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Cardiac MR assessment of myocardial iron deposition in sickle cell disease: risk factors and association with cardiac function

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Introduction

Pediatric patients with sickle cell disease (SCD) who receive regular transfusions are at risk for developing cardiac toxicity from iron overload. Although studies have demonstrated risk factors for liver iron overload in these patients, the correlation with cardiac iron deposition has not been well established.

Purpose

To assess the correlation between myocardial iron deposition as assessed by T2* sensitive MRI and several functional and biochemical measures in a pediatric SCD population.

Methods

22 patients with SCD (Hb SS) at a steady state had T2* sensitive cardiac MRI (1.5 T Avanto™, Siemens) from January 2009 to August 2009. All patients were receiving regular transfusion every 3-4 weeks and chelation therapy. For each patient, a T2* value obtained from the interventricular septum. Cardiac T2* values of > 25 ms were recorded as normal, 20-25 ms as borderline and < 20 ms as abnormal.

Results

Median myocardial T2* was 32.1 + 6.5 ms. Mean myocardial T2* values were significantly lower than T2* values measured in the paravertebral muscles in the same patients (32.1 ± 6.5 versus 48.4 ± 5.2, $p < 0.01$). 1 patient (5%) had an abnormal T2* value of 17 ms, while two patients (9%) had borderline low values. Myocardial T2* weakly negatively correlated with chronological age($r = -0.46$, $p = 0.81$). Although there was a strong correlation between liver biopsy iron concentration and transfusion burden ($r = 0.63$, $p = 0.002$) and ferritin ($r = 0.72$, $p = < 0.001$), transfusion burden was only weakly correlated with cardiac T2* ($r = -0.27$, $p = 0.29$) and ferritin levels ($r = -0.42$, $p = 0.11$). The left ventricular end-diastolic volume and ejection fraction showed a weak correlation with T2* measurements ($r = 0.17$ and, $r = 0.21$ respectively).

Conclusion

Myocardial iron deposition cannot be predicted by liver biopsy or indirect biochemical measures and therefore cardiac MRI may be indicated for serial monitoring of SCD patients.