Dear Colleagues,

It’s been one year since we moved into our Fetal Medicine Institute and what a year it has been! We are currently on track to exceed 600 consultations this year, thanks to the extraordinary support from you, our colleagues in the community. The feedback from you and our shared patients has been remarkably positive, and we will continue to strive for an even better patient experience in the next year.

This summer issue of Fetal Connections features the Fetal Heart Program, which has built up a remarkable reputation in the community under the leadership of Mary Donofrio, MD, and Anita Krishnan, MD. The Fetal Heart Program partners with our pediatric cardiac surgery program, led by world-renowned surgeon Richard Jonas, MD, and our dedicated Cardiac Intensive Care Unit, led by John Berger, MD, to provide the greatest depth and breadth of expertise in cardiac care in the region for our unborn and newborn babies. The Quarterly Case Review on page 2 highlights the outstanding support this team provides as they shepherd the critically ill fetus to a stable postnatal life.

Finally, we are in the midst of planning an inaugural conference focused on the fetal brain to take place in Washington in the fall of 2016. More to follow – stay tuned.

Hope you all get some rest this summer!

Best wishes,

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Director, Fetal Brain Program
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Our Fetal Heart Program

The Fetal Heart Program, an integral part of both the Children’s National Heart Institute and the Fetal Medicine Institute, was established in 2004 to provide innovative specialty care to fetuses diagnosed in utero with cardiovascular abnormalities, heart defects, rhythm abnormalities, and heart failure. The primary mission of the Fetal Heart Program is to understand how heart defects affect fetal well-being in utero and determine when and if fetal intervention may be possible. The Fetal Heart Program addresses the medical needs of fetuses with heart defects, from the time of diagnosis through the transition to postnatal life, while offering emotional support and care to families as they navigate an often stressful and difficult time.

Care Model

Most often, patients are referred to the Fetal Heart Program at 18 to 24 weeks gestation after an abnormality in the fetal heart is identified during a routine or high-risk obstetrical ultrasound. A patient may also be referred if there is a strong family history of congenital heart disease (CHD) or a maternal condition increasing the risk of CHD to the fetus. In high-risk pregnancies—such as those with a strong family history of CHD, an increased nuchal translucency noted on first

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The Fetal Heart Program recently cared for a high-risk infant diagnosed with D-transposition of the great arteries (TGA). TGA is a serious congenital heart defect in which the arteries carrying oxygenated and deoxygenated blood are transposed, resulting in severe hypoxia and decreased perfusion at the time of birth. To survive, the newborn must rely on the foramen ovale to allow oxygen-rich blood to perfuse the brain and the rest of the body. In addition, the ductus arteriosus, which usually closes after birth, must remain open to allow deoxygenated blood to reach the lungs. TGA is detected prenatally in less than 50 percent of cases nationwide, despite the fact that fetal diagnosis has been shown to increase survival both before and after surgery.1 Fetal echocardiography can be used to assess these patients in utero to make the diagnosis and risk-stratify the anticipated severity of compromise at delivery.2-4

In this case, at 23 weeks gestation the ductus arteriosus was noted to be very small with abnormal flow into the pulmonary arteries. By 33 weeks gestation, the foramen ovale was noted to be severely restrictive, creating right-sided heart dilation. Aortopulmonary collaterals also were noted. Their presence was believed to be due to hypoxic blood in the lungs from the abnormal circulation.5 At 38 weeks, the foramen ovale was noted to be closed, and the left ventricle had decreased function.

Because of the findings identified on fetal echocardiogram, this fetus was risk-stratified into the LOC 4 category and was delivered urgently via Cesarean section at Children’s National. Our multi-disciplinary delivery team coordinated the stabilization of the baby from the first seconds after birth, when the baby was severely hypoxic and poorly perfused. The baby underwent a balloon atrial septostomy, stabilizing the circulation and temporarily improving oxygen levels. However, the ductus arteriosus did not open with the prostaglandin infusion, so the infant was taken to the operating room urgently for repair. The baby did well and was discharged from the hospital two weeks later.

trimester screening ultrasound, or a suspected cardiac, extracardiac, or genetic abnormality—fetal cardiac imaging can be done as early as 12 weeks gestation through the Heart Institute’s Early Fetal Echocardiography Imaging Program, housed at the Children’s National Sheikh Zayed Campus for Advancing Children’s Medicine. Using fetal echocardiography, heart defects can be diagnosed accurately and with precision. Adjunct imaging, such as fetal MRI, is sometimes recommended to enable better assessment of extracardiac structures, including the brain, lung, and abdominal structures, to identify potential multi-system abnormalities that may affect overall diagnosis and care.

Common heart defects diagnosed through fetal echocardiography include: Tetralogy of Fallot, truncus arteriosus, other complex two ventricle abnormalities, ventricular septal defects, endocardial cushion defects, tricuspid valve atresia, pulmonary stenosis or atresia, hypoplastic left heart syndrome, other complex single ventricle defects, fetal arrhythmias, and fetal heart failure. Defects that are more difficult to assess include atrial septal defects, pulmonary venous anomalies, and coarctation of the aorta.

Once a fetal diagnosis is made, Children’s National fetal specialists and pediatric medical and surgical consultants counsel parents on what to expect during the remainder of the pregnancy and on the care plan for after delivery. Parents also may have the opportunity to meet with other families who have experienced similar circumstances. In some cases, Children’s National cardiologists may recommend interventions in the fetal period, such as in utero procedures or initiation of medication. Even if a fetal intervention is not warranted, prenatal diagnosis offers the advantage of planning the details of a baby’s delivery to control the impact of the heart defect on the complex transition from fetal to postnatal circulation. Being prepared for any such hazards allows the team of specialists in our cardiac intensive care unit to minimize the risk of secondary injury to other organs, particularly the brain.

**Delivery Planning and the Critical Care Delivery Program**

Delivery planning is determined by the specific features of the CHD identified during prenatal cardiology visits. Patients are assigned a level of care (LOC) from 1 to 4, with 4 being the highest severity. The LOC system was developed by Mary Donofrio, MD, in 2004, and each LOC designation reflects evidence-based medicine and multidisciplinary expert consensus, individualized to the patient.

Most newborns are assigned as LOC 1 or 2 and can be stabilized and cared for at their local delivery hospital, even if transport to Children’s National for surgery or intervention is subsequently necessary. In these cases, the local neonatologist will stabilize the infant, often in consultation with Children’s cardiologists. For fetuses with CHD, arrangements can be made for delivery at a facility that collaborates with Children’s cardiologists so that patients can receive specialty care immediately after delivery at the delivery hospital before transport to Children’s National for quaternary care.

Assignment of a LOC 3 or 4 is rare but may be necessary to allow our subspecialists to perform a cardiac intervention on the unstable infant in the delivery room. LOC 4 patients are most often delivered by the Critical Care Delivery team in the cardiac operating room at Children’s National. These deliveries are planned to include life-saving interventions such as emergent cardiac catheterization or surgery, initiation of extracorporeal membrane oxygenation (ECMO), and/or the ex utero intrapartum treatment (EXIT) procedure. Our multidisciplinary Critical Care Delivery team has been successful in delivering LOC 4 patients, with 100 percent delivery room survival and excellent outcomes in the highest risk patients.

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Our Fetal Heart Program (cont.)

Our Critical Care Delivery Program draws upon the rich multidisciplinary resources of the Fetal Medicine Institute to provide expertise and state-of-the-art transitional care for patients with the highest risk fetal diagnoses. To learn more, read our Spring 2014 Fetal Connections at www.ChildrensNational.org/Fetal-Connections.

Fetal Heart Program Team

The Fetal Heart Program is directed by Dr. Donofrio and draws on the expertise of Anita Krishnan, MD and David Schidlow, MD, along with an expanded team of pediatric cardiologists with significant cardiac imaging experience.

By caring for the fetus as a bona fide patient, treating illness in utero when possible, and coordinating specialized delivery room care and intervention when needed, we seek to minimize the risk of compromise and injury to the baby, including injury to the brain and other organs. This goal, in addition to the ongoing research endeavors underway to better understand and manage the fetal heart and cardiovascular circulation, enables us to look towards a bright future for our patients and their families.

Fetal Connections

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Fetal Connections is written for physicians and should be used for medical education purposes only. To view past issues of Fetal Connections, visit www.ChildrensNational.org/Fetal-Connections.

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