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CMR survey in Thalassemia Intermedia patients

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Background

Little is known about cardiac involvement in thalassemia intermedia (TI) using cardiovascular magnetic resonance (CMR). We investigated myocardial iron overload (MIO), biventricular parameters, and myocardial fibrosis in a large cohort of TI patients, underlying the differences between transfusion-dependent and non-transfusion-dependent patients.

Methods

We studied 252 adult TI patients (119 females, 39.5 ± 10.4 years) enrolled in the MIOT Network. MIO was assessed using a multislice multiecho T2* approach. Biventricular function parameters were quantified by cine sequences. Myocardial fibrosis was evaluated by late gadolinium enhancement acquisitions.

Results

One-hundred and eighty-eight (74.6%) patients showed no MIO in any segment, 56 (22%) had an heterogeneous distribution (52 with global heart T2*≥20 ms), and 8 (0.3%) showed an homogeneous MIO. Left ventricular (LV) and right ventricular (RV) dilatations were present in 113 (45%) and in 49 (19%) patients, respectively. LV dysfunction was present in the 18.0% of the cases while RV dysfunction in the 3.63%. High LV mass indexes were present in 22 (8.7%) patients. Fifty-two/227 (22.9%) patients showed myocardial fibrosis. Myocardial fibrosis was associated to LV dysfunction (P = 0.001) and high mass indexes (P = 0.038). One-hundred and fourteen patients were non-transfusion dependent (transfusion requirements absent or sporadic) while 138 patients were transfusion-dependent (regular transfusions). The mean age at start of chronic transfusions was 11.8 ± 12.3 years. Table 1 shows the comparison between the two groups. Non-transfusion-dependent patients showed significantly higher global heart T2* values and MIO with a global heart T2* < 20 ms was detected in two of them (one requiring occasional blood transfusions and one non transfused). Biventricular end-diastolic volume index, stroke volume index, left ventricular (LV) mass index, and LV cardiac index were significantly higher in the non-transfusion dependent group.

Conclusions

CMR plays a key role in the management of TI patients. Heart iron (global heart T2* < 20 ms) was not common, but a quarter of the patients had some pathological segments. A consistent number of patients had the stigmata of the high cardiac output state cardiomyopathy. Myocardial fibrosis was related to the high cardiac output state. The signs of the high output state were controlled in the transfusion-dependent-patients.

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Table 1

	Non-transfusion-dependent	Transfusion-dependent	P
Age (years)	39.9 ± 11.5	39.2 ± 9.4	0.922
Sex (M/F)	67/47	66/72	0.083
Global heart T2* (ms)	38.8 ± 6.7	35.5 ± 9.2	0.014
MIO pattern, N (%):			
No MIO	92 (80.7)	96 (69.6)	0.103
Heterogeneous MIO with global T2* ≥ 20 ms	20 (17.5)	32 (23.2)	
Heterogeneous MIO with global T2* < 20 ms	1 (0.9)	3 (2.2)	
Homogeneous MIO	1 (0.9)	7 (5.1)	
LV end-diastolic volume index (ml/m2)	99.4 ± 19.6	92.9 ± 19.1	0.009
LV end-systolic volume index (ml/m2)	36.6 ± 11.4	34.9 ± 10.4	0.249
LV stroke volume index (ml/m2)	62.9 ± 12.4	58.6 ± 13.1	0.007
LV mass index (g/m2)	69.9 ± 13.9	63.9 ± 12.9	0.004
LV ejection fraction (%)	63.7 ± 6.8	62.5 ± 6.6	0.163
LV cardiac index (L/min/m2)	7.6 ± 2.3	6.5 ± 2.2	0.002
LGE, N (%)	20/105 (19)	32/122 (26.2)	0.199
RV end-diastolicvolume index (ml/m2)	92.0 ± 23.3	86.5 ± 20.8	0.048
RV end-systolic volume index (ml/m2)	32.7 ± 14.9	31.8 ± 11.3	0.571
RV stroke volume index (ml/m2)	58.5 ± 14.9	54.5 ± 14.3	0.017
RV ejection fraction (%)	64.7 ± 8.3	63.3 ± 7.5	0.168

MIO = myocardial iron overload; LV = left ventricular; RV = right ventricular

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