

Liver and spleen assessment in post-Fontan pediatric patients

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INTRODUCTION

Fontan-associated liver disease (FALD) is one of the consequences of Fontan hemodynamics affecting long-term prognosis. Magnetic resonance elastography (MRE), a relatively new imaging technique measuring tissue stiffness, provides estimation of liver fibrosis and congestion. Spleen stiffness is considered to be dependent on the degree of liver fibrosis and can be also evaluated by MRE. Moreover, its degree should correlate with the risk of hypersplenism and esophageal varices bleeding. To the best of our knowledge, we describe the first reported data of spleen involvement in post-Fontan pediatric patients.

METHODS AND MATERIAL

Detailed follow up (both cardiac and multi-organ evaluation) was planned to assess long term consequences of Fontan hemodynamics in a group of 30 pediatric patients. Hepatic and spleen screening involved MRE, abdominal ultrasound and laboratory studies. Present study include a group of 11 patients with hepatic and spleen screening data available. We evaluated the presence of liver and spleen pathology and analysed their importance for post Fontan patients.

RESULTS

There were patients 5 to 12 years post-completion, 9 of them with systemic left ventricle. All patients had elevated spleen and liver stiffness values, the first being higher in all but two patients. Mean spleen stiffness was 5,8kPa with maximum 10kPa comparing to mean liver stiffness 4,34kPa with maximum 5,2kPa (reference range for liver stiffness is 1.54 to 2.87 kPa, for spleen slightly larger but not yet established). Splenomegaly and/or hypersplenism was detected in 4 patients, esophageal varices in 1 patient. Correlation between these findings and severity of spleen or liver stiffness was not confirmed. Moreover, 1 patient had splenic haemangioma, but the relation of this finding to Fontan circulation is unknown.

CONCLUSIONS

Magnetic resonance elastography is promising in assessment of liver as well as spleen in Fontan circulation. Presence of advanced spleen pathology suggests to implement screening for hypersplenism and esophageal varices in routine long-term follow up of Fontan patients. As this very first reported results are potentially important, they should be confirmed in future studies.