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Usefulness of myocardial strain imaging in Spinal Muscular Atrophy

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BACKGROUND:
In paediatric patients with Spinal Muscular Atrophy (SMA), cardiac systolic function is generally described to be within the normal range. Some studies have suggested the presence of subclinical dysfunction in congenital muscular diseases using tissue Doppler measurements and myocardial velocity gradients. This has not been described for SMA. The aim of this study was to further assess regional myocardial function in paediatric patients with SMA using myocardial velocity and deformation imaging.

METHODS:
Thirty-one patients with SMA (mean age, 7.2 years; range, 0-12 years) and 29 age-matched normal controls were studied with echocardiography. Standard echocardiographic measurements of left ventricular (LV) systolic and diastolic function were performed. Myocardial velocity and deformation data, including peak systolic and early and late diastolic myocardial velocities, peak systolic strain rate (SR), and peak systolic strain, were calculated in the radial direction in the inferolateral LV wall and in the longitudinal direction in the interventricular septum, the LV anterolateral wall, and the right ventricular (RV) free wall.

RESULTS:
Higher heart rates and increased LV end-systolic dimensions were seen in patients with SMA compared with controls. Significant decreases in radial and longitudinal peak systolic SR, peak systolic strain, and peak systolic and early diastolic myocardial velocities were found in the LV inferolateral and anterolateral walls in patients with SMA. No significant differences in longitudinal function could be found in the interventricular septum or in the RV free wall.

CONCLUSION:
In young patients with SMA who have global normal systolic function, reductions in systolic deformation parameters as well as reduced early diastolic myocardial velocities can be detected in the anterolateral and inferolateral LV walls. The prognostic significance of these findings warrants further longitudinal follow-up.