

## Emory Adult Congenital Heart Center Team

### ADULT CONGENITAL CARDIOLOGISTS

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**Fred H. (Rusty) Rodriguez III, MD**, Assistant Professor of Medicine and  
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### ADULT CARDIOTHORACIC SURGEONS

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Emory Adult Congenital Heart Center; Chief of Pediatric Cardiac Surgery

**Bahaaldin Alsoufi, MD**, Associate Professor of Surgery

### STRUCTURAL INTERVENTIONISTS

**Vasilis Babaliaros, MD**, Associate Professor of Medicine

**Dennis Kim, MD, PhD**, Associate Professor of Pediatrics

### CONGENITAL ELECTROPHYSIOLOGY SPECIALIST

**Michael Lloyd, MD**, Associate Professor of Medicine

### ADDITIONAL PROVIDERS

**Christine Peverini, MSN, ARNP-C**, Nurse Practitioner

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**Alicia Rangosch, RDCS**, Cardiac Sonographer

**Stacy Hedges, RDCS**, Cardiac Sonographer

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## Now seeing patients at Emory Saint Joseph's Hospital.

In addition to our primary location on the Emory University campus,  
the Emory Adult Congenital Heart Center also sees patients at the  
Emory Saint Joseph's Hospital campus:

Emory Adult Congenital Heart Center at Emory Saint Joseph's Hospital  
5671 Peachtree Dunwoody Rd, NE • Suite 300  
Atlanta, GA 30342

Emory Heart & Vascular Center Transfer Service: **404-778-4930** or **800-43-HEART**

Emory Physician Consult Line: **404-778-5050** or **800-22-EMORY**

Emory HealthConnection<sup>SM</sup>: **404-778-7777** or **800-75-EMORY** (Patients)

[emoryhealthcare.org/heart](http://emoryhealthcare.org/heart)

# The Chamber EMORY

Heart & Vascular Innovations at Emory  
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HEALTHCARE

## Managing Late Effects of Fontan Palliation in Adult Patients: A Comprehensive Team Approach

by Camden Hebson, MD, Assistant Professor of Medicine

An 18-year-old man with a known history of tricuspid atresia status post palliative surgeries in childhood recently presented at a south Georgia hospital with acute-onset palpitations and tachycardia. Upon further evaluation, his physicians learned that the patient was also experiencing shortness of breath with activity and had other rhythm issues, including a history of previous cardioversion for atrial flutter in 2012 and frequent shorter episodes of palpitations. He was diagnosed with atrial flutter, cardioverted and started on sotalol. He had previously been under the care of a pediatric cardiologist, but due to his age and the onset of additional complications, he was referred to the Emory Adult Congenital Heart Center.

At his initial appointment at Emory, the patient met with Maan Jokhadar, MD. Dr. Jokhadar confirmed the diagnosis of tricuspid atresia and determined that the defect had been treated in childhood with an atriopulmonary Fontan procedure, a technique that has since fallen out of favor due to an increased risk for late complications.

*continued inside*



# Emory Adult Congenital Heart Center

The Emory Adult Congenital Heart Center is an internationally recognized cardiology service that specializes in the care of adults with congenital heart defects (CHDs). The Center is the only adult congenital heart disease program in the state of Georgia and one of the largest programs in the country.

The Center employs a multidisciplinary approach to provide comprehensive management services for adults with CHDs. Our team includes both adult and pediatric cardiologists, congenital cardiac surgeons, electrophysiologists, interventional cardiologists, nurse practitioners, nurses, social workers and echocardiographers. We also work with collaborative partners from a broad range of specialties throughout Emory Healthcare who have particular expertise in managing adults with CHDs.

Our ultimate goal is to help patients manage their conditions and live active, healthy lives.

To refer a patient, call **404-778-5545** and speak with one of our administrative assistants. You can also learn more about the Emory Adult Congenital Heart Center by visiting us online at [emoryhealthcare.org/congenital-heart](http://emoryhealthcare.org/congenital-heart).

## References

1. Lasa JJ, Glatz AC, Daga A, Shah M. Prevalence of arrhythmias late after the Fontan operation. *Am J Cardiol.* 2014;113(7):1184-1188.
2. Hebson CL, McCabe NM, Elder RW, et al. Hemodynamic phenotype of the failing Fontan in an adult population. *Am J Cardiol.* 2013;112(12):1943-1947.

## Fontan Palliation continued

On physical examination, the patient's cardiac rhythm was irregular and an electrocardiogram (EKG) demonstrated ventricular bigeminy. An echocardiogram showed a massively dilated right atrium, sluggish flow through the Fontan circuit and a left ventricular (LV) ejection fraction of 35% to 40%. Further workup included cardiac catheterization and cardiac magnetic resonance imaging (MRI), which revealed considerable enlargement of the right atrium (Figure 1).

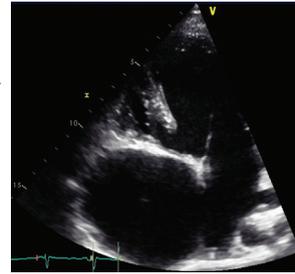


Figure 1. Pre-operative echocardiographic image, apical four-chamber view.

## Types of Fontan Palliation

In an atriopulmonary Fontan procedure, the right atrial appendage is anastomosed directly to the right or main pulmonary artery, which then directs blood from the superior and inferior vena cavae directly to the lungs without the use of a ventricular "pump." If an atrial septal defect is present, it is closed. Given the distensibility of the right atrium, a unique long-term problem is significant atrial dilation, which, along with atrial suture lines, predisposes the patient to arrhythmias. By comparison, an extracardiac Fontan procedure uses an extracardiac conduit, usually made of synthetic material, to anastomose the inferior vena cava directly to the underside of the right pulmonary artery.

In modern cardiothoracic surgery, atriopulmonary Fontans are no longer performed. However, many adults underwent this operation decades ago, so knowledge of the anatomy remains pertinent.

## Common Issues Following a Fontan Procedure

The Fontan procedure is not a complete repair, but instead a palliation with known complications that arise with time.

- Due to elevated post-hepatic pressures and relative stasis of blood flow as it passes from the liver into the Fontan circuit, fibrosis and cirrhosis develop almost universally.
- Patients can develop protein-losing enteropathy that leads to hypoalbuminemia, malnutrition, edema and an increased risk of infections due to weakening of the humoral immune response.

- Within a decade of the procedure, greater than 20% of patients will develop atrial arrhythmias; this number rises to 50% in those who undergo atriopulmonary Fontan.<sup>1</sup>
- Because patients rely on a systemic right ventricle following Fontan palliation, reduction in ventricular function is a particular concern.
- Patients can develop Fontan circulatory failure, with symptoms of dyspnea, fatigue and edema. Similar to portal hypertension, systemic vascular resistance drops, but due to the single-ventricle anatomy, cardiac output cannot adequately compensate. The result is a hemodynamic vice and spiraling symptoms.<sup>2</sup>

## A Successful Outcome

Dr. Jokhadar initiated a multifaceted treatment strategy aimed at all of the presenting problems. He started the patient on warfarin due to his history of recurrent atrial flutter and sluggish flow in the Fontan circuit, initiated lisinopril and furosemide for his ventricular dysfunction and eventually replaced sotalol with carvedilol. He referred the patient to Emory electrophysiologist Michael Lloyd, MD, who was able to successfully ablate the focus of the premature ventricular contractions, and consulted with Hepatology to obtain imaging to monitor for Fontan-related liver disease.

Finally, after the patient's ventricular function normalized with medical therapy, Dr. Jokhadar proposed a Fontan revision and discussed the case at medical/surgical case conference. There was consensus in favor of the procedure, and a few months after initial presentation, Emory congenital surgeon Bahaa Alsoufi, MD, performed a Fontan revision consisting of conversion to an extracardiac Fontan, right atrial reduction, right-sided Maze cryoablation and placement of a dual-chamber pacemaker. The surgery was successful and the postoperative course was uneventful.

At his most recent follow-up, the patient was doing quite well. His dyspnea and energy level had both improved and he was no longer having atrial or ventricular arrhythmias. Echocardiography demonstrated that his right atrium had returned to a normal size and that his LV function remained normal (Figure 2). His total bilirubin level had decreased to 1.7 mg/dL from 2.4 mg/dL at presentation. A liver ultrasound showed no significant fibrosis, and Hepatology is now following the patient regularly.



Figure 2. Post-operative echocardiographic image, apical four-chamber view.