When the tables are turned:
A unique perspective on congenital heart disease

continued from Page 2

intervene or shield them from the periodic intrusions of the clinical staff.

Adolescents are striving to establish their identity and independence. The unpredictability of care and treatment of their condition, as well as the uncertainty about prognosis, may serve as a threat to developmental accomplishments. In addition to dealing with the psychological and social stressors of puberty, adolescents with congenital heart disease may have to deal with activity restrictions. They may have the potential for a greater degree of distorted body image, self-concept and altered peer relationships than experienced by the typical teenager. The need for continued treatment may be viewed as an unwelcome intrusion or threat to the control of their own lives, potentially resulting in poor adherence to treatment recommendations.

There are numerous challenges that health care providers must overcome when treating patients with congenital heart disease. In addition to providing care for the physical disease, clinicians must watch for the patient's or family's complex coping mechanisms. Clinicians must learn to recognize individual perceptions, values and decision-making processes, and then explore and support of these patients' and families' psychosocial requirements and concerns. In addition, the entire team providing care must acknowledge their own prejudices and feelings in order to meet the needs of the patients and families. Building on the perceived strengths of the individual child and family, and supporting their efforts at coping, will help to enhance healing and adaptation.

The fact that Jack continues to do well is a testament to his resiliency and the talent of the individuals and teams who provided and continue to provide his care. We assume our children will outlive us. The extraordinary dedication of our children's caregivers means that less and less of us have to cope with the actuality that we will likely outlive our child.

Quadrox D oxygenator provides many advantages

By Heidi Hillmann, BSN, CCRN, pediatric critical care nurse, Children's Hospital of Wisconsin.

Children's Hospital first began using the Quadrox D oxygenator in June 2008. It is a membrane oxygenator composed of hollow polymethlpentene fibers that collectively provide extremely low resistance to blood flow and are highly efficient at oxygenation and carbon dioxide removal, without plasma leakage. It effectively prevents the formation of microbubbles. For optimal blood handling, the Quadrox D has a heparin bioline coating to ensure a homogeneous, highly compatible surface that effectively protects a patient's blood.

This oxygenator has a number of advantages:

- Three key factors decrease the priming volume and surface area: the design for bundling the fibers within the oxygenator, the winding technique used and the flow pattern through the device.
- By making the oxygenator more compact and optimizing the blood flow path, it is possible to decrease the surface area of the membrane and heat exchanger. This reduces its potential for thrombus formation and inflammatory activation.

Recommended pump flow for this oxygenator is between 0.5 and 7 liters per minute with sweep flows between 0.1 and 15 liters per minute.

In the near future, Jostra AG will be introducing a smaller version of the Quadrox D for patients who require less than 0.5 liters per minute of blood flow.

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Calendar

Case Review: Cardiothoracic Surgery and Interventional Cardiology review clinically significant cases with an educational and quality improvement focus. Fourth Tuesday of the month, 7:30 a.m., Herma Heart Center conference room, first floor, Children's Hospital of Wisconsin.

Journal Club: CT Surgery, Cardiology, Critical Care and Anesthesia review a topic relevant to congenital or acquired heart disease. First Tuesday of the month, 4:30 p.m., Herma Heart Center conference room, seventh floor, Children's Hospital Clinics Building.

Research Focus: Herma Heart Center staff reviews existing or new research proposals, plans for abstract/manuscript preparation, invites guest speakers. Second Tuesday of the month, 7:30 a.m., cardiology conference room, first floor, Children's Hospital.

Cath Conference: Presentation of cases scheduled for cardiac surgery, interventional cath or group discussion. Every Thursday, 7 a.m., Briggs & Stratton Auditorium, second floor, Children's Hospital.

Heart Matters, a multidisciplinary approach to cardiac education, is produced quarterly.

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Inside this issue

3-D, contrast echocardiography bring new vision to pediatric imaging

By Edward Kirkpatrick, DO, pediatric cardiologist, Herma Heart Center, Children's Hospital of Wisconsin; assistant professor, Pediatrics (Cardiology), The Medical College of Wisconsin.

The importance of 2-D echocardiography in anatomic and functional assessment of the heart has been well established. Despite very good imaging data obtained from 2-D echocardiography, it still is limited in its ability to view a 3-D structure, such as the heart, in only two dimensions. Therefore, 3-D data, such as volume and spatial orientation, can be lacking. Attempts to create 3-D ultrasound images began in the 1960s, and in the early 1990s, real-time 3-D echocardiography became possible.[1]

3-D echocardiography

3-D images initially were created from serial 2-D images through freehand imaging. Later, automated echo probes obtained rotational images at defined angles from a fixed axis.[1, 2] Further improvement of volumetric data collection utilized gated sequencing by being timed with heart rate and respirations to avoid artifacts.[1, 2] This obtained a volume set that required post-processing with specialized computer software.[1]

Current systems can obtain real-time 3-D data using probes that contain a "matrix array" with a large number of individual imaging units inside arranged in a grid fashion.[1] Images immediately are in a 3-D format that can be post-processed for specific details and volume analysis.[1] Image quality still is dependent on 2-D quality.

Clinically, 3-D echocardiography can be used for ventricular quantification of volume, mass, segmental wall motion and cardiac anatomic assessment.[1, 2] (See Figure 1.)

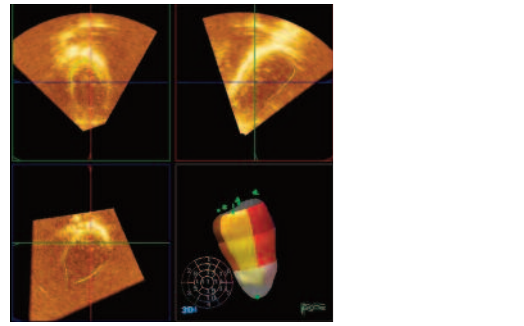

Figure 1: This graphic shows a 3-D analysis of the left ventricle in a pediatric patient, while also indicating three different planar views and a computer-generated left ventricle volume shell. This information is created from single acquisition gated over seven heart beats.

Continued on Page 4

When the tables are turned: A unique perspective on congenital heart disease

By Chris Brabant, MBA, CCP, pediatric perfusionist, Herma Heart Center, Children's Hospital of Wisconsin.

There are 11,000 babies born daily in the United States, and of those, more than 400 are born with a birth defect. Of these, 87 are cardiac anomalies, making congenital heart disease the most commonly occurring birth defect with a reported incidence of eight cases per 1,000 live births. There are approximately 35,000 new diagnoses annually, and congenital heart disease is the leading noninfectious cause of infant mortality. Furthermore, congenital heart disease is a lifelong disability that places a significant emotional and financial burden on both the patient and the family. These children are at greater risk for issues involving school performance, social adjustment, employability and quality of life.

Despite surgical intervention, only a small minority of congenital cardiac conditions are cured completely. In the majority of patients, particularly those born with complex lesions, the cardiac malformation results in a chronic condition requiring recurrent medical observation and necessitating lifelong care.

Shortly after he was born, my son Jack was diagnosed with pulmonary atresia with an intact ventricular septum, resulting in a single left ventricle and a diminutive nonfunctioning right ventricle. He was stabilized at an outlying facility and transported to Children's Hospital after his first few hours of life. On day four of life, he was brought to the operating room for a Blalock-Taussig (systemic to pulmonary) shunt. He received a bidirectional Glenn (superior vena cava to right pulmonary artery) shunt at age 4 months. At age 2, he had his third cardiac surgical procedure, which was the completion Fontan (inferior vena cava to right pulmonary artery) shunt.

As a clinical perfusionist, one of my responsibilities is to operate the heart-lung machine for children whose congenital heart disease requires the use of cardiopulmonary bypass for their surgical procedure. After Jack was born, I was placed in the unique position of having my physician and allied health colleagues care for my son.

Parents experience a spectrum of emotions during their early adjustment to a cardiac disease diagnosis. It causes the family to not only face the normal transition of having a child but also the additional stress of the disease. Parents may grieve over the loss of what was supposed to be a healthy newborn, and they may feel helpless over their inability to protect their child from harm. They may feel guilty and responsible for their child's heart defect. There also is intense dismay and dread at witnessing the pain and suffering that the child endures. The potential for marital discord may increase as a result of the stress caused by the uncertain outcome. At this difficult time, it is important to have strong personal, family and professional support.

There is significant adjustment for the siblings of a child with congenital heart disease. If the newborn's condition includes immediate hospitalization, those at home may not entirely believe there is a new baby. They may fear that they also may become sick, or that perhaps they some how caused their new brother or sister to become ill. There may be feelings of resentment because of the additional attention that an ill or recovering newborn requires. If another hospitalization is necessary after coming home, siblings may have feelings of abandonment and helplessness due to parental absence. There is additional anxiety caused by the disruption of normal routine and fear of the unknown.

The child born with congenital heart disease likely will have many challenges not experienced by a healthy child. An infant who requires immediate hospitalization and surgical intervention will have an initial loss of the normal stimuli that foster optimal growth and development. This includes the physical and physiologic protection offered by parents, such as normal touch, feeding and nurturing.

As children become older and move into the toddler and preschool years, they become increasingly aware of their environment. Hospitalization may result in extreme sensitivity to separation from parents and intrusions by strangers. Painful procedures are met with vehement resistance, and the child may be dismayed and confused by the seeming inability of the parents to rescue them from a perilous environment. Illness or hospitalization may be perceived as punishment for something they have done wrong.

Older children begin to develop an understanding of their bodies and the knowledge that the heart is a vital organ. The concept of having heart disease requires continual medical management, and periodic intervention may be particularly upsetting. Hospitalization and surgical intervention at this time may invoke fears of bodily injury and the possibility of death. Children generally perceive parents to be their ultimate protectors and become frustrated by their parents' inability to

Continued on Page 3

3-D, contrast echocardiography bring new vision to pediatric imaging

continued from Page 1

Ejection fraction is a volumetric measurement of the heart's output and often is reported on standard 2-D echo studies. Using 2-D data, the ejection fraction is derived from equations that attempt to quantify ventricular volume. Though those methods often correlate with gold standard methods of volume measurement, they do not have the same accuracy.[3]

Current 3-D echocardiography has been shown to have accuracy very similar to that of magnetic resonance imaging in adult patients.[3] Likewise, 3-D echo has shown improvement in assessment of masses in the left ventricle due to better endocardial and epicardial border delineation.[2] In addition, the use of 3-D imaging allows for a simultaneous multi-segment view of the left ventricle, which can be used to quantify regional wall motion as opposed to the subjective qualified descriptions that currently are used in 2-D wall motion assessment.[1, 2]

Intracardiac anatomy can be assessed in its full functional form with 3-D echocardiography. Images can be cropped to focus on particular details and manipulated in 360-degree rotational views to allow evaluation of areas in relation to other cardiac structures.[1, 2] This allows visualization from any perspective that may be particularly useful for surgical repair.

Contrast echocardiography

Adult studies have shown that 10-15 percent of resting echocardiograms are suboptimal and this rate can be as high as 33 percent for stress echocardiography.[4] Though pediatric patients generally allow for better echo imaging due to their body habitus, they also can have suboptimal imaging especially in postoperative states and obese patients. Contrast agents can be used to enhance echo images to allow 75-95 percent of suboptimal images to become diagnostic.[5]

Contrast agents are solutions of microbubbles filled with a soluble gas that are intravenously injected and travel into the cardiac chambers. These microbubbles create a backscatter from an ultrasound signal that is different from the surrounding blood proportional to the size, density and compressibility of the microbubble.[4] Therefore the outer shell of the microbubble and the gas inside determine its ultrasound characteristics.[4]

The use of agitated saline as a contrast agent in cardiac imaging was first reported in 1968.[6] However, its thin, large, air-filled bubble only allows filling of the right side of the heart. This has been useful for evaluating right to left shunts inside the heart or in the lungs.[6] Saline bubbles normally can't cross the pulmonary capillary bed, therefore contrast agents were engineered to be small enough to circulate into the left cardiac chambers and to be stable enough to maintain their form.[4] Currently, three second-generation contrast agents are available. Children's Hospital uses Definity®, a contrast agent made by Bristol-Myers Squibb Medical Imaging, Inc. This agent has a phospholipid shell filled with perfluoropropane gas and has an average bubble size of 1.5μm.[5]

Second-generation contrast agents generally are used for patients with limited quality echocardiography pictures, right- and left-sided ventricular opacification or endocardial border delineation.[5] (See Figure 2.) This has been shown to enhance quantification of ventricular volumes, segmental wall motion and function. In addition, contrast can better define intracardiac abnormalities such as tumors, thrombi, aneurysms and left ventricle noncompaction.[5] Newer contrast agents are in final stages of approval by the Food and Drug Administration for real-time evaluation of myocardial perfusion for detection of coronary artery disease.[4]

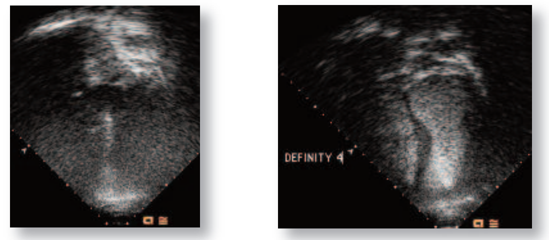

Figure 2: These graphics show a four-chamber view of the heart with poor visualization of the left ventricular endocardial border and apex. Definity® contrast greatly improved visualization of the left ventricle.

Second-generation contrast agents currently are contra-indicated in patients with any right-to-left or bidirectional cardiac shunts, therefore excluding their use in many types of unrepaired congenital heart disease. Patients with hypersensitivity to components of the contrast agents are prohibited from their use. These agents cannot be injected intraarterially.[5]

The second-generation contrast agents generally are well tolerated with Definity® contrast showing the lowest frequency of side effects at 7.6 percent.[4] These mild side effects include headache, flushing and back pain and generally are transient[4]. However, rare allergic reactions can occur at an estimated incidence of one in 10,000[5]. A Food and Drug Administration review of Definity® showed four deaths of severely ill patients and 190 adverse advents that prompted a black box warning in October 2007.[5] A detailed review showed that most of the events likely were not related to Definity®, and the FDA revised its recommendations to the previous post-marketing applications, though unstable or high-risk patients require 30 minutes post-contrast monitoring.[5]

The FDA has not approved contrast agents for use in children, so they are considered "off-label".[5] Studies have shown that these agents can be used safely in pediatrics and also improve imaging, as is the case in adults.[5, 7] Children's Hospital has a detailed contrast protocol, and 30 minutes post-contrast monitoring is performed on all pediatric patients.

Combined 3-D and contrast echocardiography

As 3-D echocardiography is limited by its 2-D quality, the enhanced imaging provided by contrast agents for 2-D echocardiography also enhances the quality of a 3-D echocardiogram.

In a study by Jenkins et al., 2-D and 3-D echocardiograms, with and without contrast agents, were compared in accuracy with MRI for left ventricle volume assessment in adult patients.[3] Accuracy improved with 2-D echocardiograms that included contrast agents and improved further with standard 3-D echocardiogram images. Accuracy was the best on contrast-enhanced 3-D echocardiograms, which had very similar values to those obtained from MRI[3]. Interobserver variability was lowest (less than 5 percent) only in the 3-D echocardiography techniques.[3]

Separately or together, 3-D and contrast echocardiography offer new tools for providing objective data to cardiac care. This will allow providers to have consistent, accurate, volumetric data and may improve anatomic assessment to plan medical and surgical therapy.

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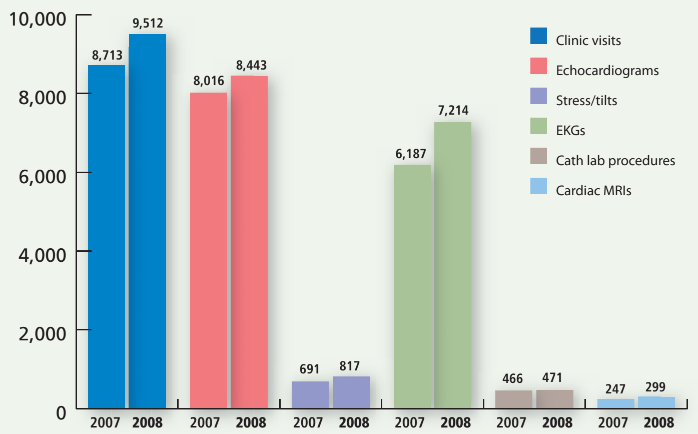
Herma Heart Center statistics



Total cases of open and closed heart surgeries increased 7.49 percent from 2007 to 2008.