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Liver damage in Sickle Cell Disease: relevance of early markers

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Introduction.Liver damage is a severe complication in Sickle Cell Disease (SCD), so far, no effective approaches to prevent or treat it are available.

Aim.To identify early predictors of liver involvement.

Patients and methods.Seventeen Sickle Cell Anemia (SCA), 38 Sickle Cell Thalassemia (HbS-βThal) and 13 HbS/HbC patients with two Stiffness data (Transient Elastography TE) (T0 and T1), were evaluated (2007-2016). Clinical manifestations, therapy, laboratory tests, abdominal ultrasound (US) and Magnetic Resonance Imaging (MRI) T2* were recorded

Results. AST, ALT, LDH were higher in SCA pts than in HbS-βThal and HbSC (ALT p<0.0001) and in HbS-βThal compared to HbSC (ALT p=0.01). GGT, ALP were higher in SCA than in HbS-βThal and HbSC (GGT p=0.013; ALP p=0.006), and without significance in HbS-βThal compared to HbSC (GGT p=0.23; ALP p=0.44). Liver synthesis indices were similar; none was HbsAg nor HCV-RNA positive. TE Stiffness was higher in SCA (KPa 8.3±6.86) than in HbSC pts (KPa 5.33±2.15; p=0.014) and in HbS-βThal (KPa 6.17±2.58) comparing SCA and HbSC pts (p=0.2). Liver Iron Concentration (LIC) was higher in HbS-βThal than in SCA and HbSC pts (HbS-βThal vs HbSC p=0.0145) and in SCA comparing HbSC (p=0.018). At univariate analysis GGT correlates with ferritin (p=0.02), TE (p=0.002), US (p=0.107) and LIC (p=0.511) in all SCD pts. A correlation between GGT and US liver echogenicity was in SCA and HbS-βThal pts; TE and US (p=0.045) correlated positively in all SCD pts. No differences were found in TE and MRI T2* at T0 and T1. US showed statistic differences at T0 compared with T1 in HbS-βThal (p=0.04) and in HbSC pts (p=0.001), but not in SCA pts (p=0.46) probably because of higher Stiffness since T0. The 76.5% of SCA, 60.5% of HbS-βThal and 30.8% of HbSC pts had >1 vaso-occlusive crisis (VOCs)/yr (SCA vs HbSC p=0.02; SCA vs HbS-βThal p=0.36; HbS-βThal vs HbSC p=00.1). Occasional transfusions (<4 RBCs Units/yr) occurred in 88.2% of SCA, 84.2% of HbS-βThal and 61.5% of HbSC pts. The 58.8% of SCA, 65.8% of HbS-βThal and 15.4% of HbSC pts took HydroxyCarbamide and 23.5% of SCA, 23.7% of HbS-βThal and none of HbSC pts took iron-chelators.

At multivariate analysis sex (male), low HbF values, high ferritin values, more severe sickle genotype as predictors of liver involvement were found independent risk factors.

Conclusions. Function liver tests with US, TE and MRI T2* taking into account sex, HbF, and SCD genotypes, are useful to early detect sickle hepatopathy.