseudohypotension is likely. I would want to know her blood pressures in the other extremities and the findings on her initial ECG and chest radiograph.

Michael H. Kim, MD
On arrival, she was intubated and given dopamine and intravenous fluids. The patient’s cardiac rhythm was rapid atrial fibrillation at a rate of 150 to 180 bpm. The systolic blood pressure was 70 to 80 mm Hg in both arms. Her hematocrit value was 39.4. Initial evaluation revealed no evidence of a leaking abdominal aortic aneurysm. Cardiology consultation was requested after ultrasonography performed in the emergency room suggested a pericardial effusion. Surface echocardiography confirmed the presence of a moderate-sized pericardial effusion with evidence of cardiac tamponade (right ventricular diastolic collapse). Mild to moderate aortic insufficiency and global left ventricular hypokinesis were present. Chest radiography revealed a widened mediastinum, an enlarged cardiac silhouette, and a tortuous thoracic aorta (Figure 1).

Eric M. Isselbacher, MD
Her blood pressure is the same in both arms, and it is now clear that her hypotension is due to cardiogenic shock from cardiac tamponade and left ventricular hypokinesis and exacerbated by rapid atrial fibrillation. The combination of a moderate pericardial effusion, aortic insufficiency, and a widened mediastinum on chest radiograph in a patient presenting with chest pain and syncope is almost certainly due to an acute type A aortic dissection. The rapid atrial fibrillation is likely secondary to hypotension and the pericardial effusion, but the origin of the left ventricular hypokinesis is uncertain. The primary therapy is to stabilize the patient and treat the hypotension caused by the cardiac tamponade with volume expansion. Given the rapid atrial fibrillation, I would also consider electrical cardioversion as a means to improve hemodynamics. Although many might be tempted to perform pericardiocentesis at this point, this procedure sometimes precipitates recurrent hemorrhage and rapid death when hemopericardium has been caused by a type A aortic dissection.

Therefore, this short window of tenuous hemodynamic stability should be used to arrange for an emergency surgical repair of the aortic dissection. A transesophageal echocardiogram (TEE) can be performed in the operating room to confirm the diagnosis and assess whether the aortic valve can be repaired.

Michael H. Kim, MD
Electrical cardioversion resulted in normal sinus rhythm with first degree atrioventricular block at a rate of 80 bpm. Nonspecific ST-T–wave abnormalities were present. Systolic blood pressure improved to 90 mm Hg. Dopamine and intravenous fluids were continued. TEE showed no clear evidence of aortic dissection. A markedly dilated ascending...
aorta, consistent with an aneurysm, and moderate aortic insufficiency were present. There was a small area of possible discontinuity at the distal ascending aorta. The descending aorta was of normal caliber. A moderate pericardial effusion was once again noted, with evidence of hemodynamic compromise. Left ventricular function had returned to normal. A pulmonary artery catheter revealed the following pressures: pulmonary artery 50/22, wedge 20, and right atrial 18 mm Hg.

Eric M. Isselbacher, MD
 Restoration of sinus rhythm with electrical cardioversion has helped to improve left ventricular systolic function and elevate the blood pressure slightly, but the patient remains hypotensive despite continued intravenous fluids and dopamine. Given our clinical suspicion of aortic dissection, and because the TEE does not show an intimal flap, we should immediately suspect the diagnosis of contained rupture of an ascending aortic aneurysm. Indeed, the echocardiogram here confirms the presence of both an ascending aneurysm and a possible region of discontinuity of the aortic wall. In instances of contained rupture of an ascending aortic aneurysm, patients often present with the classic symptoms of aortic dissection complicated by cardiac tamponade, but they lack any evidence of pulse deficits because there is no intimal flap in the distal aorta. The equalization of right and left heart filling pressures found by the pulmonary artery catheter confirms the presence of cardiac tamponade, as we had suspected clinically. Once again, with a pericardial effusion and evidence of tamponade presumably due to aortic rupture, this patient’s hemodynamic stability is tenuous, and surgical repair of the aorta should be undertaken immediately.

Michael H. Kim, MD
 Aortic angiography performed in the left anterior oblique and frontal projections disclosed no evidence of aortic dissection or rupture. There was no communication to the pericardium. At this time (4 to 5 hours after admission), the patient no longer required dopamine. Systolic blood pressure was 140 mm Hg, and her heart rate was 90 bpm in normal sinus rhythm. Plans were made to perform a diagnostic pericardiocentesis in the cardiac catheterization laboratory. Complete drainage of the pericardial fluid was to be performed only if the fluid was nonbloody. If bloody fluid was found, the patient was to go directly to the operating room.

Eric M. Isselbacher, MD
 Aortography can be falsely negative when a patient suffers an intramural hematoma of the aorta rather than a classic aortic dissection. However, based on the findings of the TEE, we might have expected to find a pseudoaneurysm of the distal ascending aorta in this patient. This finding should be reliably detected by aortography. Thus, I am surprised at the negative examination. The clinical presentation and the diagnostic imaging studies are discordant. It is certainly possible that the patient has an aortic dissection that has not been detected by either the echocardiogram or aortogram. What alternative explanation could there be? Although the possibility of a malignancy with a pericardial effusion seemed small initially, the patient does have a history of fatigue, malaise, abdominal pain, and weight loss for several months. Also, her history of paroxysmal atrial fibrillation in the presence of the pericardial tamponade suggests that she might have acutely developed atrial fibrillation, thereby precipitating an abrupt drop in blood pressure, which caused syncope. I can understand why her doctors are reluctant to proceed directly to surgery. However, her clinical presentation is so classic for aortic dissection/rupture with hemopericardium that I am unwilling to dismiss this diagnosis. The decision to try to define the nature of the pericardial fluid by performing a diagnostic pericardiocentesis is reasonable, but it might be preferable to perform the test in the operating room rather than in the catheterization laboratory in the event that she has an under-
lying aortic dissection or rupture or if the pericardiocentesis precipitates hemodynamic collapse.

Michael H. Kim, MD
As the diagnostic pericardiocentesis was being arranged, a second consultant reviewed the presentation and imaging findings. This physician defaulted to a high probability of aortic dissection based on the initial presentation. The initial 2 diagnostic tests were used to support, not refute, the possibility of an acute aortic syndrome such as aortic dissection or acute intramural hematoma of the ascending aorta. The pericardiocentesis was then cancelled owing to concerns about the risks associated with the procedure in the presence of an aortic process, and 1 additional test, spiral CT, was ordered in the hope of identifying a flap or double channel. Review of the CT revealed high-density fluid in the pericardial sac consistent with blood (Figure 2). No dissection flap was identified. The aortic root and ascending aorta up to the aortic arch were dilated.

Eric M. Isselbacher, MD
Because there is no evidence of aortic dissection, intramural hematoma, or rupture on the TEE, aortogram, or spiral CT scan, we must now consider more strongly the possibility that this patient may not have acute aortic disease. Nevertheless, the CT scan raises the possibility that the pericardial effusion contains blood. Hemopericardium secondary to a leaking aneurysm still cannot be excluded. Whereas previously, the pericardial effusion was not tapped for fear that it was secondary to aortic dissection, pericardiocentesis now seems appropriate. Given the cardiac tamponade, the effusion must be drained for therapeutic reasons. Analysis of the pericardial fluid would be useful diagnostically as well to distinguish frank blood from more typical serosanguinous fluid. If the fluid is blood, surgical repair of the large ascending aortic aneurysm should again be considered; if the fluid is serosanguinous, acute aortic dissection or intramural hematoma could be reasonably excluded, and then an appropriate evaluation of the cause of the pericardial effusion could be undertaken.

Michael H. Kim, MD
Despite the absence of a confirmed diagnosis of aortic dissection and 3 inconclusive imaging studies, the patient was taken to the operating room 15 hours after her admission.

Eric M. Isselbacher, MD
In the process of clinical diagnostic reasoning, available data are used to revise a physician’s estimate of the probability of a suspected disease until either the suspicion is sufficiently high to warrant treatment or so low that the disease is essentially excluded. Diagnostic tests are performed to increase or decrease the estimated probability of the disease, which can lead to clinical confirmation, exclusion, or the need for additional testing.

In this patient, the clinical presentation was highly suggestive of acute aortic dissection, yet despite the very high clinical probability of dissection, analysis of the results of 3 proven and widely accepted diagnostic imaging modalities did not enable the confirmation of the clinical diagnosis. Ultimately, the decision was made to discount the results of the diagnostic tests and to take the patient to surgery predominantly on an extremely strong clinical suspicion of aortic dissection and the aortic abnormalities found on the 3 imaging studies, which demonstrated findings that frequently accompany aortic dissection.

The mortality rate of untreated acute dissection of the thoracic aorta is as high as 1% to 2% per hour if left untreated. Given a high probability of proximal aortic dissection and this high risk, an argument could have been made to proceed directly to operative intervention. However, in practice, even patients with a high probability of proximal aortic dissection generally undergo additional testing to confirm the diagnosis, because thoracic surgery is risky in itself, and physicians are reluctant to expose patients to such risks if there is a chance that they do not have the disease. In
addition, the diagnostic tests provide valuable information for the surgeon. An imaging study can define anatomic details such as the extent of dissection and the degree of aortic insufficiency and localize potential entry/exit sites. TEE was performed to confirm the clinical diagnosis and to obtain such anatomic information. It was chosen as the initial diagnostic test because of its quick availability and because the patient was unstable. The “negative” or “inconclusive” result of this procedure left us with a diagnostic conundrum.

If we apply the threshold approach to clinical decision making, immediate surgical therapy may still have been warranted, because the pretest probability of aortic dissection was probably greater than the test-treatment threshold probability. The threshold probability is related to the relationship between the consequences of withholding treatment when the disease is present versus those due to administering treatment when the disease is absent. The result of the TEE study was viewed as a potential false-negative result rather than a true-negative result. A true-negative result would have led the physicians to exclude aortic dissection as the presumed diagnosis. Given the very high probability on clinical grounds of aortic dissection and a negative initial diagnostic test, the patient’s physicians had to choose whether to proceed to thoracic surgery or to perform a second diagnostic test. The decision was made to pursue a second diagnostic test, an aortogram, given that the possibility of aortic dissection remained high. A recent decision analysis supports the common practice of ordering a second diagnostic test when the probability of aortic dissection is moderate to high, provided that the delay in obtaining the test does not exceed 10 hours.

The “negative” aortogram further added to the diagnostic dilemma. Although negative or normal tests are used to exclude disease, even consecutive false-negative test results can occur and can lead to misdiagnoses that may result in poor outcomes. It was difficult for the clinicians in this case to believe that 2 accepted diagnostic tests for aortic dissection could both prove to be “negative” given the reported sensitivities and specificities for these tests.

Despite the inability to confirm the clinical diagnosis of aortic dissection with TEE and aortography, the clinicians were uncomfortable in revising the probability of aortic dissection downward to a significant degree, because neither test was truly normal. Rather, both studies showed abnormalities of the ascending aorta and/or heart that may accompany aortic dissection. The patient’s pretest probability of acute aortic dissection, based on all the data available before any of the 3 diagnostic tests were performed, was estimated to be ~90%. The case evoked comments such as “if anyone has a dissection, it’s her,” whereas the diagnostic studies proved inconclusive. Given the abnormalities on the initial 2 diagnostic tests, the clinicians could not confidently rule in or rule out aortic dissection. The probability of aortic dissection remained at least moderate.

When a preliminary review of the CT scan showed high-density fluid in the pericardial sac consistent with blood and dilatation of the aortic root and arch, this consultant felt sufficiently confident of an acute aortic process to refer the patient to surgery despite no clear diagnosis of acute dissec-

Pathological Findings (Michael H. Kim, MD)

When the chest was opened, it was apparent that bloody fluid under low pressure was present within the pericardial sac. The fluid was drained without immediate reaccumulation. The proximal aorta was enlarged, and extensive echymotic areas were present. No communication to the pericardium or intimal tear could be identified. As the operation progressed, the aorta acutely dissected through the aortic valve and the right coronary artery. The aorta was repaired, the aortic valve was replaced, and a saphenous vein was used to bypass the right coronary artery.

The cardiac surgeon thought that the patient may have had a rupture of the vasa vasorum from the intima to the adventitia, with communication to the pericardium. On review of the spiral CT scan, an area of circumferential thickening of the wall of the descending aorta and the aortic arch was thought to represent an intramural hematoma. The patient survived the operation and had a protracted hospital stay complicated by pneumonia and metabolic encephalopathy. She was discharged from the hospital 1 month after admission.

Closing Comments (Michael H. Kim, MD, and Kim A. Eagle, MD)

The clinicians were faced with conflicting or equivocal data from 3 well-accepted and discriminatory diagnostic imaging modalities regarding the likelihood of acute proximal aortic dissection. To find our way out of this clinical dilemma, we applied bayesian analysis. The bayesian approach involved the iterative updating of the clinician’s probability estimate on the basis of each additional test result. Thus, the posttest probability of disease was highly dependent on the pretest probability. If the 3 diagnostic tests had all been truly negative, the revised probability would have been low enough to sufficiently exclude aortic dissection on the basis of bayesian principles. Because prior knowledge of disease prevalence, preconceptions, and a patient’s clinical features are the basis for judging the pretest probability, some view quantitative assessments of this probability with skepticism. Nonetheless, in many instances, such estimates are extremely helpful in judging the influences of test results. However, as this case demonstrates, test results frequently are not definitively positive or negative. Such equivocal results are often confusing, because there is little or no revision of the probability for what we seek, and we become confused by other findings that may lead to blind alleys.
Clinical Diagnosis
Ascending aortic aneurysm and secondary aortic insufficiency.
Probable aortic dissection or contained aortic rupture with hemopericardium and cardiac tamponade.

Final Diagnosis
Acute intramural hematoma of the thoracic aorta progressing to aortic dissection.
Cardiac tamponade.
Ascending aortic aneurysm.

References

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Bayesian Persuasion
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