CASE PRESENTATION

A 50-year-old man with no significant past medical history presents to the emergency department with chest pain radiating to his back. He also complains of abdominal and left leg pain that started in the hip/buttock and radiated downward. He has no past surgical history. Medications include fexofenadine, fluticasone, and montelukast.

Physical examination reveals mild abdominal tenderness. Bilateral upper extremity pulses are normal, right lower extremity pulses are normal, left femoral pulse is weak, left pedal signals are present, and the left foot is cyanotic.

Laboratory results reveal a hemoglobin level of 9.8 g/dL, white blood cell count of 21,600/µL, anion gap of 23.7 mEq/L, and serum creatinine level of 2.06 mg/dL. CT shows acute type A (DeBakey I) aortic dissection with a compressed true lumen throughout the entire aorta (Figure 1). There appears to be radiographic malperfusion involving the viscera and the left kidney (Figures 2–5). There is no flow in one segment of the left common iliac artery with reconstitution of the left common femoral artery (Figures 6 and 7).

How would you proceed and why? Please elaborate on your treatment strategy.

Drs. Jackson and Bavaria: We would most likely perform a hemiaortic arch replacement if it were only radiologic malperfusion and not clinical. If there were signs of real clinical malperfusion in the visceral bed, we would proceed with either a zone 2 arch or a frozen elephant trunk (FET). FET is accomplished using either the Thoraflex device (Vascutek Ltd.), which is currently undergoing a US Food and Drug Administration trial, or antegrade thoracic endovascular aortic repair (TEVAR) at the time of open arch repair, initially described by our group at the American Association of Thoracic Surgery annual meeting in 2006. A mild bump in serum creatinine is very common, as there is the usual single renal of false lumen. Our group believes in rapid correction of the proximal lesion in type A dissection; therefore, we admit patients directly to the operating room (OR) from the helipad or emergency department, use rapid direct aortic cannulation, and proceed toward repair without delay.

Almost always, repair will accomplish reperfusion of the infradiaphragmatic peripheral vascular beds. Nevertheless, the vascular surgery team checks on the patient at the conclusion of the cardiac surgery procedure to ensure that there are palpable femoral pulses bilaterally. Assessment of visceral and renal perfusion at this point is pretty tough without opening the abdomen to check the mesenteric and renal artery pulses. The patient is often cold, coagulopathic with elevated serum lactate, and may be oliguric. But restoration of a left femoral pulse at this point is reassuring with regard to restoration of visceral and renal perfusion as well, so in general, we’d bring the patient to the intensive care unit (ICU) to monitor and warm him.

Drs. Charlton-Ouw and Estrera: The patient has a complicated acute type A aortic dissection with an aneurysm of the ascending aorta and celiac artery. Although flow into the true lumen is compromised with decreased enhancement of the celiac and superior
mesenteric arteries (SMAs), the degree of mesenteric ischemia is unclear. In our series, abdominal pain alone was a poor marker for the presence of mesenteric ischemia. Lower limb ischemia is a more ominous sign of mesenteric ischemia, but most limb ischemia resolves with repair of the proximal aorta.\textsuperscript{2,3} I would proceed with emergent open repair of the ascending aorta. Treatment of the aortic arch depends on a number of factors including the presence of aneurysm, location of entry tears and fenestrations, and distal malperfusion. Although most malperfusion resolves with proximal repair, approximately 20% of cases have persistent malperfusion. Preparing a suitable landing zone for stent grafting, either by transsternal arch debranching or in situ arch repair with an elephant trunk approach, depends on the anatomy and experience of the cardiac surgery team.

Immediate proximal repair is indicated, but the extent of the repair would depend on the tear location, tear involvement, and transverse arch diameter. Although ascending and hemiaortic arch is an acceptable approach, alleviating the distal malperfusion in as many as 80% of cases, persistent malperfusion may still remain, and concern for late aneurysmal degeneration of the proximal descending thoracic aorta exists. If the tear is extensive and involves a significant portion of the transverse arch, or if the transverse arch is enlarged (> 45 mm), then total arch replacement with FET hybrid repair would be performed. Because we have access to a hybrid arch TEVAR graft, we would use this prosthesis. If the tear is limited to the ascending aorta only and the nonenlarged transverse arch is preserved, then a zone 1 “Bavaria” repair would be performed (i.e., partial transverse arch replacement with debranching of the left common carotid and innominate arteries). After completion of the repair, selective TEVAR would then be performed if malperfusion persisted. In either case, we would perform intravascular ultrasound (IVUS) to assess the true lumen and confirm improved true lumen flow.
Drs. Fischbein, Dake, and Lee: This case highlights the importance of a collaborative aortic team to treat a life-threatening pathology that has high morbidity and mortality. At our institution, patients with type A dissections go directly to the OR for expedient repair. If the patient remains clinically stable, a 6- to 8-mm graft is anastomosed to the right axillary artery in preparation for selective antegrade cerebral perfusion during the arch reconstruction (moderate hypothermia). If the left leg is pulseless (as is suggested in this case scenario), we would also consider reperfusing the leg via a second perfusion limb “Y’d” off the arterial circuit. Depending on the anatomy, the patient would undergo either supracoronary replacement of the ascending aorta or aortic root replacement. Although dynamic malperfusion is typically resolved after aggressive partial arch replacement (removing the entire lesser curvature of the arch up to the left subclavian artery), a total arch replacement with FET would be performed, contingent on the patient’s age, primary intimal tear location, connective tissue disorder status, and aortic arch dimension. For FET, we would either deploy an antegrade stent graft (typically 10-cm length) inside a conventional elephant trunk placed into the true lumen under direct visualization or use the Thoraflex hybrid device (10-cm length to reduce the risk of paraplegia). Descending aorta true lumen flow and expansion should be confirmed with transesophageal echocardiography after flow is reestablished to the body. Before leaving the OR, we confirm the presence of peripheral pulses. Repair of the ascending and arch pathology will often improve perfusion distally, and patients are subsequently transferred to the ICU for close postoperative monitoring and resuscitation (urine output, lactate level, and peripheral pulses are closely observed).

If this method of repair does not begin to reverse the issues noted in the patient, then femoral catheterization and catheter-based imaging of the mid-thoracic aorta to the femoral bifurcations is indicated. Options at that point are dictated by adequate flow to the visceral vessels, renals, and both lower extremities. Treatment choices at this point could include extension of the FET to the diaphragm or, in extreme cases, all the way to the celiac artery if there is significant true lumen collapse there. Individual visceral stenting is indicated if dynamic or static compression is seen for each branch. Finally, a reentry site is often seen in the external iliac past the internal iliac, and nitinol stenting of the iliac segment can reestablish flow down through the affected extremity. Again, there are so many permutations of possibilities for the treatment that these cases must be individualized, and collaboration between cardiothoracic surgery, interventional radiology, and vascular surgery is often important.
Postoperatively, the patient has malperfusion involving the visceral vessels. How would you proceed?

Drs. Jackson and Bavaria: As previously indicated, it might be fairly difficult to tell while still in the OR whether renal or visceral malperfusion persists after ascending and hemiarch repair. The patient is under general anesthesia, resuscitation is ongoing so the serum lactate will still be elevated, and kidney injury from acute tubular necrosis will still be in evolution. In general, the patient is brought to the ICU from the OR, and in the ensuing hours, we take stock of the situation: Does urine output increase? Does lactic acidosis clear? If not, the possibility of ongoing visceral and renal ischemia must be entertained. If ongoing ischemia is suspected—especially if there are compromised pulses in one or both legs—we would typically bring the patient to the hybrid OR for angiography and possible TEVAR (expansion of the true lumen using TEVAR in the descending thoracic aorta can quickly ameliorate a pseudoaortic occlusion of the aorta).

In all dissection cases, we find IVUS invaluable for intraoperative guidance—avoiding excess contrast administration—and to ensure stent deployment in the true lumen. Intraoperatively, angiographic assessment of any dissection flap extending into individual visceral vessels can be performed and stenting into those vessels accomplished, although we find that is rarely necessary if the true lumen has been adequately expanded. We never hesitate to perform exploratory laparotomy to assess the viability of the intestines and to directly palpate visceral arterial or renal pulses if either is in doubt.

Drs. Charlton-Ouw and Estrera: After repair of the ascending aorta, visceral angiography and exploratory laparoscopy or mini-laparotomy should be performed to assess for bowel ischemia. If there is no evidence of bowel ischemia, the abdomen can be closed or a temporary negative pressure dressing can be placed. Resolution of the left femoral artery pulse deficit and foot cyanosis also suggests recanalization of the left common iliac artery.

If bowel necrosis or ischemia is present, a full laparotomy should be done. If there is bowel ischemia, a good pulse in the SMA, and Doppler signals in the affected territories, we can assume that the ischemia occurred prior to aortic repair. Without a palpable SMA pulse and evidence of ongoing bowel ischemia or ongoing left femoral pulse deficit, aortography should be performed. If there is evidence of ongoing visceral or limb malperfusion, I would perform TEVAR. The proximal landing zone would ideally be in the graft from either a debranched aortic arch or an elephant trunk repair.

Drs. Fischbein, Dake, and Lee: Assuming the patient has stabilized after the ascending and arch repair in the OR, if after some further resuscitation in the ICU it is determined that the visceral vessels are not optimally filling, then an immediate trip to the endovascular suite is necessary. Extension of the FET with additional TEVAR components is indicated if there continues to be true lumen collapse in the aortic segment down to the celiac. Careful angiography and determination of pressure gradients across the celiac and SMA can help determine if the origins require stenting. Use of a short self-expanding nitinol stent to equalize flow from the true lumen into the target visceral vessel is important. Consultation with general surgery can be helpful at this point if there is any clinical evidence of bowel ischemia, so they can be prepared to potentially explore and visualize the intestine. Remember that reperfusion can lead to additional problems several hours after revascularization.

Postoperatively, the patient has malperfusion of his left lower extremity. How would you proceed?

Drs. Jackson and Bavaria: In the setting of isolated unilateral lower extremity malperfusion, our default approach is to do a femoral-femoral bypass. The advantages of that approach are: (1) it can be done in the same OR as the type A dissection repair immediately at the end of that procedure, not requiring transfer to a different hybrid OR; (2) it’s quick; and (3) it doesn’t require nephrotoxic contrast administration (even in the setting of bilateral lower extremity ischemia from aortic dissection, we will sometimes perform a quick axillary-femoral and femoral-femoral bypass to reperfuse the legs, especially if there is no concern for ongoing visceral ischemia). In this case, given the initial CT findings of isolated left common iliac occlusion, we might consider percutaneous revascularization with a self-expanding stent in the common iliac artery using IVUS to be certain that we’re stenting from true lumen into true lumen, but that would generally require transfer to the hybrid OR or at least moving the patient onto a fluoroscopy-capable table, so we’d probably consider it and quickly reject it in favor of femoral-femoral bypass.
However, if either the patient is already back in the ICU when the left leg ischemia itself or its severity is recognized or there is concomitant concern for ongoing mesenteric or renal ischemia, we would typically recommend bringing the patient to the hybrid OR emergently for angiography and possible TEVAR, as previously discussed. Again, especially in the case of compromised pulses in the bilateral lower extremities, expansion of the true lumen using TEVAR in the descending thoracic aorta can quickly accomplish reperfusion of the legs. Sometimes, more distal stenting of the abdominal aorta or iliac arteries is needed; although we confess that as more distal stenting becomes necessary to reestablish pulses to the legs, we sometimes regret not having elected to do an axillary-femoral-femoral bypass in the first place. And again, endovascularly, IVUS is invaluable in treating aortic dissections of all varieties.

One must be alert to the possibility of reperfusion and compartment syndrome in this situation as well. The dictum that if you think of doing fasciotomies, then you should just do them can be a helpful guide; we generally have a low threshold for doing four-compartment fasciotomies of the reperfused calf in these clinical scenarios.

Drs. Charlton-Ouw and Estrera: If there is persistent visceral malperfusion and a proximal landing zone was not properly prepared, TEVAR can still be considered, but this is a less secure repair. If there is persistent mesenteric ischemia despite proximal open and endovascular aortic repair, we would either do retrograde SMA stenting via laparotomy or open bypass from the right common iliac artery to the SMA. If there is resolution of the visceral malperfusion but persistent left femoral pulse deficit, we would either place a left common iliac artery stent or perform right-to-left femoral-femoral bypass grafting.

Drs. Fischbein, Dake, and Lee: Some of the traditional teaching for limb ischemia in the face of aortic dissection has suggested that a femoral-femoral bypass rapidly restores flow. In our experience, we have often favored endovascular strategies after careful postoperative review of the CTA scans, as these can reveal residual distal reentry tears that compromise iailc flow. It is important to check for a diminutive iliac artery true lumen surrounded by an unopacified, often thrombosed, false lumen associated with the absence of distal false lumen reentry or exit tear. We have often been able to restore flow endovascularly via primary stenting of the iliac true lumen or by balloon fenestration of the aortic septum and, if necessary, stenting across it to create a baffle from false to true lumen if the infrarenal aortic segment is mostly false lumen. Again, the multidisciplinary approach and availability and capability of endovascular and open techniques are key to the successful revascularization of these patients. If the leg has been ischemic for a prolonged period of time and subsequent revascularization techniques are successful, the patient should be monitored for lower extremity compartment syndrome. Prophylactic fasciotomy is simple to perform and recover from and can be life and limb saving.

Postoperatively, the patient is asymptomatic. How would you proceed?

Drs. Jackson and Bavaria: Although some would consider prophylactic TEVAR to expand the true lumen, obliterate the false lumen, and/or cover any distal fenestrations between the two in the thoracic aorta, there are no level 1 data to support such an intervention. We would advocate conservative therapy and vigilance for any subsequent symptoms of peripheral malperfusion (postprandial abdominal pain, oliguria, worsening renal function, lower extremity weakness or numbness) in the immediate postoperative period. We would then continue close surveillance of the distal arch and descending thoracic aorta over the following months. If the patient experiences aneurysmal degeneration of the residual descending thoracic aortic dissection, we have a number of options for endovascular or open repair in 2017, including carotid-subclavian bypass with subsequent TEVAR or, under the auspices of clinical trials in the United States, arch branch TEVAR devices.

Drs. Charlton-Ouw and Estrera: If the patient is asymptomatic in the postoperative period, I would only observe, with no further intervention. Although several high-risk anatomic features have been identified, the benefit of preemptive TEVAR is controversial. The patient is medically optimized with blood pressure control and offered enrollment in a prospective aortic surveillance protocol. Our usual protocol for repaired DeBakey I dissections entails CT of the chest, abdomen, and pelvis at 1 month, 6 months, and then annually thereafter. If there are significant changes at 1 month, we sometimes perform a 3-month scan. For patients with renal insufficiency, noncontrast MRA offers excellent contrast-like resolution. For those who develop
complications from chronic dissection, such as aneurysm, several endovascular and open surgical options are now available.

**Drs. Fischbein, Dake, and Lee:** If the patient is recovering well in the cardiovascular ICU postoperatively, we develop a plan for postoperative imaging to follow the residual dissection that is present. Our protocol includes a gated CTA prior to discharge to risk stratify patients who might be at high risk for future problems and then close follow-up with repeat cross-sectional imaging at 1, 3, and 6 months and annually thereafter.


**Ali Azizzadeh, MD, FACS**
Director, Division of Vascular Surgery
Vice Chair, Programmatic Development
Department of Surgery
Associate Director, Vascular Therapeutics, Heart Institute
Cedars-Sinai Medical Center
Los Angeles, California
ali.azizzadeh@cshs.org
Disclosures: Consultant to Gore & Associates and Medtronic.

**Benjamin Jackson, MD, FACS**
Associate Professor and Program Director
Vascular Surgery and Endovascular Therapy
University of Pennsylvania
Philadelphia, Pennsylvania
benjamin.jackson@uphs.upenn.edu
Disclosures: None.

**Joseph Bavaria, MD, FACS**
Division of Cardiovascular Surgery
University of Pennsylvania Medical Center
Philadelphia, Pennsylvania
joseph.bavaria@uphs.upenn.edu
Disclosures: Co-Principal Investigator for Gore & Associates’ TBE trial and Medtronic’s Valiant Evo trial; Principal Investigator for Medtronic’s DISSECTION trial and Cook Medical’s TX2 postmarket trial; and consultant to Gore & Associates.

**Kristofer M. Charlton-Ouw, MD, FACS**
Associate Professor and Program Director
Vascular Surgery Fellowship and Integrated Residency Department of Cardiothoracic and Vascular Surgery
McGovern Medical School, UTHealth
Houston, Texas
kristofer.charltonouw@uth.tmc.edu
Disclosures: Consultant to Gore & Associates.

**Anthony L. Estrera, MD, FACS**
Professor and Chief of Cardiac Surgery
Department of Cardiothoracic and Vascular Surgery
McGovern Medical School, UTHealth
Houston, Texas
anthony.lestoner@uth.tmc.edu
Disclosures: Consultant to Gore & Associates.

**Michael Fischbein, MD, PhD**
Associate Professor of Cardiothoracic Surgery
Director of Thoracic Aortic Surgery
Residency Program Director
Stanford University Medical Center
Stanford, California
mfischbe@stanford.edu
Disclosures: None.

**Michael D. Dake, MD**
Thelma and Henry Doelger Professor (III)
Department of Cardiothoracic Surgery
Stanford University School of Medicine
Falk Cardiovascular Research Center
Stanford, California
mddake@stanford.edu
Disclosures: None.

**Jason T. Lee, MD**
Professor of Surgery
Director of Endovascular Surgery
Program Director, Vascular Surgery Residency/Fellowship
Division of Vascular Surgery
Stanford University Medical Center
Stanford, California
jtlee@stanford.edu
Disclosures: None.