610 Fontan associated kidney and liver disease: can we predict organ involvement with echocardiographic assessment of systolic function and atrioventricular valve insufficiency?

Marta Rotella[1], Roberta Biffanti[1], Jolanda Sabatino[1], Davide Meneghesso[2], Nicola Bertazza Partigiani[2], Alessia Basso[1], Beatrice Binda[3], Liliana Chemello[4], Luisa Cavalletto[4], Massimo Padalino[5], and Giovanni Di Salvo[1]
[1]Paediatric Cardiology, Departement of Women's and Child's Health, University of Padova Medical School, 35128 Padova, Italy, [2]Paediatric Nephrology and Dialysis Unit, Department of Women and Child's Health, University of Padova Medical School, 35128 Padova, Italy, [3]University of Padova Medical School, 35128 Padova, Italy, [4]Internal Medicine and Hepatology Unit, Clinica Medica 5, Department of Medicine-DIMED, University of Padova Medical School, 35128 Padova, Italy, and [5]Paediatric and Congenital Cardiac Surgery Unit, Department of Cardiac, Thoracic, and Vascular Science and Public Health, University of Padova Medical School, 35128 Padova, Italy

Aims: Fontan operation represents the surgical palliative option for congenital heart disease with single ventricle physiology. With the improvement of surgical and percutaneous technique, we are facing a growing population of patients with an unique pathophysiology and potential complications.
Methods and results: Patients that underwent Fontan palliation in our centre between 1993 and 2016 were included in this prospective study. We excluded patients with major congenital renal anomalies, those that underwent cardiac transplantation, and *redo*-Fontan patients. All the subjects underwent clinical evaluation, laboratory exams with complete renal and hepatic function, transient hepatic elastography, and complete cardiac evaluation. We used Schwartz equation for estimating glomerular filtration rate in patients younger than 18 years, and CDK-EPI equation for adult patients. We enrolled 35 patients, 46% female ($N = 16$), and 54% male ($N = 19$). Medium age was 17 years old, median age 15 years old (range: 10-31 years old). Medium time from Fontan completion was 160 months (range: 57-340 months). Regarding to cardiac anatomy, 10 patients had functional single left

ventricle (FSLV, 28.5%) and 21 a functional single right ventricle (FSRV, 60%); 4 patients had undetermined single ventricle (11.5%). Total cavo-pulmonary connection (TCPC) with intracardiac lateral tunnel was performed in 7 patients (20%, $N=7$), whereas 28 patients had TCPC with external conduit (80%). Data from echocardiographic evaluation showed a medium EF established with Simpson's method of 60% in patients with FSLV; patients with a FSRV or undetermined single ventricle had a medium FAC of 41.1%, with 15.1% having a reduced FAC $<35\%$. No FSLV patients had an EF $<50\%$. When using creatinine-based formula, data about renal function in our population showed a stage 2 chronic kidney disease (eGFR: 60–89 ml/min 1.73 mq) in 11% of total population ($N=4$), that became 26% when using cystatin C-based equation ($N=9$), with one patient showing a moderate reduced loss of kidney function (eGFR: 40–59 ml/min 1.73 mq). Urinalysis showed 29% ($N=10$) of patients having microalbuminuria (microalbumin/creatinine ratio between 30 and 300 mg/g). Statistical analysis demonstrated a negative correlation between systolic function (TAPSE for FSRV) and cystatin C blood levels (Pearson's R −0.428, $P=0.053$), and between systolic function (FAC and Simpson) and microalbuminuria (Pearson's R −0.414 with $P=0.049$ and Pearson's R −0.754 with $P=0.019$, respectively). Transient elastography reported 10 patients (29.4%) with abnormal hepatic stiffness for Fontan patients. That condition appeared to be more frequent in patients with higher grade of AV valve insufficiency ($P<0.05$).

Conclusions: Our population showed an higher prevalence of FSRV Fontan patients, with an expected lower systolic function compared with FSLV. 2D evaluation of systolic function showed a linear inverse correlation with renal function, suggesting that Fontan patients need a closer renal monitoring. Hepatic stiffness, which is a warning sign of potential hepatic cirrhosis need to be monitored in all Fontan patients, especially those with a worse AV valve insufficiency.