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## Hybrid Procedures in the Treatment of Congenital Heart Disease



Donald J. Hagler, MD, Frank Cetta, MD, Harold M. Burkhart, MD

The surgical and medical treatment of congenital heart disease has made remarkable progress over the past 50 years. Corrective surgery for many intracardiac defects began at Mayo Clinic and the University of Minnesota in the 1950s, with the introduction of the first cardiopulmonary bypass machines. With the use of this technology, delicate intracardiac surgical procedures could be performed, even on small infants. Over the subsequent 5 decades, more complex surgical procedures have been performed in younger and smaller infants. Currently, even low-birth-weight neonates benefit from successful complex intracardiac procedures.

In the 1980s and 1990s, catheter-based techniques evolved so that many "simple" congenital cardiac lesions could be successfully addressed with intravascular procedures rather than through surgical incisions. This approach obviates the use of cardiopulmonary bypass with its inherent risks. Examples of these procedures include transcatheter device closure of atrial septal defects (ASDs), ventricular septal defects (VSDs), and patent ductus arteriosus (PDA).

In the 2000s, the surgical and intravascular catheter techniques have been merged to perform procedures generically known as "hybrid operations." Hybrid techniques have been applied to multiple congenital heart lesions. These lesions and procedures include hypoplastic left heart syndrome (HLHS),

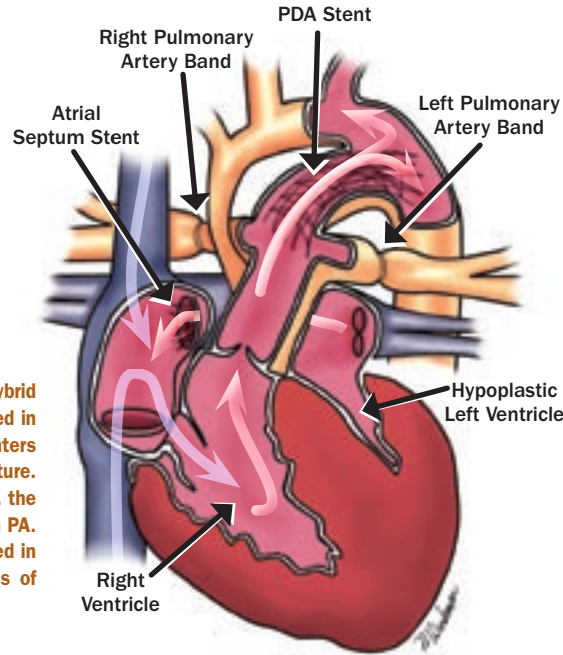
perventricular closure of VSDs, branch pulmonary artery (PA) stent placement for patients with complex right ventricular (RV) outflow tract obstruction, and relief of venous baffle stenosis in patients with transposition of the great arteries after an atrial switch operation. "In the past 5 years at Mayo Clinic in Rochester, Minnesota, the interventional congenital cardiac catheterization group has teamed with congenital cardiac surgeons to perform these procedures on a routine basis," according to Allison K. Cabalka, MD, chief of pediatric cardiac catheterization at

Mayo Clinic in Rochester.

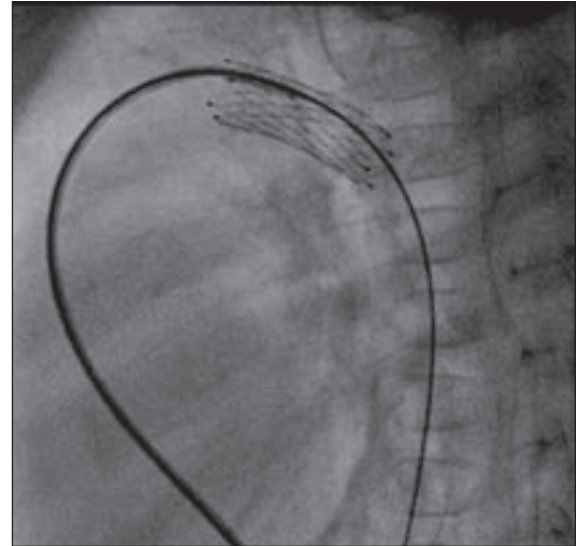
### Hybrid Procedure for HLHS

HLHS is a spectrum of disease related to hypoplasia or atresia of the left-sided cardiac structures. If untreated, this lesion is usually fatal within the first weeks of life. Neonates with HLHS have traditionally required either a Norwood operation or cardiac transplant. Although improvements have been made to the Norwood operation, the surgical management of HLHS remains challenging. Cardiac transplant in small infants is limited by donor organ availability. The Norwood operation entails reconstruction of the aortic outflow tract by using the RV and PA for systemic perfusion. Flow to the PAs is created through either a modified Blalock-Taussig shunt (systemic-to-PA connection) or a Sano shunt (RV-to-PA valveless conduit).

In 2002, a hybrid procedure for patients with HLHS was described, and it is gaining wider acceptance (Figure 1). During this procedure, the surgeon performs a pulmonary arteriotomy via a sternotomy. Cardiopulmonary bypass is not used, and the heart beats throughout the procedure. Through the arteriotomy, an interventional cardiologist inserts a delivery sheath to deploy a stent in the ductus arteriosus to ensure long-term patency (Figure 2). In this manner, antegrade flow to the descending aorta and retrograde



**Figure 1.** Elements of the hybrid procedure for HLHS. A stent is placed in the PDA via a delivery sheath that enters the PA through a purse-string suture. Once ductal patency is established, the surgeon then places a band on each PA. If needed, a stent may also be placed in the foramen ovale to ensure egress of flow from the left atrium.



**Figure 2.** Lateral fluoroscopic image of a 3-month-old infant after an HLHS hybrid procedure demonstrating the ductus arteriosus stent that ensures systemic perfusion.

flow to the aortic arch and cerebral and coronary vessels can be maintained. To restrict pulmonary blood flow, a band is placed by the surgeon on each PA (Figure 3).

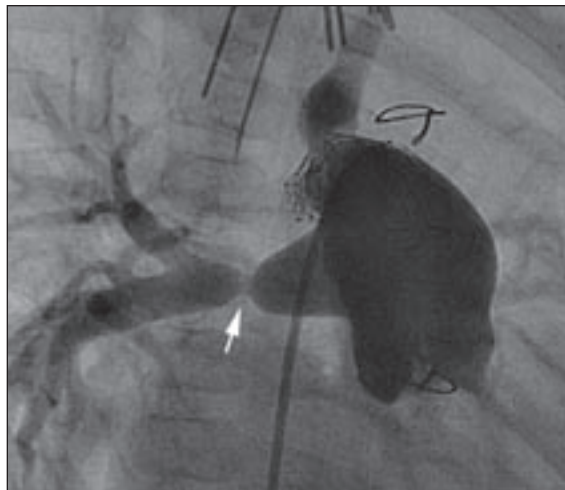
“At Mayo Clinic in Rochester, this hybrid approach is reserved for neonates with HLHS who are considered high risk for standard Norwood-Sano surgery,” says Frank Cetta, MD, chair of the division of pediatric cardiology. Preterm infants or those judged to be unsuitable for a prolonged cardiopulmonary bypass are selected for the hybrid pathway. However, some institutions now use this approach more liberally for all patients with HLHS. Long-term data regarding outcomes of the HLHS hybrid procedure are limited. “Thus far, the hybrid approach is promising,

especially because it obviates a long cardiopulmonary bypass run in a fragile neonate,” says Dr Cetta.

### Perventricular Closure of VSDs

Cardiopulmonary bypass and cardiotomy had been the standard methods for closure of muscular VSDs until the development of percutaneous muscular VSD closure devices. The effectiveness of the device and the ease of use compared to direct surgical patch closure have prompted its use in the operating room. It offers an ideal approach for closure of moderate-sized muscular VSDs in early infancy when clinical heart failure is present.

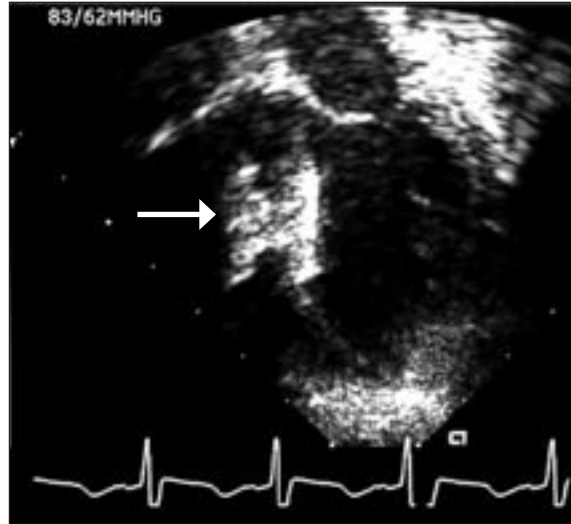
Perventricular device closure is a hybrid of operative and catheterization techniques. It is accom-



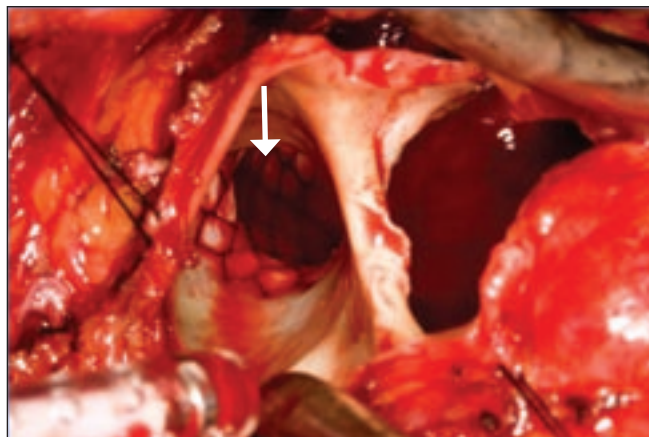
**Figure 3.** Angiogram performed retrograde via the ductal stent in a 3-month-old patient after HLHS hybrid procedure demonstrating the band (arrow) on the proximal right PA.



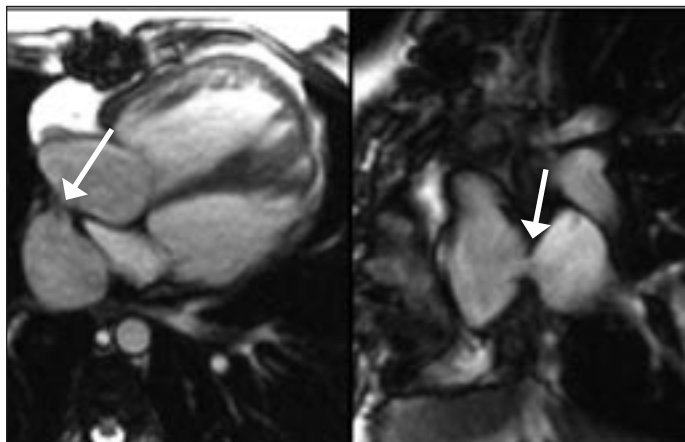
**Figure 4.** Limited sternal incision in a 6-month-old patient undergoing perventricular closure of a mid-muscular VSD.



**Figure 5.** Two-dimensional apical 4-chamber echocardiograph in a 6-month-old after successful perventricular placement of a VSD closure device (arrow).



**Figure 6.** Intraoperative photograph in an adult with tetralogy of Fallot. The patient was undergoing pulmonary valve replacement, and a proximal branch PA stenosis was relieved with intraoperative placement of a stent (arrow).



**Figure 7.** MRI scan from an adult with transposition of the great arteries after a Senning operation. A severe pulmonary venous baffle obstruction is identified (arrow). This stenosis was successfully dilated with stent placement during a hybrid procedure. The mean gradient across the stenosis was reduced from 14 mm Hg to 3 mm Hg with this procedure.

plished with a limited sternal incision to expose the lower portion of the RV (Figure 4). The surgeon then punctures the RV to insert the delivery catheter, and cardiopulmonary bypass is avoided. Transesophageal echocardiography (TEE) clearly defines the VSD location and the size of the shunt. Mid-muscular defects are ideal for this approach, but even more apical defects (below the moderator band) may be closed. “Inlet VSDs, near the atrioventricular (AV) valves at the crux of the heart, generally cannot be closed with this technique because the device may interfere with AV valve function with resultant regurgitation,” says Donald J. Hagler, MD, an interventional pediatric cardiologist at Mayo Clinic in Rochester. The TEE operator assists the surgeon and interventional cardiologist by identifying a position on the anterior surface of the RV where a direct line to the muscular VSD is possible. The RV anterior surface is punctured by the surgeon, and the cardiologist works a smooth guidewire through the defect and into the LV. The delivery sheath can then be advanced over the guidewire to thread the sheath through the VSD and into the LV. Based on the 2-dimensional echocardiographic defect size, a muscular VSD occluder is selected that has a waist diameter approximately 2 mm larger than this measurement. “The TEE guides placement of the device so that the distal disk is delivered in the LV cavity away from the mitral apparatus,” says Dr Hagler. “TEE is used to determine appropriate positioning of the device disks and to ensure that there is no interference with AV valve or ventricular function” (Figure 5).

### Hybrid Approach to Branch Pulmonary Artery Stenosis

Patients with complex RV outflow tract stenosis frequently require transcatheter placement of stents in the branch pulmonary arteries. These procedures may have a long duration of radiation exposure and become technically quite complex, especially when catheters are required to navigate multiple turns. The advantage of hybrid placement of stents is that they are performed with direct visualization by the surgeon during pulmonary valve replacement or RV outflow tract reconstruction with the assisting interventional cardiologist participating in the stent deployment.

The experience at Mayo Clinic in Rochester with intraoperative PA stent placement has demonstrated safety and efficacy. Proper sizing of the PA stent is determined by a preoperative angiogram. The surgeon usually places a stitch on the proximal stent to prevent distal migration (Figure 6). This technique successfully relieves branch PA stenosis while saving the patient from excess radiation exposure.

### Hybrid Approach for Pulmonary Venous Baffle Obstruction in Atrial Switch Patients

“In the 1960s and 1970s, patients born with transposition of the great arteries underwent Senning and Mustard operations to reroute blood flow at the atrial level to relieve cyanosis,” according to Harold M. Burkhardt, MD, a cardiovascular surgeon at Mayo Clinic in Rochester. “Most of these patients have reached adulthood, but there are consequences of these surgical techniques that have resulted in long-term residual problems.” One issue is progressive pulmonary venous baffle obstruction (Figure 7). These obstructions

may be addressed in the catheterization laboratory with stent placement, but these procedures are lengthy and require multiple 180° turns of catheter systems that are prone to kinking, with unsatisfactory results.

Similar to the periventricular muscular VSD technique described above, the surgeon begins the hybrid procedure to relieve pulmonary venous baffle obstruction by making a small atriotomy via a minimal anterior thoracotomy. Cardiopulmonary bypass is not used, and the heart beats throughout the procedure. TEE guidance is used to pass an exchange wire from the native right atrium (pulmonary venous atrium) through the pulmonary venous pathway into the superior portion of the native left atrium. A stent is deployed with TEE guidance (Figure 8). Patients are typically discharged from the hospital within 48 hours after successful hybrid stent placement for pulmonary venous baffle obstruction.

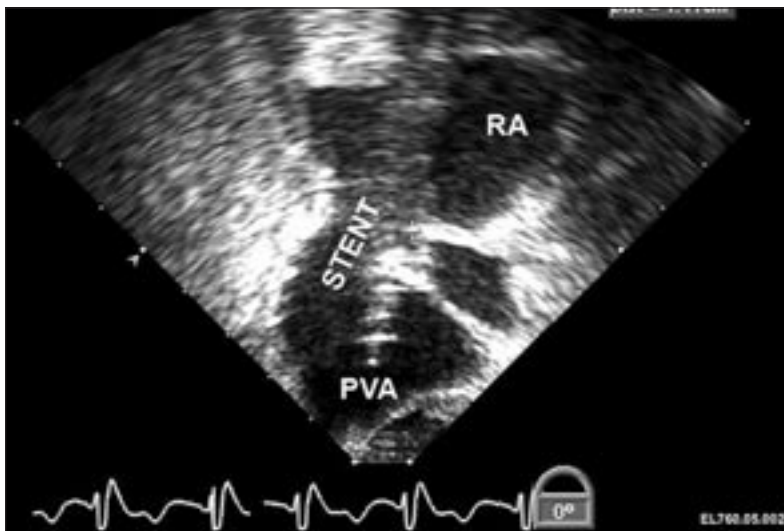


Figure 8. TEE demonstrating a stent placed via a hybrid procedure in the pulmonary venous baffle of an adult after a Senning operation. PVA, pulmonary venous atrium; RA, right atrium.

### Summary

The great advances in surgical and catheter-based techniques for patients with congenital heart disease have merged into hybrid procedures. These procedures have expanded the therapeutic options for patients with various forms of congenital heart disease. Hybrid procedures have reduced the number and duration of interventions for these patients. Many hybrid procedures are performed with the heart beating, obviating the need for cardiopulmonary bypass. The future holds great promise for expansion of collaboration between heart surgeons and interventional cardiologists in the treatment of patients with congenital disorders.

### AHA Scientific Sessions 2009: PPI's Raise Concerns

A study presented at the recent American Heart Association meeting in Orlando, Florida, suggested that patients taking the proton-pump inhibitors (PPIs) omeprazole (Prilosec) or pantoprazole (Protonix) in combination with clopidogrel (Plavix) have a higher mortality rate after percutaneous coronary intervention (PCI) than those not taking the popular anti-acid reflux drugs. The study was a retrospective review of 8,300 patients who underwent PCI between April 2003 and June 2007. Patients taking a PPI had a 30% higher risk for dying after PCI than those not taking a PPI; the highest mortality rates were found in patients taking omeprazole or pantoprazole. According to Charanjit Rihal, MD, director of the Cardiac Catheterization Laboratory at Mayo Clinic in Rochester, Minnesota, these results are not all that surprising. “Older individuals are the ones who tend to have more medical problems, including ulcers and

reflux that warrant use of PPIs,” he said. It is not clear whether these results are attributable to concomitant medical problems or to a true drug-drug interaction. “Unfortunately, the study does not indicate why patients were taking PPIs,” says Dr Rihal. He recommends that

- Patients already on PPIs for symptomatic reflux or peptic ulcer should continue these drugs.
- PPIs should be discontinued if there is no definite indication for their use.
- H2 blockers should be considered if gastrointestinal prophylaxis is indicated.

The best controlled data suggest that although in vitro interactions exist, these do not translate into a clinically significant risk. The US Food and Drug Administration is monitoring the situation closely.

## Videoscopic Denervation Therapy for Genetic Heart Diseases Predisposing to Sudden Death



Michael J. Ackerman, MD, PhD, Christopher Moir, MD

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A renaissance of an old cardiac therapy, left cardiac sympathetic denervation (LCSD), has occurred, providing another option and renewed hope for patients with malignant cardiac channelopathies, particularly long QT syndrome (LQTS) and catecholaminergic polymorphic ventricular tachycardia (CPVT). Together, these potentially lethal, highly treatable genetic heart rhythm disorders affect approximately 1 in 2,000 persons. The majority can be effectively managed with only pharmacotherapy, mainly  $\beta$ -blockers, and simple preventive measures such as avoiding exposure to medications with an unwanted QT-prolonging adverse effect. For some, however, these diseases are potently expressive, and if the patient survives sudden cardiac arrest (SCA) or experiences a breakthrough cardiac event while on pharmacotherapy, an implantable cardioverter-defibrillator (ICD) represents a life-saving and life-prolonging intervention. Unfortunately, ICD placement has become an inappropriately rapid reflex in the management of LQTS. Fortunately, the latest treatment advance with videoscopic denervation therapy has juxtaposed itself between pharmacotherapy and device therapy, where it may be more protective than pharmacotherapy with fewer complications than device therapy.

LCSD is not new but was first surgically performed in a patient with incapacitating angina and arrhythmias more than 90 years ago. LCSD involves the surgical resection of the thoracic sympathetic ganglia at T2, T3, and T4, and importantly, the resection of the lower half of the left stellate ganglion (T1). This unilateral sympathectomy is antifibrillatory. Unlike an ICD, which stands by to terminate a malignant arrhythmia, LCSD represents preventive therapy whereby denervation makes it much more

difficult for the LQTS/CPVT substrate to degenerate into ventricular fibrillation (VF) in the first place. LCSD was first performed nearly 40 years ago for the management of a patient with high-risk LQTS, and 5 years ago, analysis of its antifibrillatory effect in nearly 150 LQTS patients confirmed its very high success rate. Following this report, Michael J. Ackerman, MD, PhD, a pediatric cardiologist and director of the Long QT Syndrome Clinic at Mayo Clinic in Rochester, Minnesota, teamed up with pediatric surgeon Christopher Moir, MD, in an effort to develop videoscopic denervation therapy for the management of high-risk patients.

This procedural upgrade has occurred easily at Mayo Clinic in Rochester because of its unique combination of one of the largest LQTS specialty centers in the world, its extensive surgical experience with video-assisted thoracoscopic surgery (VATS), and the integrated teamwork between Drs Ackerman, Moir, and a team of anesthesiologists. According to Dr Moir, VATS-LCSD represents that natural next step in the maturation of denervation therapy. “In June 2009, we reported the perioperative and short-term outcomes associated with our first 20 VATS-LCSD procedures, and to date, we have performed more videoscopic denervations (more than 35) for the management of patients with LQTS/CPVT than any center in North America,” he says. Like the experience regarding traditional surgical approaches to LCSD, the videoscopic approach has taken patients at extreme risk of future episodes of SCA and lowered their risk quite dramatically. A case in point is one of Mayo’s first LCSDs, which was performed in a young woman with highly symptomatic, type 2 LQTS. She experienced approximately 15 VF-terminating ICD shocks

### Table. Considerations for Videoscopic Denervation Therapy for Patients With LQTS/CPVT

- History of appropriate VF-terminating ICD shocks
- Treatment intolerance or breakthroughs
- High-risk infants and children where treatment not deemed “protective enough” but ICD-related morbidity seems excessive (“bridge to ICD”)

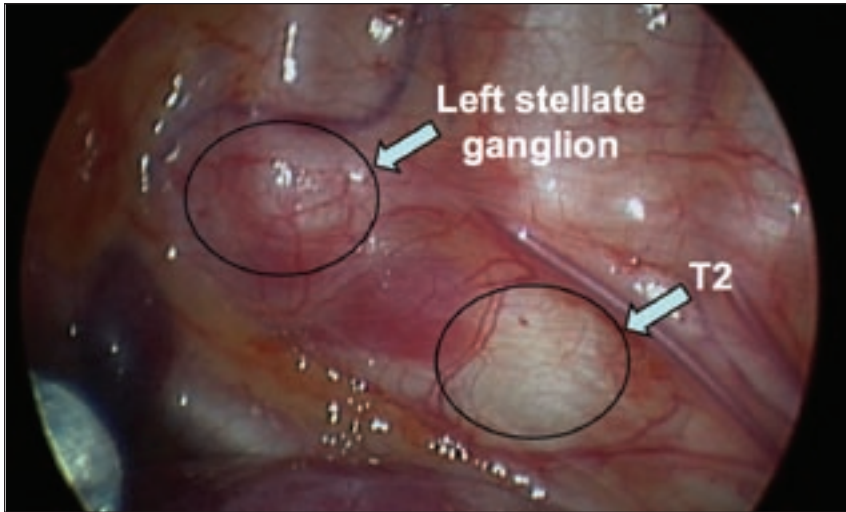


Figure. Videoscopic view of the left stellate ganglion and the thoracic sympathetic ganglion at T2.

during the year before LCSD. Now, nearly 4 years later, she has received only a single ICD shock, and in her own words, she has “gotten her life back.” Significant reduction in risk has been realized. To date, none of the VATS-LCSD has required conversion to an open surgical approach. Perhaps secondary to the outstanding anatomic exposure of the sympathetic chain in general and the left stellate ganglion specifically that is provided by the videoscopic approach

(Figure), complications such as ptosis of the eyelid or the Horner syndrome have not been observed.

Approximately half the patients referred for videoscopic denervation therapy have represented extreme phenotypes of LQTS/CPVT. Here, VATS-LCSD has been performed as “secondary prevention” because of either recurrent VF-terminating ICD therapies or recurrent cardiac events despite adequate pharmacotherapy. The other half have involved cases as so-called primary prevention in which videoscopic denervation therapy was provided to patients with  $\beta$ -blocker intolerance or for patients assessed to need more protection than just pharmacotherapy. The Table summarizes the current clinical indications for videoscopic denervation therapy at Mayo Clinic in Rochester. “LCSD’s antifibrillatory effect also appears to be relatively substrate-independent, as patients with each of the 3 major LQTS genotypes and patients with CPVT have undergone denervation with both perioperative and short-term success,” says Dr Ackerman. However, LCSD must not be viewed as curative. Recurrent events have been observed following denervation. Instead, videoscopic denervation therapy should be viewed as a minimally invasive approach to LCSD from which most patients are dismissed within 48 hours after surgery, complications are negligible, and risk of SCA is reduced significantly but not eliminated completely. In this context, a patient with LQTS/CPVT must undergo careful risk assessment to determine the right therapy for the right patient to be provided at the right time. More than 1,000 patients have been evaluated in the past decade in the LQTS Clinic at Mayo Clinic in Rochester.

In addition to its clearly established therapeutic role in LQTS/CPVT, Dr Ackerman and Samuel Asirvatham, MD, an electrophysiologist at Mayo Clinic in Rochester have joined a multicenter study to evaluate the role of LCSD for more common heart diseases associated with fatal ventricular arrhythmias. The study, PREVENT-VT, will randomly assign high-risk patients with ischemic heart disease to standard clinical therapy (ICD) or to ICD plus LCSD. PREVENT-VT will likely begin patient enrollment in summer 2010. If LCSD’s antifibrillatory effect is demonstrated among patients with ischemic heart disease in a similar manner to its protective role in LQTS/CPVT, then the number of patients who might benefit from this therapy will increase markedly. It is possible that any patient with a history of VF-terminating ICD therapies may benefit from videoscopic denervation therapy regardless of the particular cardiac pathology that has predisposed the heart to potentially lethal ventricular arrhythmias.

## RECOGNITION



Cary W. Akins, MD, clinical professor of surgery at Harvard Medical School, was the 2009 John W. Kirklin Visiting Professor at Mayo Clinic in Rochester. Dr Akins (left) is pictured with Hartzell V. Schaff, MD, chairman of the Division of Cardiovascular Surgery at Mayo Clinic in Rochester.

## Mayo Clinic Establishes Collaboration With Samsung Medical Center in South Korea



Figure 1. Samsung Medical Center.



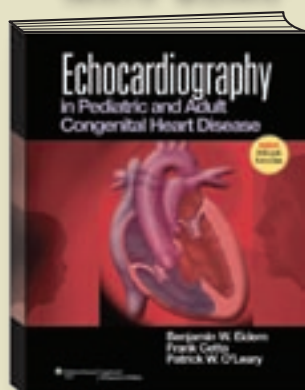
Figure 2. Dr Oh (front left) reviewing a cardiac imaging study with collaborators at Samsung Medical Center in Seoul, South Korea.

Mayo Clinic in Rochester, Minnesota, celebrated its collaborative arrangement with Samsung Medical Center in Seoul, South Korea, at the grand opening ceremony of the Samsung Cardiovascular Imaging Center in Collaboration With Mayo Clinic on September 5, 2009. The collaboration will continue Mayo Clinic's mission of patient-oriented medical practice, research, and education worldwide. Samsung Medical Center is one of the largest medical centers in South Korea and is well known throughout Southeast Asia for its top ratings for quality of care and patient satisfaction.

Jae K. Oh, MD, a cardiologist at Mayo Clinic in Rochester, is the codirector of the Samsung Cardiovascular Center. He has directed the renovation and expansion of clinical and research areas as well as the training and integration of personnel. The center integrates the disciplines of cardiology, radiology, and nuclear medicine to provide comprehensive anatomic and functional cardiac evaluations. Additional facilities include suites for interventional and surgical procedures and an imaging analysis laboratory. Virtual consultation is conducted using satellite video-conferencing facilities.

Cardiovascular researchers from Mayo Clinic in Rochester and Samsung Medical Center have jointly written Clinical Research Development Project grant applications for the study of heart failure imaging. The first Mayo Clinic-Samsung Cardiovascular Symposium took place on December 5, 2009, in Seoul, with participants from both institutions.

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Mar 7-11, 2010, Vail, CO

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Echocardiographic Symposium at Vail: New Technologies, Live Scanning, and Clinical Decision Making  
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Mayo Echocardiography Review Course for Boards and Recertification  
Aug 21-24, 2010, Rochester, MN

Electrophysiology Review for Boards and Recertification  
Aug 25-28, 2010, Rochester MN

Pediatric Cardiology 2010 Board Review Course  
Aug 29-Sep 3, 2010, Dana Point, CA

Mayo Cardiovascular Review Course for Cardiology Boards and Recertification  
Sep 25-30, 2010, Rochester, MN

Mayo Interventional Cardiology Board Review  
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Coronary Artery Disease: Prevention, Detection and Treatment  
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International Stroke Conference  
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ACC.10: 59th Annual Scientific Session  
Mar 14-16, 2010, Atlanta, GA  
Web: [ACC10.acc.org](http://ACC10.acc.org)

The Heart Rhythm Society's 31st Annual Scientific Sessions  
May 12-15, 2010, Denver, CO  
Web: [www.hrsonline.org/Sessions/](http://www.hrsonline.org/Sessions/)

ASE 21st Annual Scientific Sessions: Practical Applications of Cardiovascular Ultrasound in the 21st Century  
Jun 12-15, 2010, San Diego, CA  
Phone: 919-297-7157; e-mail: [abuff@asecho.org](mailto:abuff@asecho.org)

## RECOGNITION



Rick A. Nishimura, MD, has been named a Master of the American College of Physicians and will be inducted next spring at Internal Medicine 2010 in Toronto.



David R. Holmes Jr, MD, has been elected vice president of the American College of Cardiology. Rick A. Nishimura, MD, and Carole A. Warnes, MD, are on the American College of Cardiology Board of Trustees.

### Mayo Clinic Cardiovascular Update

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