Does Fontan circulation engender progressive liver dysfunction?


Division of Pediatric Cardiology, Citta’ della Salute e della Scienza, Turin, Italy (1)
Division of Pediatric Gastroenterology, Citta’ della Salute e della Scienza, Turin, Italy (2)
Division of Pediatric Cardiac Surgery, Citta’ della Salute e della Scienza, Turin, Italy (3)

Introduction
Total cavopulmonary connection (TCPC) forces systemic venous blood into the lungs, equalizing caval and pulmonary pressure. Chronic hepatic stasis generates a progressive liver dysfunction, eventually leading to cirrhosis. We investigated prospectively the hemodynamic changes and the liver status after TCPC.

Methods
From March 2013 to December 2014, 64 TCPC patients (pts) underwent cardiac catheterization and liver examination (blood tests, ultrasound and gastroscopy) at our center.

Results
Median age was 10 (5-32) yrs, median distance from TCPC was 10 (1-19) yrs. Catheterization showed the following data: pulmonary arterial pressure (PAP) 11.6±2 mmHg (>15 mmHg in 10 pts), ventricular end-diastolic pressure (VEDP) 6.67±2.58 mmHg, pulmonary vascular resistances (PVR) 2±1 WUm2 (>2 in 18 pts); cardiac index (QSI) 3.15 ±1.27 ml/min/m2; systemic O2 saturation 94±4% (<95% in 18 pts), QP/QS 0.9±0.2.

37 interventions were performed in 27 patients. We found the following significant correlations: interval from TCPC vs QSI (-0.30, p<0.001), interval from TCPC vs VEDP (0.3, p<0.01), PVR vs QSI (-0.81, p<0.001). Nor PAP, PVR or QP/QS were significantly related with age or interval from TCPC. Trans-hepatic gradient was 2.33+1.10 mmHg; 20 pts had major venous collateral vessels from the liver. Gastroscopy showed oesophageal varices in 6 pts (0.09%) having PVR >2UWm2. Conversely, all pts with PAP >15 mmHg had either venous collaterals or oesophageal varices.

Liver function was normal in all pts. Hepatomegaly was found in 23 pts; the liver was nodular and/or inhomogeneous in 10 and 35 pts. Stiffness was 16.63 + 5.96 KPa and significantly related to time from TCPC (r:0.33, p<0.01).

A subgroup of patients showed a negative trend very early after TCPC.

Conclusions
This is the largest prospective series showing that TCPC engenders a progressive decrease of QSI and increase in VEDP, with a tendency to PAP and PVR to raise. This is balanced by the progressive development of venous collateral vessels, from both caval and hepatic systems. The hepatic stiffness increases with time, but cirrhosis and esophageal varices are found in few patients. Only a multidisciplinary approach will be able to identify patients at risk.