

The staff of the Cardiac Center at The Children's Hospital of Philadelphia is making significant advances in patient care and research. We're proud of the work highlighted here because, like our colleagues across the country, we're devoted to the children to whom we provide care. We hope you find this update interesting and useful.

Contact us at cardiac@email.chop.edu or visit heart.chop.edu for more information.

Today at CHOP: Cardiac Update

To Survive and to Thrive

Single Ventricle Care and Research Program

Improving the quality and duration of life for children with surgically repaired single ventricle defects is one of the great challenges in pediatric cardiology today.

This fall, the Cardiac Center established the Single Ventricle Care and Research Program to focus on this challenge through coordinated, comprehensive clinical care and research investigations. Directed by Jack Rychik, M.D., the program is one of the first of its kind.

As patients who have had surgical repair of hypoplastic left heart syndrome, tricuspid atresia, double-outlet left ventricle and other single ventricle lesions age, they face problems such as exercise intolerance, heart rhythm disturbances, abnormal clot formation with risk of stroke, and loss of protein in the stool.

The program includes a multidisciplinary team of cardiologists, nurses, cardiothoracic surgeons, gastroenterologists, hematologists, radiologists and other specialists who provide disease-focused opinions, as well as consultative services. The team hopes to establish clinical practice standards and management protocols to share with other institutions.

Program staff will educate community healthcare providers, and help families coordinate the complex care often required of single ventricle children. A single ventricle survivorship program, modeled after pediatric cancer survivorship programs, will be established to care for and track patients for years.

Because of its large patient cohort, the Single Ventricle Care and Research Program will be able to develop a detailed database and conduct focused research. Potential investigations include: clinical trials in areas such as drug therapies and catheter interventions; collaboration with industry partners for the development of new treatments; and basic science research into the genetic origins and molecular basis of the disease and its operative sequelae.

Forty years ago, when the Fontan procedure was introduced, the survival of an infant with a single ventricle defect was considered miraculous. Today, most survive. The Single Ventricle Care and Research Program hopes to play a role in the next leap: full, healthy lives for each.

The NeuroCardiac Care Program

As survival rates for complex congenital heart defects consistently improve, clinicians are focusing increased attention on neuro-developmental outcomes in affected patients, many of whom are now entering primary and secondary school.

As a group, children with complex congenital heart disease, particularly those with conditions severe enough to require surgery in the first months of life, have a higher incidence of academic, behavioral and coordination problems than children without complex CHD.

To devote appropriate resources and develop a comprehensive approach to these issues, the Cardiac Center at CHOP has created the NeuroCardiac Care Program (NCCP). Under the direction of Gil Wernovsky, M.D., the program began seeing patients in November 2009.

Children enrolled in the NCCP receive screening, evaluation and coordination of services by CHOP specialists in cardiology, neurology, developmental pediatrics, social work, nutrition and speech, and physical and occupational therapy. This interdisciplinary approach fosters a

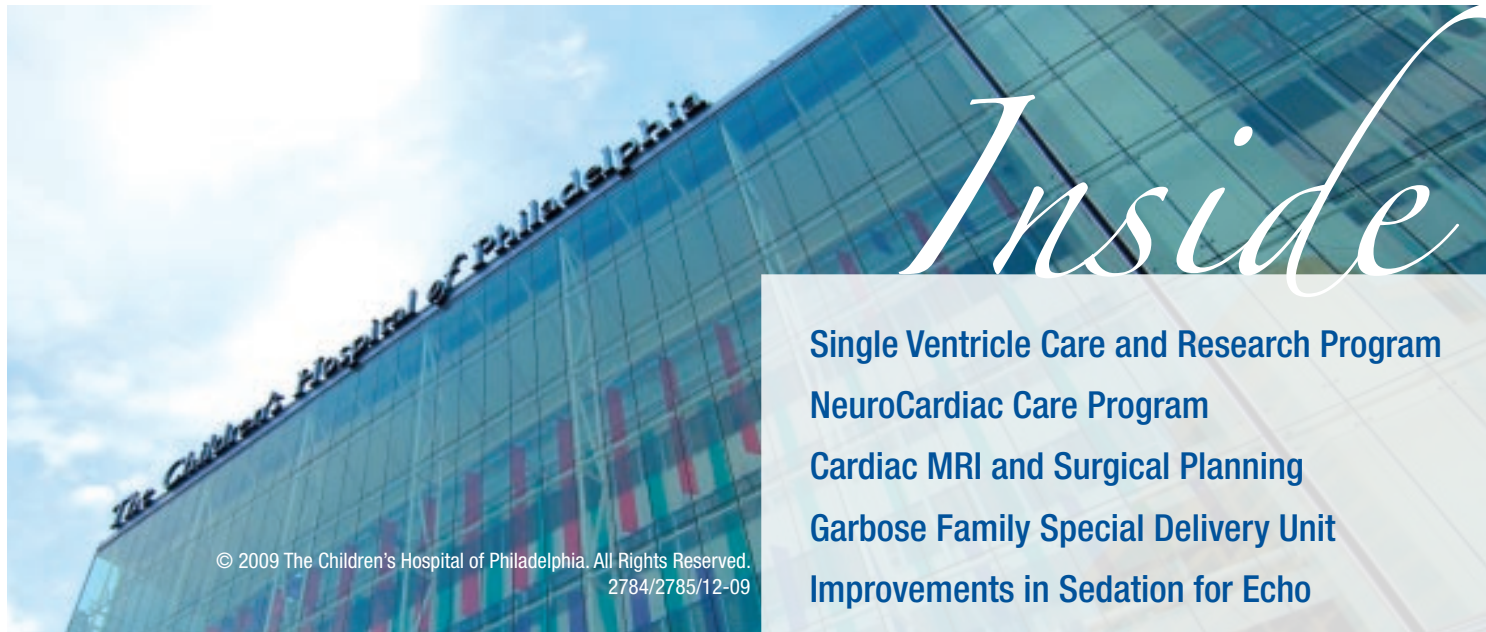
fuller understanding of developmental issues as they affect the entire child. Regular follow-up takes place at milestones throughout the first 18 months of life and then annually through adolescence.

The NCCP also serves as an educational resource for families, and for primary care pediatricians and other community healthcare providers, who are seeing growing numbers of CHD survivors in their practices.

Research is a critical component of the NCCP's efforts. The program works closely with the Cardiac Center's NeuroCardiac Research Group, formed in 1998, to investigate the causes and progression of neurodevelopmental sequelae. Studies conducted through the program will ultimately benefit its patients, while observations of the patient population will suggest new directions for research.

Initially the NCCP is enrolling neonates and infants with complex CHD who have been cared for in CHOP's Cardiac Intensive Care Unit. In subsequent phases, the program will expand to include older children and those not previously seen at CHOP.

News from your colleagues at the Cardiac Center



Single Ventricle Care and Research Program
NeuroCardiac Care Program
Cardiac MRI and Surgical Planning
Garbose Family Special Delivery Unit
Improvements in Sedation for Echo

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A Virtual Approach

Cardiac MRI and Surgical Planning

Cardiologists and surgeons at CHOP, in conjunction with bioengineers at the Georgia Institute of Technology, have developed a tool using cardiac magnetic resonance and advanced engineering techniques to do “virtual surgery” and plan out and predict the effects of different surgical approaches by computer. They described their initial experience in MRI and surgical planning methodology in the August, 2009 issue of the *Journal of the American College of Cardiology: Cardiovascular Imaging*.



“Using state-of-the-art cardiac MRI and sophisticated engineering approaches, the team can assess different surgical options to achieve the optimum blood flow from the liver and other parts of the body to the lungs with the lowest power losses to maximize the heart’s energy efficiency,” says co-author Mark A. Fogel, M.D., director of cardiac MRI at CHOP.

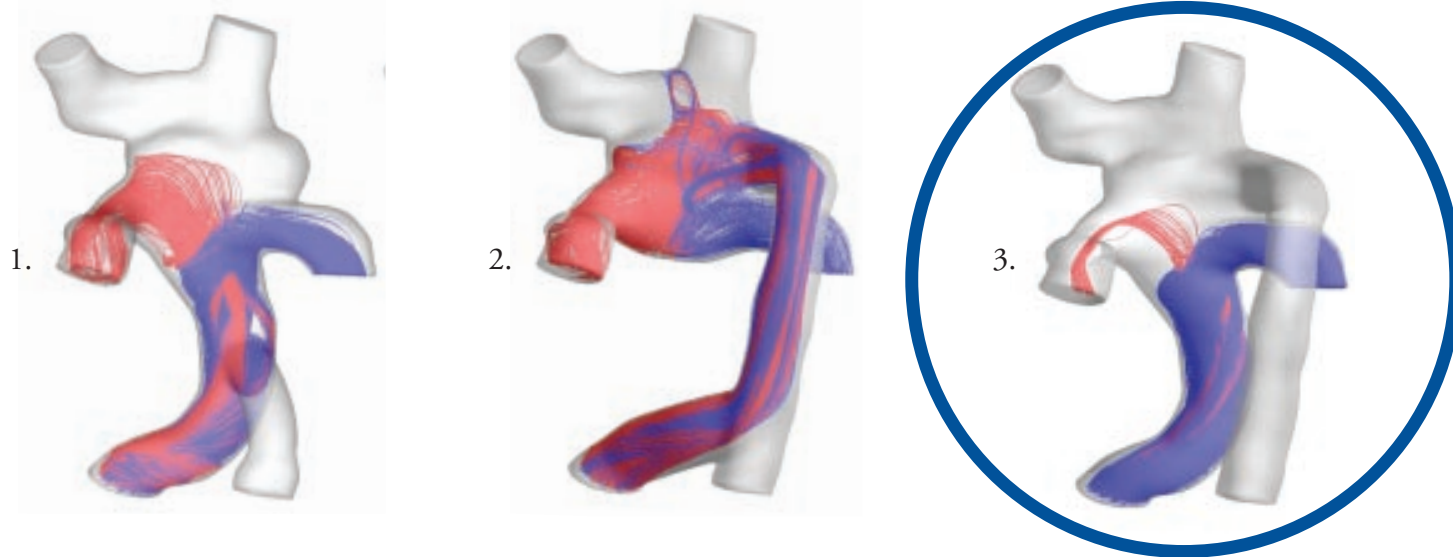
The paper details surgical planning for a 4-year-old with heterotaxy syndrome who was after the Fontan procedure but had developed left lung pulmonary arteriovenous malformations with a systemic oxygen saturation of 72 percent. Surgical planning included: acquiring cardiac magnetic resonance images; modeling the preoperative heart anatomy and blood flow; performing virtual surgeries; using computational fluid dynamics to model the proposed postoperative flow; and measuring the distribution of liver-derived hormonal factors and other clinically relevant parameters.

The cardiologists and engineers saw a highly uneven flow distribution: The left lung received about 70 percent of the cardiac output but only 5 percent of the hepatic blood. After analyzing the 3-D anatomy and flow, three surgical options with computational fluid dynamic modeling were investigated, similar to the diagrams below, to predict how much hepatic blood would flow to each lung and the associated power losses with each geometry.

Thomas L. Spray, M.D., chief of the Division of Cardiothoracic Surgery at CHOP, favored an option (No. 3 below) that demonstrated a slightly higher power loss but exhibited the better hepatic factor distribution to both lungs. Five months after surgery, the child showed a dramatic improvement in clinical condition, and oxygen saturation levels increased to 94 percent.

Cardiologists at CHOP and the engineers at GA Tech have completed seven image-based surgical planning cases to date and are leaders in the use of cardiac MRI and other highly developed technologies in the single ventricle and tetralogy of Fallot populations.

THREE SURGICAL OPTIONS WITH COMPUTATIONAL FLUID DYNAMIC MODELING



Advanced Cardiac MRI at CHOP

Cardiac magnetic resonance imaging at CHOP offers a full array of services that include 3-D cardiovascular anatomy; coronary imaging; ventricular function; blood flow; tissue characterization, such as myocardial scarring and tumor identification; and combined catheterization-MRI studies (XMR). CHOP was a leader in developing functional fetal cardiac MRI and measurement of collateral flow in Fontans. A new exercise cardiac MRI service is also available.

IN ONE PLACE • *The Garbose Family Special Delivery Unit*

True story: Brie, after a prenatal diagnosis of complete heart block, was delivered by c-section at 9:04 a.m. Induction of anesthesia took place at 9:06 and echocardiogram at 9:17. Pacing wires were placed at 9:27. She was paced at 70 beats per minute and admitted to the Cardiac Intensive Care Unit at 10:16 a.m.

More than 250 babies with birth defects diagnosed *in utero* have been born in the Garbose Family Special Delivery Unit (SDU), the world’s first delivery unit exclusively for babies with diagnosed birth defects, since it opened at CHOP in June 2008. A delivery unit in a pediatric hospital completes the spectrum of care that begins with prenatal diagnosis and monitoring. Every baby and mother has an expert team, comprised of numerous subspecialties, through delivery and from the moment of birth.

Approximately 40 percent of the babies born in the SDU are patients of CHOP’s Fetal Heart Program — including Brie. The SDU is on the Hospital’s sixth floor, which also includes all cardiac inpatient services. A newborn can be in the Cardiac Operative and Imaging Complex in seconds.

The prevalence of fetal diagnosis of heart defects continues to grow: The annual volume of fetal echocardiograms by the Fetal Heart Program has doubled in the past eight years to more than 2,000, and approximately 70 percent of newborns admitted to CHOP’s Cardiac ICU are diagnosed prenatally. A delivery unit adds an invaluable dimension to cardiac care.

SEDATED ECHO • *An Effective Alternative to Chloral Hydrate*

Oral chloral hydrate has long served as the mainstay for sedating children for transthoracic echocardiography (TTE) and other forms of noninvasive imaging. However, clinicians have become dissatisfied with chloral hydrate due to unpredictable onset and offset of sedation and concern for its safety profile.

A team of cardiologists and cardiothoracic anesthesiologists at CHOP decided to examine an alternative. In July 2007, the staff began offering sedated echo using face mask anesthesia.

A retrospective chart review comparing the two strategies found significant advantages to face mask anesthesia. Among the findings:

- In the chloral hydrate group, the study was not completed and was considered a failure in 6 percent of patients. In the anesthesia group, there were no failures.
- Time from onset of sedation to the beginning of TTE was significantly less (31 minutes less, on average) for the anesthesia group.
- Time from onset of sedation to discharge was significantly less (43 minutes less, on average) for the anesthesia group.

The cost of TTE with face mask anesthesia is higher. However, the costs are partially offset by the elimination of failed studies, the cost of a repeat study, improved efficiency in preparation and recovery of the patient, and potential for improved patient safety. Further details will be published in the *Journal of the American Society of Echocardiography*.

“Face mask anesthesia offers numerous advantages, and we’ve found it is preferred by both families and the medical team,” says study co-author Susan C. Nicolson, M.D., chief, Division of Cardiothoracic Anesthesia. “It has become the standard for TTE sedation at our institution. We hope other institutions will examine our findings and consider making the same change.”

Recent Publications

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Atrial septal defect devices used in the cardiac catheterization laboratory. Gervasi L, Basu S. *Progress in Cardiovascular Nursing*. 2009 24(3):86-9.

What do cardiovascular nurses know about the hematological management of patients with Eisenmenger syndrome? Moons P, Fleck D, Jaarsma T, et al. *Eur J of Cardiovascular Nursing*. 2009 8(4):246-50.