EXTRAMEDULLARY HEMATOPOIESIS IS ASSOCIATED WITH A THALASSAEMIA INTERMEDIA-LIKE PATTERN OF MYOCARDIAL AND LIVER IRON LOADING IN REGULARLY POLYTRANSFUSED THALASSAEMIA PATIENTS

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Introduction. Extramedullary hematopoiesis (EMH) is an incidental finding in regularly and historically polytransfused thalassaemia patients[1] but no study has evaluated if it is a marker of a peculiar pattern of iron loading. We studied the relationship between EMH and Magnetic Resonance Imaging (MRI) findings.

Methods. 1266 thalassemia patients (pts) regularly transfused (655 F; 31.25 ± 8.86 years) consecutively enrolled in the Myocardial Iron in Thalassemia (MIOT) Network[2] were considered.
MRI was used to assess the presence of EMH by SPGR sequences, to quantify cardiac and hepatic iron overload by a multiecho T2* approach,[3] and to assess cardiac function, volumes[4] and pulmonary diameter by SSFP sequences. Myocardial fibrosis was evaluated by LGE technique.[5]

Results. EMH was detected in 167 pts (13.2%). No significant differences were found in the chelation regimens between the two groups.
EMH+ pts had significant less cardiac iron overload than EMH- patients (13.2 vs 28.3% of pts with global heart T2*<20 ms, P=0.003; Figure 1).
The MRI liver iron concentration (LIC) was significantly lower in the EMH+ patients than EMH- pts (6.23 ± 8.13 vs 9.23 ± 11.71 mg/g/dw, P=<0.0001; Figure 2). Considering the 482 (38.1%) patients with MRI LIC≥7 mg/g dw, the EMH+ group had a significant lower frequency of global heart T2*<20 ms (18.4% vs 40.8% p=0.007).
Biventricular volumes indexed by body surface area, cardiac index, ejection fractions, atrial areas and presence of myocardial fibrosis were comparable between the two groups. EMH+ patients had a significantly higher LV mass index (62.3±13.2 vs 58.63±13.19 g/m2, P=0.001; Figure 3) and a significantly higher pulmonary artery diameter (24.7±4.2 vs 23.6±3.8 mm; P=0.002) (Figure 4).

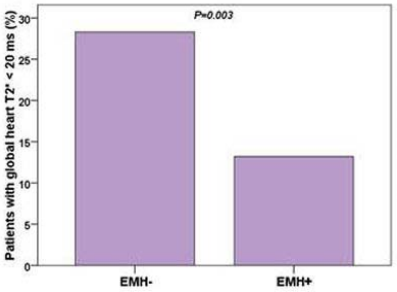

Figure 1

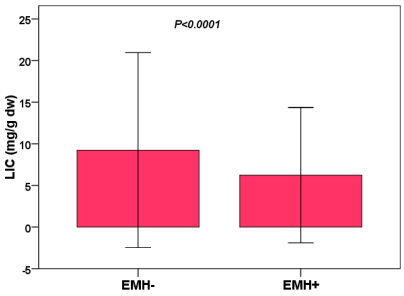

Figure 2

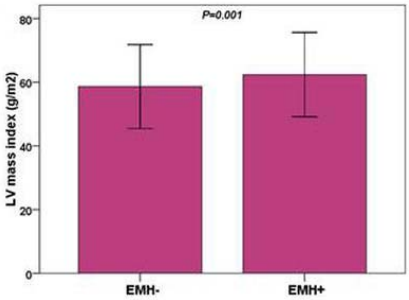

Figure 3

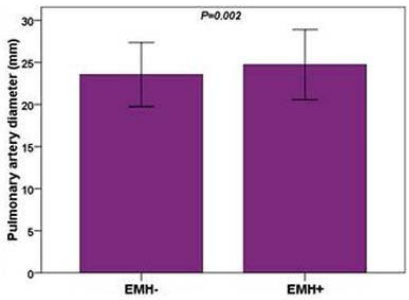

Figure 4

Conclusions. In this large cohort of regularly transfused thalassemia patients, EMH was not rarely observed and was associated to a heart thalassemia intermedia like pattern (reduced cardiac iron loading and stigmata of high cardiac output state) despite the transfusional regimen.

References. [1] Taher AT et al. Blood 2010;115:1886-92. [2] Meloni A et al. Int J Med Inform 2009;78:503-12. [3] Pepe A et al. JMRI 2006;23:662-8. [4] Marsella et al. Haematologica 2011;96:515–20. [5] Pepe A et al. Heart 2009;95:1688-93.