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Long-term longitudinal prospective CMR study in patients with thalassemia major

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Introduction. We aimed to determine longitudinal changes in cardiac iron and function assessed by cardiovascular magnetic resonance (CMR) over 6 years in a large cohort of thalassemia (TM) patients.

Methods. We considered 426 TM patients (205 males; 30.87±8.21 years) consecutively enrolled in the MIOT (Myocardial Iron Overload in Thalassemia) Network with a CMR follow-up (FU) study at 72 months (6 years).

The T2* in all 16 myocardial segments was quantified and 4 patterns of myocardial iron overload (MIO) were identified: no MIO (all segments with T2*≥20 ms), heterogeneous MIO (some segments with T2*≥20 ms and other segments with T2*<20 ms) and global heart T2*≥20 ms, heterogeneous MIO and global heart T2*<20 ms, and homogeneous MIO (all T2*<20 ms). Risk classes were defined on the basis of the patterns of MIO from worst to normal: homogeneous MIO → heterogeneous MIO with global T2*<20 ms → heterogeneous MIO with global T2*≥20 ms → no MIO.

Biventricular function was quantified by cine images.

Results.TheFigure shows the frequency of the 4 patterns at both scans

In the 254 patients with baseline MIO (at least one segment with T2*<20 ms), the following changes were detected at the FU:

- improvement, defined as a transition to a better risk class, in 182 (71.7%);

- stabilization, defined as no change in the risk class, in 62 (24.4%);

- worsening, defined as a transition to a worse risk class, in 10 (3.9%).

Among the 172 patients without baseline MIO, 30 (17.4%) worsened, that is developed MIO at the FU.

Biventricular end-diastolic volume indexes (EDVI) were significantly lower at the FU CMR. In patients with significant baseline MIO (global heart T2*<20 ms) a significant decrease in all biventricular volumes and a significant increase in left ventricular ejection fraction (EF) (mean difference: 3.83±8.48%, P<0.0001) as well as in right ventricular EF (mean difference: 1.79±9.04%, P=0.042) were detected with a concordant improvement of MIO status.

The 50.7% of the patients changed the type of chelator during the FU based on CMR results. The percentage of patients who changed the chelation therapy was significantly higher in patients with significant MIO than in patients without MIO (60.2% vs 46.2%; P=0.008).

Conclusion. Over a period of 6 years, the continuous monitoring of cardiac iron levels and a tailored chelation therapy allowed an improvement in more than 70% of patients with baseline MIO and a consequent improvement of biventricular function.