Early-onset late gadolinium enhancement is a prognostic factor for young cardiomyopathy patients with Duchenne Muscular Dystrophy

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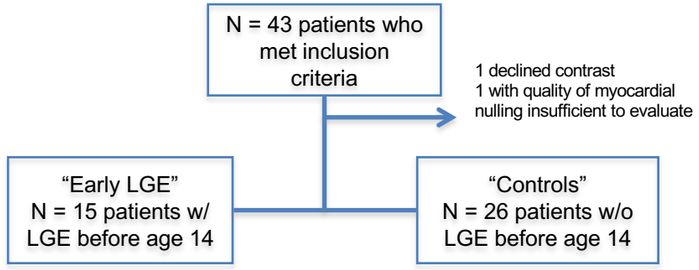
Disclosures

- No conflicts of interest to disclose

Background

- Duchenne Muscular Dystrophy (DMD) involves mutations which cause lack of functional dystrophin protein, leading to weakness in all muscle cells
- In the heart, progressive weakening of muscle leads to ventricular dilation, dysfunction and, ultimately, heart failure
- Late gadolinium enhancement (LGE) on cardiac MRI (cMRI) has been shown to increase with age and correlates with progression to left ventricular (LV) dysfunction
- We hypothesize that those with early LGE have worse prognosis than those who develop LGE later

Methods



- Retrospective lifetime chart review
- Inclusion criteria: pediatric patients with DMD in our center seen between January 2009 and July 2013 with at least one MRI
- Exclusion criteria: inability to determine group identification

Imaging Data

- Primary Outcome: fractional shortening (FS)
- Secondary Outcome: LV dimensions by echo and MRI

Clinical Data

- Patient age, genetic testing, cardiac medications, ambulation status
- Heart rate, BMI

Results

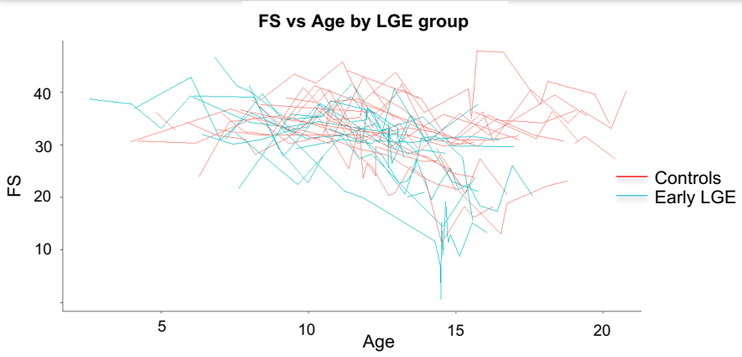


Figure 1: Controls experienced a significantly different average change of FS per year compared to "early LGE" patients (-0.57% vs -1.11%, **p-value = 0.004**)

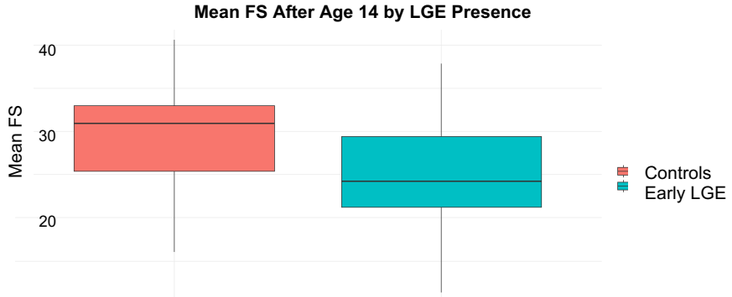


Figure 2: Mean FS was significantly lower for patients with "early LGE" compared with the control group (24.89% vs 29.78%, **p-value = 0.028**)

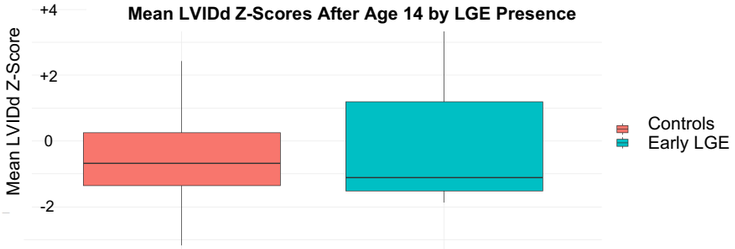


Figure 3: Mean LV internal diameter in diastole Z-scores were not significantly higher for patients with "early LGE" (**p-value = 0.3466**)

Results

- No significant differences between the "early LGE" and control groups regarding age of ambulation loss (**p-value = 0.31**), presence of obesity (**p-value = 0.32**), average heart rate (**p-value = 0.8**) or genetic etiology (**p-value = 0.3**)

Median Age at Initiation of Cardiac Medications

	"Early LGE"	Controls	P-value
ACE Inhibitor	8.4	10.56	0.025
Mineralocorticoid receptor antagonist	13.4	19.1	0.0024
Beta Blocker	13.4	19.1	0.0017

Table 1: Compared to controls, patients with "early LGE" were started on Angiotensin-Converting Enzyme (ACE) inhibition, mineralocorticoid antagonism, and beta blockade at a younger age

Discussion

- Pre-clinical changes are detectable by cardiac MRI, even if systolic function via echo appears normal
- Early onset of LGE, before adolescence, is associated with earlier progression of cardiac dysfunction
- Genetic etiology and severity of skeletal muscle weakness did not correlate with earlier progression of cardiomyopathy
- In our study, detection of LGE by MRI likely led to earlier initiation of ACE inhibitors, mineralocorticoid receptor antagonists, and beta blockade

Conclusion

- **Early onset of cardiac myocardial fibrosis as indicated by LGE on cardiac MRI is associated with earlier progress to cardiac dysfunction**
- **Our data supports the standard use of cMRI as part of a personalized medical approach in the preadolescent stage for DMD patients prior to the onset of overt cardiomyopathy**

References

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