MEET STEVEN BIBEVSKI, MD, PHD

Dr. Steven Bibevski is a pediatric and congenital cardiac surgeon with expertise in complex congenital heart defect repair.

A native of Melbourne, Australia, Dr. Bibevski completed fellowships in both adult and pediatric cardiac surgery at the University of Michigan, one of the busiest congenital heart programs in the United States. He also completed a visiting fellowship at the Royal Children’s Hospital in Melbourne, the southern hemisphere’s largest pediatric heart center.

Both pediatric fellowships offered him experience in extracorporeal life support, post-operative intensive care unit management and heart transplantation, as well as all aspects of managing patients with adult congenital heart disease.

Dr. Bibevski’s specific clinical interests are in single ventricle physiology, transposition of the great vessels and atrio-ventricular canal defects, and adult congenital re-operative surgery. He also has a strong interest in reducing the need for repeat operations by utilizing innovative conduits and preserving native tissue and valves where possible.

Dr. Bibevski’s research history includes a PhD in cardiovascular physiology from Case Western Reserve University in Cleveland. He currently serves as the lead investigator in an international clinical study of the autonomic nervous system in infants and children with congenital disease.

The goal of the Adult Congenital Heart Defects (ACHD) program at Memorial Regional Hospital and Joe DiMaggio Children’s Hospital is to serve referring physicians of patients with complex congenital cardiac abnormalities.

These patients often have anatomic and physiologic abnormalities that are very different from the general population. Providing them with the best care requires knowledge about their histories, as well as the predictable cardiovascular problems that they may encounter as adults. By offering intermittent monitoring and guidance, the program is an excellent resource for congenital cardiac patients and their physicians.

At Memorial Regional Hospital, we work with hospitalists, maternal-fetal specialists and a full range of subspecialty physicians for inpatient care. We strive to ensure that patients continue to see their primary physicians for all ongoing care between the evaluations and treatments that we provide.

In this and upcoming issues of Adult Congenital Cardiology Today, we’ll briefly highlight a member of our team, include noteworthy news about the Adult Congenital Cardiology field, and present a brief case. We hope that the case presentations will serve to illustrate issues or procedures with which we routinely deal and that you’ll find interesting.

We’re always trying to improve our service – so we welcome any comments or suggestions from you, our readers, referring physicians and patients.
CASE STUDY: TYPICAL ISSUES AFTER A FONTAN OPERATION

This is a 31-year-old male who presented to the emergency room with cyanosis and peripheral edema. He knew that he had some type of congenital heart defect, but was unclear about the details.

The ACHD service was consulted. Echocardiography demonstrated that the patient had a type of single ventricle, double inlet left ventricle, and had undergone a Fontan operation. Because he was in atrial fibrillation, he was seen by Ming Young, MD, who specializes in electrophysiologic problems in congenital heart disease patients of all ages. After the patient was cardioverted, his signs of congestive heart failure gradually improved.

With a great deal of effort, we were able to obtain more details about the patient’s malformation and previous treatments. At one day old, he had undergone a Blalock-Taussig shunt because of pulmonary atresia in addition to his single ventricle. He underwent a classic Fontan operation at age 7. He did reasonably well clinically, but had remained cyanotic. Persisting cyanosis is usually caused by a leak in the Fontan pathway that allows unoxygenated systemic venous blood to enter the left atrium.

Two previous outside catheterizations had failed to identify the site of a leak, and the patient was subsisting with systemic saturations averaging about 85 percent. His exercise tolerance was definitely limited by his low oxygen saturations, but he was remarkably functional and worked in a demanding job as a hotel manager.

The patient had not recently seen anyone about his congenital heart disease because his pediatric cardiology program was unable to continue to care for him as an adult. He was thrilled to know that the congenital heart defect specialists in our group understood his condition and could provide follow-up and treatment.

After he recovered from his arrhythmia, the patient underwent an MRI by Michael McClearly, MD, and Liliam Valdez-Cruz, MD, the radiology and congenital heart imaging specialists in our group. His overall anatomy, chamber volumes and function were well-delineated, but a suspected complex leak between the trabeculations of his atrial-pulmonary connection could not be easily visualized on the MRI. He was therefore taken to the Congenital Heart Defects Catheterization Laboratory.

Detailed intraprocedural transesophageal echocardiographic and angiographic studies indicated that the leak was originating in the right atrial appendage. The tract through the trabeculations was successfully cannulated and closed with a catheter occlusion device by Larry Latson, MD.

Thanks to this treatment, the patient’s saturations increased to the low 90s, and he has had a dramatic improvement in his exercise capacity. He recently became engaged and is looking forward to his wedding later this year.

The Fontan operation is a procedure that has revolutionized the outcomes of patients who were born with a single ventricle. The procedure routes all of the systemic venous blood directly to the pulmonary arteries. Patients can get along surprisingly well without a ventriculopulmonary pump for pulmonary blood flow. This type of circulation, however, is not as good as the normal circulation, and requires high systemic venous pressures.

The Fontan operation has numerous variations that can all have unique later issues. The cyanosis and arrhythmias exhibited by this patient are common in those individuals with a classic atrio-pulmonary connections. A more modern variation utilizing either an intracardiac or extracardiac tunnel can improve the flow dynamics – and reduce the incidence of baffle leaks and, possibly, late arrhythmias. In cases of significant revision of the Fontan pathway is recommended if they are having significant difficulties.

Although the average exercise capacity of a patient following the Fontan operation tends to be in the low-normal range at best, many treated patients can live happy and productive lives. Periodic evaluations in a center specializing in adult congenital heart defects are helpful for providing anticipatory guidance and additional specialized procedures if necessary.