1550157

Quantification of Myocardial Iron Deposition by Two-dimensional Speckle Tracking in Patients with β-Thalassemia Major and Blackfan-Diamond Anemia

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Background

Cardiac disease related to transfusional iron overload is the leading cause of death in patients with β-thalassemia major. Identifying early myocardial iron deposition is important since significant cardiac involvement may predate a decrease in left ventricular systolic function. Cardiac magnetic resonance (CMR) is currently the only noninvasive examination to quantitate iron deposition within the myocardium.

Methods

We retrospectively analyzed myocardial mechanics by echocardiography using two-dimensional speckle tracking in 26 chronically transfused patients with β -thalassemia major or Diamond Blackfan Anaemia and compared it to cardiac magnetic resonance T2 star (T2*) calculations . We divided our population in two groups: patients with an interventricular T2* value <=20 msec (low T2*) and patients with >20 msec (normal T2*), which indicates a clinically insignificant degree of iron deposition. They were compared to a control group of 18 patients, age and gender matched, with a normal echocardiogram and no history of hemoglobinopathy.

Results

Patients with significant myocardial iron deposition (low T2*) had a uniform decrease in longitudinal and circumferential strain compared to normal controls (-15±2% vs -19±2% and -20±5% vs -25±4%, respectively, p<0,007). Furthermore, peak twist and peak apical rotation were significantly lower in low T2* vs either normal T2* or normal control patients. Conversely, no significant difference was observed when comparing normal T2* to normal controls. There was a strong and direct correlation between average global longitudinal strain and T2* values (r=-0.75, p=0.0001). Using a cut-off of <-17 %, global longitudinal strain predicted a T2* value of less than 20 msec with a sensitivity of 92% and a specificity of 77%.

Conclusion

Myocardial mechanics, asensitive marker of myocardial dysfunction offers a simple alternative to cardiac MRI for assessing patients for significant myocardial iron dep