of redundant collateral vessels. No pre-operative cardiac catheterization was performed. The 3D print model was available in the cardiac catheterization laboratory during percutaneous coiling of 2 of 3 known aortopulmonary collaterals. The third was found to be atretic as shown in Figures 1E and 1F. This process helped focus catheter-based intervention and reduced the amount of fluoroscopy time and contrast exposure. The central shunt remains in place to promote growth of confluent native pulmonary arteries. The patient is awaiting complete repair. Historically, CT angiography has been used infrequently because it does not measure hemodynamics or provide images with high enough resolution to allow for pre-operative planning (3). However, as seen in Figure 1, 3D printing of CT datasets may provide significant advantages in pre-operative and pre-procedural planning. This method can allow for reductions in general anesthesia exposure, fluoroscopy time, and cardiopulmonary by pass time. Three-dimensional prints can also be used as didactic tools to help educate parents and patients about patient-specific cardiac anatomy and educate trainees about specific cardiac anomalies. Integration of this rapidly developing technology within cardiovascular centers needs further study but has the potential to revolutionize diagnosis and therapy.

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Mechanical Dispersion by Strain Echocardiography: A Predictor of Ventricular Arrhythmias in Subjects With Lamin A/C Mutations

Sudden death is the first symptom of heart disease in many patients. Prevention of sudden death must therefore be achieved by screening groups of high-risk individuals and offering an implantable cardioverter-defibrillator (ICD) as the primary prevention therapy in selected individuals. Selecting patients for primary ICD therapy remains challenging. Patients with mutations in the lamin A/C gene (LMNA) constitute a small but important part (5% to 8%) of those with familial dilated cardiomyopathy. The cardiac phenotype is malignant and characterized by atrioventricular block, as well as supraventricular and ventricular arrhythmias, which often precede cardiac dilation (1). General ICD guidelines for dilated cardiomyopathy patients, recommending primary prevention ICD at ejection fraction (EF) <30% to 35%, are not appropriate in patients with LMNA mutations, and we have recently reported on risk predictors in this population (1). Dispersed myocardial contraction by strain echocardiography, mechanical dispersion, is an excellent marker of ventricular arrhythmias and is independent of EF (2,3). We hypothesized that mechanical dispersion by strain echocardiography may be a marker of ventricular arrhythmias beyond EF in patients with LMNA mutations and therefore serve as a risk marker of ventricular arrhythmias before EF decrease.

We included 33 LMNA mutation-positive subjects (35 ± 16 years), of which 13 (39%) were probands and 20 (61%) were mutation-positive family members. At presentation, 9 (27%) fulfilled criteria of dilated cardiomyopathy whereas 24 (73%) had normal or mildly dilated cardiac dimensions. Ventricular arrhythmias were defined as nonsustained (ns) ventricular tachycardia (VT) if ≥3 consecutive ventricular beats ≥120 beats/min lasting <30 s (n = 6). Sustained VT was defined as VT lasting >30 s, VT with hemodynamic deterioration or ventricular fibrillation (VF) (n = 5). No VT were detected in 22 subjects. All subjects gave written informed consent in this institutional review board-approved study.

By speckle tracking echocardiography, peak longitudinal negative strain was assessed in 16 left ventricular segments and averaged to global longitudinal strain (GLS). Time intervals from start Q/R on electrocardiogram to peak negative strain during the
The cardiac cycle was assessed. Mechanical dispersion was defined as the standard deviation of this time interval from 16 left ventricular segments, reflecting myocardial contraction heterogeneity (4,5).

Mean EF was 51 ± 11%, GLS -20.4 ± 3.8%, and mechanical dispersion 42 ± 12 ms. As expected, subjects with sustained VT/VF had reduced cardiac function by EF and GLS when compared with patients without sustained VT/VF (p = 0.001 and p = 0.01, respectively) and mechanical dispersion was pronounced (p = 0.001) (Figure 1). Only mechanical dispersion was increased in those with any arrhythmic events (nsVT + sustained VT, n = 11) versus patients free of arrhythmic events (n = 22) (49 ± 14 ms vs. 38 ± 10 ms, p = 0.02), whereas EF and GLS were not different (EF: 47 ± 15% vs. 52 ± 7%, p = 0.14; GLS: -19.2 ± 5.3% vs. -21.0 ± 2.9%, p = 0.22). There was a significant linear trend toward an increase of mechanical dispersion by severity of arrhythmias (p = 0.001) (Figure 1). By receiver-operating characteristic analyses, mechanical dispersion showed good discrimination of those with nsVT + sustained VT (area under the curve [AUC]mech disp: 0.74, 95% confidence interval [CI]: 0.55 to 0.93; AUCEF: 0.64, 95% CI: 0.41 to 0.86, and AUCGLS: 0.54, 95% CI: 0.30 to 0.78).

Mechanical dispersion may be an additive marker of ventricular arrhythmias in patients with relatively preserved ventricular function early in LMNA disease. This finding is in line with our previous results demonstrating mechanical dispersion as a predictor of ventricular arrhythmias in patients with EF >35%. Mechanical dispersion may overcome the limitation of EF, which predicts poor outcome only in those with obviously reduced ventricular function.

**FIGURE 1** Mechanical Dispersion in Lamin A/C Mutation Positive Subjects

(Top) Strain curves from apical 4-chamber view in a lamin A/C mutation-positive subject without (left) and with (right) ventricular tachycardia (VT). Arrows are indicating time from start Q/R on electrocardiogram to peak longitudinal strain. Mechanical dispersion is pronounced in the subject with VT (right). (Bottom left) Bar charts of relation between echocardiographic parameters and severity of VT. Patients with sustained VT/ventricular fibrillation (VF) (pink bars) had significantly higher values of mechanical dispersion and lower left ventricular ejection fraction (LVEF) and absolute global longitudinal strain (GLS) than did those with no VT (green bars). Mechanical dispersion showed a positive linear trend along with severity of VT (p = 0.001). (Bottom right) Receiver-operating characteristic (ROC) curves in 33 lamin A/C mutation positive subjects for the ability of mechanical dispersion, LVEF and GLS to discriminate between those without (n = 22) and with nonsustained VT/sustained VT/VF (n = 11).
Kawasaki disease (KD) is a generalized systemic vasculitis, although the coronary artery system is typically involved. Coronary artery lesions (CAL) develop in 25% of children during the acute stage of KD and may lead to infarction, sudden death, or chronic coronary artery insufficiency (1). In about 50% of cases, complete, spontaneous resolution occurs within 1 year after onset (2).

The purpose of this study was to evaluate myocardial perfusion reserve with the use of perfusion cardiac magnetic resonance (CMR) in children with a previous history of KD and coronary involvement and correlate it with coronary morphologic abnormalities.

Fourteen asymptomatic patients with history of KD and coronary involvement (male patients, n = 8; mean age 10.2 ± 7.2 years) were prospectively examined with CMR using a 1.5-T magnetic resonance (MR) unit (Intera, Philips Healthcare, Best, the Netherlands). Electrocardiography-gated 2-dimensional steady-state free precession images were acquired for function assessment. Stress/rest first-pass perfusion was assessed using a turbo fast low-angle shot sequence (in-plane spatial resolution: 2.5 × 2.5 × 10.0 mm; 3 short-axis slices during intravenous contrast medium infusion (gadobutrol 0.1 mmol/kg body weight) after administration of 140 μg/kg/min adenosine for 4 min and at rest, with a 15-min interval. To assess fibrosis, late gadolinium enhancement (LGE) images were acquired 10 min after (inversion recovery turbo fast low-angle shot). A 3-dimensional steady-state free precession sequence with T2 and fat saturation pre-pulses was used for MR angiography. Imaging was performed under anesthesia, if necessary, with continuous intravenous infusion of remifentanil and controlled ventilation.

Mean and segmental myocardial perfusion reserve index (MPRI), as defined by the ratio of stress to rest myocardial signal intensity relative upslope, were measured quantitatively (QMass MR 7.5, Medis, Leiden, the Netherlands). Left ventricular endocardial and epicardial boundaries of the left ventricle were automatically outlined and manually edited for through-plane motion on a 16-segment model, in time series of short-axis cine MR images. An abnormal coronary artery was defined according to criteria established by the Japanese Ministry of Health. A 5-SD threshold above the mean remote myocardial signal was used to study LGE images.

Comparisons of continuous variables with a normal distribution were performed using the independent-sample Student t test. Correlation analysis was assessed using Pearson correlation. The coefficient of variation was calculated to study the variability of the measurements. Data are expressed as mean ± SD unless otherwise specified. Values of p < 0.05 were considered statistically significant.

In 8 of 14 patients, CMR was performed under anesthesia. Persisting CAL were identified with MR angiography in 5 patients. Inducible perfusion defect by visual assessment was detected in 1 patient. LGE identified myocardial scar in 1 patient. Mean MPRI was significantly impaired in all patients, compared with historical pediatric control subjects (0.86 ± 0.256 vs. 2.46 ± 0.3, p < 0.001, 1 sample Student t test) (3). No significant difference in mean MPRI was identified between patients with regressed CAL (9 of 14) and persistent CAL (5 of 14) (1.0 ± 0.3 vs. 0.79 ± 0.225; p = 0.31). In patients with persisting CAL, no differences in MPRI were demonstrated between segments subtended by arteries with regressed and persistent CAL. Patients’ clinical and CMR characteristics are presented in Table 1.

Previous histologic studies in the acute and chronic phase of KD have confirmed inflammatory cell