A surveillance for advanced liver disease and characterization of hyperechogenic lesions after Fontan surgery in a prospective cohort of cases with congenital univentricular heart

L. Chemello¹, C. Giraudo¹, R. Motta¹, R. Biffanti¹, C. Tessari¹, M. Padalino¹, L. Cavalletto¹

¹University of Padua, Padova, Italy

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Background: The congenital univentricular heart (UH) is a rare disease corrected with the palliative Fontan’s intervention, that restores the survival of most child and young patients up to adulthood. Unlike, these patients develop a series of complications, among all, the Fontan associated liver disease (FALD), that causes the occurrence of liver cirrhosis and hepatocellular carcinoma (HCC), that negatively impacts the life expectancy in the majority of cases.

Purpose: Our primary aim was to identify the presence of severe FALD in our patients by liver fibrosis and portal hypertension scores (FORNS, liver and spleen-platelet score or LSPS and SSPS), and by liver (LS) and spleen (SS) stiffness measurement in relation to time since Fontan, and secondarily to characterize all the abdominal-US hyperechogenic lesions (US-HL) according to the LI-RADS (LR) CT/MRI categories.

Method: One-hundred-46 outpatients (83 m/63 f, aged 25.2±11.0 yrs) born with UH corrected with Fontan circuit have been recruited prospectively from 2019 to 2023 (FU mean 24.5±15.1 mos). Each patient underwent a complete clinical history and instrumental examination, by imaging obtained with US, CT and MRI, a specific lab-test profile, and the LS and SS evaluation by transient elastography (VCTE, Fibroscan). ROC curves were built to select the scores cut-off proved to be the most accurate to diagnose and grade FALD. Based on the presence of US-HL found in 43 cases (29,5%), we grouped: 23 cases with LR-1 or 2 (benign), 15 with LR-3 (suspect) and 5 with LR-4 or 5 (malign).

Results: Features for definition of failing-Fontan appeared in 17.5%, 47.5% and 80% of cases with <15; 15-25 or >25yrs since Fontan surgery, respectively (p<0.01). All the scores used (FORNS with cut-off ≥4, LSPS ≥1.4, SSPS ≥2.3, LS ≥20.2 and SS ≥30.7) showed high sensitivity and specificity, improving FALD diagnostic accuracy and staging (all p<0.001). Moreover by identification of selective criteria related to advanced liver disease and portal hypertension, we applied a novel risk-score for failing-Fontan, that predicted a low-moderate risk profile (HR 0.30;CI 0.18-0.52) in 72 patients and a high risk profile (HR 3.29,CI 1.93-5.60) in 74 (51%). During FU, 13 patients (8.9%) died, 5 received OHT (3.4%) and 5 had HCC occurrence (3.4%). Finally, cases with US-HL stratified by classes LR-3-4-5 (20 cases, 46.5%) appeared significantly associated with a more advanced FALD (16/4, 75% vs. 68/58, 46%; p=0.004) and a longer time since Fontan (26.3±9.4 vs. 19.8±10.8; p=0.025).

Conclusion: The novel risk score proposed may help to predict the development of cirrhosis in high risk patients and focussed on the need for surveillance by imaging methods to characterized the US-HL, that appeared at risk of malignity (nodule size >20mm and/or presence of wash-out into portal-hepatocellular phase or into LR ≥3 classes).