

## Blinded Late Gadolinium Enhancement Quantification of Age Matched Adolescent and Young Adult Becker and Duchenne Muscular Dystrophy Cardiomyopathy

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### Type of submitter

Fellow in Training

### Abstract

**Background:** Becker Muscular Dystrophy(BMD) cardiomyopathy varies widely in regards to onset, severity, and progression. Evidence-based treatment of myocardial fibrosis attenuates fibrosis progression associated with left ventricular ejection fraction(LVEF) decline. However, a lack of early natural history complicates risk stratifying this orphan cardiomyopathy population. Cardiac magnetic resonance imaging(CMR) detects precursor myocardial fibrosis using late gadolinium enhancement(LGE) and is established in Duchenne Muscular Dystrophy(DMD) cardiomyopathy surveillance. We hypothesized that a sub-population of adolescent and young adult BMD patients will display similar LGE compared with age matched DMD patients.

**Methods:** We retrospectively reviewed age-matched CMR reports of 35 DMD and 33 BMD patients. Demographic data, initial volumetric analysis, and dichotomized LGE status were collected. In LGE positive studies, the LGE percentage of a basal, papillary, and apical segment was blindly quantified by the Full-Width Half Maximum technique. Nine studies without all three segments of appropriate quality for quantification were deemed inadequate. We quantified LGE in 10 BMD and 20 DMD studies.

**Results:** There was no significant difference in age(BMD  $17.4 \pm 6$ ; DMD  $18.3 \pm 3.9$  years), height, weight, left ventricular ejection fraction(LVEF)(BMD  $55.1 \pm 12\%$ ; DMD  $53.3 \pm 10.7\%$ ) or right ventricular ejection fraction(BMD  $59.7 \pm 5.9\%$ ; DMD  $59.5 \pm 5.7\%$ ) between the BMD and DMD groups. 12/33(36%) BMD patients and 25/35(71%) DMD patients were LGE positive. The youngest LGE positive BMD patient was 15 years. The percent LGE for BMD and DMD patients was  $26.3 \pm 10.6\%$  and  $23.0 \pm 9.5\%$  respectively(p-value 0.865). The Pearson correlation coefficient for the relationship of percent LGE to LVEF for all patients was -0.52(p-value < 0.005).

**Conclusions:** This study is the youngest and largest reported cohort of BMD cardiomyopathy CMR data. One third of BMD adolescent and young adults have myocardial fibrosis detectable by CMR. The fibrosis burden of affected BMD patients is similar to affected DMD patients and inversely related to LVEF. CMR based risk-stratification of adolescent BMD patients should be considered.

### Categories

Advanced Fellow: Research

### Program Name

Nationwide Children's Hospital

## Left Ventricular Outflow Tract Obstruction Leads to Decreased Myocardial Strain in Right Ventricles: Comparison of Coarctation of the Aorta, Aortic Stenosis, Hypoplastic Left Heart Syndrome and Normal Patients

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### Abstract

**Background:** Myocardial strain has been used in acquired and congenital heart disease to detect occult ventricular function in the setting of normal ejection fraction. Left ventricular (LV) strain abnormalities have been shown to be abnormal in various left-sided obstructive disease. There is however, paucity of right ventricle (RV) strain data in this population. Recently, RV strain was shown to be abnormal in patients with hypoplastic left heart syndrome (HLHS) at various stages of repair. Early detection of dysfunction may improve understanding of peri-operative management. LV size is variable in patients with left-sided obstructive disease including HLHS, coarctation of the aorta (CoA) and aortic stenosis (AS). The size and function of the LV as a result of the ventricular-ventricular interaction can impact RV function. The effect of the spectrum of LV outflow tract obstruction on RV strain has not been previously evaluated.

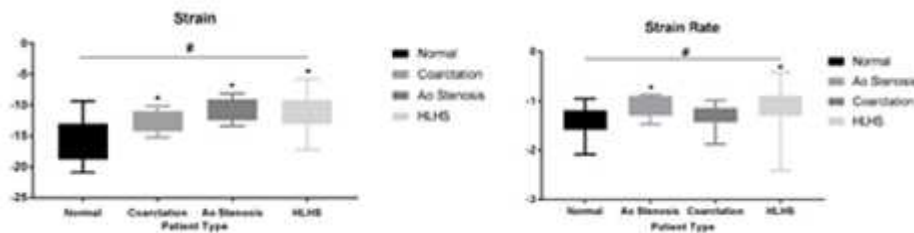
**Objective:** We sought to determine if there is progressive RV strain decline with degrees of LV outflow tract obstruction.

**Methods:** Retrospective review of pre-operative echocardiograms from patients with CoA, AS, HLHS and normal patients were analyzed using TomTec<sup>®</sup> strain software from a long axis (4 chamber) based on previously described technique. Statistical analysis was performed to compare longitudinal strain and strain rate values across all four groups using t-tests and one-way ANOVA via Graph Pad<sup>®</sup> software (Prism<sup>®</sup>).

**Results:** A total of 80 patients were analyzed (8 CoA, 8 AS, 40 HLHS, 24 normal) and all echocardiograms were obtained within the first week of life. Longitudinal strain and strain rate were significantly decreased ( $p < 0.0001$ ,  $p = 0.018$ ) across all populations (Figure 1). Strain magnitude was lower in CoA, AS and HLHS compared to normal ( $p = 0.03$ ,  $p = 0.0007$ ,  $p < 0.0001$ ). Similarly, strain rate was decreased in the AS and HLHS groups ( $p = 0.02$ ,  $p = 0.006$ ), but not CoA, compared to normal. There were, however, no differences in strain or strain rate between the three groups.

**Conclusions:** These results show a correlation of LV outflow tract obstruction with lower strain, despite normal function. As RV function is an independent risk factor for survival in lesions with LV outflow tract obstruction, having a pre-operative myocardial strain abnormality suggests that the at-risk RV may benefit from initiation of cardio-protective therapy. Future longitudinal studies are needed to determine the utility of myocardial strain to predict outcomes.

**Figure 1:** Strain and strain rate are significantly decreased across all populations of LV outflow tract obstruction, as denoted by # ( $p < 0.05$ ). A significant difference between individual groups compared to normal is designated by \* ( $p < 0.05$ ).



**Categories**

3rd year Fellow: Research

**Program Name**

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