

Evaluation of Fontan failure using the severity of FALD as secondary organ disease

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Introduction:

Despite improved survival, Fontan palliated patients are prone to failure of the Fontan circulation and second organ dysfunction in the form of Fontan-associated liver disease (FALD). In this study we established a graduation of FALD based on laboratory and ultrasonographic liver assessment and analyzed its applicability for definition of Fontan failure.

Methods:

Liver assessment was performed in 90 consecutive patients with a median age of 17.3 years [IQR 14.9]. The extend of FALD was graded as mild, moderate and severe due to laboratory parameters (liver enzymes, Fibrotest®), liver ultrasound and liver stiffness measurement (Fibroscan®).

Hemodynamic assessment was performed using echocardiography and cardiac catheterization.

Fontan failure was defined as active protein-losing enteropathy, impaired cardiopulmonary exercise capacity measured by spiroergometry with a VO₂max below 45 % of the age adjusted standard value or more than 2 hospitalisation periods due to cardiopulmonary decompensation within 12 months.

Results:

FALD was graded as mild, moderate and severe in 46, 34 and 10 patients (51.1 %; 37.8 %; 11.1 %).

The extend of FALD significantly correlated with exercise capacity ($p < 0.001$) and systolic ventricular function based on echocardiography ($p = 0.003$). Invasive pressure measurement revealed a strong correlation between the extend of FALD and Fontan pressure ($p < 0.001$), mean pulmonary artery pressure ($p = 0.002$) and end-diastolic ventricular pressure ($p = 0.003$). Fontan failure was detected in 17 patients (18.9 %) and correlated with severity of FALD ($p < 0.001$). Death occurred in 4 patients with failing Fontan circulation, FALD was graded moderate in 1 and severe in 3 patients ($p = 0.004$).

Conclusion:

Detection of FALD is mandatory for Fontan surveillance and monitoring of the failing Fontan. Severity of FALD significantly correlates with exercise capacity, ventricular function and hemodynamics of Fontan palliated patients. FALD graduation might be a useful diagnostic for definition of Fontan failure and evaluation for heart transplant.