

## Background

Patients with Duchenne muscular dystrophy (DMD) gradually develop myocardial fibrosis, believed to indicate worse prognosis. This has been demonstrated by late gadolinium enhancement (LGE) on cardiac MRI (CMR).

## Objective

Describe the utilization of CMR in patients with DMD for identification of left ventricular (LV) fibrosis, ventricular size, and systolic function.

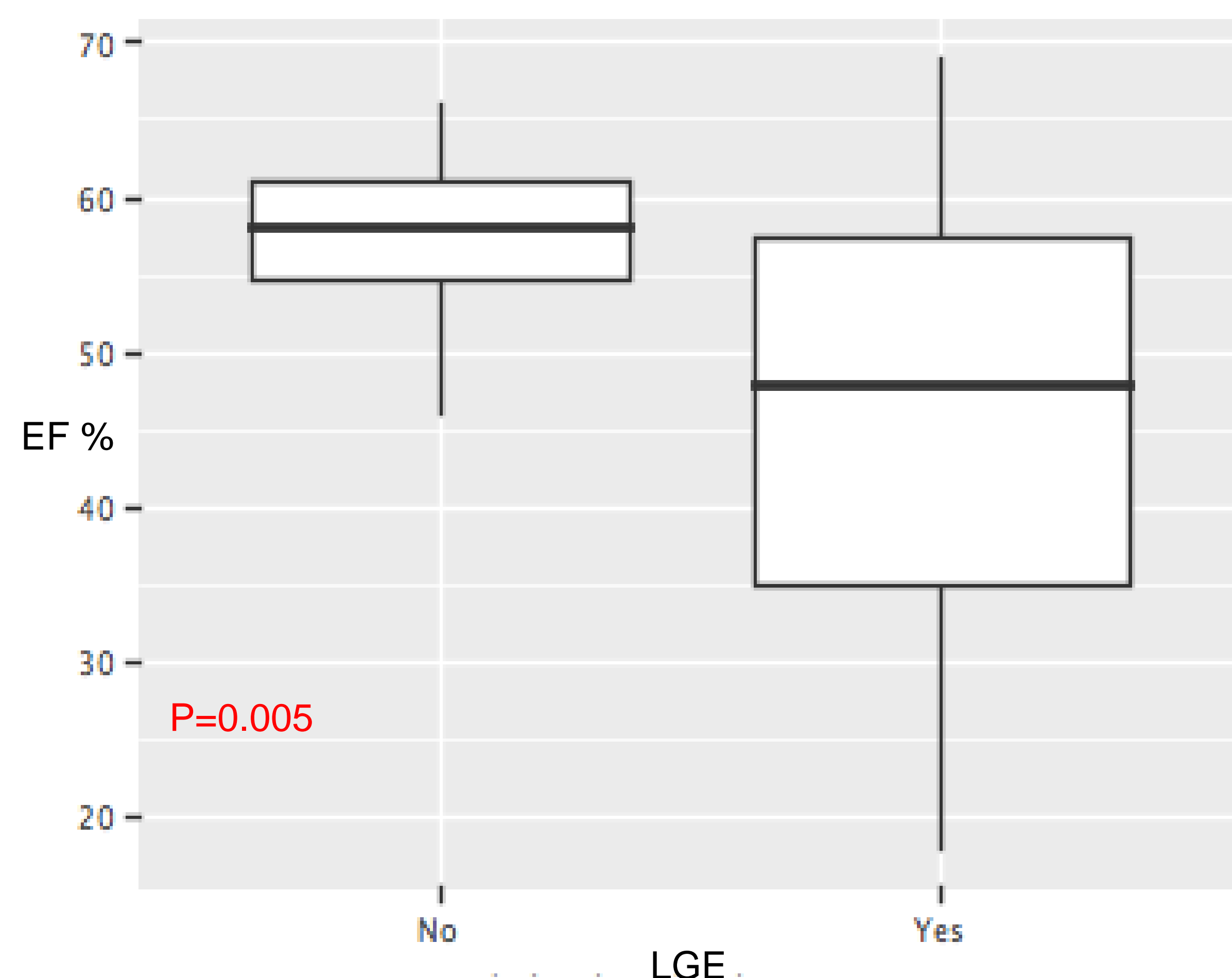
## Methods

Retrospective chart review of DMD patients at 17 tertiary pediatric hospitals (U.S. and Canada) between 2005-2015 (Diagnosis by 2012). Females, patients without CMR, and those without LGE documentation were excluded. Variables are reported as medians with interquartile range (IQR) and counts. Univariable and multivariate analyses were performed to evaluate the effect of LGE on CMR parameters. Pearson correlation coefficient was performed to measure the correlation between ejection fraction by CMR and echocardiography. An alpha < 0.05 was considered statistically significant.

## Results

- 436 patients with DMD
- 61 (14%) underwent 78 CMRs (12 patients had 2 CMRs, 5 pts had 3 CMRs)
- LGE presence or absence documented for 51 patients
- Age of diagnosis for those undergoing CMR compared to those who did not [(4 (3-6) vs. 5 (3-6); p=0.66].
- Median age at 1<sup>st</sup> CMR 14 (11-16)
- LGE present in 27 patients (44%)
- 36 (71%) were on steroids
- 29 (57%) were on cardiac medications.
- ACE inhibitors/angiotensin II receptor blockers (26) and beta blockers (10) were most common
- Multivariate analysis for LGE shows a higher EF is associated with lower odds of LGE (OR 0.87, CI 0.78-0.96, p=0.009)

Figure 1. CMR EF (%) by presence of LGE



## Results

Table 1. Clinical and study data for 1st CMR by presence of LGE

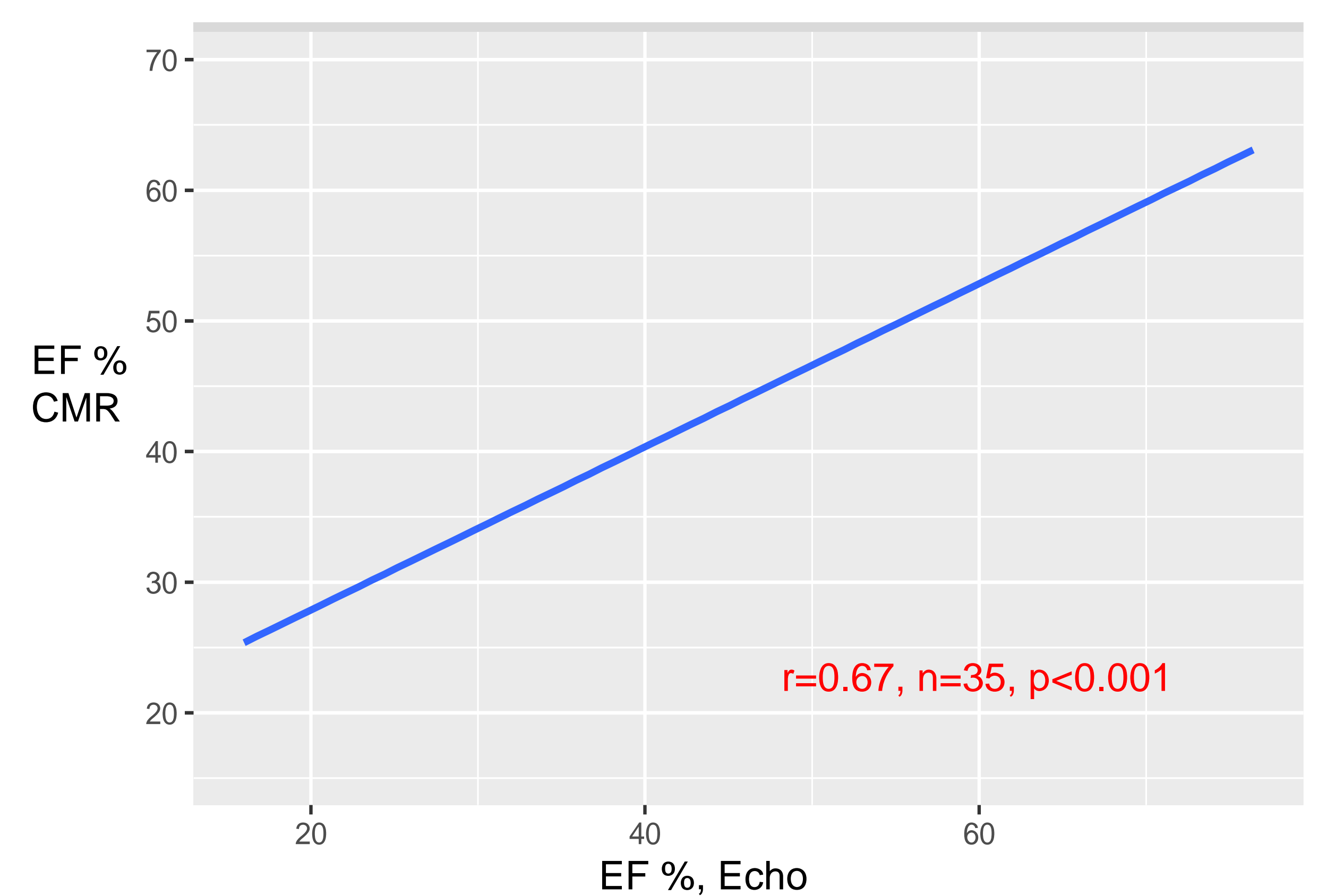
	No LGE n=24	LGE n=27	p value
Age at CMR	11 (11-14)	14 (12-16)	0.0195
Height	137 (130-144)	152 (129-156)	0.2
Weight	46 (36-57)	50 (44-70)	0.1
BSA	1.37 (1.09-1.48)	1.42 (1.22-1.7)	0.1
Deceased/Lost to Follow-up*	2* (8%)	3 (11%)	0.09
LVEDV indexed	68 (63-77)	80 (64-118)	0.048
LVEDV Z score	-2.8 (-3.6, -1.13)	-1.74 (-4.3, +2.22)	0.3
RVEDV indexed	64 (59-71)	65 (58-76)	0.8
RVEDV Z score	-4.2 (-4.9, -2.2)	-3.94 (-5.8, -2.3)	0.4
LVESV indexed	30 (25 - 36)	40 (29- 63)	0.024
LVESV Z score	-0.3 (-1.7, +0.44)	0.84 (-1.45, +4.21)	0.081
RVESV indexed	29 (23 -33)	31 (27 - 36)	0.4
RVESV Z score	-1.6 (-3.32, -1.36)	-2.03 (-3.6, -0.9)	0.9
LV EF	58 (55-61)	48 (35-58)	0.005
RV EF	57 (56-61)	55 (50-59)	0.1

CMR- cardiac MRI, BSA- body surface area, LV- left ventricle RV-right ventricle, eDV- end diastolic volume, eSV- end systolic volume, EF- ejection fraction

Table 2. Medication data by presence of LGE

	No LGE n=24	LGE n=27	p value
Steroids	17 (71%)	19 (70%)	1
Cardiac Medications	12 (50%)	17 (63%)	0.35
ACEi/ARBs	11 (46%)	15 (55%)	0.5
Beta blocker	3 (12%)	7 (26%)	0.2
Aldosterone antagonist	0	4 (15%)	0.05
Loop Diuretic	0	1 (4%)	0.3

Figure 2. Correlation between EF (%) CMR vs Echocardiogram



## Participating Pediatric Hospitals

- Ann and Robert H. Lurie Children's Hospital of Chicago (Chicago, IL)
- Arkansas Children's Hospital (Little Rock, AR)
- Mayo Clinic Children's Center (Rochester, MN)
- Primary Children's Hospital (Salt Lake City, UT)
- C.S.Mott Children's Hospital (Ann Arbor, MI)
- Morgan Stanley Children's Hospital (New York, NY)
- Children's Healthcare of Atlanta (Atlanta, GA)
- Children's Hospital of Pittsburgh (Pittsburgh, PA)
- Seattle Children's Hospital (Seattle, WA)
- Kravis Children's Hospital at Mount Sinai (New York, NY)
- Nemours Alfred I Dupont Hospital for Children (Wilmington, DE)
- Cincinnati Children's Hospital (Cincinnati, OH)
- Loma Linda University Children's Hospital (Loma Linda, CA)
- Children's Health at UT Southwestern Medical Center (Dallas, TX)
- University of Virginia Children's Hospital (Charlottesville, VA)
- Golisano Children's Hospital (Rochester, NY)
- Stollery Children's Hospital (Edmonton, Alberta, Canada)

## Conclusions

- Cardiac MRI is not widely used in the assessment of patients with DMD.
- CMR is an accurate screening tool that can detect fibrosis and dysfunction without LV volume changes
- CMR can potentially help with intensification of heart failure treatment.
- Ventricular dysfunction was more often seen with fibrosis.
- ACE inhibitors, aldosterone antagonists, and beta blockers are underutilized in patients with DMD.
- A long term multi-center prospective study is warranted to evaluate the effect of cardiac medications on the LGE load.