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Hypertension in the Adult Congenital Heart Disease Population: A Review of the Literature

By Mark Townsend, MD; Robert Battle, MD;
Alexander Ellis, MD; Sam Lee, MD; and
William Moskowitz, MD

As medical and surgical techniques to repair or palliate congenital heart lesions continue to improve, the number of patients surviving to adulthood continues to expand rapidly. The total adult congenital heart disease (ACHD) population surpassed one million in 2005.¹ As the number of patients with ACHD increases, so, too, does the likelihood that adult cardiologists and primary care physicians are involved in their care. Familiarity with the anatomy and physiology of the individual lesions as well as late-onset complications is vital for all physicians caring for this group of patients. Medical management of adult comorbidities in the ACHD population however, lacks uniformity in basic health issues such as hypertension, which is still often considered to be a "psychosocial issue."²

As younger patients with congenital heart disease grow up, the onset of 'typical' adult diseases further complicates their care, adding additional variables to already complicated cardiovascular profiles. Risk factors for hypertension are amplified in this population by the usual lifestyle choices confounded by intrinsic physical limitations in some patients. Exercise intolerance and activity restriction are known to increase the risk for obesity, which in itself is a significant risk factor for the development of hypertension. Emotional

challenges such as anxiety and mood disorders are also prevalent in the ACHD population. The link between depression and enhanced sympathetic activation cannot be ignored in the ACHD population with hypertension.³ Lastly, even elements as basic as target blood pressures for a given congenital heart lesion and its surgical repair remain poorly understood.

We undertook a systematic review of the current literature for information pertaining to the diagnosis and management of hypertension in the ACHD population.^{4, 5} We discuss hypertension care within the limitations of relevant evidence-based data in the current literature.

Methods

Medline / PubMed was searched for citations with the keywords "hypertension" or "systemic hypertension" paired with all keywords listed in the appendix. Available MeSH headings were then used to search for all congenital conditions and pharmacologic classes discussed. Given the scarcity of pertinent literature, all citations were reviewed and included based on relevant content, regardless of format or trial design. References of included articles were scanned for additional publications to check the completeness of our review. No other review articles were available for analysis. Only articles written in English and French were reviewed. Articles were not excluded based on date of publication.

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Tetralogy of Fallot: Natural History and Late Complications

The ACHD population with TOF is adversely affected by the natural history of the resultant pulmonary insufficiency. The late morbidity of pulmonary insufficiency has been increasingly appreciated in recent years, such as progressive right ventricular dilation and fibrosis or scar around the ventriculotomy predisposing to lethal arrhythmias.⁶ Patients are “asymptomatic” until RV dysfunction is severe and irreversible; sudden death may be the presenting feature in up to 6% of patients.⁷ Right ventricular dysfunction is further compounded by preexisting pulmonary hypertension created by the use of various central shunts as an early bridging procedure before a complete repair, in an era before early primary repair was possible. Central shunts often distort the architecture of the pulmonary arteries, leading to distal pulmonary arterial obstruction.

Biventricular failure is not uncommon in this group, and is not well tolerated. Predisposition for left ventricular failure in this population is partially driven by the kinetics of abnormal right ventricular function, the association of which is not well understood.^{8,9} In addition, an association between TOF and aortic medial abnormalities exists which can lead to progressive aortic root dilation and aortic insufficiency.^{10,11} Given this underlying vascular medial abnormality, systemic hypertension serves as an additional insult in this population.

“The significance of adult onset comorbidities in the congenital heart disease population as it ages has been predicted and lamented for years.”

Our review of the literature found no studies that addressed the prevalence or management of hypertension in patients with repaired TOF at any age. However, the natural history of this disease indeed suggests a predisposition to systemic hypertension. In an uncontrolled series of 147 uncorrected TOF adults evaluated with cardiac catheterization, 9.5% of patients had overt systemic hypertension.¹² This population shares histological similarities to systemic hypertension at baseline in terms of myosin composition of ventricular myocardium, the significance of which is not completely clear.¹³ Patients with surgically repaired ventricular septal defects and TOF were shown together to have a mildly higher, but significant systolic blood pressure during exercise than age-matched controls.¹⁴

The use of calcium channel blockers for hypertension in TOF has been suggested, given the possible benefit of simultaneously reducing pulmonary vascular resistance. The post-operative use of nifedipine has been reported to be efficacious in children after cardiothoracic surgery, including TOF repair, with a target systolic blood pressure of ≤ 110 mm Hg in patients 5 years or older. No adverse effects were noted in a total of 337 hours of intravenous use.¹⁵ The risk of using calcium channel blockers is that sinus node slowing may be significant enough to precipitate hemodynamically-significant bradycardia or the emergence of an ectopic atrial pacemaker.¹⁶ This is especially germane given that underlying sinus node dysfunction may occur in one third of



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TOF patients, increasing the potential complications with use of calcium channel blockers in this population.¹⁷ Thus, in practice, calcium channel blockers are not often used in patients with repaired TOF.

Both beta-1 and beta-2 receptors are down-regulated in TOF. The use of beta blockers is known to increase both types of beta receptors.^{18, 19, 20} Beta blockers are often used for aortic root dilation in Marfan Syndrome without clear consensus; by extension, there is no current consensus on their use in TOF.^{21, 22} Research needs to be done to establish the potential role of beta blockers in the populations with: (1) pre-clinical and, (2) overt alterations of right and left sided structures and function, including aortic root dilation. The effects of beta blockers on right ventricular function in addition to potential modification of RV mediated arrhythmias in this population remain relative unknowns, and require investigation. In practice, beta blockers are often used in the management of hypertension in TOF given the associated aortopathy and the risk of ventricular arrhythmias precipitating sudden death.

Coarctation of the Aorta

The association between coarctation of the aorta and hypertension is well established in an extensive body of literature. Regardless of timing or method of repair, it is recognized that this population is plagued by persistent arterial stiffness despite a widely patent repaired aorta. This is well-demonstrated by ambulatory blood pressure monitoring studies.²³ In the current adult population, late hypertension is observed in at least 25% of patients.^{24, 25} Almost two thirds of patients have been reported to be hypertensive 15-30 years after coarctation repair.^{26, 27} Age at the time of initial repair is the most important predictor of late hypertension, which is relevant to the adult population since later repair occurred more frequently in the past.^{28, 29} Of interest, the presence of a commonly associated bicuspid aortic valve does not appear to effect blood pressure, but does pose a risk for general aortic complications such as aneurysms, dissection and rupture.^{30, 31} Risk factors for the late onset of hypertension are listed in Table 1.

Table 1. Risk Factors for the Late Onset of Hypertension in Coarctation in the Absence of Re-coarctation

- "Gothic" geometry of the aortic arch³² - limited by difficult reproducibility³³
- Residual descending aortic narrowing³⁴
- Repair with a subclavian patch³⁵
- Polytetrafluoroethylene patch aortoplasty³⁶
- Reduced vascular reactivity³⁷
- Late repair^{38, 39, 40}
- Maximum exercise systolic BP > 193 mm Hg⁴¹

The etiology of hypertension after coarctation repair is multifactorial. Decreased baroreflex sensitivity to changes in arterial pressure has been shown in 6 control matched children after repair.⁴² Histologic changes of the carotid wall with increased intima medial thickness has been reported.^{43, 44} Renin levels were shown to increase after captopril challenge in a small number of coarctation patients when compared to patients with essential hypertension, suggesting a renin mediated effect.⁴⁵ A follow-up study, however, showed antidotal evidence that in spite of the extensive activation of the renin-



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angiotensin-aldosterone axis in this population, the effect of angiotensin-converting enzyme (ACE) inhibition is quickly lost.⁴⁶ This has not been reproduced in a large trial.

In spite of the clear predisposition of this population to systemic hypertension, the benefits of medical intervention have not yet been studied on a large scale. Large registries still have not been designed to standardize treatment regimens.⁴⁷ Many patients with a history of previous coarctation repair are not receiving optimal care.⁴⁸

Per ACC/AHA guidelines, beta blockers, ACE inhibitors and angiotensin receptor blockers should be used as first-line medications with the choice of medication influenced in part by aortic root dimensions, aortic insufficiency and valvar aortic stenosis.⁴⁹ In tandem with medical management, hypertensive patients should be evaluated for possible recoarctation defined as a peak gradient of more than 20 mm Hg across the coarctation repair site via cardiac catheterization.^{50, 51} Screening for hypertension is performed by measuring the blood pressure in both arms and a leg keeping in mind the historical use of subclavian flaps for surgical repair.

Single Ventricle Physiology with a Morphologic Left Ventricle

Tricuspid atresia makes up the large majority of patients with single ventricle physiology, a morphologic left ventricle and a Fontan anastomosis. Adult patients who have undergone some variation of a Fontan procedure are prone to atrial arrhythmias, protein-losing enteropathy, and thrombus formation.⁵² Specifically, patients with a Fontan for tricuspid atresia have a 10 to 20% risk of atrial tachycardia or flutter-fibrillation, and a 13% risk of developing protein-losing enteropathy as a result of elevated systemic venous pressures. NYHA class is I or II in as many as 90% of patients. There is a low incidence of left ventricular dysfunction.^{53, 54, 55}

Neurohormonal activation after a Fontan occurs both acutely and chronically with elevation of renin, angiotensin II, vasopressin and endothelin-1.^{56, 57, 58} This cascade theoretically predisposes this population to ventricular hypertrophy, which in the setting of a Fontan is poorly tolerated because forward flow through the lungs is passive and dependent on good diastolic function of the single ventricle. A small series of 23 patients with a mean age of 19.4 years looked for LV hypertrophy but reported normal LV masses as assessed by MRI, albeit with diminished ejection fractions compared to a normal control population.⁵⁹ The elucidation of the risk of hypertrophy and hypertension will be a requisite as this population ages.

Medical management in this population is often first initiated for Fontan dysfunction with minimal attention paid to blood pressure ranges. The use of ACE inhibitors has been widespread as a front line agent along with diuresis in patients with Fontan dysfunction, in spite of the fact that this management does not affect pulmonary vascular resistance significantly.⁶⁰ Data on the use of ACE inhibitors is limited. Two very small trials have compared patients with Fontans who are treated with ACE inhibitors to untreated patients, with the outcome being response to exercise. There was no change in cardiac autonomic or hemodynamic response to exercise with the addition of ACE inhibition.^{61, 62} This topic deserves a more thorough investigation. The use of other classes of medications is limited, but the potential role of newer medications is exciting. For example, endothelin-1 is believed to play a significant role in the elevation of pulmonary vascular resistance as



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evidenced by muscularization of the pulmonary arterioles, but has not yet been applied clinically to this population.^{63, 64}

The range of normative blood pressures for this population is unknown, and needs to be established with large, multi-center trials. The incidence and effect of systemic hypertension in this adult population is undocumented. In our practice, an LV mass index of 90 to 100 g/m² measured by trans-thoracic echocardiography is arbitrarily used as an independent indication for medical management, rather than blood pressure alone.

Systemic Morphologic Right Ventricles

Three conditions account for the majority of patients with systemic right ventricles: congenitally corrected transposition of the great arteries (L-TGA), atrial switch operations (Senning and Mustard repairs) for transposition of the great arteries (D-TGA), and Hypoplastic Left Heart Syndrome (HLHS). The relevance of the ability of systemic right ventricles to generate systemic hypertension as defined by left ventricular standards is an unknown since normal systemic right ventricular (RV) pressure ranges have not been defined.

Replete in the hypertension literature is the improvement of hypertension and left ventricular function with exercise. Systemic right ventricular function after a Mustard repair does not, however, improve with exercise.^{65, 66} Patients with either L-TGA or atrial switch operations were noted to have chronotropic incompetence and an impaired stroke volume response during exercise.⁶⁷ It is not clear whether this information can be extrapolated to other systemic right ventricles given our lack of understanding of their natural history.

Congenitally Corrected Transposition of the Great Arteries (L-TGA)

Nature's corollary to surgically created systemic right ventricles is L-TGA. The natural history of this condition is not benign, even in the absence of associated conditions. These patients are prone to systemic RV dysfunction, systemic / tricuspid valve regurgitation, conduction abnormalities, complete heart block, arrhythmias and sudden death.^{68, 69, 70} The morphologic RV is usually supplied by a morphologic right coronary. As the myocardium hypertrophies to accommodate its work load, it is prone to ischemia, further worsening ventricular oxygenation and inducing further hypertrophy.^{71, 72} Surgical options depend on associated conditions. The double switch operation (incorporating an atrial switch along with a great artery switch) has been used, but is not widely implemented as the standard of care and is a high risk procedure in adult populations.⁷³

Defining acceptable blood pressure ranges that strike a balance between compensatory and excessive hypertrophy will be important in the future. With the assertion that 90 – 97% of these 'systemic' tricuspid valves are abnormal and prone to regurgitation, the preload of the morphologic right ventricle is rarely normal.⁷⁴ Afterload reduction with ACE inhibition as of yet has no proven role in the management of L-TGA. In the face of moderate tricuspid regurgitation, preload alteration in the form of diuresis is often added, also without proven benefit. If systemic AV valve regurgitation is severe, valve replacement is performed before the onset of right ventricular failure. No large clinical trials to date have studied the medical management of L-TGA. The balance between management



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of preload and afterload reduction needs to be investigated, along with the definition of goal blood pressure ranges.

Hypoplastic Left Heart Syndrome

The afterload of a systemic right ventricle in HLHS is not normal on several accounts, even in the presence of a normal blood pressure. The pathophysiology of HLHS entails aortic arch hypoplasia, and presumably invokes the natural history of coarctation itself. Surgical correction via completion of a Fontan operation, (putting the pulmonary and systemic circulations in series), inherently increases afterload on a single ventricle by increasing the vascular length of the circuit, and decreasing the vascular cross-sectional area.⁷⁵ Neurohormonal activation after a Fontan occurs both acutely and chronically as discussed above, including activation of endothelin-1. Forearm vascular resistance has been shown to be increased along with diminished vascular endothelin function in these patients.^{76, 77} Under normal circumstances, an increase in left ventricular afterload leads to an increase in ventricular contractility, but in patients with a Fontan, this compensatory mechanism is altered and does not occur. These patients are left with a mismatch in ventricular contractility and afterload with resultant reduction in mechanical efficiency and limitation in ventricular functional reserve.⁷⁸

Afterload reduction therapy should be considered in all patients with a systemic right ventricle as a result of HLHS, although the benefit remains unproven. Cardiac dysfunction in this population is poorly tolerated, evidenced by gradual elevation of Fontan pressures and eventual Fontan dysfunction. The corollary use of ACE inhibitors to prevent myocardial remodeling and hypertrophy of the left ventricle is an established tenant in general practice, the significance of which is not clear in HLHS.^{79, 80} Diastolic dysfunction secondary to compensatory hypertrophy is difficult to accurately quantitate in an outpatient setting, but if left to progress unchecked, will inevitably be detrimental.

Research needs to be done in this field beginning with a definition of a normal blood pressure range in a Fontan patient with HLHS. The potential use of newer drugs such as endothelin receptor antagonists in this population is an exciting option that may be beneficial in maximizing pulmonary blood flow, thereby further improving Fontan hemodynamics.⁸¹

Atrial Switch Operations

The coronary circulation of the systemic RV in both the Mustard and Senning procedures is supplied by a right coronary system, creating the potential for a myocardial perfusion supply / demand mismatch. This population is known to be at a significant risk for supraventricular tachyarrhythmias, notably atrial flutter which is associated with CHF.⁸² In a series of 448 survivors of the Mustard procedure, only 40% remained in sinus rhythm at 20 years post surgery.⁸³ Patients have been reported to have a 2.4% loss of sinus rhythm per year.⁸⁴ The risk of sudden death is as high as 7% without identifiable risk factors in a series of 113 patients with Mustard repairs.⁸⁵

Systemic vascular resistance (SVR) after an atrial switch has been shown to be abnormally increased during exercise.⁸⁶ Empiric use of losartan in a small number of patients with a mean systolic blood pressure of 117 showed improvement in systemic RV ejection fraction, tricuspid regurgitation and functional capacity in terms of exercise



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time. The mean systolic blood pressure dropped to 107 mm Hg on 50 mg/day.⁸⁷ Enalapril may not affect SVR in this population. In a study of 8 patients, blood pressure decreased over one year on enalapril without a decrease in systemic vascular resistance, exercise capacity and cardiac index at peak exercise.⁸⁸ A pilot study of a similar patient population before and after ACE inhibitor therapy for > 6 months showed no difference in exercise and MRI determined variables.⁸⁹ The benefit of the seemingly widespread and routine use of ACE inhibitors or angiotensin receptor blockers remains controversial and unproven after an atrial switch.

Conclusion

The significance of adult onset comorbidities in the congenital heart disease population as it ages has been predicted and lamented for years. However, the ongoing inadequacy of our current knowledge base as it pertains to the ACHD population is evidenced by the paucity of research that has been conducted on one of the most basic of adult health concerns, hypertension. Our review of the literature in common subpopulations with ACHD reveals little in terms of the incidence, prevalence, and implications of hypertension. Without a full understanding of the natural history of hypertension in the ACHD population, evidence based management algorithms seem frustratingly distant. Future research at its most basic level needs to consist of defining normal blood pressure ranges specific to the congenital cardiac malformation and the variations of each surgical correction.

Appendix

Keywords used along with "hypertension" and "systemic hypertension:"

- Adult Congenital
- Grown Up Congenital Heart Disease
- GUCH
- Tetralogy of Fallot
- coarctation of the aorta
- tricuspid atresia
- congenitally corrected transposition
- L-TGA
- atrial switch
- Mustard
- Senning
- systemic right ventricle
- right ventricle
- hypoplastic left heart

Relevant MeSH terms available that were used:

- aortic coarctation
- hypoplastic left heart syndrome
- transposition of great vessels
- Fontan
- Tetralogy of Fallot
- tricuspid atresia
- angiotensin converting enzyme inhibitors
- adrenergic beta-antagonists
- calcium channel blockers

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The Division of Pediatric Cardiology at UNC - Chapel Hill is recruiting for an open rank clinical faculty position.

Primary responsibilities will be at the North Carolina Children's Heart Center affiliate site in Greensboro, NC providing outpatient and inpatient care for a regional medical center and participating in the education of UNC medical students and residents.

Additional opportunities include hospital-based roles at the Children's Hospital, Chapel Hill. Candidates must be Board Certified or Board Eligible in Pediatric Cardiology.

Applicants should forward a CV and cover letter to:

Scott Buck, MD
Division of Pediatric Cardiology,
North Carolina Children's Heart
Center
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CONGENITAL CARDIOLOGY TODAY

Medical News, Products and Information

Novel Gene Found for Dilated Cardiomyopathy

Researchers in the Heart Institute at Cincinnati Children's Hospital Medical Center have discovered a novel gene responsible for heart muscle disease and chronic heart failure in some children and adults with dilated cardiomyopathy (DCM).

Mutations in the ANKRD1 gene may cause DCM, which is the most common cause of chronic heart failure in young people and the most common reason for heart transplant. ANKRD1 is a gene that encodes a protein that plays a role in the structure and functional ability of the heart.

The internationally conducted study was published in the July 21, 2009 issue of the *Journal of the American College of Cardiology*.

"Our study indicates that variants in ANKRD1 result in dysfunction of the contraction apparatus and signaling machinery of the heart – the method by which cells communicate to influence heart function," says Jeffrey Towbin, MD, Co-Director of the Heart Institute and Director of Cardiology at Cincinnati Children's. "This clarifies the mechanisms by which these inherited mutations cause disease in a subset of DCM patients."

DCM is a condition in which the heart becomes weakened and enlarged and cannot pump blood efficiently. The decreased heart function can affect the lungs, liver, kidneys and other body systems. DCM is one of the cardiomyopathies, a group of diseases that primarily affect the heart muscle. Cardiomyopathies have different causes and affect the heart in a variety of ways. In DCM the major pumping chamber of the heart, the left ventricle, is dilated, often without any obvious cause.

DCM occurs more frequently in men than in women and is most common between the ages of 20 and 60 years, although it also occurs in fetuses, newborns and children. About one in three cases of congestive heart failure is due to DCM, which also occurs in children.

Dr. Towbin and his colleagues screened 208 patients, mostly children and young adults, with DCM for gene mutations. They found three, disease-associated variants of the ANKRD1 gene. All four patients carrying the variants were male. This prevalence rate is consistent with prevalence data for most of the other known genes associated with DCM. This finding confirms previous gene discoveries by Dr. Towbin's group. It also "provides us with a better understanding of the causes and mechanisms involved in the development of this disease and will enable better genetic testing and new treatments to be devised to improve outcomes of this serious disease," according to Dr. Towbin.

The study was funded by grants from the National Institutes of Health, the Children's Cardiomyopathy Foundation and the Abby Glaser Children's Heart Fund. Collaborating institutions included Texas Children's Hospital and Baylor College of Medicine in Houston, the Medical Faculty Mannheim at the University of Heidelberg in Germany, the Institute of Cardiovascular Science and University College in London in the United Kingdom, and the Tokyo Medical and Dental University in Japan.

Dr. Towbin is co-author of another study in the same issue of *JACC* showing that the ANKRD1 gene also causes a different clinical form of cardiomyopathy.

Registration is Open for Cardiology 2010

Cardiology 2010 will take place February 10-14, 2010 at the Contemporary Resort and Convention Center, Walt Disney World, Lake Buena Vista, Florida, USA.

This annual post-graduate course is designed for physicians, nurses, perfusionists, administrators, clinical pharmacists, respiratory therapists, and all others involved in the care of neonates, infants, children and young adults with cardiovascular disease.

The International course faculty will present over 300 talks in Plenary

Sessions, as well as small group and subspecialty breakout sessions covering all areas necessary for comprehensive care of patients. Each year, the course faculty, topics and format are chosen following a careful assessment of prior attendee's comments and reviews. *Cardiology 2010* also invites new young investigators from around the globe to present new science and ideas.

Based upon the positive feedback they received following *Cardiology 2009* in Nassau last year, they have modified the course format to allow attendees the opportunity to enjoy more free time without a strict meeting agenda. In addition, in response to the realities of the current economic climate, they have reduced the cost of the meeting for all attendees, while still providing over 30 contact hours including hot topics, basic reviews, subspecialty breakouts and much more!

Abstract Submission Deadline is October 19th, 2009; Notification of Acceptance will be October 30, 2009. Course participants are encouraged to submit abstracts for consideration for the *Annual Outstanding Investigator and Nursing Scientist Awards*. The top six abstracts will be presented as oral presentations; the remaining will be presented in one of three poster sessions. The international course faculty will select the recipients of the *7th Annual Outstanding Investigator Award*, and the *5th Annual Nursing Scientist Awards*; the winners will receive an award at the featured plenary session on February 13th, 2010, of a complimentary registration for *Cardiology 2011* and a \$500 travel grant.

Abstracts will be accepted in all aspects of cardiovascular care, including cardiac issues in the neonatal and pediatric intensive care units, cardiovascular nursing, inpatient and outpatient cardiology, surgical, anesthesia and perfusion research, as well as basic cardiovascular science.

There is a Special Track for Residents, Fellows and Junior Faculty, Including a Pre- and Post-Conference Seminar. Expanding on a very popular feature in previous years, a special course-long track is planned for trainees and junior faculty

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Supervisor Pediatric and Fetal Echocardiography Laboratory

The Scott and Laura Eller Congenital Heart Center at St. Joseph's Hospital and Medical Center in Phoenix, Arizona is recruiting for a SUPERVISOR for its PEDIATRIC AND FETAL ECHOCARDIOGRAPHY LABORATORY. The successful candidate will be joining a new rapidly expanding group of full-time academically-driven pediatric cardiologists, pediatric cardiac intensivists, and pediatric cardiac surgeons. A state-of-the-art brand-new 24-bed Pediatric Cardiac Intensive Care Unit, unique to the state, was recently opened.

The echocardiography laboratory, under the direction of Ernerio T. Alboliras, MD, FACC, FAAP, FASE, currently has six sonographers and performs more than 7,000 studies per year, including 650 fetals. All images are digitally archived. There is great opportunity to experience a broad spectrum of imaging congenital and acquired heart defects, from simple to complex; to include echocardiographic involvement during interventional catheterization, Hybrid operative procedures and open heart surgery. All aspects of ultrasound imaging – transthoracic, transesophageal, fetal, intracardiac, and stress – are performed. Tele-echocardiography transmission from other hospitals is routine. Ample opportunity for participation in echocardiography research and education is available.

REQUIREMENTS: Five years' experience in pediatric echocardiography, BCLS, RDMS or RDCS certification, ability to follow a consistent Pediatric Echo Lab imaging protocol, efficiently manage a busy Lab and work and communicate effectively with physicians, sonographers and other staff required. Strong congenital heart and previous supervisory experience in a Pediatric and Fetal Echocardiography Laboratory highly preferred.

HOSPITAL SUMMARY: St. Joseph's Hospital and Medical Center has been a symbol of quality healthcare in the Valley of the Sun for more than 110 years. With more than 740-beds and 5,200 employees, we are extremely proud to announce that we are the only Arizona hospital to be voted a "Best Place to Work in the Valley" six years in a row. We are also proud to be named a Top 25 Workplaces for Women, a top hospital by Ranking Arizona Magazine and routinely recognized as one of the country's best neuroscience centers by U.S. News & World Report. The hospital is part of Catholic Healthcare West, which has more than 40 hospitals in Arizona, California and Nevada.

For questions regarding The Scott and Laura Eller Congenital Heart Center please visit http://www.stjosephs-phx.org/Medical_Services/Congenital_Heart_Center/index.htm or email ernerio.alboliras@chw.edu

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early in their career to provide guidance on career choices, financial security, work-life balance and other practical guidelines for long-term success in cardiovascular medicine.

For more information, to download the program, to submit abstracts, or to register, go to: www.chop.edu/cardiology2010.

MRI Simulation of Blood Flow Helps Plan Child's Delicate Heart Surgery

Researchers at the Georgia Institute of Technology, collaborating with pediatric cardiologists and surgeons at The Children's Hospital of Philadelphia, have developed a tool for virtual surgery that allows heart surgeons to view the predicted effects of different surgical approaches. By manipulating three-dimensional cardiac magnetic resonance images of a patient's specific anatomy, physicians can compare how alternative approaches affect blood flow and expected outcomes, and can select the best approach for each patient before entering the operating room.

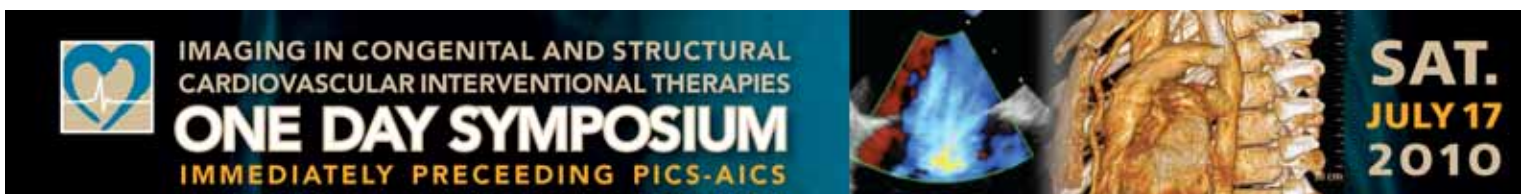
"This tool helps us to get the best result for each patient," said co-author Mark A. Fogel, MD, an Associate Professor of Cardiology and Radiology, and Director of Cardiac MRI at The Children's Hospital of Philadelphia. "The team can assess the different surgical options to achieve the best blood flow and the optimum mixture of blood, so we can maximize the heart's energy efficiency."

In the August issue of the *Journal of the American College of Cardiology: Cardiovascular Imaging*, the researchers describe the surgical planning methodology, detailing how the tool helped them to plan the surgery of a four-year-old girl who was born with just one functional ventricle, or pumping chamber, instead of two.

Two in every 1,000 babies in the United States are born with this type of single ventricle heart defect. These children typically suffer from low levels of oxygen in their tissues because their oxygen-rich and oxygen-poor blood mix in their one functional ventricle before being redistributed to their lungs and body.

To correct this, the children undergo a series of three open-heart surgeries – called the staged Fontan reconstruction – to reshape the circulation in a way that allows oxygen-poor blood to flow from the limbs directly to the lungs without going through the heart. While these vascular modifications can eliminate blood mixing and restore normal oxygenation levels, surgeons and cardiologists must ensure that the lungs will receive proper amounts of blood and nutrients after the surgery so that normal development occurs.

"Preoperatively determining the Fontan configuration that will achieve balanced blood flow to the lungs is very difficult and the wide variety and complexity of patients' anatomies requires an approach that is very specific and personalized," said Ajit Yoganathan, PhD, Regents' Professor in the Wallace H. Coulter Department of Biomedical Engineering at Georgia Tech and Emory University. "With our surgical planning framework, the physicians gain a better understanding of each child's unique



heart defect, thus improving the surgery outcome and recovery time."

The patient described in this paper, Amanda Mayer, age four, of Staten Island, NY, had previously undergone all three stages of the Fontan procedure at The Children's Hospital of Philadelphia, but developed severe complications. Her oxygen saturation was very low – only 72%, compared to normal levels of at least 95% – which indicated the possibility of abnormal connections between the veins and arteries in one of her lungs. Normally, the liver releases hormonal factors that prevent these abnormal connections, so the presence of the malformations indicated a low supply of hepatic blood to the lung.

To improve the distribution of these hormonal factors to both lungs, the surgeons needed to re-operate and reconfigure the patient's cardiovascular anatomy. Georgia Tech's surgical planning framework helped Thomas L. Spray, MD, Chief of the Division of Cardiothoracic Surgery at Children's Hospital, to determine the optimal surgical option.

"MRI acquires images of the child's heart without using radiation," said Spray. "Then we use the computerized technology to model different connections to simulate optimum blood flow characteristics, before we perform the surgery."

The image-based surgical planning consisted of five major steps: acquiring magnetic resonance images of the child's heart at different times in the cardiac cycle, 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 as feedback to the surgeon.

Fogel collected three different types of magnetic resonance images, and Yoganathan, along with graduate students Kartik Sundareswaran and Diane de Zélicourt, generated a three-dimensional model of the child's cardiovascular anatomy. From the model they reconstructed the three-dimensional pre-operative flow fields to understand the underlying causes of the malformations.

For this particular patient, the team saw a highly uneven flow distribution – the left lung was receiving about 70% of the blood pumped out by the heart, but only five percent of the hepatic blood. Both observations suggested left lung malformations, but closer examination of the flow structures in that particular patient revealed that the competition between different vessels at the center of the original Fontan connection effectively forced all hepatic factors into the right lung even though a vast majority of total cardiac output went to the left lung.

To facilitate the design of the surgical options that would correct this problem, Jarek Rossignac, PhD, a professor in Georgia Tech's School of Interactive Computing, developed Surgem, an interactive geometric modeling environment that allowed the surgeon to use both hands and natural gestures in three-dimensions to grab, pull, twist and bend a three-dimensional computer representation of the patient's anatomy. After analyzing

Pediatric Cardiology Faculty Position



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The Division of Pediatric Cardiology at Loyola University Chicago (LUC) Stritch School of Medicine's Department of Pediatrics is seeking a pediatric cardiologist to join its growing practice. The position requires excellent clinical skills in general pediatric cardiology. The ideal candidate should be BE/BC in pediatric cardiology with demonstrated expertise in multimodality noninvasive imaging of congenital and structural heart disease. Candidates should have excellent judgment, good work ethic, and interact well with peers, other medical and support personnel, and community physicians. Faculty also have important roles in student and resident education and have the opportunity to conduct research.

Based in the western suburbs of Chicago, Loyola University Health System (LUHS) is a quaternary care system with a 61-acre main medical center campus and 14 off-site facilities in Cook, Will and DuPage counties and the Gottlieb Memorial Hospital campus in Melrose Park. On the medical center campus is Loyola University Hospital, a 570-bed licensed facility; it houses a Level 1 Trauma Center, a Burn Center and the Ronald McDonald® Children's Hospital of Loyola University Medical Center. Also on campus are the Cardinal Bernardin Cancer Center, Loyola Outpatient Center, Center for Heart & Vascular Medicine, and Loyola Oral Health Center as well as the LUC Stritch School of Medicine, LUC Niehoff School of Nursing and a fitness center. LUHS is a nationally recognized leader in providing specialty and primary health-care services as well as in conducting groundbreaking research in treatment of heart disease, cancer, organ transplantation and neurological disorders.

Loyola's Ronald McDonald Children's Hospital, located in Loyola University Hospital, is a "hospital-within-a-hospital" and is comprised of 36 general inpatient, 20 newborn nursery, 14 pediatric intensive care, and 50 neonatal intensive care beds. The hospital is staffed by a full complement of pediatric subspecialty services and a 46-member residency program.

Please send a letter of interest and CV to: Joel Hardin, MD, Division Director of Pediatric Cardiology, jhardin@lumc.edu, or Holly Nandan, Director of Faculty Recruitment, hnandan@lumc.edu.

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The Loyola University Health System is an affirmative action/equal opportunity educator and employer. The University undertakes affirmative action to assure equal employment opportunity for underrepresented minorities, women, and persons with disabilities.



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Dear Colleagues,

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Almost all types of items are useful, but the most needed items are standard right heart catheters like GL's and Lehman's, and guidewires, especially exchange length "glide" or stiff wires.

Valve and angioplasty balloons 10-30 mm are also in very short supply.

I am happy to cover shipping costs.

Other folks doing foreign work are welcome to contact me since we may be able to trade or share material to mutual advantage...maybe share some stories as well.

Thanks as always for the support!

-Michael S. Womack MD

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the three-dimensional reconstruction of the failing cardiovascular geometry, the team considered three surgical options.

The research team then performed computational fluid dynamics simulations on all three options to investigate for each how well blood would flow to the lungs and the amount of energy required to drive blood through each connection design. These measures of clinical performance allowed the cardiologists and surgeons to conduct a risk/benefit analysis, which also included factors such as difficulty of completion and potential complications.

Of the three choices, Spray favored the option that showed a slightly higher energy cost, but exhibited the best performance with regards to hepatic factor distribution to the left and right lungs. Five months after the surgery, Mayer showed a dramatic improvement in her overall clinical condition and oxygen saturation levels, which increased from 72% to 94%. Mayer is breathing easier, and is now able to play actively like other children, according to her cardiologist, Donald Putman, MD, of Staten Island, NY.

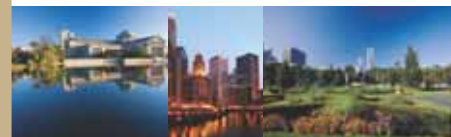
"The ability to perform this work is a team effort," Fogel added. "State-of-the-art three-dimensional cardiac MRI married to modern biomedical engineering and applied anatomy and physiology enabled this approach. With the advanced pediatric cardiothoracic surgery we have here at The Children's Hospital of Philadelphia, patients can benefit from this new method."

Additional authors on the paper include Shiva Sharma from Pediatric Cardiology Services, Kirk Kanter from the Division of Cardiothoracic Surgery at Emory University, and Fotis Sotiropoulos from the Department of Civil Engineering at the University of Minnesota.

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The **Division of Pediatric Cardiology at the University of Utah School of Medicine** and Primary Children's Medical Center is recruiting additional pediatric cardiologists with major interests in:

- **noninvasive imaging,**
- **transplant/heart failure, or**
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The candidate should be BC/BE in pediatric cardiology and have a strong clinical background with expertise and interest in at least one of the areas listed above. The division currently consists of 21 division members and has a very active, growing clinical program, an active, thriving fellowship training program, and a very active clinical research program; it is one of the participating centers in the Pediatric Heart Disease Clinical Research Network funded by the NIH. Protected time and mentoring for clinical research will be available within the Division for clinical research studies.

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Interested individuals should contact:

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Do you or your colleagues have interesting research results, observations, human interest stories, reports of meetings, etc. that you would like to share with the congenital cardiology community?

Submit your proposed article to Congenital Cardiology Today at: RichardK@CCT.bz

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