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Noncompaction Cardiomyopathy: A Not So Rare Cardiomyopathy in Children

Jeffrey A. Towbin, MD; Jason N. Johnson, MD; Gary S. Beasley, MD; Karine Guerrier, DO, MP

Characterized by excessive and unusual trabeculation of the mature left ventricle (LV), left ventricular noncompaction cardiomyopathy (LVNC) is thought to occur due to arrest of the final phase of cardiac development, the compaction phase, where the compact myocardium fully forms. A spongy morphologic appearance of the myocardium is typical, with abnormal trabeculations and intertrabecular recesses typically being most evident in the apical portion of the LV.^{1, 2} In 2006, the American Heart Association Scientific Statement on the classification of cardiomyopathies formally classified LVNC as its own disease entity.³ The European working group on myocardial disease, however, continues to consider LVNC as a trait rather than a specific disease, but recommends genetic analysis when a diagnosis is made. ^{4, 5} Multiple forms of LVNC occur, each with different features and outcomes.

Pathology of Left Ventricular Noncompaction

In the early embryo, the heart is a loose interwoven mesh of muscle fibers.⁶ The developing myocardium gradually condenses, and large spaces within the trabeculation meshwork disappear, condensing and compacting the ventricular myocardium and solidifying the endocardial surfaces. Trabecular compaction is normally more complete in the LV than in the right ventricular (RV) myocardium. The situation in which this compacting pathway fails is thought to be due to an arrest in endomyocardial morphogenesis and result in postnatal LV noncompaction.^{6,7} The gross pathological appearance of LVNC is characterized by numerous, excessively prominent trabeculations and deep intertrabecular recesses resembling RV endomyocardial morphology. Histologically, the recesses and their troughs are lined with endothelium, indicating that these recesses are not sinusoids. In some cases, zones of fibrosis and elastic tissue are found scattered on the endocardial surfaces with extension into the recesses. The abnormal endocardial fibrous and elastic tissue will result in abnormal mechanics, causing the apex and bases to rotate in the same direction, without the normal twisting contraction of the heart. 8-10 The endomyocardial morphology of LVNC lends itself to the development of mural thrombi within the recesses, which can embolize and cause stroke.¹¹⁻¹³ The coronary arterial circulation is usually normal; however, intramural perfusion could be adversely affected by the prominent trabeculations and intertrabecular recesses, particularly in the subendocardium. While the precise mechanism for malignant ventricular arrhythmias in LVNC patients is not known, impaired flow reserve, causing intermittent ischemia, has been proposed as having a role. In addition, myocardium around the deep intertrabecular recesses may serve as zones of slow conduction and substrate for reentry.

Incidence of Left Ventricular Noncompaction Cardiomyopathy

The incidence and prevalence of LVNC is uncertain because of changing diagnostic criteria and predilection for LV hypertrabeculation in certain populations¹⁴⁻²⁰ and absence of symptoms in many patients.²¹ In the 1990's, the reported prevalence of isolated LVNC was 0.05% of all adult echocardiograms in a large institution, whereas more recently the prevalence was reported as less than 0.14%. In contrast, Sandhu et al.¹⁴ demonstrated a 3.7% prevalence of definite or probable LVNC by echocardiography in adults with LV ejection fraction (EF) \leq 45% and a 0.26% prevalence for all patients referred for echocardiography. In a registry of pediatric cardiomyopathy patients, noncompaction was discovered in five.²²

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Clinical Features and Diagnosis of Left Ventricular Noncompaction

The clinical presentation of LVNC is highly variable, ranging from asymptomatic to end-stage heart failure, lethal arrhythmias, or thromboembolic events.^{2, 3, 12,13,23-25} LVNC may exist as isolated disease, but it may also be associated with hypertrophic and dilated cardiomyopathies (HCM; DCM).

In infancy, LVNC commonly develops with signs and symptoms of heart failure. Childhood LVNC can occur with heart failure, rhythm abnormalities, or sudden death.^{12,13, 16,21} Concurrent systemic disease has been reported with worse outcomes.² When patients receiving transplants were added, the five-year survival free of death or transplantation was 75%.²

The same specter of arrhythmia, heart failure, and thrombosis is described in adults. However, the majority of patients are asymptomatic and identified serendipitously on echocardiography. It may be advisable to use multi-modality imaging (valid echocardiographic criteria, speckled tracking, and cardiac magnetic resonance imaging [CMR]) to confirm the diagnosis based on the ratio of compacted and non-compacted layers and on the non-trabeculated mass.²⁶⁻²⁹

Patients with LVNC are known to develop ventricular arrhythmias including ventricular tachycardia (VT) and ventricular fibrillation (VF), atrial fibrillation (AF), or conduction abnormalities (sinus bradycardia or complete heart block). Ventricular pre-excitation is also common.^{2, 7, 26-32}

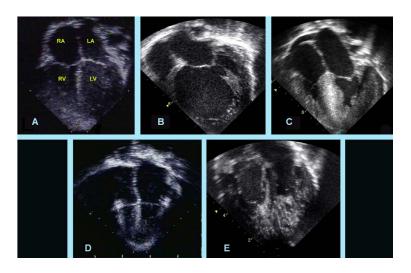


FIGURE 1

Echocardiographic Left Ventricular Noncompaction Phenotypes The heterogeneous features that cover the spectrum of patients with the different forms of LV noncompaction. Four-chamber views demonstrate normal atrial and ventricular sizes and thickness with a hypertrabeculated LV wall and apex (arrow a); normal atrial sizes with a dilated and hypertrabeculated LV wall and apex (arrow b); hypertrophic and hypertrabeculated ventricular walls and apex with a hypertrophic septum (arrow c); normal to small ventricular sizes with dilated atria bilaterally (arrows) and a hypertrabeculated LV (d); normal atrial sizes with severely hypertrabeculated LV and RV wall and apices (arrows e); normal atrial sizes with tricuspid atresia (no tricuspid valve) and a hypertrabeculated LV wall and apex (arrow f), and Ebstein's anomaly with LVNC, which demonstrates a normal left atrial size with a large right atrium, displaced tricuspid valve towards the RV (closed arrowhead), and a severely hypertrabeculated LV wall and apex (open arrowhead).

Inducibility of sustained VT during electrophysiological studies has demonstrated little value as a tool for risk stratification in LVNC.^{23,33} An electrocardiographic finding suggestive of potential for reentry is the presence of QRS notches (QRS fragmentation) or J waves (early repolarization).^{23, 24, 34,35} Additionally, the high proportion of patients without previously documented arrhythmias have the same amount of spontaneous arrhythmias (premature ventricular complexes [PVCS] or non-sustained VT) on Holter monitor as those who survived an arrest, suggesting that this might be a trigger for more serious events.

Subtypes of Left Ventricular Noncompaction

One of the important issues in the diagnosis and outcome of these patients, particularly in childhood, is the specific LVNC phenotype expressed, as each may have different outcomes. A special feature is that the phenotype may vary between DCM and HCM forms ("undulating phenotype") in which the heart changes from one form to the other, potentially on multiple occasions.² In these situations, the final "undulation" is typically an end-stage disease with severe heart failure. Subtypes of Left Ventricular Noncompaction (**Figure 1**)^{36,37} include the following:

 Isolated LVNC, in which abnormal trabeculations are associated with normal LV size, thickness, and systolic and diastolic function in the absence of other structural heart disease and with no evidence of arrhythmias (Figure 1A).

- Clinically, this subgroup appears to be benign during childhood and affects approximately 25%-35% of all subjects. These individuals do well unless they exhibit ventricular arrhythmias or if there is a family history of symptomatic LVNC. This subgroup, which is sometimes called the "benign form" of LVNC, usually undergoes yearly follow-ups in the outpatient clinic as infants and young children. As adolescents, they are seen every three years if there is no symptomatic change or family history of LVNC. These patients are not treated with medications or restricted from activities.^{7,26,32,36}

2. Isolated LVNC with arrhythmias (arrhythmogenic form of LVNC).

- These patients appear to have an elevated risk of sudden life-threatening events and require closer follow-up and therapeutic intervention. The arrhythmias noted include VT, VF, atrioventricular block, supraventricular tachycardia [SVT], and atrial fibrillation [AF].^{23-25,38} This subtype is currently designated as a form of arrhythmogenic cardiomyopathy (ACM).³⁸

- 3. The dilated cardiomyopathy form of LVNC clinically mimics DCM, with a dilated hypertrabeculated LV with depressed systolic function and appears to have worse outcomes than isolated DCM (Figure 1B).
 - The follow-up and treatment for these patients is similar to those with pure $\rm DCM.^{22,37}$
- The HCM form of LV noncompaction mimics HCM with LV hypertrophy, commonly with asymmetric septal hypertrophy, and a small, trabeculated LV cavity (Figure 1C).
 - Hypercontractile systolic function and diastolic dysfunction also occur. This form of LVNC appears to have worse outcome than in pure HCM.^{22,37}
- The mixed form of LVNC includes HCM with DCM, HCM or DCM with restrictive physiology (Figure 1C) and commonly has an "undulating phenotype" going from hypertrophic to dilated back to hypertrophic and commonly and finally ending at a DCM form of disease.
 - This subtype may be associated with neuromuscular

disease and hypotonia. Young children, particularly infants and neonates, can succumb to heart failure or arrhythmias, especially if they have metabolic derangement.^{2,11, 36} Electrocardiograms (ECGs) of these patients have the same abnormalities as the other forms. In some cases, extreme QRS forces are notable.

- 6. The restrictive form is a rare and clinically challenging subtype of LVNC because it mimics the clinical behavior of restrictive cardiomyopathy (RCM) with dilated atria and diastolic dysfunction associated with normal LV dimension, thickness, and systolic function (Figure 1D).³⁹
 - Like children with RCM, this subgroup is typically considered a transplant candidate early after diagnosis if they have presented with syncope as they are at risk for degeneration of tachyarrhythmia and conduction disease.
- In the biventricular cardiomyopathy form of LVNC in which both LV and RV are hypertrabeculated, systolic dysfunction and diastolic function are common (Figure 1E).
 Heart failure is commonly seen as well.³⁹
- In RV noncompaction, an uncommon form, the RV is hypertrabeculated while the LV appears normal.⁴⁰
 - RV filling can be marginalized, and arrhythmias may occur.
- In the congenital heart disease (CHD) form of LVNC, any form of CHD may be associated, but right-heart obstructive forms are most typical.¹⁵
 - The most common forms of CHD include atrial septal defects (ASD), ventricular septal defects (VSD), pulmonic stenosis (PS), pulmonary atresia, tricuspid atresia, Ebstein's anomaly, heterotaxy syndrome, and hypoplastic left heart syndrome (HLHS), among others.^{7,26,32,41}

Outcomes depend on the specific form of CHD, but it is noted that these patients can have worse postoperative outcomes than patients with the same CHD but without LVNC.

Imaging of Left Ventricular Noncompaction

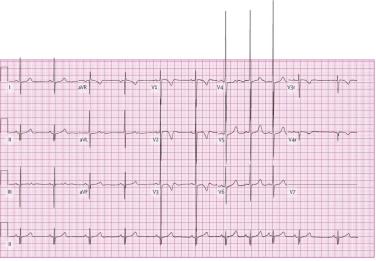
The diagnostic criteria for LVNC are based on imaging.^{18-20,22,27-29,38} Echocardiography has been the most common modality utilized to diagnose and describe LVNC (Figure 1 & Figure 2). The diagnostic criteria were recently revisited by some of the original authors, and the use of multi-modality imaging (the validated echocardiographic criteria, speckled tracking, and CMR) was suggested to confirm the diagnosis.^{18-20,22,27} The initial echocardiographic criteria were based on the ratio of noncompacted and compacted layers (which should be >2:1 in LVNC) and on measurement of the non-trabeculated mass versus the mass of the global LV with CMR.³⁸ Speckled tracking may reveal rigid body rotation, in an objective way, with the absence of the normal twist.^{8.10,42-45} These findings are present in children and their family members and is associated with lower functional status.^{8, 45}

Thuny et al compared two-dimensional echocardiographic images obtained at end-diastole and end-systole with the images obtained using CMR at end-diastole to validate the diagnosis of LVNC (**Figure 2**).²⁸ Sixteen adults (48 +/- 17 years) with LVNC underwent echocardiography and CMR within the same week and were compared to assess noncompaction in seventeen anatomical segments. All segments were analyzed using CMR, but only 87% could be analyzed using echocardiography. A two-layered structure was observed in 54% of patients by CMR and 43% by echocardiography. Therefore, CMR was thought to be superior to standard echocardiography in assessing the extent of noncompaction and providing supplemental morphological information; however, this has not been validated in children. CMR also assesses the presence of extensive fibrosis that correlates with late gadolinium enhancement (LGE).⁴⁶

Electrocardiography in Left Ventricular Noncompaction

The ECG for patients with LVNC is typically abnormal and, in infants and young children, commonly shows excessive voltage.^{2,32} Approximately 30% of these young subjects with LVNC have extreme mid-precordial voltages and mimic the ECG seen in Pompe disease (Figure 3). The childhood forms of LVNC may be associated with ventricular pre-excitation as well.

Adult patients often also have LV hypertrophy as well as atrioventricular and intraventricular conduction delay. Left bundle branch block (LBBB) is



mm/s 10 mm/mV 150 Hz 7.1.1 ISL 241 HD

FIGURE 2

Electrocardiogram in Left Ventricular Noncompaction ECG Note the prominent precordial voltage.

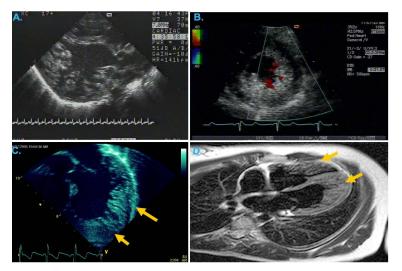


FIGURE 3

Imaging Approaches in Left Ventricular Noncompaction

A). Parasternal long axis echocardiogram of LVNC demonstrates the appearance of the spongiform myocardium/hypertrabeculation;
B). Parasternal short axis echocardiogram of LVNC demonstrates the appearance of a hypertrophied hypertrabeculated myocardium with color Doppler demonstrating blood in the intratrabecular recesses;
C). Apical four-chamber view with severe hypertrabeculation at the apex and posterior wall LVNC seen with colorized myocardium;
D). Cardiac MRI demonstrating biventricular noncompaction (arrows).

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- Michael G. Earing, MD, MS Healthcare Management

Member of IAC Echocardiography Board of Directors, Representing ACHA / Medical Director of the Chicago Adult Congenital Heart Disease Alliance

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often present.^{23,24} Early repolarization, as described by Haissaguerre and colleagues, was more often present in those who had a history of VT or VF, but also occurred in 23% of patients without such history.^{24,45} The same findings were reported by Steffel and colleagues.²³

Arrhythmias in Left Ventricular Noncompaction

Ventricular tachyarrhythmias, including cardiac arrest owing to VF, are reported in 38%-47% and sudden death in 13%-18% of adult patients with LVNC.^{24,25,33,47} Caliskan and coworkers investigated ICD therapy indications and outcomes in 77 patients with LVNC (mean age 40 +/-14 years), 44 (57%) of whom had an ICD implanted using standard ICD guidelines for non-ischemic cardiomyopathy. Implantation for secondary prevention occurred in 12 patients (seven with VF, five with sustained VT) and for primary prevention (heart failure or severe LV dysfunction) in 32 patients.⁴⁸ During a mean follow-up of 33 +/- 24 months, the patients presented with appropriate ICD shocks owing to sustained VT after a median of 6.1 months (range 1 to 16 months), including four of 32 (13%) of patients in the primary prevention group and four of 12 (33%) in the secondary prevention group (p=0.04). During a mean follow-up of four months (range 2-23 months), an additional nine patients had received inappropriate ICD therapy: six (19%) in the primary prevention group and three (25%) in the secondary prevention group. The relatively high percentage of appropriate shocks for sustained VT in this population suggest that patients with LVNC are at high-risk for sudden cardiac death and that implanting an ICD in these adult patients is appropriate, although no patients with LVNC were included in the previous trials that were the basis of the current guidelines. These observations support the current guidelines to implant an ICD for primary prevention based on the presence of heart failure or severe LV dysfunction in combination with other (presumed) high-risk factors in patients with LVNC. (Cl 14-

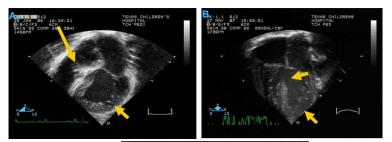




FIGURE 4

Left Ventricular Noncompaction Associated with Congenital Heart Disease

A). LVNC with tricuspid atresia. The top arrow demonstrates the tricuspid valve abnormality and the bottom arrow points to the apical hypertrabeculation

B). LVNC with a large ventricular septal defect. The top arrow demonstrates the ventricular septal defect and the bottom arrow points to the apical hypertrabeculation;

C). LVNC with Ebstein anomaly. The top arrow demonstrates the Ebstein's tricuspid valve abnormality and the bottom arrow points to the apical hypertrabeculation.

28%) (Figure 2A and Suppl. Figures 4A and 4B). The heart rhythm abnormalities were either reported at first presentation or occurred during the follow-up period. Interestingly, some families with distinct gene mutations showed predominantly one specific type of arrhythmia, such as sinus node disease in HCN4 gene carriers [51].⁴⁹⁻⁵³

Brescia et al evaluated two hundred forty-two children between January 1990 and January 2009 that were diagnosed with isolated left ventricular noncompaction.54 The mean age at diagnosis was 7.2±6.9 years, although 95 patients (39%) presented within the first year of life. Thirty-one (12.8%) of the children died and 13 (5.4%) received a transplant. One hundred fifty (62%) patients presented with or developed cardiac dysfunction. The presence of cardiac dysfunction was strongly associated with mortality (hazard ratio, 11; P<0.001). ECG abnormalities were present in 87%, with ventricular hypertrophy and repolarization abnormalities occurring most commonly. Repolarization abnormalities were associated with increased mortality (hazard ratio, 2.1; P=0.02). Eighty children (33.1%) had an arrhythmia and those with arrhythmias had increased mortality (hazard ratio, 2.8; P=0.002). Forty-two (17.4%) had ventricular tachycardia, with five presenting with resuscitated sudden cardiac death. There were 15 cases of sudden cardiac death in the cohort (6.2%). Nearly all patients with sudden death (14 of 15) had abnormal cardiac dimensions or cardiac dysfunction. No patient with normal cardiac dimensions and function without preceding arrhythmias died.

Howard et al evaluated data on 348 patients with LVNC and found that thirty-eight (11%) had ventricular pre-excitation on ECG, with 84% of those with pre-excitation and LVNC having cardiac dysfunction.⁵⁵ Kaplan-Meier analysis demonstrated significantly higher risk of developing significant dysfunction in patients with WPW and LVNC versus LVNC alone (hazard ratio 4.64 [2.79, 9.90]). Twelve patients (32%) underwent an ablation procedure with an acute success rate of 83%. Four patients with cardiac dysfunction were successfully ablated, with three demonstrating an improvement in cardiac function post ablation.

Clinical Genetics of Left Ventricular Noncompaction

Ichida and colleagues reported that 44% of isolated LVNC patients had inherited LVNC, with 70% having autosomal dominant (AD) and 30% having X-linked (XL) inheritance.³² This was confirmed by Hoedemakers and coworkers who showed by echocardiographic screening that in 194 family members from 50 LVNC probands, 2/3 of families had a familial cardiomyopathy; however, not only LVNC but HCM and DCM were also detected.⁵⁶ Pleiomorphism was confirmed by others.^{22,36,37,57} Nonpenetrance of detected genetic defects was high, which means that affected family members have to be followed over time. In XL recessive LVNC, female carriers have not been found to develop frank clinical disease and are echocardiographically normal. Consistent with XL inheritance, no male-to-male transmission of the disease occurs. Autosomal dominant inheritance occurs in some familial cases of LVNC without CHD and in most, if not all, cases associated with CHD. In some families with AD LVNC associated with CHD, affected members can be identified in whom no CHD is evident at the time of evaluation because their cardiac defects include minor forms of CHD, such as small ASDs, VSDs, or patent ductus arteriosus (PDA) that have spontaneously closed, along with other individuals with severe CHD, such as HLHS. Autosomal recessive (AR) and mitochondrial inheritance is seen in cases dominated by mitochondrial and metabolic derangement.^{11,37}

Pediatric Cardiologist

Orlando, FL, United States



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Molecular Genetics of Left Ventricular Noncompaction

Barth syndrome is a clinical association of myocardial dysfunction, cyclic neutropenia, cardiolipin deficiency, skeletal myopathy, abnormal mitochondria, organic aciduria (primarily 3-methylglutaconic aciduria), growth retardation, and cholesterol abnormalities.^{37,58-60} It is an XL disorder and has been thought to be allelic to several phenotypically different disorders on chromosome Xq28, such as LVNC and DCM.^{36,37,57,58} Pathogenic variants in the tafazzin gene (TAZ) were among the first to be identified in male patients and carrier females with isolated LVNC and with Barth syndrome, but not in the adult population with AD transmission. This gene codes a protein family called tafazzins that participate in the metabolism of cardiolipin, the signature phospholipid of mitochondria, and is also responsible for other forms of infantile cardiomyopathy. ⁵⁹⁻⁶¹

In AD LVNC, multiple genes have been identified. In general, genetic heterogeneity is found. Ichida et al identified pathogenic variants in α -dystrobrevin as causative in children and young adults with LVNC with or without CHD.⁶² Subsequently, pathogenic variants in sarcomere protein- and Z-line-encoding genes were identified, with mutations in β -myosin heavy chain (MYH7), α -cardiac actin (ACTC), cardiac troponin T (TNNT2), and ZASP/LBD3.63-68 Sarcomeric gene mutations in the LVNC population imply that LVNC is part of a broader spectrum of cardiomyopathies, including HCM, RCM, and DCM. In addition to sarcomere-encoding genes and cytoskeletal genes, there are pathogenic variants in ion channels, which have been shown to cause LVNC and rhythm disturbance. These include the sodium channel gene SCN5A, HCN4, transient receptor potential melastatin 4 (TRPM4) gene. This channel mediates a Ca21-activated nonselective cationic current (INSCca).468-470. In the heart, the TRPM4 channel represents the cardiac Ca2+-activated transient inward current (Iti) and plays a key role in the cardiac conduction system. 50-52, 69-71

Dystrophin is another cytoskeletal protein that has been associated with LVNC and boys with Duchenne and Becker muscular dystrophy.^{41,72,73} Skeletal muscle biopsy has, in some cases, identified mitochondrial abnormalities, suggesting a nuclear import protein is the primary abnormality.⁴² Mutation analysis of the mitochondrial genome has identified mutations as well. Consideration of other potential genetic causes needs to account for the known molecular defects resulting in congenital heart anomalies, as well as those molecular abnormalities resulting in diseases of myocardium itself.

It is notable that the same gene mutation can result in different phenotypes (DCM, HCM, or RCM), even in the same family. Pedigree analysis, family evaluation, and genetic counseling are necessary, in collaboration with the cardiologist. Genetic testing is recommended when a pathogenic variant is identified in the proband. In the 2019 HRS expert consensus statement on evaluation, risk stratification, and management of arrhythmogenic cardiomyopathy, Towbin and colleagues made a class I recommendation with level of evidence B-NR, stating "If the proband has a disease-causing gene variant, it is recommended that first-degree relatives of individuals with LVNC undergo clinical screening for the disease along with genetic counseling and genetic testing". In addition, they made a IIa B-NR recommendation and stated, "In individuals with the clinical diagnosis of pathologic LVNC, genetic counseling and genetic testing are reasonable for diagnosis and for gene-specific targeted cascade family screening."³⁸

PRDM16, a transcription factor, which has been implicated as a cause of LVNC in 1p36 syndrome patients and in dilated cardiomyopathy, promotes expression of genes required for growth of the compact

myocardium, while genes involved in trabecular growth are suppressed. Cellular respiration for the generation of ATP occurs predominantly using the aerobic pathway under these conditions. When PRDM16 is deleted, genes required for compact myocardium growth demonstrate a decreased level of expression, whereas those required for trabecular growth are released from PRDM16-regulated suppression. Wu et al performed gene expression analysis and suggested a possible shift away from aerobic toward anaerobic cellular respiration.^{74,75} Other studies have suggested that disturbances in the Notch pathway might be involved in LVNC development as well.

Therapy and Outcome

Heart Failure

Heart failure in LVNC reduces longevity.⁷⁶⁻⁸⁰ The specific therapy depends on the clinical echocardiographic or CMR findings. In patients with systolic dysfunction and heart failure, anti-congestive therapies identical to those used in patients with DCM are appropriate. In particular, angiotensin converting enzyme inhibitors (ACEi) and β -adrenergic blocking agents have been useful. Consideration for the use of sacubitril/valsartan, an Angiotensin II Receptor Blocker-Neprilysin Inhibitor Combination (ARNi) treatment, instead of the ACEi, has become increasingly popular in children. Diuretics may also be needed; however, among patients exhibiting findings more consistent with an HCM or diastolic dysfunction (heart failure with preserved EF; HFpEF) physiologic phenotype (HCM, RCM), β -blocker therapy alone may be more appropriate in children. In children and adults with the HCM form of LVNC and symptomatic, myectomy may be appropriate. Cardiac resynchronization therapy can be helpful, certainly for LBBB with septal flash. Finally, in patients with heart failure associated with any form of LVNC, mechanical circulatory support and transplant may be necessary.

Specific Conditions

In patients with LVNC and associated mitochondrial or metabolic dysfunction, some investigators add a "vitamin cocktail" to the cardiac therapy, with coenzyme Q10, carnitine, riboflavin, and thiamine commonly used alone or in combination. There is no good data to support this approach, however.

In patients having associated CHD, appropriate therapeutic approaches can include simple pharmacological therapy with diuretics for volume overload associated with left to right shunts, more complex pharmacological therapy for patients with restrictive physiology and pulmonary hypertension, or invasive therapy with catheter intervention or surgical repairs, depending on the lesions.

Antithrombotic Therapy

Anticoagulation is needed in patients with depressed LV function in the presence of AF. The CHADS2-VASC score, which is widely used by adult medicine clinicians to assess the risk of cardioembolic event among patients with AF, may be helpful. Previous studies have shown its ability to identify low-risk individuals who can be managed without antithrombotic therapy, as well as to assess the risk of stroke among others and this might be helpful in decision-making, also when AF is not present. When there is evidence of thrombi in the LV, oral anticoagulation with anti-vitamin K antagonists or heparin is warranted. To date, we have no evidence for the efficacy of the direct oral anticoagulants in patients





ACUTE CARE CARDIOLOGIST

As one of the nation's leading pediatric health care systems, the Heart Institute at UPMC Children's Hospital of Pittsburgh provides a comprehensive service line; with CT surgery, interventional cardiology, electrophysiology, advanced imaging (MRI/CT), heart failure/PHT, ACHD, acute care, perinatal and preventative cardiology programs. <u>The</u> <u>Heart Institute</u> provides comprehensive pediatric and adult congenital cardiovascular services to the tri-state region and consists of 35 pediatric cardiologists, 5 pediatric cardiothoracic surgeons, 8 pediatric cardiac intensivists and 9 cardiology fellows along with 19 physician extenders and a staff of over 100. We are honored to be ranked **#2 nationally** and **#1 in Pennsylvania** for pediatric cardiology and heart surgery by *U.S. News and World Report*. Our Cardiac surgical program is one of the top in the country, having held a 3-star rating from Society of Thoracic Surgery (STS) consistently over the years.

The Division of Cardiology within the Heart Institute of UPMC Children's Hospital of Pittsburgh / University of the Pittsburgh School of Medicine is recruiting a full -time board certified pediatric cardiologist to join our robust program with expertise or subspecialty training in inpatient hospital care. We are seeking an exceptional individual with proficient skills in general pediatric cardiology, with a focus on acute care. Our Acute Care Unit, lead by Dr Evonne Morell, is staffed by general cardiologists and advanced practice providers, and has a long history of active participation in national QI initiatives.

This position comes with a competitive salary and faculty commensurate with experience and qualifications at the University of Pittsburgh School of Medicine. The University of Pittsburgh is an Equal Opportunity/Affirmative Action Employer. Interested individuals should forward a letter of intent curriculum vitae, and three letters of reference. Informal inquiries are also encouraged.

Contact information:

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with LVNC.^{81,82} In children, the use of aspirin daily has been promoted by some investigators.²¹ In the 2019 HRS expert consensus statement on evaluation, risk stratification, and management of arrhythmogenic cardiomyopathy, Towbin and colleagues made a class I recommendation with level of evidence B-NR, stating "Anticoagulation is recommended in individuals with LVNC with atrial fibrillation and in those with previous embolic events."³⁸ In addition, they made a IIb B-NR recommendation and stated "Anticoagulation may be reasonable in individuals with LVNC with evidence of ventricular dysfunction."

Arrhythmias

In patients with any form of LVNC who have arrhythmias or syncope, prolonged ambulatory monitoring or electrophysiological study may be warranted, and device therapy (pacing, ICD, or cardiac resynchronization) may be indicated. Catheter ablation is appropriate for WPW syndrome as well as for other arrhythmias, including atrial fibrillation. It may be challenging to address ventricular arrhythmias with catheter ablation, but this has been done as well. In adults and a small cohort of at-risk children, primary prevention with an ICD is commonly considered.

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References

- Engberding R, Yelbuz TM, Breithardt G. Isolated noncompaction of the left ventricular myocardium: a review of the literature two decades after the initial case description. Clin Res Cardiol. 2007; 96: 481–488.
- Pignatelli RH, McMahon CJ, Dreyer WJ, et al. Clinical characterization of left ventricular noncompaction in children. A relatively common form of cardiomyopathy. Circulation. 2003; 108: 2672-2678.
- Maron BJ, Towbin JA, Thiene G, et al. Contemporary definitions and classification of the cardiomyopathies: an American Heart Association Scientific Statement from the Council on Clinical Cardiology, Heart Failure and Transplantation Committee; Quality of Care and Outcomes Research and Functional Genomics and Translational Biology Interdisciplinary Working Groups; and Council on Epidemiology and Prevention. Circulation. 2006; 113: 1807-1816.
- Elliott P, Andersson B, Arbustini E, et al. Classification of the cardiomyopathies: a position statement from the European Society of Cardiology Working Group on Myocardial and Pericardial Diseases. Eur Heart J. 2008; 29: 270-276.
- Rapezzi C, Arbustini E, Caforio AL, et al. Diagnostic work-up in cardiomyopathies: bridging the gap between clinical phenotypes and final diagnosis. A position statement from the ESC Working Group on Myocardial and Pericardial Diseases. Eur Heart J. 2013; 34: 1448-1458.
- 6. Sedmera D, McQuinn T. Embryogenesis of the heart muscle. Heart Fail Clin. 2008; 4: 235-238.
- Chin TK, Perloff JK, Williams RG, et al. Isolated noncompaction of left ventricular myocardium. A study of eight cases. Circulation. 1990; 82: 507-513.

- Kauer F, Geleijnse ML, van Dalen BM. Role of left ventricular twist mechanics in cardiomyopathies, dance of the helices. World J Cardiol. 2015; 7: 476-482.
- Peters F, Khandheria BK, Libhaber E, et al. Left ventricular twist in left ventricular noncompaction. Eur Heart J Cardiovasc Imaging. 2014; 15: 48-55.
- Sabatino J, Di Salvo G, Krupickova S, et al. Left ventricular twist mechanics to identify left ventricular noncompaction in childhood. Circulation: Cardiovascular Imaging. 2019;12. doi.org/10.1161/ CIRCIMAGING.118.007805.
- Scaglia F, Towbin JA, Craigen WJ, et al. Clinical spectrum, morbidity, and mortality in 113 pediatric patients with mitochondrial disease. Pediatrics. 2004; 114: 925-931.
- 12. Stöllberger C, Blazek G, Dobias C, et al. Frequency of stroke and embolism in left ventricular hypertrabeculation/noncompaction. Am J Cardiol. 2011; 108: 1021-1023.
- Greutmann M, Mah ML, Silversides CK, et al. Predictors of adverse outcome in adolescents and adults with isolated left ventricular noncompaction. Am J Cardiol. 2012; 109: 276-281.
- Sandhu R, Finkelhor RS, Gunawardena DR, Bahler RC. Prevalence and characteristics of left ventricular noncompaction in a community hospital cohort of patients with systolic dysfunction. Echocardiography 2008; 25: 8-12.
- Hughes ML, Carstensen B, Wilkinson JL, Weintraub RG. Angiographic diagnosis, prevalence and outcomes for left ventricular noncompaction in children with congenital cardiac disease. Cardiol Young. 2007; 17: 56-63.
- Aras D, Tufekcioglu O, Ergun K, et al. Clinical features of isolated ventricular noncompaction in adults long-term clinical course, echocardiographic properties, and predictors of left ventricular failure. J Card Fail 2006; 12: 726–733.
- 17. Kovacevic-Preradovic T, Jenni R, Oechslin EN, et al. Isolated left ventricular noncompaction as a cause for heart failure and heart transplantation: a single center experience. Cardiology 2009; 112: 158-164.
- Kohli SK, Pantazis AA, Shah JS, Adeyemi B, Jackson G, McKenna WJ, Sharma S, Elliott PM. Diagnosis of left-ventricular noncompaction in patients with left-ventricular systolic dysfunction: time for a reappraisal of diagnostic criteria? Eur Heart J. 2008; 29: 89-95.
- Gati S, Rajani R, Carr-White GS, Chambers JB. Adult left ventricular noncompaction: reappraisal of current diagnostic imaging modalities. JACC Cardiovasc Imaging. 2014; 7: 1266-1275.
- 20. Gati S, Chandra N, Bennett RL, et al. Increased left ventricular trabeculation in highly trained athletes: do we need more stringent criteria for the diagnosis of left ventricular non-compaction in athletes? Heart. 2013; 99: 401-408.
- Caliskan K, Michels M, Geleijnse ML, van Domburg RT, van der Boon R, Balk AH, Simoons ML. Frequency of asymptomatic disease among family members with noncompaction cardiomyopathy. Am J Cardiol. 2012;110: 1512-1517.
- 22. Jefferies JL, Wilkinson JD, Sleeper LA, et al. Cardiomyopathy phenotypes and outcomes for children with left ventricular myocardial noncompaction: Results from the Pediatric Cardiomyopathy Registry. J Card Fail. 2015; 21: 877-884.
- Steffel J, Kobza R, Oechslin E, et al. Electrocardiographic characteristics at initial diagnosis in patients with isolated left ventricular noncompaction. Am J Cardiol. 2009; 104: 984-989.
- 24. Caliskan K, Ujvari B, Bauernfeind T, et al. The prevalence of early repolarization in patients with noncompaction cardiomyopathy presenting with malignant ventricular arrhythmias. J Cardiovasc Electrophysiol. 2012; 23: 938-944.
- 25. Celiker A, Ozkutlu S, Dilber E, Karagöz T. Rhythm abnormalities in children with isolated ventricular noncompaction. Pacing Clin Electrophysiol. 2005; 28: 1198-1202.
- 26. Oechslin E, Jenni R. Left ventricular non-compaction revisited: a

distinct phenotype with genetic heterogeneity? Eur Heart J. 2011; 32:1446-1456.

- Punn R, Silverman NH. Cardiac segmental analysis in left ventricular noncompaction: experience in a pediatric population. J Am Soc Echocardiogr. 2010; 23: 46-53.
- Thuny F, Jacquier A, Jop B, et al. Assessment of left ventricular noncompaction in adults: side-by-side comparison of cardiac magnetic resonance imaging with echocardiography. Arch Cardiovasc Dis. 2010; 103: 150-159.
- Rao K, Bhaskaran A, Choudhary P, Tan TC. The role of multimodality imaging in the diagnosis of left ventricular noncompaction. Eur J Clin Invest. 2020; 00:e13254.
- Howard TS, Valdes SO, Hope KD, et al. Association of Wolff-Parkinson-White with left ventricular noncompaction cardiomyopathy in children. J Card Fail 2019; 25: 1004-1008.
- 31. Yaksh A, Haitsma D, Ramdjan T, et al. Unexpected finding in an adult with ventricular fibrillation and an accessory pathway: Non-compaction cardiomyopathy. Neth Heart J. 2014; 22: 182-185.
- Ichida F, Hamamichi Y, Miyawaki T, et al. Clinical features of isolated noncompaction of the ventricular myocardium: Long-term clinical course, hemodynamic properties and genetic background. J Am Coll Cardiol. 1999; 34: 233-240.
- Caliskan K, Kardos A, Szili-Torok T. Empty handed: A call for an international registry of risk stratification to reduce the 'sudden-ness' of death in patients with non-compaction cardiomyopathy. Europace 2009; 11: 1138-1139.
- Das MK, Maskoun W, Shenet C, et al. Fragmented QRS on twelvelead electrocardiogram predicts arrhythmic events in patients with ischemic and nonischemic cardiomyopathy. Heart Rhythm. 2010; 7: 74-80.
- Cetin MS, Cetin EHO, Canpolat U, et al. Usefulness of fragmented QRS complex to predict arrhythmic events and cardiovascular mortality in patients with noncompaction cardiomyopathy. Am J Cardiol. 2016; 117: 1516-1523.
- 36. Towbin JA. Left ventricular noncompaction: a new form of heart failure. Heart Fail Clin. 2010; 6: 453-469.
- Towbin JA, Jefferies JL. Cardiomyopathies due to left ventricular noncompaction, mitochondrial and storage diseases, and inborn errors of metabolism. Circ Res. 2017; 121: 838-854.
- Towbin JA, McKenna WJ, Abrams DJ, et al. 2019 HRS Expert consensus statement on evaluation, risk stratification, and management of arrhythmogenic cardiomyopathy. Heart Rhythm 2019; 16: e373-e407.
- 39. Miura F, Shimada J, Kitagawa Y, et al. MYH7 mutation identified by next-generation sequencing in three infant siblings with bi-ventricular noncompaction presenting with restrictive hemodynamics: A report of three siblings with a severe phenotype and poor prognosis. J Cardiol Cases. 2019; 19: 140-143.
- Rangathan A, Ganesan G, Sangareddi V, Pillai AP, Ramasamy A. Isolated noncompaction of right ventricle – a case report. Echocardiography. 2012; 29: E169– E172.
- Finsterer J, Stöllberger C, Fazio G. Neuromuscular disorders in left ventricular hypertrabeculation/noncompaction. Curr Pharm Des. 2010; 16: 2895-2904.
- van Dalen BM, Snelder SM, Geleijnse ML. Left ventricular twist: An often ignored but crucial determinant of left ventricular function. J Am Coll Cardiol. 2018; 71: 584.
- Stokke TM, Hasselberg NE, Smedsrud MK, et al. Function: comparison between ejection fraction and strain. J Am Coll Cardiol. 2017; 70: 942-954.
- 44. van Dalen BM, Caliskan K, Soliman OI, et al. Diagnostic value of rigid body rotation in noncompaction cardiomyopathy. J Am Soc Echocardiogr. 2011; 24: 548-555.
- 45. Udink ten Cate FE, Schmidt BE, Lagies R, et al. Reversed apical rotation and paradoxical increased left ventricular torsion in children

with left ventricular non-compaction. Int J Cardiol. 2010; 145: 558-559.

- 46. Wan J, Zhao S, Cheng H, et al. Varied distributions of late gadolinium enhancement found among patients meeting cardiovascular magnetic resonance criteria for isolated left ventricular non-compaction. J Cardiovasc Magn Reson. 2013; 15: 20. doi:10.1186/1532-429X-15-20.
- 47. Steffel J, Duru F. Rhythm disorders in isolated left ventricular noncompaction. Ann Med. 2012; 44: 101-108.
- 48. Kayvanpour E, Sedaghat Hamedani F, Gi W-T, et al. Clinical and genetic insights into non-compaction: a meta-analysis and systematic review on 7598 individuals. Clin Res Cardiol. 2019; 108: 1297–1308.
- Oechslin EN, Attenhofer Jost CH, Rojas JR, Kaufmann PA, Jenni R (2000) Long-term follow-up of 34 adults with isolated left ventricular noncompaction: a distinct cardiomyopathy with poor prognosis. J Am Coll Cardiol. 36: 493–500.
- Schweizer PA, Koenen M, Katus HA, Thomas D. A distinct cardiomyopathy: HCN4 syndrome comprising myocardial noncompaction, bradycardia, mitral valve defects, and aortic dilation. J Am Coll Cardiol 2017; 69: 1209–1210.
- Schweizer PA, Schröter J, Greiner S, et al. The symptom complex of familial sinus node dysfunction and myocardial noncompaction is associated with mutations in the HCN4 channel. J Am Coll Cardiol. 2014; 64: 757-767.
- 52. Milano A, Vermeer AMC, Lodder EM, et al. HCN4 mutations in multiple families with bradycardia and left ventricular noncompaction cardiomyopathy. J Am Coll Cardiol. 2014; 64: 745-756.
- 53. Towbin JA. Ion Channel dysfunction associated with arrhythmia, ventricular noncompaction, and mitral valve prolapse: A new overlapping phenotype. J Am Coll Cardiol. 2014; 64: 768-771.
- 54. Brescia ST, Rossano JW, Jefferies JL, et al. Mortality and sudden death in pediatric left ventricular noncompaction in a tertiary referral center. Circulation 2013; 127: 2202-2208.
- 55. Howard TS, Valdes SO, Hope KD, et al. Association of Wolff-Parkinson-White with left ventricular noncompaction cardiomyopathy in children. J Card Fail. 2019; 25: 1004-1008.
- 56. Hoedemaekers YM, Caliskan K, Michels M, et al., The importance of genetic counseling, DNA diagnostics, and cardiologic family screening in left ventricular noncompaction cardiomyopathy. Circ Cardiovasc Genet. 2010; 3: 232-239.
- 57. Towbin JA, Lorts A, Jefferies JL. Left ventricular noncompaction cardiomyopathy. Lancet. 2015; 386: 813-825.
- 58. Aprikyan AA, Khuchua Z. Advances in the understanding of Barth syndrome. Br J Haematol. 2013; 161: 330-338.
- 59. Chin MT, Conway SJ. Role of tafazzin in mitochondrial function, development and disease. J Dev Biol. 2020; 8: E10.
- Bertero E, Kutschka N, Maack C, Dudek J. Cardiolipin remodeling in Barth syndrome and other hereditary cardiomyopathies. Biochim Biophys Acta Mol Basis Dis. 2020;1866: 165803.
- Schlame M, Xu Y. The function of tafazzin, a mitochondrial phospholipid-lysophospholipid acyltransferase. J Mol Biol. 2020. Mar 29:S0022-2836(20)30259-X. doi: 10.1016/j.jmb.2020.03.026. Online ahead of print.
- 62. Ichida F, Tsubata S, Bowles KR, et al. Novel gene mutations in patients with left ventricular noncompaction or Barth syndrome. Circulation. 2001; 103:1256-1263.
- 63. Hoedemaekers YM, Caliskan K, Majoor-Krakauer D, et al. Cardiac beta-myosin heavy chain defects in two families with non-compaction cardiomyopathy: Linking non-compaction to hypertrophic, restrictive, and dilated cardiomyopathies. Eur Heart J 2007; 28: 2732-2737.
- Teekakirikul P, Kelly MA, Rehm HL, Lakdawala NK, Funke BH. Inherited cardiomyopathies: molecular genetics and clinical genetic testing in the postgenomic era. J Mol Diagn. 2013; 15: 158-170.
- 65. Klaassen S, Probst S, Oechslin E, et al. Mutations in sarcomere



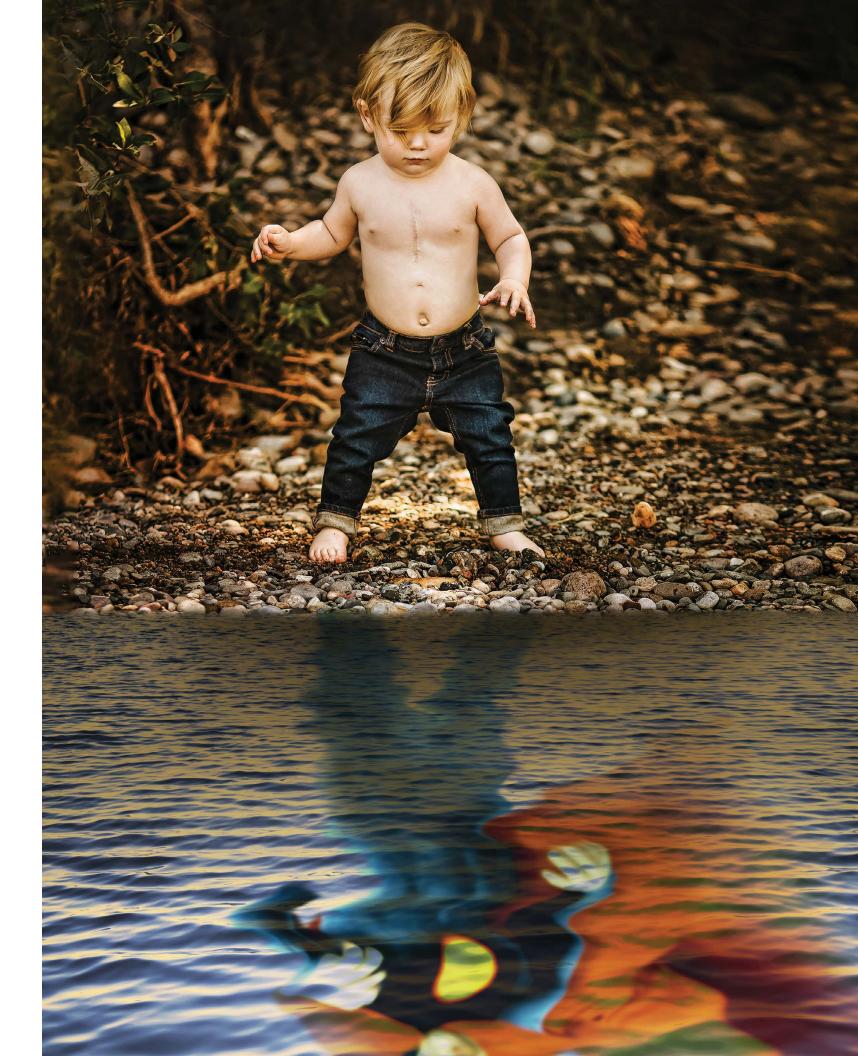




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Pediatric Cardiologist Wichita, Kansas

The Ward Family Heart Center at Children's Mercy Kansas City seeks a candidate at the assistant or associate professor level who would join our team as a pediatric cardiologist based at the CMKC multi-specialty clinic in Wichita, KS. The successful candidate would join an existing group of 28 cardiologists (25 in Kansas City, 2 in Wichita, KS and 1 in Topeka, KS), 3 CV surgeons, 30 APNs. Our Wichita office consists of two cardiologists, an APRN and dedicated sonographers and nursing staff. The position has minimal inpatient duties but will require monthly travel to support our outreach clinics in central and western Kansas. Experience and interest in outpatient cardiology and echocardiography is a must. There are opportunities also for development of a Fetal Echocardiography program in the community if interested. Trainees in their final year are welcome to apply.

Candidates must be board-certified or board-eligible in Pediatric Cardiology. Strong communication skills are key. There are opportunities for teaching medical students and residents with faculty appointment at the University of Kansas School of Medicine – Wichita campus. Salary and academic rank are commensurate with experience.

Wichita is a prosperous, thriving community with a metropolitan population of 650,000, the largest city in Kansas. Located 180 miles southwest of Kansas City, the city is known for its central role in American aviation design and manufacturing. It is an affordable and very livable city of historic neighborhoods, highly-rated schools and several colleges and universities.

Our Heart Center serves a population of over 5 million in the heart of the U.S.A. We perform over 500 cardiac operations, 600 cardiac catheterizations including over 200 invasive EP procedures, 18,000 outpatient visits, and more than 20,000 echocardiograms annually. Our two state-of the art catheterization labs are both hybrid labs and equipped with the latest 3D imaging and EP technology. Telehealth is available and facilitates our outreach clinics. We have video-conferencing capabilities that are routinely used by providers from distant locations to dial into our conferences for patient care and education. In 2021, the Ward Family Heart Center program was ranked # 24 nationally by USNWR.

Our Kansas City-based super-specialty resources include Electrophysiology (which includes Clinical EP, pacing and Genetic Arrhythmia), Cardiac Transplantation/Heart Failure, Interventional Cardiology and Advanced Cardiac Imaging (fetal echo, 3D echo, trans-esophageal echo, CT, MRI and 3D printing). We also provide specialized, team-based care in Fetal Cardiology (with on-site delivery services for high-risk neonates in Kansas City), Interstage Monitoring (CHAMP), Preventive Cardiology, Cardiac Genetics, Cardio-oncology, Single Ventricle Survivorship, Pulmonary Hypertension, a dedicated POTS clinic and Cardiac Neurodevelopmental Services.

The successful applicant must share our unwavering commitment to excellence, integrity, collegiality, antiracism, and respect for inclusion of individuals with diverse backgrounds.

Please submit CV and cover letter to:

https://faculty-childrensmercykc.icims.com/jobs/18126/physician/job

Or, for additional information please contact: Stephen Kaine, MD Interim Co-Director, The Ward Family Heart Center Children's Mercy Kansas City physicianjobs@cmh.edu

Children's Mercy Kansas City is an independent, non-profit, 390-bed pediatric health system, providing over half a million patient encounters each year for children from across the country. Children's Mercy is ranked by U.S. News & World Report in nine specialties. We have received Magnet® recognition five times for excellence in nursing services. In affiliation with the University of Missouri-Kansas City and the University of Kansas, our faculty of nearly 800 pediatric specialists and researchers are actively involved in clinical care, pediatric research and educating the next generation of pediatricians and pediatric subspecialists.

The Children's Mercy Research Institute (CMRI) integrates research and clinical care with nationally recognized expertise in genomic medicine, precision therapeutics, population health and health care innovation. In 2021, the CMRI moved into a nine story, 375,000 square foot space emphasizing a translational approach to research in which clinicians and researchers work together to accelerate the pace of discovery that enhances care. We invite you to visit Children's Mercy Hospital virtually at <u>cmkc.link/PhysicianTour</u>

For more information about Children's Mercy Kansas City and about Kansas City itself, visit <u>cmkc.link/TakeYourPlace</u>.

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protein genes in left ventricular noncompaction. Circulation. 2008; 117: 2893-901.

- Takasaki A, Hirono K, Hata Y, et al. Sarcomere gene variants act as a genetic trigger underlying the development of left ventricular noncompaction. Pediatr Res. 2018; 84: 733–742.
- Wang C, Hata Y, Hirono K, et al. A wide and specific spectrum of genetic variants and genotype–phenotype correlations revealed by next-generation sequencing in patients with left ventricular noncompaction. J Am Heart Assoc. 2017; 6: e006210. doi: 10.1161/ JAHA.117.006210.
- 68. van Waning JI, Caliskan K, Hoedemaekers YM, et al. Genetics, clinical features, and long-term outcome of noncompaction cardiomyopathy. J Am Coll Cardiol. 2018; 71: 711-722.
- Shan L, Makita N, Xing Y, et al. SCN5A variants in Japanese patients with left ventricular noncompaction and arrhythmia. Mol Genet Metab. 2008; 93: 468-474.
- Saito Y, Nakamura K, Nishi N, et al. TRPM4 mutation in patients with ventricular noncompaction and cardiac conduction disease. Circ Genom Precis Med. 2018; 11: e002103. doi.org/10.1161/ CIRCGEN.118.002103.
- Abriel H, Syam N, Sottas V, Amarouch MY, Rougier JS. TRPM4 channels in the cardiovascular system: physiology, pathophysiology, and pharmacology. Biochem Pharmacol. 2012; 84: 873–881.
- Finsterer J, Stöllberger C, Towbin JA. Left ventricular noncompaction cardiomyopathy: cardiac, neuromuscular, and genetic factors. Nat Rev Cardiol. 2017; 14:224–237. doi: 10.1038/ nrcardio.2016.207.
- 73. Finsterer J, Stöllberger C. Primary myopathies and the heart. Scand Cardiovasc J 2008; 42: 9-24.
- 74. Wu T, Liang Z, Zhang Z, et al. PRDM16 Is a compact myocardiumenriched transcription factor required to maintain compact myocardial cardiomyocyte identity in left ventricle. Circulation 145:586–602. 2022. PMID: 35188799.
- Mably JD, Wu JC, Wang D-Z. New insights into the molecular underpinnings of LVNC. Circulation 2022; 145(8): 603-605, DOI: (10.1161/CIRCULATIONAHA.121.058371).
- Vaidya VR, Lyle M, Miranda WR, et al. Long-term survival of patients with left ventricular noncompaction. J Am Heart Assoc. 2021;10:e015563. DOI: 10.1161/JAHA.119.015563.
- Purevjav E, Chintanaphol M, Orgil B-O, Alberson NR, Towbin JA. Left ventricular noncompaction cardiomyopathy: From clinical features to animal modeling. 2022. DOI: http://dx.doi.org/10.5772/ intechopen.101085.
- Rohde S, Muslem R, Kaya E, Dalinghaus M, van Waning JI, Majoor-Krakauer DF, Towbin JA, Caliskan K. State-of-the art review: Noncompaction cardiomyopathy in pediatric patients. Heart Fail Rev. 2022 Jan;27(1):15-28. Doi: 10.1007/s10741-021-10089-7.
- Femia G, Semsarian C, Ross SB, Celermajer D, Puranik R. Left ventricular non-compaction: Review of the current diagnostic challenges and consequences in athletes. Medicina 2020, 56(12), 697; https://doi.org/10.3390/medicina56120697.
- Lorca R, Martín M, Pascual I, Astudillo A, Díaz Molina B, Cigarrán H, Cuesta-Llavona E, Avanzas P, Rodríguez Reguero JJ, Coto E, Morís C, Gómez J. Characterization of Left Ventricular Non-Compaction Cardiomyopathy. J Clin Med. 2020 Aug 5;9(8):2524. doi: 10.3390/ jcm9082524. PMID: 32764337; PMCID: PMC7464545.
- Stöllberger C, Wegner C, Finsterer J. CHADS2- and CHA2DS2VASc scores and embolic risk in left ventricular hypertrabeculation/ noncompaction. J Stroke Cerebrovasc Dis. 2013; 22: 709-712.
- Berkovitch A, Mazin I, Younis A, et al. CHA2DS2-VASc score performance to predict stroke after acute decompensated heart failure with and without reduced ejection fraction. Europace. 2019; 21: 1639-1645.



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It Takes a Team - The Pivotal Role of Allied Health Professionals

Kamel Shibbani

This month's article from The PICS Society highlights the efforts of the entire cath team - namely the highly skilled and hard-working nurses, radiologic technologists, nurse practitioners, physician assistants and all our other invaluable allied health care providers. We were fortunate enough to sit down with four who represent quintessential components of the cath team: Sharon Cheatham, PhD, ACNP-BC, FPICS (Chair of the PICS Nurses and Allied Health Professionals Committee), Emily Kish, BSN, RN (Cath Lab Assistant Nurse Manager at University Hospitals Rainbow Babies and Children's Hospital), Jason Swinning, RT(R)(CI), RCIS (Radiologic Technologist and Imaging Specialist at Nationwide Children's Hospital) and Kathleen Nolan, RT(R)(CI) (Radiologic Technologist and Imaging Specialist at Rush University).

KS: Thank you for taking time for today's discussion. We want to highlight the essential roles that allied health care providers play in the cath lab and to stress that the cath team is truly a **team**. A team that, for it to function properly, relies on more than physicians. We're hoping you can talk about the pivotal role your professions play in the cath lab, and how the PICS Society can help bring your contributions to the forefront. *Let's talk a bit about the roles allied health care professionals perform in the cath lab. SC:* Thank you to everyone here! My name is Sharon Cheatham, and I have more than 20 years' experience in interventional cardiology. I actually helped with the first PICS meeting in Boston – we had about 86 attendees! Things have changed quite a bit through the years!

Regarding the role of the allied health professionals, as they say, "It takes a village!". Well, for the cath lab, it takes a team. We spend a lot of time before the patient is brought into the lab where we do intensive and detailed huddles. We make sure the entire team is informed, from patient history to previous procedures to the anesthesia plan, as well as catheters and devices that may be used. All of this is to ensure the best possible patient outcome.



Emily Kish



Dr. Sharon Cheatham

Jason Swinning



Kathleen Nolan



Dr. Kamel Shibanni

EK: Talking about a team, both Jason and I work in pediatric cath labs that greatly focus on teamwork and being comfortable playing different roles within the team. I think it's very important to have a physician who can embrace the mentality that it doesn't matter what the credentials are behind a name because very often our roles are interchangeable. We also bring to the table (as nurses, techs and NP's) diversity in training and skill sets. You can have staff with neonatal ICU experience, others with adult

experience, and rad techs with CT and 3D rendering experience. This allows the team to take care of everyone from patients who are just born to 87-year-old patients with Congenital Heart Disease. It is very important to use that experience and lean in on each other's skill sets.

JS: I couldn't agree more. As a matter of fact, if we have a job opening, we post it for either a nurse or a rad tech and simply take the best candidate out of the pool of applicants.

KN: In today's lab, technologists can be brought in wherever necessary! Data collection for research, scrubbing in and assisting on cases, preparing for cases, hemodynamics monitoring, assisting nurses that are scrubbed in, really anything that's needed.

"I think the more involved staff can be, that will only lead to better patient outcomes because everyone is working together as one cohesive team."

Jason Swinning, RT(R)(CI)

KS: Are there any changes that you can think of that can promote your roles and help with patient care?

JS: One of the things we do well, and can build on, is involving all of our staff and making sure that all team members are engaged in the procedure. For example, staff interested and trained in 3D reconstruction can do that while the physician scrubbed in is getting ready for the next step. We've moved away from the traditional roles of a circulating nurse only opens packages or a scrub tech that only wipes wires. I think the more involved staff is in the procedures, the better patient outcomes we will see because everyone is working together as one cohesive team. **EK:** I completely agree with that! I would also emphasize the need to cross-train staff to different roles because that really helps you appreciate the other roles and allows you to anticipate what your teammates need next. No one can do their role and succeed without the person next to them, including the physician.

SC: As a nurse practitioner, I had something similar to an interventional cardiology fellowship because my mentor took the time and effort to train me in how he wanted the cardiac catheterization to be. That training and education served me throughout my career. And I, in turn, taught the nurses that were scrubbed at the table. I think that by training others you're only going to make your life easier and improve your outcomes. So, if I could do one thing, it would be to train more people in a way similar to how I was trained. I would encourage interventional cardiologists to find someone they can invest in and train, because in the long run, outcomes will improve, and it will be well worth the effort.

KS: I would love to hear your thoughts on the role you see yourselves playing beyond the cath lab - things like patient advocacy, safety, quality standards...etc.

EK: I think our role as nurses and techs definitely expands beyond the four walls of the cath lab. One of the things I can do as a lab manager is to foster a "speak-up" culture. This not only improves the safety and quality of the lab, but also allows me and the team to be patient advocates. We also try to engage the team in quality initiatives and make sure they are always asking "Why?". I think every team member needs to find their

THE PICS SOCIETY

passion outside of circulating or scrubbing, and they should be encouraged to pursue that. Research is another example! I'll let Jason chime in there because that is an area he is very passionate about.

JS: I think it's very important to have staff involved in research because it allows you to understand why we're making the decisions we make in the cath lab. It also gives you a chance to be involved in cutting-edge technology and gives you the chance to see something go from the bench to clinical practice! And to echo what Emily said, the ability to speak up in a procedure is tremendously important. Being able to speak up ensures that everyone is on the same page and that we are all doing what's best for the patient.

KS: Jason, you brought up non-physician staff involvement in research. What role do you all think the PICS Society can play in promoting that?

JS: Well, one of the ways that PICS Society can do that is through the Nurse/Tech break out sessions at PICS. These can be opportunities to focus on nurse and rad tech-driven research. I think there's a role for PICS Society to support nurses and techs that would like to do their own research, beyond just data mining or involvement in animal studies.

EK: I think it's also important for physicians to think of nurses and rad techs as team members that can play an important role in research. Physicians tend to default to fellows to help with research, but I think it would be great to have physicians involve both the fellows and the nurses/rad techs! As far as what PICS Society can do to help with research, the most exciting aspect for me is the networking opportunities. If you're thinking of doing a purely nursing / tech driven research project, I think you'll get more out of that if you can use your networks from the PICS Society to make it a multi-center study.

"No one can do their role and succeed without the person next to them, including the physician."

Emily Kish, RN

KS: Outside of research, how can the PICS Society promote the roles that you all are playing? And how can we also promote more participation from our allied healthcare members within PICS Society?

SC: Marketing and education! Making sure that allied health care professionals know that the Society exists and understand the benefits of being a member. Knowing that they can reach out to other members to ask about availability of various devices or techniques being used at

other centers. I think we need to be marketing to the allied health professionals to join and to make sure we have educational material immediately available to them.

EK: That's exactly where I would start – with marketing. There are nurses and techs that don't even know that PICS or PICS Society exists! That can start with the physicians – encouraging the physicians to bring in more of their team. I think it's also important to keep it affordable to staff. Not a lot of hospitals can afford to send their staff. As for the meeting itself, the nursing breakout sessions are amazing! We could also have a round-table discussion for nurses where we discuss new topics or share experience from different centers.

JS: If PICS Society can help to spread the word that the PICS can be virtual for those who aren't able to participate due to funding, I think that would be huge. Also, some of the hands-on vendor sponsored activities like heart dissection or ICE-catheter simulation are very helpful for staff! It really helps us understand the procedure and be more engaged.

KN: Even though the PICS meeting provides a lot of continuing education credits for radiation technologists, not a lot of technologists attend the meeting. If the PICS Society can give a scholarship or a grant for techs to attend the meeting, that would be great. Also, continuing education for technologists does not focus on congenital cardiac interventions, whereas these meetings are specifically focused on what we do every day. So, there's a lot of value for technologists to attend. Oddly, one of the benefits of this terrible pandemic has been that PICS was accessible to a lot more people! A virtual component of the meeting would definitely help attendance for technologists, even if it's just to attend the live cases. Another option could be to provide the opportunity for folks to watch taped cases after the fact.

SC: I agree, if we can figure out how to make the meeting virtual and more accessible to people that couldn't attend in person, that would be very important. For example, making that a perk for being a member of the PICS Society to encourage people to join.

EK: Another place that PICS Society can help with: physicians have always had the opportunity to bounce ideas off each other in different platforms, and I think that's something we can help allied health professionals do!

JS: I agree! A list-serve would be hugely helpful! It could be for both research projects and to ask others how they handle day to day issues in the lab.

KS: Thank you all for this fantastic opportunity and this great conversation! I appreciate everyone's time and I look forward to continuing this conversation, in person, in Chicago!







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CICU Medical Director - Pediatric Cardiac Intensivist, Associate Professor or Professor

The Division of Pediatric Cardiology and the Department of Pediatrics at LSU Health Sciences Center in New Orleans is accepting applications for a Cardiac Intensivist academic faculty position at the rank of Associate Professor or Professor (non-tenure, clinical track) to serve as Medical Director of the Cardiac Intensive Care Unit at Children's Hospital New Orleans. Rank will be determined by the candidate's credentials and experience.

As Medical Director for the Cardiac Intensive Care Unit, incumbent will work with the Heart Center Co-Directors to coordinate and integrate a team approach, unifying cardiovascular surgeons, intensivists, anesthesiologists, cardiologists, and advanced practice providers. Additional responsibilities include recruitment, training, and development of current and new CICU staff as well as fostering academic productivity and professional development of its faculty and trainees and continue our participation in national registries such as the Pediatric Cardiac Critical Care Consortium (PC4).

Required: MD or equivalent, trained in pediatric intensive care and/or cardiology with an additional year of dedicated training in cardiac ICU, OR have a minimum of 5 years of CICU experience; BC/BE (or equivalent) in either Pediatric Cardiology or Pediatric Critical Care; licensed to practice medicine in the State of Louisiana before start date.

Please apply here:

https://lsuhsc.peopleadmin.com/postings/10579

Contact: Thomas Kimball, MD Chief of Pediatric Cardiology, Co-Director of the Heart Center 504.894.5445, <u>Thomas.Kimball@LCMChealth.org</u>



Pediatric Cardiac Intensivist, Assistant Professor or Associate Professor

The Division of Pediatric Cardiology and the Department of Pediatrics at LSU Health Sciences Center in New Orleans is accepting applications for an additional Cardiac Intensivist. This is an academic position and appointment will be at the rank of Assistant Professor or Associate Professor (non-tenure, clinical track) and will be determined by the candidate's credentials and experience. The cardiac intensive care team will work in conjunction with pediatric cardiology, cardiothoracic surgery, and other subspecialists, in the management of cardiac patients.

The Heart Center has an ACGME-accredited LSUHSC pediatric cardiology fellowship training program. There are excellent opportunities for clinical, translational and basic research, particularly with the LSUHSC Cardiovascular Center of Excellence as well as the Heart Center's Research Core. The successful candidate is expected to have strong clinical skills, have an interest in research, and participate in teaching of residents and fellows.

Minimum Qualifications: MD or equivalent; trained in pediatric intensive care and/or cardiology with an additional year of dedicated training in cardiac ICU, OR have a minimum of 5 years of CICU experience; BC/ BE (or equivalent) in either Pediatric Cardiology or Pediatric Critical Care; licensed to practice medicine in the State of Louisiana before start date.

Please follow this link for additional information and application instructions: <u>https://lsuhsc.peopleadmin.com/postings/10578</u>

Contact: Thomas Kimball, MD Chief of Pediatric Cardiology, Co-Director of the Heart Center 504.894.5445 Thomas.Kimball@LCMChealth.org



Pediatric Cardiologist (MRI)

The Department of Pediatrics, Section of Pediatric Cardiology at Tulane University School of Medicine in New Orleans, in partnership with the Children's Hospital of New Orleans (CHNOLA), is seeking a full-time academic Pediatric Cardiologist with particular expertise in advanced non-invasive cardiac imaging in CT and MRI technologies to join our faculty group. The Heart Center team includes 22 physician member with expertise in non-invasive imaging, electrophysiology, cardiac intervention, adult congenital heart disease, pediatric coronary artery disease, heart failure and cardiac transplantation, pulmonary hypertension and fetal cardiology. The team includes three cardiac surgeons and five cardiac intensivists. The Heart Center is based at CHNOLA and includes not only Tulane faculty but also faculty in the LSU Health Sciences Center. The selected candidate must have demonstrated excellence in advanced non-invasive imaging involving CT and MRI modalities as well as echocardiography. Incumbent will be involvement in developing and leading a comprehensive state of the art cardiac CT and MRI program at CHNOLA.

The Department of Pediatrics at Tulane is committed to providing excellence in education, clinical care, and research. The selected candidate will become actively involved in all aspects of teaching including training medical students and residents.

As a University employed faculty member, you will receive a negotiable salary commensurate with experience, and a fully comprehensive benefits package, and relocation allowance. Academic appointment will be at the rank of Assistant, Associate or Full Professor determined by the candidate's credentials and experience.

For qualifications, application instructions, and more: apply.interfolio.com/85904

CAREER OPPORTUNITY

MEDICAL NEWS



Practice with Leading Integrated, Healthcare System in Arizona!

Banner Health and the Division of Pediatric Cardiology at Banner Children's Specialists (BCS) is seeking a board certified/board eligible Pediatric Cardiologist with fellowship training in advanced imaging and fetal imaging to join a thriving and growing practice of six providers in Phoenix Metropolitan Area in Arizona.

This position is open to both experienced and new graduates. We will support a provider with an interest in exercise physiology. Based at Banner Children's at Desert Medical Center, the candidate will practice general cardiology, with routine clinic and call. The group covers all of Banner Health's hospitals and shares call 1:8. **On-site you will have access to echocardiography, catheterization laboratory, exercise lab, ultrasound, and more.**

Banner Children's at Desert Medical Center in Mesa, a

state-of-the-art, 206-bed children's hospital opened in 2009, along with some of our other hospitals, provides comprehensive pediatric care from general to emergency, as well as services for specific childhood diseases. Highly trained physician specialists, social services staff and numerous integrative therapy programs ensure a complete range of care for children. We have a child-friendly atmosphere accented by soothing colors and dedicated play areas, as well as a volunteer dog therapy program that helps cheer up young patients during their stays.

- Phoenix metro Pediatric population of 350,000+
- Child friendly pediatric EDs (90,000 visits in system yearly)
- Minimally invasive & robotic pediatric surgery
- PICU with 24/7 Pediatric IntensivistsFull-time hospitalists
- State-of-the-Art Pediatric Cancer Care Unit
- 24 bed PICU, level 3 NICU staffed 24 hours by neonatologists and neonatal nurse practitioners
- Member of NACHRI

Banner Health is one of the largest non-profit healthcare systems in the country with thirty hospitals, six long term care centers, valley-wide urgent care centers and an array of other services, including family clinics, home care services and home medical equipment, in six Western states. We offer physicians highly integrated and innovative environments, a collaborative team-oriented workplace and clinical settings that focus on patient care excellence.

PLEASE SUBMIT YOUR CV FOR IMMEDIATE CONSIDERATION, TO: doctors@bannerhealth.com For questions, please call Tiffany Lewis, Sourcing Director at 602-747-4578. Visit our website at: www.bannerhealth.com Please, No Agency Solicitations!

The safety of our team members and patients is of utmost importance, so Banner is requiring the COVID-19 vaccine for all team members. As members of the health care field, we are in the business of caring for people, so we take seriously our commitment to ensure our patients and teams are safeguarded from this rapidly changing and dangerous disease.

As an equal opportunity and affirmative action employer, Banner Health recognizes the power of a diverse community and encourages applications from individuals with varied experiences and backgrounds. Banner Health is an EEO/AA - M/W/D/V Employer.



atHeart Medical Receives FDA Approval for the Second Phase of the ASCENT ASD US IDE Trial

Company's reSept™ ASD Occluder Aims to Evolve Septal Closure with its Novel Metal-Free Frame Design

atHeart Medical, a medical device company dedicated to establishing the new standard of care for closure of atrial septal defects (ASD), today announced it has received approval for the start of the second phase of its ASCENT ASD U.S. Investigational Device Exemption (IDE) pivotal trial.

The prospective, single-arm study is evaluating the safety and efficacy of the reSept[™] ASD Occluder, the first occluder with a metalfree, bioresorbable frame, for the treatment of patients with clinically significant, isolated ASDs. Primary endpoints will be compared with established performance goals for previously FDA-approved transcatheter ASD occluders.

"I am pleased the first phase of enrollment in our pivotal trial progressed smoothly and according to plan. This is an exciting milestone for the company," said Laurent Grandidier, CEO of atHeart Medical. "As we initiate the second phase, our team is focused on adding clinical sites across the U.S. and expanding internationally to include several enrolling sites in France. I commend the team's diligence to further validating the safety and efficacy of the reSept ASD Occluder, a critical step in our journey to evolve septal closure and provide a better solution for patients."

The reSept ASD Occluder aims to address the limitations of current occluders which have metallic frames that can place patients at risk of complications associated with long-term presence of metal in the heart and may limit future transseptal interventions, such as mitral valve interventions. Initial clinical experience demonstrates positive safety and performance in the closure of the ASDs treated with the company's device.¹

"The low-profile reSept Occluder is a dynamic system that allows for versatile physician control during the intervention, potentially adapting to the different patient anatomies to address ASDs," commented Dr. Scott Lim, Professor of Medicine & Pediatrics at the University of Virginia in Charlottesville, VA, and a leading enrolling site in the trial. "Over time, reSept's metal-free frame resorbs, leaving a minimal implant behind. This is an exciting advancement that provides the potential to preserve future treatment options and potentially do better for our patients."

About ASCENT-ASD Investigational Device Exemption (IDE) Trial

ASCENT-ASD is a prospective, single-arm, global multi-site clinical investigation study that will enroll a total of up to 250 patients. The study aims to demonstrate the safety and efficacy of the reSept[™] ASD Occluder for treating clinically significant secundum ASD with a transcatheter approach as compared to pre-defined performance goals from other commercially available occluder devices. For more information, please visit <u>www.clinicaltrials.gov</u> – Trial Identifier: NCT04591392.

About atHeart Medical

atHeart Medical is a medical device company with offices in Switzerland and the United States committed to establish a new standard of care for treatment of atrial septal defects (ASD).

For more information, please visit: www.atheartmedical.com.

About ASD

Commonly described as a "hole in the heart", an ASD is an opening in the septum between the left and right atria. Most ASDs are congenital defects, affecting six in 10,000 births.² They can also be the result of a procedure that requires transseptal crossing. A large atrial septal defect can cause extra blood to overfill the lungs and overwork the right side of the heart. If not treated, the right side of the heart eventually enlarges and weakens. The blood pressure in the lungs can also increase, leading to pulmonary hypertension. When ASDs require closure, the current standard of care is to implant a septal occluder with a metallic frame through a minimally invasive procedure.

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CAREER OPPORTUNITY



Interventional Pediatric Cardiologist

Successful Pediatric Cardiology Practice - Tropical Florida Coast

Pediatric Cardiology Associates, located in Tampa Bay on Florida's Gulf Coast, is seeking a BC Interventional Pediatric Cardiologist with advanced fellowship training and experience in Congenital Intervention.

- Ideally seeking candidates with a minimum of 5 years of experience post-fellowship
- Large, experienced, well-established team of 14 pediatric cardiologists and 3 NPs with offices and clinics located throughout the Tampa/St. Petersburg area
- Offer comprehensive congenital cardiac care from fetal life through adulthood
- The team includes members of all pediatric cardiology sub-specialties including: fetal, advanced imaging (CT, MRI, 3D echo), intervention, electrophysiology, cardiomyopathy/heart failure, prevention, and adult congenital
 - The interventional team performs over 400 catheterizations per year, about 60% of which are interventions
- Recent interventional team accomplishments include:
 - Implanting their 150th transcatheter pulmonary valve, Summer 2019
 - First program in Florida to implant the Gore Cardioform ASD Occluder, Fall 2019
 - Only program in Tampa Bay currently offering PDA device closure for premature newborns, first implant, Winter 2016
- Sub-specialty clinics include general pediatric cardiology, intervention, pulmonary hypertension, cardiomyopathy, ACHD, electrophysiology, and prevention
- Our ACHD program is the ONLY certified Adult Congenital Heart Association program in central Florida
- This position also offers:
 - Full time interventional duties with expected procedural volume of 200+ catheterizations per year
 - No expectation of inpatient service coverage
 - 24/7 collaboration with our excellent pediatric cardiac surgical and pediatric cardiac intensive care teams at St Joseph's Children's Hospital
- Our center offers a unique depth of hospital infrastructure:
 - Two state of the art 1000+ square foot hybrid capable catheterization labs/ORs (one biplane, one single plane)
 - Two additional biplane catheterization labs
 - Two EP labs
- We have the added benefits of a children's hospital inside a large tertiary adult hospital simplifying care across all patient ages with easy access to consultants from all pediatric and adult specialties
- Pediatrix, as a national pediatric cardiology group with over 125 pediatric cardiologists, provides opportunities for quality initiatives that can have national impact
- · We offer an attractive schedule allowing freedom to enjoy a great quality of life
- Generous compensation package offered

Tampa Bay's warm weather affords plenty of opportunities to relish the great outdoors year round. You will live in a region others only get to enjoy on vacation. Golf at one of nearly 100 courses or relax on one of the many pristine white-sand beaches. The area offers an assortment of family venues such as zoos, aquariums, theme parks, and state parks. Additionally Tampa Bay offers access to world-class museums, professional sporting events and the performing arts. There is a wide range of residential choices to fit every budget and lifestyle - whether you are looking for big city downtown living, golf course communities, waterfront lifestyle, majestic horse farms or historic neighborhoods.

Effective November 1 st , 2021 Pediatrix will require all employees and new hires to be vaccinated against COVID-19, unless they qualify for an approved medical and/or religious exemption.

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Our clinicians enjoy a competitive compensation package with many locations offering sign on bonuses, relocation and tuition reimbursement. *Our benefits include:

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- Leadership training and advancement opportunities
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*These benefits are for full time employees, employees in other types of employment classifications may be eligible for some of these benefits.

Mednax, Inc. and its affiliated practices operating as Pediatrix[®] Medical Group (Pediatrix) are one of the nation's leading providers of highly specialized health care for women, babies and children. Since 1979, Pediatrix-affiliated clinicians have been committed to providing coordinated, compassionate and clinically excellent services across the continuum of care, both in hospital settings and office-based practices. Specialties including obstetrics, maternal-fetal medicine, and neonatology are complemented by 18 pediatric subspecialties and a newly expanded area of primary and urgent care clinics. The group's high-quality, evidence-based care is bolstered by investments in research, education, quality-improvement and safety initiatives.

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A Non-Invasive Way to Predict Heart Attack and Stroke

Pitt's Kang Kim Receives Nearly \$3 Million in NIH Funding to Detect Dangerous Atherosclerotic Plaques Using Super-Resolution Ultrasound

Acute coronary syndromes (such as heart attacks) and strokes are a leading cause of morbidity and mortality in the U.S. and Europe. About 80 percent of those events occur because a buildup of plaque in the arteries—or atherosclerotic plaque (AP)—ruptures.

Research led by Kang Kim, PhD, Associate Professor at the University of Pittsburgh, recently received \$2,996,514 over four years from the National Institutes of Health for work that will use super-resolution ultrasound (SRU) imaging to noninvasively detect APs that have a high chance of rupturing so that doctors can intervene.

"Over the past decade, researchers have been pushing for a way to identify rupture-prone APs," said Kim, who holds appointments in both the Swanson School of Engineering's Department of Bioengineering and the School of Medicine's Department of Medicine. "One thing that plays a critical role in AP rupture is when a new, dense network of blood vessels begins growing into the AP from the vasa vasorum (VV), or the smaller vessels that supply larger arteries and veins. However, we don't yet have the noninvasive tools to assess abnormal microvascular expansion in vivo, or inside the living body. That's the problem this project is meant to solve."



The technology developed through this project seeks to shift the current imaging approach in identifying

microvessels of AP from "intravascular," or an ultrasound that uses a catheter to view the inside of the artery, to a "fully noninvasive transcutaneous" imaging approach, which instead can noninvasively see microvasculature.

If successful, the technology could be incorporated into current ultrasound scanners already in use to identify plaques at high risk of rupturing, allowing doctors to prescribe lifesaving interventions and treatments to prevent a stroke.

"In addition to further evaluating our super resolution ultrasound imaging technology, I'm excited about determining if characteristics of the VV can be a predictive biomarker of AP rupture," said Kim.

Kim's team includes:

 Flordaliza Villanueva, MD, Associate Chief of Cardiology Translational Research and professor of medicine and bioengineering

- Edith Tzeng, MD, Chief of VA Vascular Surgery and Professor of Surgery and Bioengineering
- Julie Phillippi, PhD, UPMC Pellegrini Chair in Cardiothoracic Surgery and Associate Professor of Cardiothoracic Surgery and Bioengineering

For this research, the Pitt team is also partnering with Dr. Alkystis Phinikaridou from King's College in London, and Dr. Renu Virmani, President and Medical Director at CVPath Institute.

The four-year project is titled "Super Resolution Ultrasound Imaging of Vasa Vasorum to Characterize the Progression of Atherosclerotic Plaques and Predict Rupture Vulnerability" and began Feb. 1, 2022. The work was funded through the competitive funding opportunity titled NIH Research Project Grant (Parent R01). Read more about the project here:

https://reporter.nih.gov/search/ RIHs6jejJECD9RW3IKjTRg/projectdetails/10374343



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- NewYork-Presbyterian KipS

Komansky Children's Hospital

Pediatric Cardiologist

The Division of Pediatric Cardiology is seeking a Pediatric Cardiologist to join our team at New York-Presbyterian/Komansky Children's Hospital-Weill Cornell Medicine.

Candidates must be board certified or board eligible in Pediatric Cardiology. Experience in pediatric cardiac MRI is desirable but not mandatory, and suitable candidates would be offered the director position for pediatric cardiac MRI. Candidates wishing to pursue a physician scientist career with a strong research portfolio are encouraged to apply. Candidates must have excellent skills in non-invasive imaging, in particular transthoracic, transesophageal, and fetal echocardiography.

The successful candidate will be appointed under the Division of Pediatric Cardiology and will work predominantly at our main Upper East Side campus. In addition, the successful candidate may be covering our general and fetal outreach clinic in Lower Manhattan (presently 1 day a week).

The pediatric cardiac program at Weill Cornell Medicine is an integral part of New York-Presbyterian Hospital (NYP), which is one of the largest pediatric cardiac programs in the country and ranked 11th in the 2021 US News and World Report of best hospitals for Pediatric Cardiology & Heart Surgery. Our surgeons perform cardiac procedures at NewYork Presbyterian - Weill Cornell Medical Center, as well as Columbia University Medical Center.

Interested candidates please send CV to: Ralf Holzer, MD, MSc rjh3001@med.cornell.edu

Diversity is one of Weill Cornell Medicine's core values and is essential to achieving excellence in patient care, research, and education. We welcome applications from candidates who share our commitment to fostering a culture of fairness, equity, and belonging. Weill Cornell Medicine is an Equal Employment Opportunity Employer, providing equal employment opportunities to all qualified applicants without regard to race, sex, sexual orientation, gender identity, national origin, color, age, religion, protected veteran or disability status, or genetic information.



Pediatric Cardiac Interventionist Position

UC Davis Children's Hospital School of Medicine Department of Pediatric Cardiology

The Department of Pediatrics at the University of California, Davis School of Medicine is recruiting 1 full-time academic Pediatric Cardiac Interventionist within the Pediatric Cardiology specialty. We are recruiting at the Assistant/Associate/Full Professor level in the Clinical X or Health Science Clinical Series in the Section of Pediatric Cardiology, Department of Pediatrics and specifically trained and experienced in Pediatric Cardiac Interventions. Candidates must possess an M.D. or D.O. degree, be board certified in Pediatric Cardiology and must possess or be eligible for licensure in the State of California.

The interventional cardiology candidate must have at least 3 years of experience in interventions in congenital heart disease and have the desire and ability to build and grow a practice in interventional cardiology volume.

Work distribution will depend on the specific skillset of the candidate. It is expected that the candidate will share in the on-call and weekend/ holiday coverage schedule.

In addition to the clinical responsibilities, the ideal candidate will be expected to participate in teaching of medical students, residents and fellows, research activities of the Department of Pediatrics, and serve on departmental committees.

The candidates must have the following qualifications:

- M.D. or D.O.
- Successful completion of an approved pediatric residency training program.
- Successful completion of an approved Pediatric Cardiology fellowship training program.
- Successful completion of a Pediatric Interventional Cardiology advanced fellowship training program.
- Board certification in Pediatric Cardiology.
- Eligibility for a California Medical License.
- Demonstrated proficiency in the teaching of students and housestaff.
- Demonstrated proficiency to perform clinical research.
- Ability to foster collegiality and work collaboratively in a diverse environment, including working closely with Adult Congenital Heart Disease services.
- Service including committees, leadership ability, and community outreach.
- Have Board Certification in Adult Congenital Heart Disease (preferred).

The Pediatric Heart Center at UC Davis Children's Hospital is inland Northern California's only full-service cardiac care facility for children and young adults, offering the latest tests and treatments for a range of congenital or acquired cardiovascular conditions. Our integrated multidisciplinary team of surgeons, specialists, physicians, nurses and researchers offer Northern California's most sophisticated specialized diagnostic, interventional and surgical expertise in comprehensive diagnostic, therapeutic, and surgical procedures for children with heart defects.

For full consideration applications should be received by December 27, 2021. However the position will remain open until filled. Completed applications include CV, Cover Letter, Statement of Contributions to Diversity, Equity, and Inclusion and contact information for 3-5 references.

Candidates should submit their application online at: https://recruit.ucdavis.edu/JPF04580.

CAREER OPPORTUNITY

MEETING CALENDAR





Pediatric Cardiologist

The Department of Pediatrics at Southern Illinois University School of Medicine is recruiting an MD/DO for a fourth pediatric cardiologist position at the Assistant or Associate Professor level. Faculty will join a rapidly expanding cardiology program at our Children's Hospital, an 80-bed CHA affiliated pediatric referral center for Central and Southern Illinois with a referral base of almost 2 million. The current program includes stateof-the-art noninvasive imaging in TTE, TEE, fetal echocardiogram, and advanced MRI imaging. We have developed a highly successful collaborative clinical and research program with a nationally recognized pediatric cardiology center. Opportunities exist to participate in resident and medical student education receive an advanced and degree in medical education. Candidates must be board eligible in Pediatrics and Pediatric Cardiology. Illinois licensure is required prior to official start date. Travel in central Illinois to outreach clinics is required.

Applications are accepted online at: https://www.siumed.edu/hr

For additional information, please contact:

Ramzi Nicolas, MD 217.545.9706 rnicolas@siumed.edu

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JUNE

10-13

ASE 2022 – 33rd Annual Scientific Sessions Seattle, Washington, USA https://www.asescientificsessions.org/register-now/

22-25

CSI Frankfurt Frankfurt, Germany https://www.csi-congress.org/conferences-courses/conferences/csifrankfurt

23-25

ACHA's 9th National Conference: Leading the Way for 1 in 100 Virtual only https://www.achaheart.org/get-involved/events/2022/9th-nationalconference/

AUGUST

03-06

NeoHeart: Cardiovascular Management of the Neonate Anaheim, California, USA <u>https://web.cvent.com/event/f5efadb3-8886-4c5b-</u> 9944-c41980940049/summary

05-06

International PDA Symposium 2022 Anaheim, California, USA https://pdasymposium.com/

21-26

Pediatric & Adult Congenital Cardiology Review Huntington Beach, California, USA https://ce.mayo.edu/cardiovascular-diseases/content/2022-pediatricand-adult-congenital-cardiology-review-course



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