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Recent Publications:

Lee N, Das A, Taylor M, **Hor KN**, Banerjee RK. Energy transfer ratio as a metric of right ventricular efficiency in repaired congenital heart disease. *Congenital Heart Disease*. 2013 Jul-Aug;8(4):328-42. Epub 2013 Jan 21. PMID: 23331703

Ryan TD, Taylor MD, Mazur W, Cripe LH, Pratt J, King EC, Lao K, Grenier MA, Jefferies JL, Benson DW, **Hor KN**. Abnormal circumferential strain is present in young Duchenne muscular dystrophy patients. *Pediatric Cardiology*. 2013 Jun;34(5):1159-65. Epub 2013 Jan 29. PMID: 23358912

Statile CJ, Taylor MD, Mazur W, Cripe LH, King E, Pratt J, Benson DW, **Hor KN**. Left ventricular noncompaction in Duchenne muscular dystrophy. *Journal of Cardiovascular Magnetic Resonance*. 2013 Aug 1;15:67. PMID: 23914774

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Toro-Salazar OH, Gillan E, O'Loughlin MT, Burke GS, Ferranti J, Stainsby J, Liang B, Mazur W, Raman SV, **Hor KN**. Occult cardiotoxicity in childhood cancer survivors exposed to anthracycline therapy. *Circulation Cardiovascular Imaging*. 2013 Nov 1;6(6):873-880. Epub 2013 Oct 4. PMID:24097420

**Referrals and Consultations**

The Cardiac MRI program at Nationwide Children's Hospital accepts referrals from across the country.

**Online:** NationwideChildrens.org

**Fax:** (614) 722-4000

**Phone:** (614) 722-6200 or 1(877)722-6220

**Physician Direct Connect Line for urgent physician consultations:**

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# Case Study: Cardiac Magnetic Resonance

## The Heart Center

*Kan Hor, MD, director of Cardiac MRI, cardiology division*

At Nationwide Children's Hospital, cardiac magnetic resonance (CMR) has become an important diagnostic tool in the care of all patients, especially pediatric cardiology patients, and is the centerpiece of the Advanced Cardiac Imaging Laboratory, which combines the clinical and technical expertise of clinicians, radiologists and technicians from The Heart Center and the Department of Radiology. Here we profile a complex case of a patient with Duchenne muscular dystrophy (DMD) who presented to Nationwide Children's and was referred to the Advanced Cardiac Imaging Laboratory. With the aid of the program's advanced, high-resolution CMR, the team was able to visualize heart structures at a level not possible with conventional imaging technology. Plans for advanced CMR services and new research initiatives are also discussed.



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## PATIENT: MYOCARDIAL FIBROSIS

### PHASE I: Presentation

An 11-year-old male with Duchenne muscular dystrophy (DMD) was taken to a local emergency room with complaints of nausea and sharp chest pain that radiated to the left shoulder. Physicians noted an elevated troponin level and ST segment change on his electrocardiogram. The patient was transferred to Nationwide Children's Hospital and admitted to the cardiac catheterization laboratory.

### PHASE II: Testing and Diagnosis

The patient underwent a cardiac catheterization, which demonstrated normal coronary arteries and hemodynamic data. An echocardiogram showed decreased ventricular function with mild enlargement of the left ventricular chamber size. Cardiac MRI further demonstrated the presence of sub-epicardial myocardial fibrosis unique to the Duchenne muscular dystrophy patient population, as well as areas of edema that may be consistent with active inflammation beyond his baseline disease. (Figure 2) The cause of the chest pain was secondary to myocardial damage resulting in myocardial fibrosis.

### PHASE III: Treatment and Follow-up

Despite the new findings of myocardial fibrosis, patient's left ventricular function was only mildly depressed. The patient was treated with carvedilol, aspirin, lisinopril and deflazacort and discharged home after three days. The patient re-presented three months later with similar but less severe chest pain and underwent a repeat CMR, demonstrating no further progression of left ventricular function and myocardial fibrosis burden. Patient was discharged home within 24 hours. The patient is being followed by the Muscular Dystrophy Clinic at Nationwide Children's and will undergo annual CMR imaging to monitor progression of DMD-associated heart disease.

Cardiac magnetic resonance (CMR) imaging has many capabilities beyond traditional noninvasive diagnostic modalities to help physicians better assess the health status of their patients and provide a more precise road map for intervention.

The 3-D imagery produced by CMR offers unprecedented graphic details of anatomical defects in the cardiovascular system, the burden of scar tissue caused by heart disease and a range of other cardiac conditions that affect pediatric patients. CMR even makes it possible to create 3-D images of blood vessels, a process called magnetic resonance angiography (Figure 1), to give physicians an accurate visual of defects prior to surgery or heart catheterization. CMR also has advantages over other non-invasive technologies, such as transthoracic echocardiogram. Whereas CMR can accommodate patients of all sizes, as patients get older, the acoustic windows of the echocardiogram make it difficult to see important structures in complex patients.

### Case Elements: Visualization of myocardial tissue

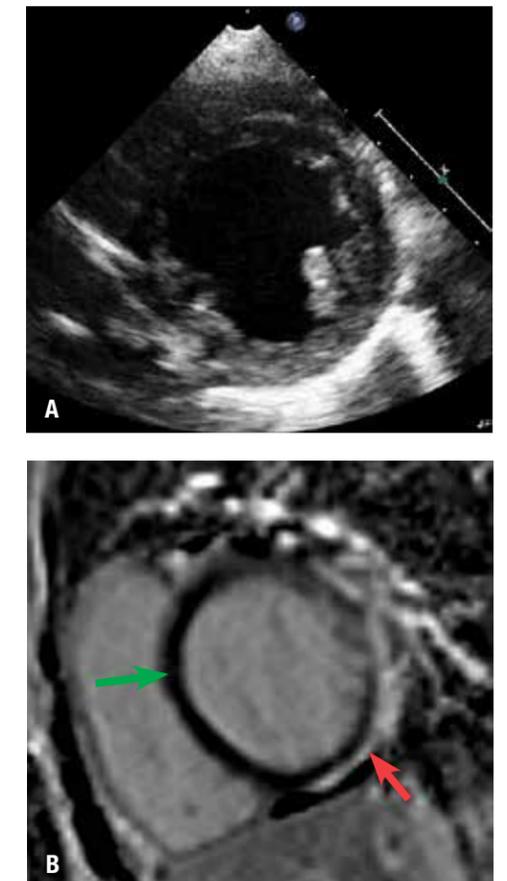
CMR allows superior assessment of not only the anatomy, but also of some physiologic parameters, such as flow-through structures, and quantitative data for chamber size and cardiac function by ejection fraction. CMR also provides key information on myocardial tissue characteristics and go beyond squeeze (shortening fraction or ejection fraction) by looking at inflammation, scar burden, myocardial mechanics and other features. This capability is especially helpful in diagnosing complex cardiac problems, such as those experienced by a patient with DMD (see details above) who presented recently to Nationwide Children's with chest pain. The added value of cardiac magnetic resonance imaging in this patient (Figure 2) was demonstrating the area of myocardial fibrosis with areas of inflammation that could not be elucidated by cardiac catheterization or echocardiogram, thus allowing the care team to optimize the treatment regimen and ensure the best outcome for the patient and family.

### Case Elements: Importance of CMR on re-presentation

This patient re-presented approximately two months later with similar but less severe chest pain. Given his previous cardiac magnetic resonance findings, he was taken to the cardiac magnetic resonance suite, bypassing an invasive cardiac catheterization study. Fortunately, the cardiac magnetic resonance study did not demonstrate further myocardial injury and the patient was able to be discharged home within 24 hours of admission. The added value of cardiac magnetic resonance study at this visit led to accurate diagnosis, decreased cost and length of hospitalization and harm to the patient. For pediatric patients with chest pain that could be symptomatic of myocardial damage, CMR imaging should be considered as a noninvasive alternative for accurate diagnosis that may not be possible by either echocardiography or invasive cardiac catheterization. This is particularly important in the DMD patient population, as demonstrated here.



**Figure 1:** These images offer examples of the value of (A) magnet resonance angiography (MRA) and (B) computerized tomographic angiography (CTA). Patients with Loey-Dietz syndrome have a high risk of aortic aneurysm and rupture and death around the second decade of life. (A) Patient presented for a six-month follow-up MRA due to progression dilation of abdominal aorta (red arrows) and was noted to have an intramural thrombus (green arrow) flattening the front of the abdominal aorta. His doctors were preparing him for elective but urgent surgery due to the high risk of aneurysm rupture. (B) One week later, he presented with severe chest pain and underwent emergent CTA, which indicated rupture of the aneurysm. The patient was transferred to an adult cardiac unit for further care.



**Figure 2:** Sample short axis at the base of the heart from (A) echocardiogram (TTE) and (B) cardiac magnetic resonance imaging (CMR). Like TTE, CMR can show chamber size but more accurate assessment of global heart function by ejection fraction. CMR can go above and beyond what TTE can and includes but is not limited to myocardial fibrosis imaging. As in many boys with DMD, such as the patient profiled here, there is evidence of myocardial scar indicative of damaged heart tissue in the typical region (white areas indicated by the red arrow compared to normal heart tissue dark areas indicated by the green arrow) that can not be shown by TTE or cardiac catheterization procedures.

### Research and Future Directions

Nationwide Children's CMR program has grown by 60 percent in just 18 months. Patients come not only from the regional referral base, but also from across the country. The program's growth will be supported with construction of a third CMR scanner, due for completion in early 2014. The new MRI scanner will be a Siemens 3 Tesla Skyra magnet with the most current software and hardware, improving not only the quality but the speed of the scan as well as providing new clinical and research software not currently available. The new CMR features a higher field strength that will heighten our ability to capture detailed cardiac images and the equipment's larger design will allow us to accommodate older and larger pediatric patients. The new scanner will be utilized for both conventional and CMR studies.

Faculty with the Advanced Cardiac Imaging Laboratory are also engaged in a number of research projects designed to further enhance the value of CMR in the diagnosis and treatment of a range of cardiac disorders in children. Studies underway include efforts to identify the best way to reduce the rate of cardiac scar tissue formation in patients with DMD; development of clinical interventions designed to reduce the cardiac effects of certain chemotherapeutic agents in pediatric cancer patients; and an evaluation of tissue engineered vascular grafts in congenital heart surgery.