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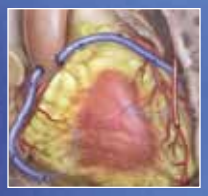


Cleveland Clinic

Cardiac Consult

Heart and Vascular News from Cleveland Clinic | Winter 2017

› SPECIAL ISSUE

COMPLEX CASES

Dear Colleagues:

If the past decade didn't make it clear enough, 2016 has left no doubt: Continuing change is the surest constant we can count on in U.S. healthcare in the years ahead.

The inevitability of that change is something we all must grapple with, particularly when change increasingly comes in the form of tightening reimbursements. Those sorts of changes can make it all too tempting for organizations to hunker down and play it too safe, avoiding the most complex cases that might sometimes skew outcomes numbers or draw disproportionate staff time and resources.

That's a temptation Cleveland Clinic's Miller Family Heart & Vascular Institute is committed to never give in to. This special issue of *Cardiac Consult* is devoted exclusively to brief reports of complex cases managed at Cleveland Clinic in the past few years — years of unprecedented healthcare system change.

These case studies show how our institute's interdisciplinary corps of clinicians — cardiothoracic surgeons, vascular surgeons and all manner of cardiovascular medicine subspecialists — pool their expertise in creative ways to solve problems for patients who've often been told elsewhere that their cases are beyond intervention. We find that this collaboration is facilitated by our institute's 12 umbrella teams under which 43 specialized centers are organized to best serve the needs of the many complex patients we see.

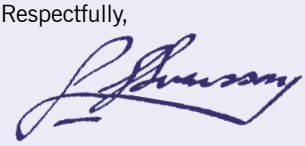
At the most fundamental level, we take on these complex cases for the individual patients themselves, who bring needs that we must attempt to meet if we have any degree of experience and expertise on which to offer them hope.

On another level, we welcome extraordinarily challenging cases because they sow the seeds for the therapies of tomorrow. If our predecessors had been less bold in tackling the daunting cases of their day, how many techniques and treatments that we now take for granted would have never materialized?

As we all step into the changing landscape ahead, our institute's commitment to our core principles will remain steadfast. These include pledges to (1) change and innovate as needs require; (2) preserve our practice, research and education; and (3) keep untouchable our high-quality patient care. The final principle is the most sacrosanct and applies whether a patient's needs are routine and straightforward or unprecedented and daunting.

If you have a complex case that falls in the latter category and requires referral, Cleveland Clinic would be honored to collaborate with you on your patient's care. We pledge to devote the same dedication, resourcefulness and coordination of care as you will see illustrated in the cases on the pages that follow.

Respectfully,



Lars G. Svensson, MD, PhD

CHAIRMAN | Sydell and Arnold Miller Family Heart & Vascular Institute

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Image of the Issue

By Patcharapong Suntharos, MD, and Hani Najm, MD

THREE-DIMENSIONAL PRINTING IN CONGENITAL HEART DISEASE

Ethan is a 9-year-old boy with heterotaxy syndrome that was characterized by complex congenital heart disease involving situs inversus, complete common atrioventricular canal, total anomalous pulmonary venous return (TAPVR), left superior and inferior vena cavae to the left-sided atrium, two good-sized D-loop ventricles with the aorta arising from the right ventricle remote from the ventricular septal defect, and pulmonary atresia with multiple major aortopulmonary collateral arteries (MAPCAs).

As an infant, he underwent TAPVR repair with anastomosis of the pulmonary venous confluence to the right side of the common atrium followed by unifocalization of his MAPCAs to a 12-mm aortic homograft-valved conduit connected to the right ventricle. He later underwent nine cardiac catheterizations, mainly to alleviate significant pulmonary artery branch stenoses and augment pulmonary artery blood flow. Nevertheless, he still had profound cyanosis.

In July 2016, we needed to innovate a procedure to improve oxygen saturation values in this deeply cyanosed child, but standard imaging modalities were inadequate for surgical planning. Building on Cleveland Clinic's experience developing 3-D-printed heart replicas to plan transcatheter pulmonary valve implantations in patients with congenital defects, we decided to generate a 3-D-printed model of Ethan's heart to assist our planning.

Using contrast-enhanced MRIs and CTs of his heart, we developed the 3-D-printed model above, which allowed us to examine the probability of whether a procedure could improve his oxygen saturations. When the surgeon (co-author Hani Najm, MD) was able to hold the exquisitely detailed heart replica in hand and make an incision in it, the feasibility of our proposed novel procedure was demonstrated.

That procedure was executed in real life exactly as planned on the 3-D replica, and the outcome was excellent. Ethan is now doing very well, with saturation percentages in the low 90s — a level he had never achieved before.

3-D-printed replicas are exceptional tools for planning percutaneous and surgical interventions for complex congenital heart defects. Our team looks forward to more opportunities to apply this technology in previously untreatable cases. ■

Dr. Suntharos (sunthap@ccf.org) is a pediatric and congenital interventional cardiologist, and Dr. Najm (najmh@ccf.org) is Chair of Pediatric and Congenital Heart Surgery.

Bilateral Lung Transplant, CABG and Tricuspid Valve Repair for CAD with End-Stage Lung Disease

BY MICHAEL TONG, MD, MBA, AND SHINYA UNAI, MD

Presentation and History

A gentleman in his mid-60s with a history of smoking presented to an outside institution with worsening shortness of breath, orthopnea and lower extremity edema. CT showed severe emphysematous changes and basilar scarring compatible with advanced idiopathic pulmonary fibrosis with a chronic obstructive pulmonary disease component. V/Q scan revealed mismatched ventilation-perfusion defects suggesting chronic pulmonary embolism that led to severe pulmonary hypertension. Echocardiogram showed normal left ventricular function, but the right ventricle was severely dilated and the function was moderately poor; he also had severe tricuspid regurgitation.

Over the next three months, the patient became increasingly symptomatic, requiring up to 8 liters of oxygen with activity. He was referred for lung transplantation at his local transplant program but was turned down when it was discovered that he also had severe triple-vessel coronary artery disease (CAD). Given his age, pulmonary hypertension, tricuspid regurgitation and severe CAD, his case was considered too complex and too high-risk.

He was referred to Cleveland Clinic for a second opinion. Undeterred by his advanced disease complexity, he remained motivated and worked hard to maintain his level of fitness despite the need for escalating doses of oxygen. Our committee accepted him for combined double lung transplantation, coronary artery bypass grafting (CABG) and tricuspid valve repair, and he was placed on the transplant waitlist.

The Surgery in Brief

About four weeks after the patient's listing, a suitable lung donor was identified and the patient was brought to the OR. Following median sternotomy, left internal thoracic artery (LITA) was harvested in a skeletonized fashion. Saphenous vein was harvested endoscopically. He was heparinized and placed on cardiopulmonary bypass. Antegrade and retrograde cardioplegia was induced to achieve cardiac arrest.

The vein grafts were anastomosed to the obtuse marginal and posterior descending arteries, and the proximal ends were then anastomosed to the ascending aorta. The cross-clamp was removed and the heart began to beat again. The hilar structures of both lungs were dissected free, and when the organs arrived in the OR, the left lung was removed and the donor left lung was implanted; the same was done on the right side. Finally, grafting of the LITA to the left anterior descending artery (LAD) was performed, and the tricuspid valve was repaired with a 30-mm tricuspid rigid annuloplasty ring.

CABG: Numbers of Note (2016)*

815	Isolated CABG surgeries
0.5%	Isolated CABG operative mortality (vs. 1.9% predicted [STS])
1.6%	Operative mortality for CABG + aortic valve replacement (N = 247) (vs. 4.5% predicted [STS])
3.3%	Operative mortality for CABG + mitral valve replacement (N = 30) (vs. 10.5% predicted [STS])
0.0%	Operative mortality for CABG + mitral valve repair (N = 73) (vs. 3.8% predicted [STS])

*Here and throughout, 2016 data represent the 12 months from 12/1/2015 to 11/30/2016, as full-year 2016 data were not available at press time.
STS = Society of Thoracic Surgeons

Outcome

The patient fared well postoperatively. Initial fluid overload was treated with diuretics. He was extubated on postoperative day 5 and transferred to the step-down unit on day 8. His postsurgical recovery was rapid, presumably thanks to his level of fitness, and he was discharged home on postoperative day 15. He continues to do well one year postoperatively.

Discussion: Not Uncommon, Still Complex

The combination of CAD and end-stage lung disease (Figure 1) is common as well as a predictor of complexity and risk. Typically a patient with CAD waiting for lung transplantation would be offered percutaneous coronary intervention prior

to transplant, but three to six months of dual antiplatelet therapy (aspirin and clopidogrel) would then be required and the patient could be listed only after stopping the clopidogrel, due to a prohibitive risk of bleeding during surgery. Given our patient's rapid rate of decline, he probably would not have had six months to live under such a scenario. Similarly, patients with end-stage lung disease may have other cardiac abnormalities such as aortic stenosis and tricuspid regurgitation. Our patient had developed secondary pulmonary hypertension that in turn caused increased right ventricular strain and tricuspid valve regurgitation.

Various operative sequences are possible when it comes to performing combined CABG and lung transplantation. To shorten the operation's duration, we typically would perform the CABG first while waiting for the donor lungs to arrive (Figure 2, with the completed combination procedure shown in Figure 3). This also provides increased coronary blood flow, which can be protective against intraoperative myocardial infarction. However, if the LITA-to-LAD bypass is performed with an in situ LITA graft, it can make the left lung explant and implant more difficult. Therefore, the LITA-to-LAD bypass can be done after the left lung is implanted or the LITA can be used as a free graft. Likewise, if the patient needs a mitral valve replacement, this should be done after the left lung transplant to avoid the risk of atrioventricular groove disruption when lifting the heart to access the left chest.

The Value of Versatility

At most centers, the presence of concomitant cardiac disease during lung transplantation requires two separate surgical teams: a thoracic surgeon for the lung and a cardiac surgeon for the heart. Given the unpredictable nature and timing of transplantation, this can pose a logistical challenge, which prompts many centers to turn down such patients. Currently, 15 to 20 percent of Cleveland Clinic lung transplant recipients have been turned down by other programs, in many cases due to the presence of CAD.

At Cleveland Clinic, we have a team of three cardiac surgeons and one thoracic surgeon performing lung transplantation, which gives us the versatility to offer combined surgeries to patients without additional surgical risk and without logistical challenges. This versatility, together with cardiac surgery and lung transplant volumes that are among the very highest in the nation, makes our team uniquely well prepared to offer surgery to complex patients like this who have no other options. ■

Dr. Tong (tongz@ccf.org) is a staff surgeon and Dr. Unai is a cardiothoracic surgery fellow, both in the Department of Cardiothoracic Surgery.

Figure 1. Illustration of fibrotic lungs with concomitant coronary artery disease.

Figure 2. CABG performed prior to bilateral lung transplantation.

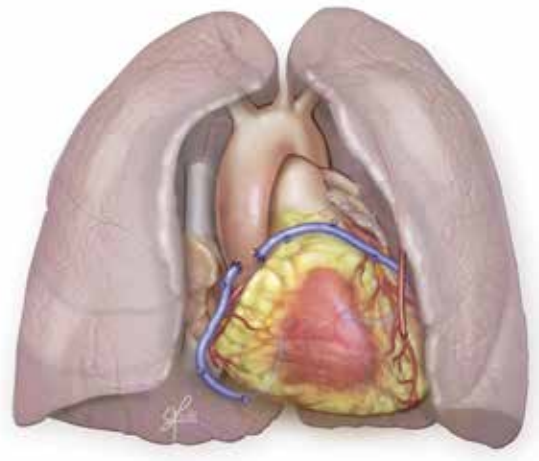


Figure 3. Completed combined CABG/double lung transplant.

The 'Hemi-Commando' Arrives: Incorporated Aortomitral Homograft for Bivalvular Endocarditis

BY JOSE NAVIA, MD

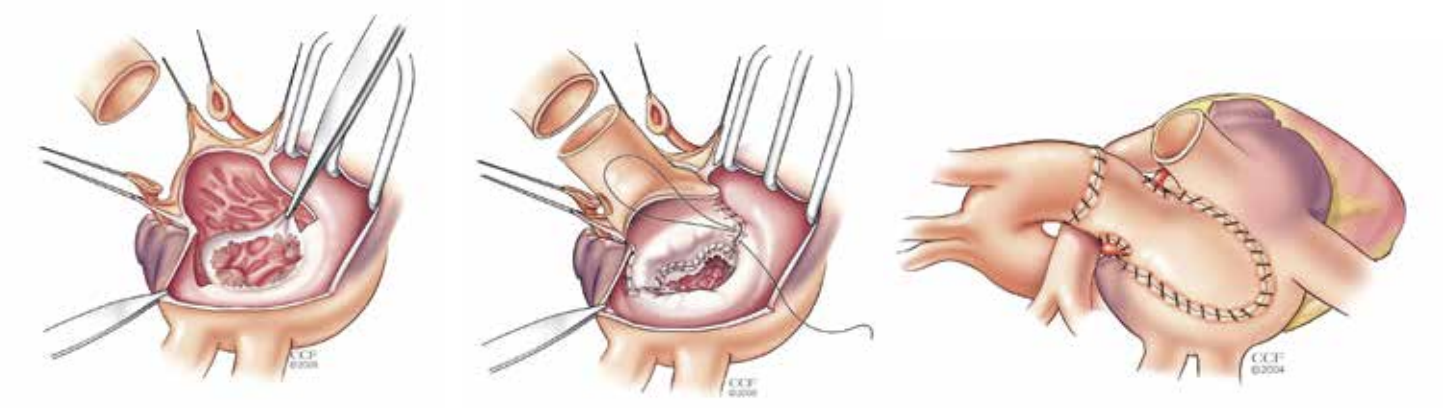


Figure 1. Illustrations of three stages of the hemi-commando procedure. **Left:** Surgeon's view of the heart after complete removal of infected tissue prior to placement of the homograft. **Middle:** Implantation of the aortomitral homograft as a unit. **Right:** The completed repair.

Case Vignette

A 64-year-old woman with a mechanical aortic valve presented to Cleveland Clinic with fever, shortness of breath and congestive heart failure. Blood cultures showed her to be positive for *Escherichia coli*. Echocardiography suggested the presence of dense bacterial colonies and vegetation around the aortic prosthesis, intervalvular fibrous body and mitral valve, with abscesses around the aortic root. Moderate mitral valve regurgitation was detected.

These findings were borne out in the OR, where the prosthetic valve was removed and involvement of the aortic root, intervalvular fibrous body and mitral valve was noted.

Easing the Challenges of Surgical Endocarditis Treatment

Severe infective endocarditis (IE) and its treatment can devastate the tissues around the heart. Historically, bivalvular IE has been treated with antibiotics, complete debridement of infected tissue, aortic valve homograft, and mitral valve repair or replacement.

But surgical repair and replacement of IE-affected tissues around the aortic and mitral valves — especially the intervalvular fibrous body in cases of prosthetic valve endocarditis — is a great challenge. Some surgeons call it a "commando operation."

Most commando operations are reoperations, often in patients with prosthetic aortic valve IE in which the infection aggressively extends to and destroys not just the aortic annulus but also the intervalvular fibrous body and the anterior leaflet of the mitral valve. The intervalvular fibrous body, an essential piece of structural tissue between the valves, is reconstructed using either autologous or bovine pericardial tissue, which often proves difficult.

Our team at Cleveland Clinic has recently developed a new technique to come to the relief of the commando operation. Instead of replacing both infected valves, the technique replaces the aortic valve and repairs the mitral valve using a transplanted segment of donor heart aorta and mitral valve, thus addressing the need to restore the intervalvular fibrous body in a less technically challenging manner.

This procedure — formally known as the incorporated aortomitral homograft and referred to at Cleveland Clinic as the "hemi-commando" — shows great promise as a more satisfactory solution that incorporates tissue of almost the same quality as the resected tissue.

Case Continued: The Hemi-Commando in Action

We used this hemi-commando technique in the patient from the case vignette above. In this instance, the homograft included the aortomitral membrane, the ascending aorta, the dome of the left atrium and part of the mitral valve annulus. The homograft was transplanted intact, as a unit (Figure 1). An annuloplasty ring was placed around the mitral valve.

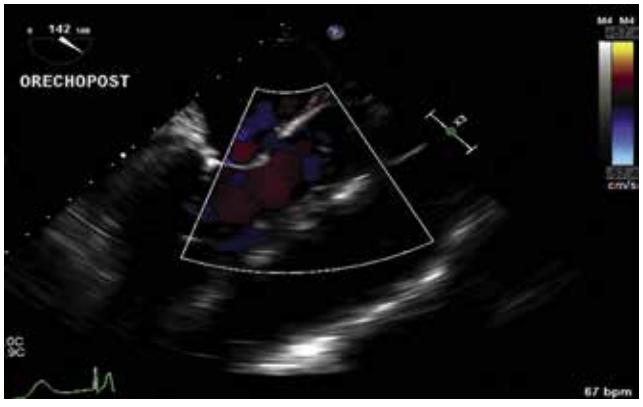
Postoperative echocardiography (Figure 2) showed normal aortic valve homograft function and trivial mitral valve regurgitation.

Experience with the Hemi-Commando to Date

By preserving the native mitral valve with its subvalvular apparatus, chordae and papillary muscle, the incorporated aortomitral homograft is designed to reduce trauma, increase resistance to infection and preserve native mitral valve function.

The technique has proved reliable and safe in our experience to date. Twenty-five patients have undergone the incorporated aortomitral homograft procedure here as of mid-2016, with a good survival curve. Eighteen cases were reoperations, and four were second reoperations.

Figure 2. Postoperative echocardiograms of the case patient showing the position and function of the homograft.



VALVE SURGERY: Numbers of Note (2016)

0.5%	Operative mortality for isolated aortic valve replacement (N = 377) (vs. 2.1% predicted [STS])
1.2%	Operative mortality for isolated mitral valve replacement (N = 84) (vs. 4.9% predicted [STS])
0%	Operative mortality for isolated mitral valve repair (N = 314) (vs. 0.7% predicted [STS])
2.0%	Operative mortality for combined aortic and mitral valve replacement (N = 51)
2.2%	Operative mortality for other valve surgeries* (N = 457)
0.3%	Procedural mortality for transcatheter aortic valve replacement (N = 365)

*Excludes those combined with CABG, which are reported on page 4.
STS = Society of Thoracic Surgeons

The Payoffs: High-Quality Tissue, Native Mitral Valve

In cases of extensive prosthetic aortic valve IE that involves the intervalvular fibrous body and mitral valve, radical complete debridement of all infected and devitalized tissue is the main principle of surgical treatment. The goals are to ensure secure fixation of the new prosthetic valve and avoid recurrent or residual infection, periprosthetic leak, dehiscence or subannular aneurysm formation. This occasionally leads to resection of the intervalvular fibrous body and, consequently, disruption of the fibrous skeleton of the heart, creating a tissue defect that is successfully repaired with an aortic valve homograft and its anterior mitral valve leaflet.

At Cleveland Clinic, we recommend consideration of the hemi-commando technique in these challenging IE cases, as it provides the advantage of incorporating tissue of almost the same quality as the native tissue and reconstructs the intervalvular fibrous body with the invaluable preservation of the native mitral valve. ■

Dr. Navia (naviaj@ccf.org), who developed the hemi-commando technique, is a staff surgeon in the Department of Cardiothoracic Surgery and a member of Cleveland Clinic's multidisciplinary Endocarditis Center.

Multistage Repair Featuring a Branched Frozen Elephant Trunk Technique for Aortic Dissection

BY ERIC E. ROSELLI, MD, AND BRIAN GRIFFIN, MD

History

A 42-year-old man from North Carolina referred himself to the Aorta Center in Cleveland Clinic's Miller Family Heart & Vascular Institute in 2015. At age 8, he had been diagnosed with Marfan syndrome, a genetic disorder that led to an imbalance in the maintenance and turnover of his connective tissues.

In early adulthood, he developed an aneurysm of the root of his aorta. In 2001, at age 28, he suffered an acute type A aortic dissection, a tear in the intimal layer of his ascending aorta that led to complete separation of the layers of the aorta from the root to the bifurcations of both femoral arteries. He underwent an emergency operation to replace his aortic root and ascending aorta, including reimplantation of the coronary arteries and replacement of the aortic valve with a mechanical prosthesis (Bentall procedure with a composite valve graft). The operation was limited to addressing the most life-threatening segments of the aortic dissection even though the dissection had extended all the way down through the femoral arteries.

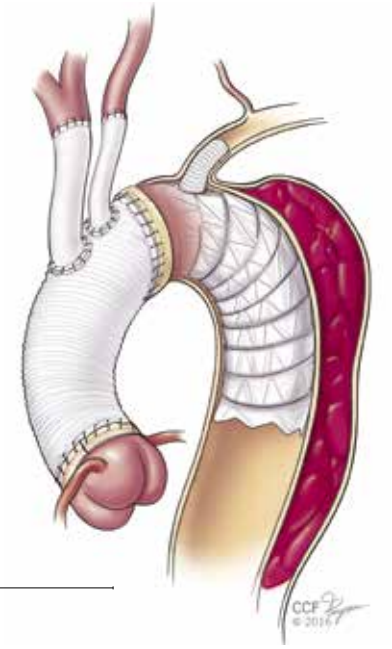


Figure 2. Illustration demonstrating results of the branched frozen elephant trunk procedure.

Figure 1. Volume-rendered three-dimensional CT reconstruction of the contrast-enhanced images from outside institutions, reformatted at the Aorta Center. The top arrow denotes the maximally dilated segment of the chronically dissected aorta in the distal aortic arch. The bottom arrows indicate the aneurysmal femoral arteries on both sides.

Several years later, when the patient was 35, his providers noted that the residually dissected and untreated segments of his aorta had begun to dilate, and regular imaging was performed to monitor the dilation.

Over the two years before he came to Cleveland Clinic, the patient's aorta had grown to a size that prompted consideration of additional aortic replacement. But the surgical options recommended at outside institutions carried high risk for death, stroke and paralysis, so the patient put off further surgery until learning of the multidisciplinary expertise of Cleveland Clinic's Aorta Center and Marfan Syndrome/Connective Tissue Disorders Clinic.

Presentation

Upon the patient's presentation at Cleveland Clinic, his CT scans were retrieved for review. Their raw data were downloaded to our servers, and reconstruction software was used to perform detailed image analysis. At presentation, his maximum aortic diameter was greater than 7 cm, and the aneurysmal degeneration extended from the end of his previous repair through the entirety of the aorta and into both femoral arteries, each of which measured nearly 3 cm (Figure 1).

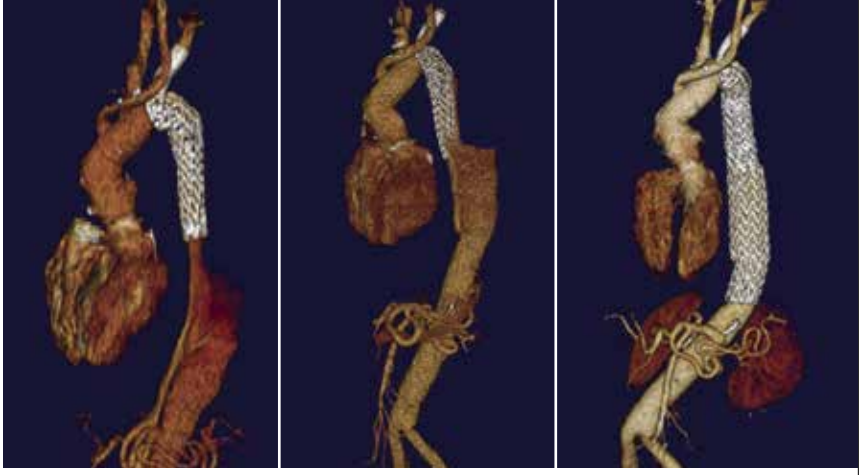


Figure 3. Volume-rendered 3-D CT reconstructions demonstrating (left) the aorta after the first but before the second stage of the aortic repair, (middle) the period between the second and third stages, and (right) follow-up at three months after repair completion.

Management Planning

With these image analyses as our basis, our team developed a comprehensive staged treatment plan to include multiple operations over a two-week period. The plan was fleshed out with a series of additional evaluations, as follows:

- The patient was evaluated by a genetic counselor to confirm the specifics of his diagnosis, address questions on the heritability of his connective tissue disorder and determine the needs of family members.
- He was seen by our ophthalmology colleagues at Cleveland Clinic's Cole Eye Institute to evaluate a previous lens replacement in one eye related to his Marfan syndrome and to assess the other eye.
- Echocardiography and evaluation by a cardiologist with expertise in Marfan syndrome (co-author Griffin) revealed moderately severe mitral regurgitation with annular dilatation and large redundant mitral leaflets with preserved biventricular function and ventricular dilatation.

After discussion with the team, mitral repair was added to the plan for the first-stage operation. The patient was admitted for warfarin reversal with heparin bridging, and coronary catheterization via a right radial approach demonstrated preserved coronary anatomy without occlusion or aneurysm. This approach allowed for more direct access to the coronaries and avoided both the aneurysmal femoral arteries and navigation through the dissected distal aorta.

Staged Surgical Treatment

The first operation was a redo sternotomy with right axillary artery cannulation to provide the safety of selective antegrade brain perfusion. The mitral valve was repaired, and the aortic arch was replaced using a branched modified frozen elephant trunk technique developed by one of the authors (Dr. Roselli) that includes the following (Figure 2):

- Direct placement of a commercially available thoracic stent graft
- Creation of an opening in the stent graft device
- Direct stent grafting of the left subclavian artery
- Circumferential sewing of the stent graft into the mid-arch region of the aorta during the selective brain perfusion phase
- Sewing of a separate bypass graft to the left common carotid and innominate arteries

After a week and a half of recovery, the patient underwent the second-stage operation: open thoracoabdominal repair, including repair of both femoral artery aneurysms, separate branch grafting to each of the visceral and renal arteries, and preservation of the internal iliac arteries, which are important for spinal cord protection. The repair was limited to a level just above the diaphragm to maintain important intercostal branches and provide a period of ischemic conditioning to the spinal cord before the third-stage operation, which involved finalizing treatment with complete replacement of his aorta from the left ventricle to the superficial femoral arteries (Figure 3).

Three days after the open thoracoabdominal repair, he underwent completion with a thoracic endovascular aortic repair with access through the recent femoral artery interposition graft.

Outcome

The patient recovered well from this tour-de-force reconstruction and returned home to North Carolina by car with no serious complications and fully neurologically intact. He has since returned for three-month follow-up and will continue to undergo annual imaging to monitor his repairs and native vasculature. ■

Dr. Roselli (roselle@ccf.org) is a cardiothoracic surgeon and Director of the Aorta Center. Dr. Griffin (griffib@ccf.org) is Head of the Section of Cardiovascular Imaging and a staff cardiologist in the Marfan Syndrome/Connective Tissue Disorders Clinic.

AORTA SURGERY: Numbers of Note

1,156	Aorta surgeries performed (2016)
2.9%	Aorta surgery operative mortality (2016)
4,629	Outpatient visits to Aorta Center (2015)
6-fold	Increase in thoracic aorta surgical volumes over past 20 years

LV Outflow Tract Obstruction After Mitral Valve Replacement: Two Cases, Same Keys to Success

BY NICHOLAS G. SMEDIRA, MD, AND ROBERT J. STEFFEN, MD

Left ventricular outflow tract obstruction (LVOTO), both fixed and dynamic, can be caused by many factors. Two of our recent cases display a fixed pattern of severe obstruction secondary to completely retained in situ anterior mitral valve leaflet.

Case 1

A 58-year-old woman underwent attempted mitral valve repair for degenerative mitral disease at an outside institution. Multiple attempts at valve repair were followed by bovine mitral valve replacement with complete retention of the anterior leaflet. Her postoperative course was prolonged because of cardiogenic shock requiring temporary mechanical support and the need for tracheostomy. A year later, she had persistent heart failure symptoms including fatigue, dyspnea on exertion, fluid retention and weight gain.

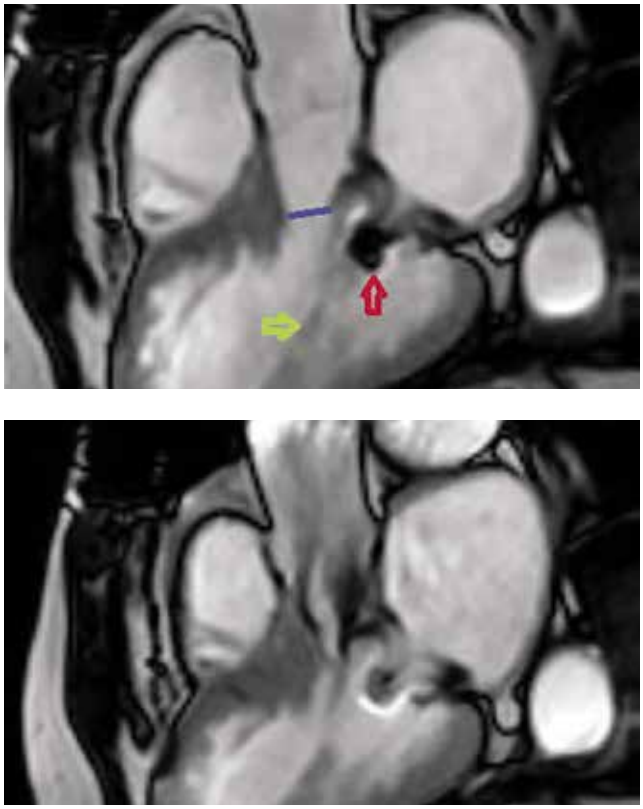


Figure 1. Cardiac MRIs from the first patient. Top image demonstrates a narrow left ventricular outflow tract (LVOT) (blue line) in diastole. Its diameter is 9 mm at the narrowest point, in contrast to 24 mm at the level of the aortic valve. Red arrow indicates the prosthetic mitral valve strut; yellow arrow indicates chordae to the retained anterior mitral leaflet. Bottom image demonstrates turbulent flow through the LVOT in systole.

An echocardiogram performed at Cleveland Clinic showed a fixed, elevated gradient across her LVOT secondary to retained anterior mitral leaflet. Her resting LVOT gradient was 52 mm Hg. The gradient increased to 56 mm Hg with Valsalva maneuver and 70 mm Hg with stress. The interventricular septum was noted to be 1.5 cm. There was no paravalvular leak or mitral regurgitation. Cardiac MRI confirmed the outflow obstruction secondary to a completely retained leaflet and subvalvular apparatus (Figure 1).

Case 2

A 69-year-old woman had undergone mechanical mitral valve replacement as well as coronary bypass at an outside institution in 2002. She had known for a few years that she had an elevated LVOT gradient. Recently she had developed worsening chest pain and dyspnea on exertion with occasional rest symptoms. Cardiac catheterization confirmed that her bypass grafts were patent without any new coronary artery disease.

A transesophageal echo (TEE) demonstrated turbulent flow in the subaortic area, with a peak gradient of 155 mm Hg. Her aortic valve was normal. There was no paravalvular leak or mitral regurgitation. The width of her septum was 1.5 cm. The echo demonstrated the cause of her gradient to be the retained valve and subvalvular apparatus, with chordal attachments extending into the LVOT.

SEPTAL MYECTOMY: Numbers of Note (2016)

201	Septal myectomies performed
0.5%	Septal myectomy operative mortality
1967	Year the first septal myectomy was performed at Cleveland Clinic
3,030	Total septal myectomies performed through 2016

Figure 2. Photo of the explanted anterior mitral leaflet, chordae and papillary muscle that was retained at the time of mitral valve replacement in the first patient.

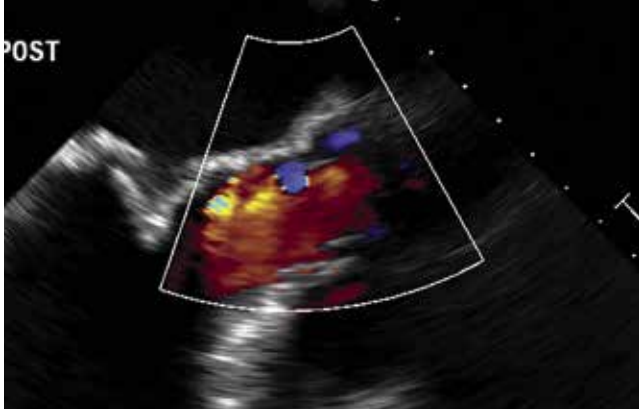


Figure 3. Postoperative transesophageal echocardiogram of the first patient showing laminar flow through the left ventricular outflow tract.

Two Cases, Similar Surgical Approach

Although the presentation of each patient was unique, the challenge of widening the outflow tract without removing the prosthetic valve was similar. Both procedures were performed using the transaortic approach we use to treat patients with hypertrophic obstructive cardiomyopathy.

Through a transverse aortotomy just above the sinotubular junction, the LVOT and its contents were assessed. This view also allowed for visualization of the antero- and infero-interventricular septum, papillary muscles, chordae and anterior mitral leaflet.

In the first patient, the anterior leaflet of the mitral valve was completely removed from around the sewing cuff and struts of the biologic prosthesis (Figure 2). The anterolateral papillary muscle was found to have a prominent head, which was removed. Lastly, a 3-g myectomy of the anteroseptum was performed. Postoperative TEE demonstrated a wide-open LVOT with no dynamic obstruction on administration of dobutamine and nitroglycerine (Figure 3). No paravalvular leak was noted around the prosthesis. The patient was extubated on postoperative day 0 and discharged on day 5.

Similarly, in the second patient the remnant of the anterior mitral leaflet was resected. A severely hypertrophied head was removed from both anterolateral and posteromedial papillary muscles. Lastly, a 5-g septal myectomy was performed. This patient had developed atrial fibrillation as well, so a concomitant CryoMaze procedure was performed. Postoperative TEE demonstrated a wide-open outflow tract with no dynamic obstruction or paravalvular leak.

Discussion: Insights Gleaned Through Volume and Experience

These cases demonstrate key aspects of investigation and operative care of patients with LVOTO after previous mitral valve replacement. Both reports detail complete in situ retention of the anterior leaflet, suggesting a possible mechanism for the obstruction. Maintaining continuity between the annulus and left ventricle through chordal-sparing techniques is important in preserving late left ventricular function in patients undergoing mitral replacement. These cases show, however, the importance of completely detaching the anterior leaflet from the annulus, excising the central portion and positioning valve remnants at or below the commissure level (below the 3 and 9 o'clock positions as viewed through the left atrium).

Identifying LVOT pathology in patients with a mitral prosthesis in place can be very difficult using traditional transthoracic echocardiography. In our first patient, MRI was used as an adjunct to identify retained chords from the in situ anterior mitral leaflet. In our second patient, TEE was used to visualize the retained subvalvular apparatus and identify it as the cause of the high gradient.

Performing these operations through a transverse aortotomy greatly simplifies the surgery. Direct visualization of the outflow tract allows the surgeon to confirm the cause of obstruction and intervene as indicated on the hypertrophied septum, papillary muscles and anterior mitral leaflet. Both patients were treated using limited cardiac dissection and without re-replacing the mitral prosthesis, saving significant time and reducing the risk of complications. As a result, the cross-clamp time without the maze procedure was only 20 minutes. Both patients were extubated within 24 hours, left the ICU one day after surgery and had an uneventful recovery.

LVOTO entails a complex interplay of all components of the ventricle. Surgeons must be cognizant that the anterior mitral leaflet and its subvalvular apparatus can lead to LVOTO if not detached and moved at the time of mitral valve replacement. Surgery for LVOTO in the setting of a mitral prosthesis can be done safely and effectively with good outcomes. ■

Dr. Smedira (smedirn@ccf.org) is a staff surgeon and Dr. Steffen is a cardiothoracic surgery fellow, both in the Department of Cardiothoracic Surgery.

TAH as Bridge to Transplant for a Patient with Biventricular Heart Failure and Myriad Comorbidities

BY NADER MOAZAMI, MD; EDWARD SOLTESZ, MD; AND MIRIAM JACOB, MD

Presentation

A 57-year-old man diagnosed in 2008 with viral cardiomyopathy subsequently developed progressive nonischemic cardiomyopathy. In early 2015, he reported symptoms consistent with atrial fibrillation and underwent ablation and implantable cardioverter-defibrillator (ICD) placement. In mid-April 2015, he was admitted to his local hospital with cardiogenic shock and treated with dobutamine and diuretics. Nevertheless, his condition continued to deteriorate rapidly. He developed biventricular heart failure, was placed on extracorporeal membrane oxygenation (ECMO) support and was transferred to Cleveland Clinic in late April for further management.

Initial Management

On arrival, the patient was cachectic and in renal and liver failure. Although he was intubated, his neurological function was intact, and he was able to understand and consent to a plan of care. One week after his arrival, ECMO and his ICD were removed and his heart was replaced with a SynCardia temporary Total Artificial Heart (TAH) (illustrated at right) as a bridge to transplantation.

Over the succeeding weeks, the patient had a complicated recovery that included respiratory failure, pneumonia, leukocytosis and dysphagia. With aggressive resuscitation and multidisciplinary management, he continued to overcome these complications, and his liver and renal function improved with the TAH therapy. With aggressive additional in-hospital physical therapy, he improved to the point that in late July he was listed as UNOS status 1A for heart transplantation.

In mid-August, the TAH driver was changed to SynCardia's portable Freedom® driver, with the goal of allowing the patient to wait for a donor heart at home. Ultimately, he and his wife felt insecure about leaving the hospital and decided to wait at Cleveland Clinic.

Transplantation and Post-Transplant Outcome

In early October, five months after his admission to Cleveland Clinic, the patient's TAH was explanted and replaced with a healthy, well-functioning donor heart. Although this was a complex reoperation, he quickly improved after transplantation and was discharged home in mid-November

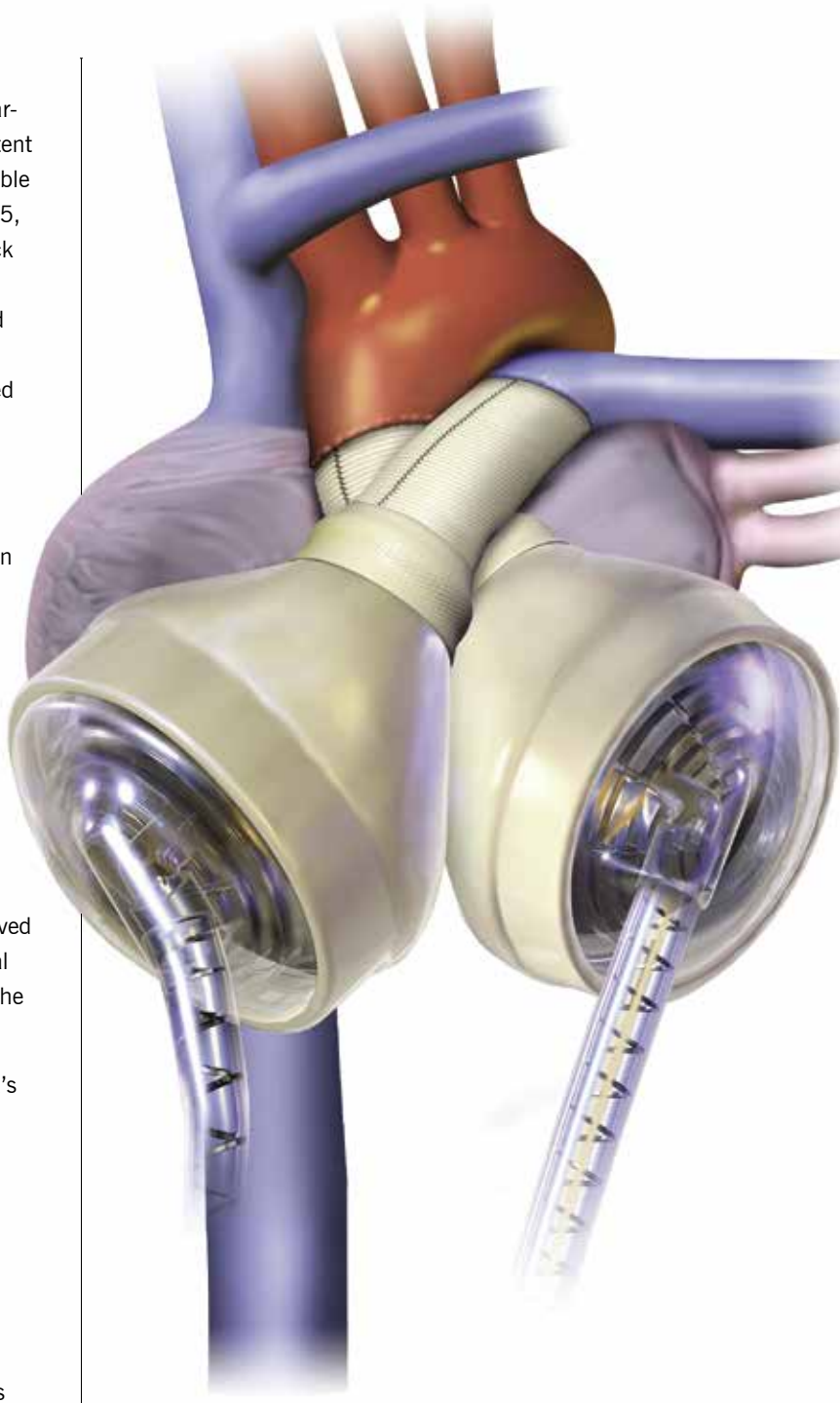


Image courtesy of syncardia.com

2015 on typical post-transplant medications with prescriptions for home occupational, physical and speech therapy.

One year later, the patient returned for a checkup. He was back to normal activity, eating well, symptom-free and in good spirits.

Discussion: When TAH Is the Only Option

This patient presented multiple challenges. He was initially too sick to transplant, with severe biventricular failure. A TAH was his only salvage option. These devices have proved highly effective as rescue therapy in patients with biventricular failure. However, because patients like this typically spend months in the hospital to recuperate, success with a TAH requires cooperative, resilient and motivated patients (and families) who are able and willing to work hard to improve and strengthen themselves in preparation for transplant.

Heart transplantation following removal of a TAH is challenging, in view of the significant inflammatory response to the device and the extensive fibrosis that obliterates all surgical planes.

Complex, but Typically So

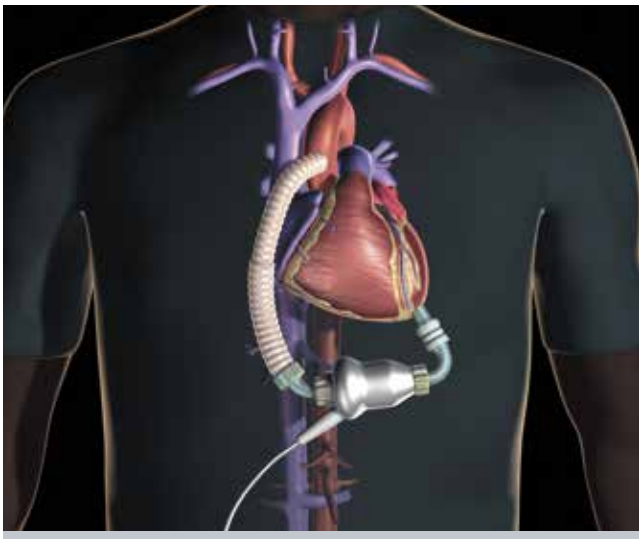
Although this patient's case appears dramatic, it is typical of the complex patients we see every day and illustrates the dual strengths of Cleveland Clinic's Cardiac Transplantation and VAD Program: (1) an expert, well-trained, fully integrated multispecialty team and (2) more than 20 years' experience with all types of mechanical support devices. These qualities enable us to tailor therapy for very sick patients with varying physiologies and indications.

Our heart transplant team comprises three surgeons and 12 cardiologists. The cardiologists rotate weekly, which allows them to become familiar with every patient. They are supported by a superb staff of mechanical support nurse practitioners who manage day-to-day care. The transplant team meets daily to discuss how best to achieve our common goal of stabilizing patients, transplanting them and sending them home.

This patient's array of serious comorbidities underscores the importance of supportive staff and multidisciplinary specialty expertise for advanced heart failure care. Our cardiac anesthesiologists are comfortable with the challenges of anesthetizing patients on mechanical support devices and those undergoing heart transplant. Their expertise in the surgical ICU is invaluable. We likewise count on expert colleagues from Cleveland Clinic's distinguished infectious disease, nephrology, gastroenterology, pulmonary medicine and other specialty programs to help resolve the multiple medical issues that so often accompany advanced heart failure.

By working together, we aim to ensure that these extremely fragile patients receive optimal care around the clock during extended hospitalization and after discharge. ■

Dr. Moazami (moazamn@ccf.org) and Dr. Soltesz (soltese@ccf.org) are cardiothoracic and heart transplant surgeons in the Department of Cardiothoracic Surgery. Dr. Jacob (jacobm2@ccf.org) is a cardiologist in the Section of Heart Failure and Transplant Medicine.



VADs & TRANSPLANT: Numbers of Note

169	Heart transplants performed, 2014-2016
86.7%	3-year adult heart transplant patient survival (N = 105) (vs. 84.8% expected)*
156	Adult durable ventricular assist devices (VADs) or TAHs implanted, 2014-2016
76.3%	2-year survival for primary VAD recipients, 2012-2016**

*Scientific Registry of Transplant Recipients (Jan. 2017 report)
**Intermacs report through Sept. 30, 2016

A Complex Case of Arterial Thoracic Outlet Syndrome

BY SEAN LYDEN, MD

Presentation

In 2015, a 56-year-old woman was transferred to Cleveland Clinic for management of acute onset of left arm ischemia. Three-dimensional (3-D) CT-reconstructed imaging showed a left subclavian aneurysm with embolization, a rare and potentially dangerous complication of arterial thoracic outlet syndrome (Figure 1).

Figure 1. 3-D CT reconstruction at presentation showing the left subclavian aneurysm with embolization.

Initial Interventions

Because of the embolic burden in the outflow of the arm and hand, the initial strategy was catheter-directed pharmacologic thrombolysis. We used a dual-level infusion for thrombolysis with tissue plasminogen activator (t-PA). Progress of the therapy was checked eight hours after initiation. Minimal progress of the lysis was noted, and there was significant residual clot burden in the arm, so mechanical thrombectomy was performed with the Possis AngioJet™ device.

After thrombectomy, several areas of stenosis were uncovered in the brachial and radial arteries. These were treated with percutaneous transluminal angioplasty, which improved flow to the arm. The patient was kept on therapeutic anticoagulation with planned resection of the subclavian artery aneurysm.

Second Ischemic Event

Despite anticoagulation, rethrombosis of the brachial and axillary arteries occurred six hours later. The patient was returned to the OR, where she underwent open thrombectomy of the arteries from a brachial cutdown and local instillation of t-PA into the radial and ulnar arteries to improve outflow. The forearm was noted to be tense. Clinically, compartment syndrome was present and was the likely etiology of the rethrombosis. This was treated with forearm fasciotomy and carpal tunnel release. The hand perfusion was markedly improved by the repeat operation.

Resection of Ribs and Aneurysm

Elevation of the subclavian artery over a cervical rib leads to local arterial trauma, aneurysmal formation with thrombus deposition, and embolization to the distal extremity. For this reason, surgical correction of the arterial thoracic outlet pathology was performed a few days later. This included resection of the left cervical and first ribs, resection of the aneurysm and reconstruction with a left subclavian-to-axillary artery graft using the right femoral vein. The operation was notable for a very friable and small proximal subclavian artery just distal to the left vertebral artery.

Duplex ultrasonography before discharge and at six-month follow-up intervals during the first year showed a widely patent bypass and equal upper extremity brachial pressures.

Figure 2. 3-D CT reconstruction demonstrating stenosis at the proximal anastomosis one year after initial presentation.

A Year Later: Stenosis Found at Proximal Anastomosis

Although the patient had no symptoms, surveillance duplex ultrasonography after one year showed increased left subclavian artery velocities from 60 to 187 cm/s at the proximal anastomosis suggestive of a greater than 50 percent stenosis. Pulse volume recordings were consistent with new mildly dampened waveforms in the left arm, and a pressure drop of 10 mm Hg was noted in the left brachial artery relative to the right. Findings were confirmed by CT angiography, and 3-D CT reconstruction clearly demonstrated a stenosis at the proximal anastomosis (Figure 2).

Weighing the Treatment Options

Multiple options were available to treat this problem. Surgical revision was an option, but the reoperative nature and the small friable subclavian artery at the original operation made this the least appealing approach. Endovascular revision, the minimally invasive approach, was appealing to the physicians and patient. Several interventional options were considered:

- Angioplasty has been a successful strategy in lower extremity bypass stenoses. Cutting or scoring balloons typically have had better outcomes than plain balloons due to the ability to allow dilation at lower pressures and deal with the intimal hyperplastic nature and the "watermelon seed" movement of plain balloons over these smooth stenoses. This approach was not chosen due to the large diameter mismatch of the artery to the vein and the lesion being primarily of the artery.

Figure 3. CT reconstruction showing the widely patent bypass upon elevation of the patient's arm as well as proximal anastomotic stenosis (arrow).

- Stents placed across the thoracic outlet typically are at risk for stent fracture if the thoracic outlet has not been surgically decompressed with rib removal. The CT was done with the patient's arms above the head and confirmed adequate surgical decompression of the space, making stenting an option (Figure 3).
- Self-expandable nitinol stents have had good outcomes in the treatment of subclavian artery occlusive disease. The large vein diameter would have required a very large self-expanding stent. We felt the risk of distal migration would make a stent of this type a less-than-ideal choice.
- Self-expandable drug-eluting stents have had good patency in the femoral arterial segment. The shortest commercially available stent is 40 mm long, which would have been too large for this short lesion. The largest diameter is 8 mm and would have had the same risk of distal migration as a plain nitinol stent.
- Balloon-expandable drug-eluting stents have had excellent results in the coronary circulation and come in short lengths but were not available in large enough diameters for this vessel.

VASCULAR SURGERY: Numbers of Note (2015)

3,555	Vascular surgeries, including bypass and arteriovenous access procedures
0%	In-hospital mortality for femoral endarterectomy with stenting (N = 120)
1,877	Patient visits to the Lower Extremity Wound Clinic

- Drug-eluting peripheral balloons were also considered, but the balloons are long and would require treatment of a long segment of normal vessel, and there are no data on the efficacy of this approach.
- We decided to use a peripheral balloon-expandable stent to allow flaring of the distal portion to accommodate the diameter mismatch from the subclavian artery to the vein bypass. We felt this had the lowest chance of migration.

Treatment, Outcome and Follow-Up

A 6-mm-diameter, 17-mm-long stainless steel stent was used and was flared to 10 mm distally. The completion angiogram was normal (Figure 4).

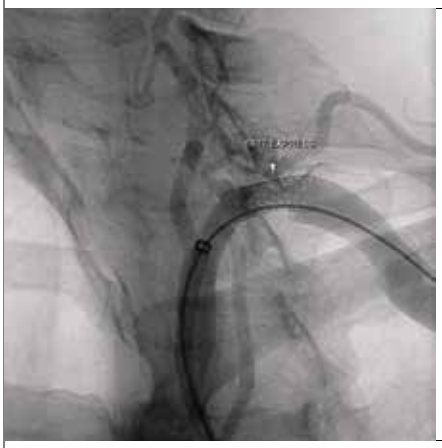


Figure 4. Angiogram taken after stenting and flaring of the distal stent. Note the absence of residual stenosis.

The patient was discharged the same day on dual antiplatelet therapy and returned for office follow-up six weeks later with normalization of the duplex velocities, normal plethysmography and equal brachial pressures. We expect to continue to follow her for this problem for many years to come. ■

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When Nonresponse to CRT Demands Systematic, Multidisciplinary Troubleshooting

BY JOHN RICKARD, MD, MPH

History and Presentation

Mr. JL is a 48-year-old African-American male who until one year ago had a medical history significant only for obstructive sleep apnea, for which he intermittently used continuous positive airway pressure therapy. He noted at his job as a construction worker that he was becoming progressively short of breath during routine tasks and was eventually referred to a cardiologist in his local community. Echocardiography showed significant left ventricular dysfunction, with a left ventricular ejection fraction (LVEF) of 20 percent. Left heart catheterization showed normal coronary arteries. He was started on carvedilol 12.5 mg twice daily, lisinopril 10 mg daily and furosemide 80 mg daily. He continued to have significant symptoms but was able to return to work.

Repeat echocardiography six months after diagnosis revealed his LVEF to still be 20 percent despite optimal medical therapy. As a result, he was referred for implantation of a biventricular defibrillator. A percutaneous left ventricular lead could not be placed, so he was referred for surgical left ventricular lead placement, which he underwent successfully at an outside institution. He was discharged in stable condition.

Over the next six months, his symptoms continued to worsen. He could no longer work and was hospitalized for heart failure. When a follow-up echocardiogram showed that his LVEF had dropped to 15 percent, he was referred to Cleveland Clinic's Cardiac Resynchronization Therapy Optimization Clinic.

CRT Optimization Clinic at a Glance

This specialized clinic employs an algorithmic approach for patients who have not responded to cardiac resynchronization therapy (CRT) as much as was expected in terms of echocardiographic and/or symptomatic improvement. The clinic uses a multidisciplinary model — with input from electrophysiologists, heart failure and imaging cardiologists, cardiothoracic surgeons and sometimes other subspecialists — to discuss findings from the algorithm-driven examination, determine potential reasons for inadequate CRT response and establish a treatment plan to optimize response and long-term outcomes.

Case Continued at Cleveland Clinic

Physical examination revealed volume overload, a nonischemic cardiomyopathy of only one year's duration, grade 2+ lower extremity edema and faint bibasilar rales. Comprehensive device evaluation showed 99 percent biventricular pacing with excellent function of all leads. Twelve-lead ECG was performed (Figure 1, left panel) and showed a paced pattern with a deep S wave in lead V1, raising potential questions about lead position. Adequate capture of the left ventricular lead was confirmed, and no anodal stimulation was noted.

Suppression of pacing revealed a wide underlying left bundle branch block (Figure 1, right panel), suggesting a highly conducive substrate for response to CRT. When chest X-ray showed an anterior/anterolateral surgically placed left ventricular lead (Figure 2), this was determined to be the likely culprit for CRT nonresponse following discussion at a multidisciplinary conference.

Figure 1. The patient's ECGs upon presentation (left) and with suppression of biventricular pacing (right).

Figure 2. Lateral chest X-ray showing an anterior/anterolateral left ventricular pacing lead.

Figure 3. Coronary sinus venogram showing potential targets for percutaneous coronary sinus lead placement at the lateral and posterolateral branches.

A copy of the patient's coronary sinus venogram was then obtained (Figure 3) to determine whether another attempt at percutaneous placement would be reasonable. The venogram showed two potential targets:

- The lateral branch was relatively small and tortuous at the proximal portion with a trifurcating system at the ostium that could be accessed but apparently would not accept a lead.
- The posterolateral branch was moderately sized, but a stenosis in its midsection made it unable to accept a lead, although a wire could pass beyond it.

A repeat attempt at surgical lead placement was considered, but consultation with cardiothoracic surgery colleagues determined that the patient was not an ideal candidate due to anatomical constraints. A CRT optimization echocardiogram with three-dimensional strain imaging was performed, showing worsened cardiac performance with CRT on versus off.

RHYTHM DISORDERS: Numbers of Note (2015)

5,175	Electrophysiology lab procedures
1.62%	Rate of major complications from ablation for atrial fibrillation
1,038	Lead extractions
97.9%	Lead extraction clinical success rate
0.97%	In-hospital risk-adjusted complication rate for ICD implants (vs. 90th percentile rate of 1.09% from NCDR ICD Registry™)

In view of these collective findings, the CRT Optimization Clinic's multidisciplinary team made a final recommendation to proceed with a repeat attempt at percutaneous coronary sinus lead placement targeting the posterolateral branch. The recommendation calls for interventional cardiology assistance to perform venoplasty of the stenotic midsection of the vessel to allow passage of the lead. As of this writing, the procedure has been scheduled but not yet performed.

Discussion: The Case for CRT Optimization

CRT has been a godsend for individuals with advanced heart failure, but as many as one-third of patients fail to respond to CRT. In the absence of a thorough multidisciplinary assessment, these patients are at increased risk for poor exercise tolerance, repeat hospitalizations and early death.

At Cleveland Clinic, we recommend that all patients who receive a CRT device be re-evaluated with an echocardiogram within six to nine months. At that point, patients who have realized little to no benefit should undergo "troubleshooting" of their care to systematically identify possible reasons for nonresponse. We have created our CRT Optimization Clinic, one of the few programs of its type in the nation, to deliver that troubleshooting in a way that ensures a collaborative, multidisciplinary approach guided by a thorough, algorithm-directed evaluation.

The clinic's collaborative ethos extends to fully include patients and their referring cardiologists. If a major intervention is required (e.g., reoperation to move a lead), the recommendation is discussed with the referring clinician and a plan is formulated. If only minor device changes are needed or no significant interventions are possible, patients are followed by their referring cardiologist with Cleveland Clinic's CRT Optimization Clinic acting in a consultative role. ■

Dr. Rickard (rickarj2@ccf.org) is an electrophysiologist and Director of Cleveland Clinic's CRT Program.

Advanced Multilayer Cardiac Disease from Prior Thoracic Radiation: Why Options Matter

BY BRIAN GRIFFIN, MD; SAMIR KAPADIA, MD; AND DOUGLAS JOHNSTON, MD

Presentation

A 77-year-old man presented to Cleveland Clinic for a second opinion on his dyspnea that had begun two years earlier and had worsened significantly over the prior eight months. His complex medical history included thoracic radiation therapy for Hodgkin's lymphoma in 1979. He subsequently developed coronary artery disease, which had been treated with stenting in 2009. He also suffered from renal insufficiency, due to having only one kidney, and Crohn's disease.

We conducted a series of tests:

- ECG showed normal sinus rhythm and right bundle branch block, but no Q waves.
- Pulmonary function testing confirmed moderate obstructive lung disease and moderate to severe restrictive disease.
- Echocardiography showed severe aortic stenosis and a moderate degree of mitral valve calcification and stenosis. (See Figures 1 and 2 for representative findings.)
- Chest CT showed extensive calcification of both valves, as well as moderate pulmonary fibrosis and adhesions, all of which were attributed to his prior radiation.
- Coronary angiography revealed only mild to moderate coronary disease and patent stents.

Based on these test results, we determined the patient's dyspnea was not caused by coronary disease but rather by calcific valve disease.

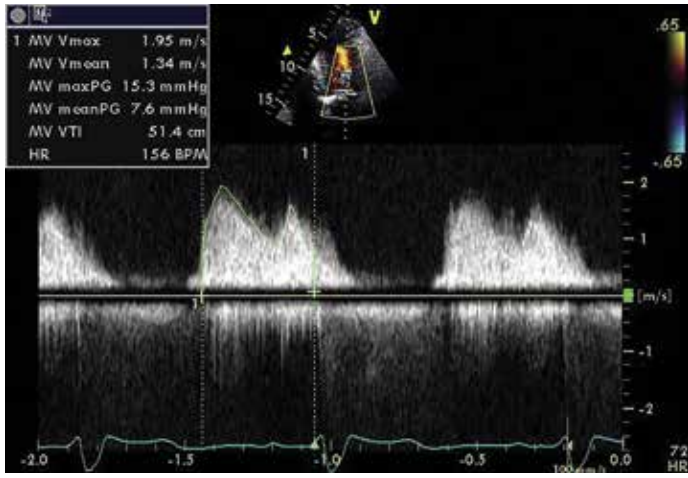

Figure 2. Continuous-wave Doppler echocardiogram of the patient's mitral valve upon presentation.

Management Options

With complex patients like this, management decisions are best made by a multidisciplinary team including a cardiac imaging specialist, an interventional cardiologist and a cardiothoracic surgeon. Three treatment options were discussed in this case:

1) Surgical replacement of the aortic and mitral valves. Issues included:

- Whether the aortic valve should be debrided or replaced
- Whether mitral valve replacement would be necessary
- Whether the patient was a good surgical candidate
- What postoperative complications could be anticipated, considering his medical history and comorbidities

2) Medical treatment only. Questions included:

- Could the patient handle diuretics with a single kidney?
- Was palliative care his only option?

3) Transcatheter aortic valve replacement (TAVR). Questions included:

- Was this option feasible?
- Would treating his aortic valve, but not his mitral valve, relieve his symptoms?

The patient's Society of Thoracic Surgeons (STS) risk score was calculated to be 17.4 percent, which made him a high-risk surgical candidate. Moreover,

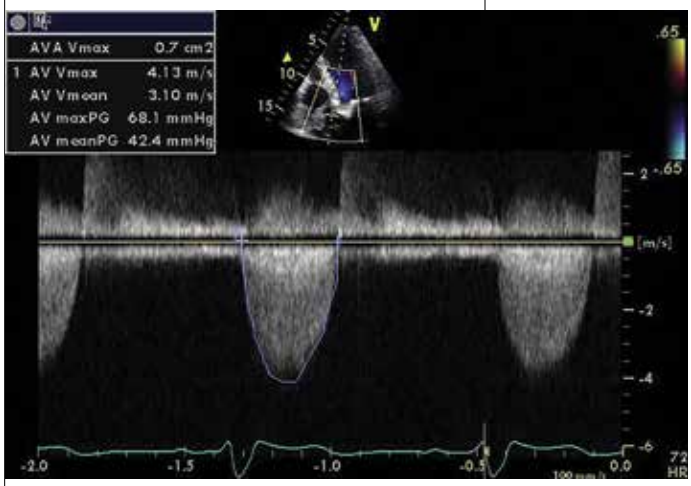

Figure 1. Echocardiography findings showing aortic velocity upon the patient's presentation.

his brachiocephalic vein, distal ascending aorta, proximal arch and right ventricular myocardium all were located at an unsafe distance (4-5 mm) from the retrosternal midline. This significantly increased the risk of damage to his heart during surgery.

We believed the patient's dyspnea could be relieved by improving blood flow through his aortic valve alone, given the significant stenosis present, so we determined he would be a good candidate for TAVR. Ideally, his mitral valve would have been replaced too, but this would have required open surgery, which we had eliminated as an option due to his STS risk score and the anatomical risk noted above.

Treatment and Outcome

TAVR was performed via transfemoral access without incident following balloon aortic valvuloplasty. The patient was left with a mild paravalvular leak, which improved on its own without intervention.

CARDIAC IMAGING: Numbers of Note (2015)

78,162	Echocardiograms
7,747	Cardiac CTs
3,286	Cardiac MRIs
2,027	Patient visits to Center for Pericardial Disease, a doubling of annual volume since 2011

At his one-year checkup (see Figure 3), the patient was breathing better and feeling better, and he said he felt he was "back to normal." His mitral valve, although still mildly narrowed, was not affecting his ability to exercise.

Discussion

Radiation therapy is known to cause extensive fibrosis and damage to the heart, lungs and great vessels. Common effects include calcific valve disease, constrictive pericardial disease, restrictive pattern of cardiomyopathy, myocardial fibrosis/scarring, coronary artery disease (ostial), microvascular dysfunction, carotid disease, conduction system disease and fibrotic lung disease.

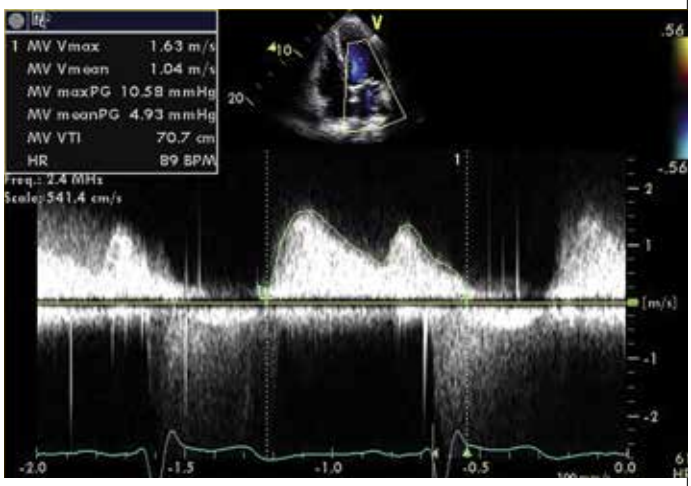

Figure 3. The patient's echocardiography findings one year after TAVR.

Radiation-associated heart disease is most common in patients who have received radiation for Hodgkin's lymphoma or breast cancer. In addition to varying by volume of heart irradiated and time since radiation exposure, risk is further increased by the following factors:

- Total dose exceeding 30-35 Gy
- Dose exceeding 2 Gy/day
- Younger age at time of radiation
- Concomitant cardiotoxic chemotherapy
- Use of cobalt as the radiation source
- Other cardiovascular risk factors

In many ways, our patient illustrates the problems inherent in treating radiation-associated heart disease. First, clinicians must be alert to the possibility of radiation as an etiologic factor, which is best determined by a thorough medical history. Due to the extremely delayed nature of the side effects (our patient had been radiated 35 years earlier), any history of chest radiation should be considered a potential underlying cause.

Once the possibility of radiation-associated heart disease is identified, multidisciplinary team input should be sought to ensure the best treatment choice. These are difficult patients whose treatment requires much thought, and diverse expertise and experience can be invaluable.

Experience shows that radiation-associated heart disease portends increased long-term mortality following cardiothoracic surgery, as demonstrated by a retrospective Cleveland Clinic cohort study (*Circulation*. 2013;127:1476-1484) of patients undergoing cardiac surgery for radiation heart disease versus other etiologies of cardiovascular disease. Our findings showed that radiation heart disease was associated with significantly worse survival. The reasons for this result are not well understood, though the combination of cardiac surgery and prior radiation may lead to compromised lung function and thereby compromise long-term outcomes, though immediate surgical mortality is low.

A key conclusion of our cohort study was that alternatives to surgery may be required in radiation heart disease to improve long-term survival. This case underscores the imperative for treatment options in this risky patient population. It also demonstrates that for patients with severe aortic stenosis and a functioning mitral valve, TAVR can represent an excellent solution. ■

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Treatment of a Rare Cardiac Tumor: The Value of Second-Opinion Case Review for Affiliated Providers

BY MICHAEL J. LAZAR, MD, MHA

Case Vignette

In July 2016, a 56-year-old woman presented to her community hospital in central Pennsylvania complaining of indigestion. Workup suggested a potential heart problem, and she was sent to an interventional cardiologist. Cardiac catheterization demonstrated normal coronaries, but a large tumor was found to be present in the right ventricle immediately below the tricuspid valve. This was confirmed with an echocardiogram, which also revealed a moderately sized pericardial effusion. The patient was referred to UPMC Susquehanna in Williamsport, Pennsylvania, for management.

We saw the patient in clinic. Because primary cardiac tumors are rare, one in this location is often secondary to a systemic process such as lymphoma. We ordered a whole-body PET scan and excluded this possibility.

Although our team has removed many cardiac tumors, the location of this one was unsettling, as it was encroaching on the junction between the atria and the ventricle. It was likely that a major reconstruction would be necessary following resection of the mass.

In view of UPMC Susquehanna's affiliate relationship with Cleveland Clinic's Miller Family Heart & Vascular Institute, I immediately called Cleveland Clinic cardiothoracic surgeon Edward Soltesz, MD, MPH, for his opinion on whether the tumor could be safely resected.

Dr. Soltesz and I reviewed the patient's PET scan and echo together via secure image-sharing technology. Because her angiogram was available only on a disc, it was sent by overnight mail. The following day, Dr. Soltesz and his colleagues reviewed the films and confirmed that the patient was an appropriate candidate for surgery. They also agreed that her tumor was rare enough to merit referral to Cleveland Clinic for surgery. The patient agreed to travel to Cleveland and underwent a successful surgery by Dr. Soltesz shortly thereafter. She has recovered well and returns to UPMC Susquehanna for follow-up care.

The Value of a Collegial Affiliation

As an affiliate of Cleveland Clinic's Heart & Vascular Institute, UPMC Susquehanna is able to provide all its cardiovascular physicians a direct pipeline to their counterparts at Cleveland Clinic for free second-opinion consultations on complex cases like this one.

Since our affiliation began in February 2014, we have called Cleveland Clinic for a second opinion on 30 to 40 patients. The service is overwhelmingly a consultation service, as only three or four of these patients ended up traveling to Cleveland Clinic for surgery as this patient did.

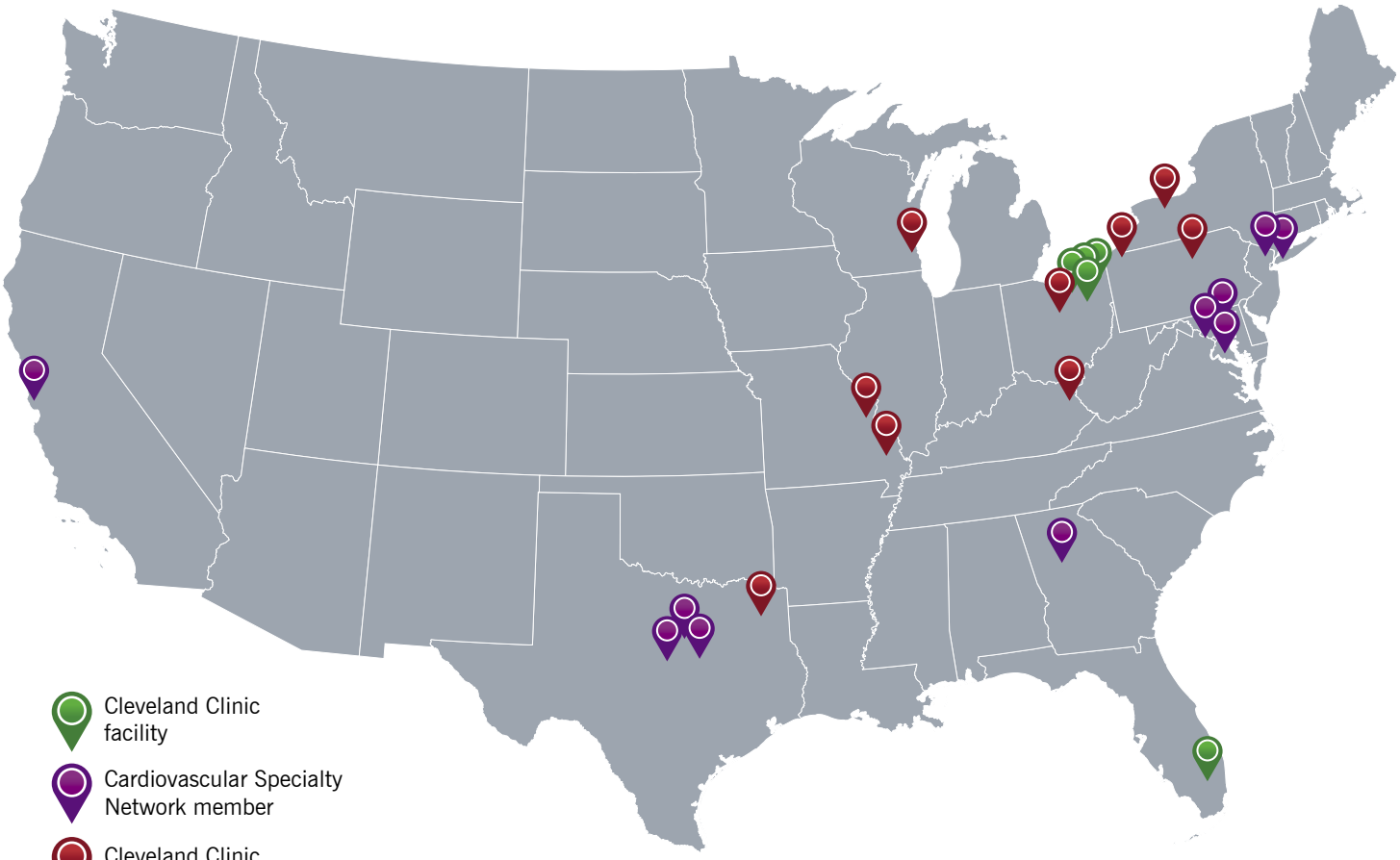
Our referring physicians tell patients that if they come to UPMC Susquehanna for a second opinion, we can actually offer them a *third* opinion from Cleveland Clinic, if necessary. If a patient does not need surgery, this additional level of expert consultation further reassures the patient that everything is being done that should be done.

As a referral hospital for complicated patients that smaller hospitals cannot handle, UPMC Susquehanna draws comfort from knowing our Cleveland Clinic colleagues are there for us when needed. The first patient for whom I sought a second opinion under this affiliation was a 73-year-old with a congenital cardiac abnormality. Cleveland Clinic cardiothoracic surgeon and Heart & Vascular Institute Chair Lars Svensson, MD, PhD, was taking consults that day and proved to be a wealth of expertise and experience. He agreed that, among the possible options, ligation of the anomaly would be a suitable approach. Surgery went well, the patient stayed local and the family was impressed by the result and the speed of service.

How It Works

Most of the cases for which we seek consultation with Cleveland Clinic involve surgical questions — e.g., whether the patient is a surgical candidate or the best surgical approach to use. Often these questions involve cutting-edge technologies present only at quaternary centers like Cleveland Clinic.

For the case patient above, I called Dr. Soltesz directly. The usual practice, however, is for my nurse to gather the patient information, call the number provided for affiliate physicians, and submit documents and images via secure electronic transfer as needed. Cleveland Clinic forwards the information for review by the appropriate subspecialist physician, who then provides direct feedback to the requesting physician



Cleveland Clinic has partnered with hospitals and health systems across the U.S. to promote improved clinical and operational aspects of cardiovascular care, including the provision of second-opinion case reviews. This map shows the locations of current affiliated provider organizations (such as UPMC Susquehanna) as well as members of Cleveland Clinic's Cardiovascular Specialty Network, a coalition of provider organizations that have entered into deeper collaborative relationships with Cleveland Clinic.

at the affiliated provider organization. This service helps us determine whether surgery is warranted, the timing of the procedure, the recommended approach and whether escalation of care is required.

Our cardiologist colleagues follow a similar process, often asking their Cleveland Clinic counterparts to read echocardiograms or assist with cath lab metrics.

At Cleveland Clinic's end, case reviews are categorized as emergency, urgent or nonurgent for appropriate triage. Emergency review allows for the physician at the affiliated provider to contact Cleveland Clinic physicians on a 24/7 basis via an internal paging system or direct phone call to the appropriate specialist. In some instances, multiple Cleveland Clinic physicians take part in a second-opinion case review, as dictated by case complexity and the requirement for subspecialty collaboration.

Our discharge data across the board confirm that our affiliate relationship with Cleveland Clinic — including these second-opinion case reviews — enhances the quality of care we are able to provide our patients. We look forward to continued collaboration. ■

SECOND-OPINION CASE REVIEW: Numbers of Note

125	Second-opinion case reviews in 2015 (77 surgical, 48 medical)
149	Second-opinion case reviews in 2016 (84 surgical, 65 medical)

Dr. Lazar is a cardiothoracic surgeon and medical director of the Heart & Vascular Institute at UPMC Susquehanna, an affiliate of Cleveland Clinic's Miller Family Heart & Vascular Institute.

For more on cardiovascular affiliation opportunities with Cleveland Clinic, visit affiliatenetwork.clevelandclinic.org.

RESEARCH Roundup

Quick Takes on Studies Presented at Recent Cardiovascular Meetings

Upending Assumptions on NSAIDs and the Heart

Despite fears to the contrary, the sole remaining selective COX-2 inhibitor in the U.S., celecoxib, confers no excess cardiovascular (CV) risk compared with two widely used nonselective NSAIDs. In fact, celecoxib is associated with fewer CV adverse events than both naproxen and ibuprofen and appears to have lower rates of GI and renal adverse events and all-cause mortality. So reported investigators with the 24,000-patient PRECISION trial at the American Heart Association's (AHA) Scientific Sessions 2016.

"PRECISION shows unequivocally that the CV risk of celecoxib was not greater than the risk of the older nonselective NSAIDs naproxen and ibuprofen," says Cleveland Clinic's Steven Nissen, MD, who chaired the trial. "In fact, celecoxib was arguably the safest of the three NSAIDs studied." He adds that while the trial has broad implications, clinicians must provide patients with proper context: "We included only individuals taking chronic daily prescription-strength NSAIDs for at least 18 months; we didn't study occasional use of over-the-counter NSAIDs." For more, see **consultqd.clevelandclinic.org/precisiontrial**.

First IVUS Study of a PCSK9 Agent Boosts Hopes for Outcomes Trial

The PCSK9 inhibitor evolocumab produced unequivocally greater atheroma reductions on intravascular ultrasound (IVUS) compared with placebo in coronary artery disease patients receiving optimal background statin therapy. That's the word from the 968-patient randomized GLAGOV trial — the first IVUS study of a PCSK9 agent — as presented by study chair Steven Nissen, MD, at the AHA Scientific Sessions 2016.

After 18 months of therapy in the multicenter trial, mean LDL cholesterol was 93.0 mg/dL in patients receiving statin+placebo versus 36.6 mg/dL in those receiving statin+evolocumab. "No one's ever reached levels that low in a clinical trial," notes Dr. Nissen. The LDL levels were associated with a reduction in percent atheroma volume for evolocumab (−0.95 percent) but not placebo (+0.05 percent) and a greater share of patients showing plaque regression in the evolocumab group (64.3 vs. 47.3 percent). "While we await definitive outcome trials of evolocumab, this is strong evidence that reducing LDL cholesterol to unprecedentedly low levels can reverse disease progression," says Dr. Nissen. For more, see **consultqd.clevelandclinic.org/glagov**.

MILANO-PILOT Dashes Hopes for ApoA-1 Milano

A therapy once touted for its promise in treating coronary disease by targeting HDL cholesterol seems to have met its demise, based on results of the MILANO-PILOT study presented at the AHA Scientific Sessions 2016. The agent, known as MDCO-216, contains ApoA-1 Milano, a variant of the ApoA-1 HDL lipoprotein discovered among residents of a village in Italy. ApoA-1 Milano carriers had levels of coronary atherosclerosis far lower than would be expected based on their very low levels of HDL cholesterol and high triglycerides.

Initial studies of MDCO-216 were promising, prompting the randomized international MILANO-PILOT trial, which compared MDCO-216 with placebo (plus optimal medical therapy) among 126 patients with recent acute coronary syndrome. "Five weekly IV infusions of MDCO-216 had no favorable impact on plaque compared with placebo," says lead investigator Stephen Nicholls, MBBS, PhD, a consultant to the Cleveland Clinic Coordinating Center for Clinical Research, which coordinated the trial. "This is more bad news for the HDL story." For more, see **consultqd.clevelandclinic.org/milano**.

Paclitaxel-Coated Balloon Outperforms PTA in SFA Disease

The investigational Stellarex™ drug-coated balloon (DCB) is superior to standard balloon percutaneous transluminal angioplasty (PTA) in a population with complex superficial femoral artery (SFA) disease. So show one-year findings of the ILLUMENATE pivotal trial, presented by Cleveland Clinic's Sean Lyden, MD, at TCT 2016. The 43-site study randomized 300 patients with SFA disease to the paclitaxel-coated DCB or standard balloon PTA. The main efficacy end point — primary patency and freedom from clinically driven target lesion revascularization at 12 months — was met by significantly more DCB recipients (76.3 percent) than PTA recipients (57.6 percent). The primary safety end point — freedom from device/procedure-related death at 30 days and from target limb amputation and clinically driven target lesion revascularization at 12 months — was likewise met by significantly more patients in the DCB group (92.1 vs. 83.2 percent).

The study is notable for its large share of patients (43 percent) with severe calcification, says Dr. Lyden: "This low-dose DCB has shown superiority over standard balloon angioplasty in one of the most complex patient groups studied in a DCB device trial." For more, see **consultqd.clevelandclinic.org/illumenate**.

RESOURCES FOR PHYSICIANS

Stay Connected with Cleveland Clinic's Heart & Vascular Institute

Consult QD — Heart & Vascular

Online insights and perspectives from Cleveland Clinic experts:

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C. Martin Harris, MD

About Cleveland Clinic

Cleveland Clinic is an integrated healthcare delivery system with local, national and international reach. At Cleveland Clinic, more than 3,400 physicians and researchers represent 120 medical specialties and subspecialties. We are a main campus, more than 150 northern Ohio outpatient locations (including 18 full-service family health centers and three health and wellness centers), Cleveland Clinic Florida, Cleveland Clinic Lou Ruvo Center for Brain Health in Las Vegas, Cleveland Clinic Canada, Sheikh Khalifa Medical City and Cleveland Clinic Abu Dhabi. In 2016, Cleveland Clinic was ranked the No. 2 hospital in America in *U.S. News & World Report's* "Best Hospitals" survey. The survey ranks Cleveland Clinic among the nation's top 10 hospitals in 13 specialty areas, and the top hospital in heart care for the 22nd consecutive year.

Cleveland Clinic

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Cardiac Consult

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events.medtelligence.net/CV17.html

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Sat.-Mon., March 4-6, 2017
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Information/registration:
ccfcme.org/echo17

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Information/registration:
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These activities have been approved for *AMA PRA Category 1 credit™*.

NON-CME EVENT

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Thu.-Fri., May 18-19, 2017
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