Congenital Heart Center of Georgia Leadership Team

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Children's Healthcare of Atlanta

EMORY HEART & VASCULAR CENTER

Emory Healthcare
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Decatur, Georgia 30030

Research

The Emory+Children's Pediatric Research Center is one of only three sites in the country awarded the Centers for Disease Control and Prevention grant, Surveillance of Congenital Heart Disease (CHD), Focusing on Adolescents and Adults. The associated with this grant will provide the most comprehensive snapshot of the care and needs of those with CHD.

Referrals*

Emory Adult Congenital Heart Center
Call the Emory Physician Consult Line at 404-778-5050 or 800-22-EMORY.

Children's Sibley Heart Center
Call 800-542-2233 or 404-256-2593 to refer a patient.

** Patients 21 years old and older should schedule appointments at the Emory Adult Congenital Heart Center and patients under 21 should schedule at Children's Sibley Heart Center.*

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The Chamber

Heart & Vascular Innovations
Summer 2014

Children's Healthcare of Atlanta
EMORY HEART & VASCULAR CENTER

Complications of Repaired Congenital Heart Defects in Adults

By Wendy M. Book, MD, Fred H. Rodriguez, III, MD, and Laura E. Thomason, MD

Congenital heart disease (CHD) complicates approximately 1% of live births in the general population and affects about 40,000 births each year in the United States. Though many of these cardiac malformations were once incompatible with life, medical and surgical advances now help more than 90% of affected children reach adulthood.[1] Between 650,000 and 1.3 million adults are currently living with CHD.[2] The majority of CHD repairs are palliative rather than curative, and additional interventions may be needed later in life.

Patient Presentation

A 41-year-old woman with a history of CHD sought medical attention for back pain. Her cardiac history consisted of a perimembranous ventricular septal defect (VSD) repaired in 1971, with re-repair two weeks later due to patch dehiscence. She was also born with coarctation of her aorta, which was diagnosed when she was a teenager and repaired with a C-graft from the left subclavian artery to the descending aorta in 1985.

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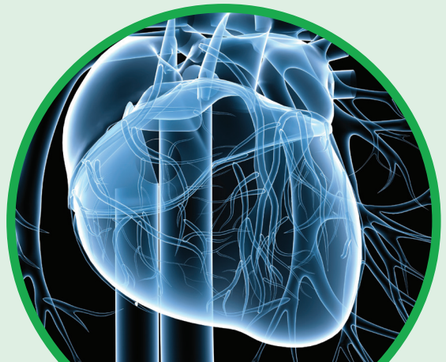

For the next 25 years, she received inconsistent cardiology follow-up and no imaging of the coarctation repair. She had a known residual VSD, described as left ventricle (LV) to right ventricle (RV), for which no intervention had previously been recommended. Due to an incidental finding on her lumbar spine during magnetic resonance imaging (MRI), an abdominal computed tomography (CT) scan was performed that demonstrated a cirrhotic nodular morphology of the liver and features of portal hypertension. The patient had no history of alcohol abuse, and testing for viral hepatitis was negative. Liver biopsy was consistent with congestive hepatopathy and stage 3 fibrosis, findings suggestive of right heart failure. The patient was referred to the Emory Adult Congenital Heart Center for further evaluation.

The patient's physical examination was remarkable for elevated jugular venous pressure and hepatomegaly, findings consistent with increased right atrial (RA) pressure. Her S2 was normal, and there was no RV heave. Her echocardiogram demonstrated marked dilation of the RA, but not the RV, and a residual VSD with shunting from the LV to the RA. No features of pulmonary hypertension were noted. Cardiac MRI/MRA (magnetic resonance arteriography) revealed a 5.1-cm aneurysmal dilatation at the insertion site of the subclavian graft into the descending aorta and residual coarctation, with narrowing of the native aorta to 12 mm. Cardiac catheterization showed an RA pressure of 17/18 (14) mmHg, normal pulmonary artery pressure and pulmonary capillary wedge pressure, and a pulmonary-to-systemic blood flow ratio (Qp:Qs) of 2:1.

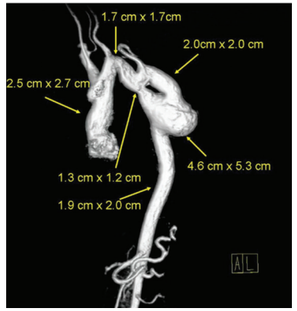

Figure 1. Pre-operative MRI of aortic anatomy.

Following review of her imaging in our multi-disciplinary conference, the patient was referred for staged repair. Endovascular exclusion of the aneurysmal segment was followed by surgical VSD closure and ascending-to-descending aortic bypass. The patient tolerated the procedures with no complications.

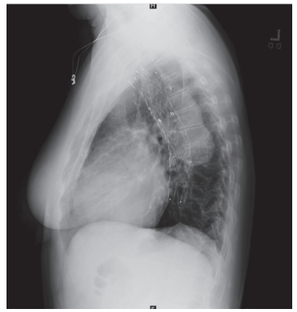

Figure 2. Pre-operative chest X-ray with aortic aneurysm visible posterior to the coarctation repair graft.

This case illustrates the importance of ongoing specialty adult CHD care for surveillance of late complications from childhood repairs. While this patient's residual VSD was known, the degree of left-to-right shunting was underestimated and the shunt location misjudged, eventually leading to end-organ dysfunction (cirrhosis). Previous case reports have described the incidental discovery of late aneurysm formation in patients with repaired coarctation.[3] However, this case illustrates two uncommon late complications of CHD surgical repairs: aneurysm formation following coarctation repair and right heart failure as a result of residual shunting around a repaired VSD. For patients with repaired coarctation, guidelines recommend lifelong cardiology follow-up, including consultation with a cardiologist with expertise in adult CHD and evaluation of the repair site by MRI or CT at least every five years.[4]

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Why Choose the Congenital Heart Center of Georgia

A collaboration between the **Children's Healthcare of Atlanta Sibley Heart Center** and the **Emory Heart & Vascular Center**, the Congenital Heart Center of Georgia is one of the largest programs in the country – and the only one in Georgia* – specializing in the treatment of children and adults with congenital heart disease (CHD). Led by Robert Campbell, MD, Brian Kogon, MD, and Wendy Book, MD, the comprehensive center unites physicians from Children's Healthcare of Atlanta and Emory to coordinate the transition from pediatric to adult care. Together, we help individuals with congenital heart defects receive appropriate lifelong care.

* *Congenital Heart Center of Georgia is not a legal entity, and instead is a marketing name used by the separate companies Children's Healthcare of Atlanta, Inc. ("Children's") and Emory Healthcare, Inc. ("Emory"). Children's and Emory are individual and separate legal entities and are each independently and financially responsible for the goods and services that each provides. Children's Healthcare of Atlanta Sibley Heart Center is a service of Children's.*

References

1. Child JS, Aboulhosn J. Chapter 236. Congenital Heart Disease in the Adult. In: Longo DL, Fauci AS, Kasper DL, Hauser SL, Jameson JL, Loscalzo J, eds.Harrison's Principles of Internal Medicine. 18th ed. New York: McGraw-Hill; 2012.
2. Go AS, Mozaffarian D, Roger VL, et al. Heart disease and stroke statistics – 2013 update: a report from the American Heart Association. Circulation. Jan 1 2013;127(1):e6-e245.
3. Horlick EM, McLaughlin PR, Benson LN. The Adult with Repaired Coarctation of the Aorta. Curr Cardiol Rep. 2007 Jul;9(4):323-30.
4. Warnes CA, Williams RG, Bashore TM, et al. American College of Cardiology/American Heart Association (ACC/AHA) 2008 Guidelines for the Management of Adults with Congenital Heart Disease: a report of the ACC/AHA Task Force on Practice Guidelines (writing committee to develop guidelines on the management of adults with congenital heart disease). Circulation. 2008 Dec 2;118(23).