

Fontan-associated liver and renal disease: a prospective single center study to early detect long-term complications



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BACKGROUND :

Fontan operation represents the surgical palliative option for congenital heart disease with single ventricle physiology. With the improvement of surgical techniques, we are facing a growing population with multiorgan complications: in particular, hepatic and renal dysfunction remain mostly unknown.

MATERIALS AND METHODS:

We enrolled patients that underwent Fontan palliation in our Centre between 1985 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 aimed at 1) evaluate the cardiovascular, renal and hepatic dysfunction and 2) show the relationship between renal and hepatic results and cardiovascular status and the effect of time on multiorgan involvement.

RESULTS:

Cardiological

We enrolled 45 patients, 60% male. Medium time from Fontan completion was 13 years 6 months (167 months), SD $\pm$ 6.8 years (82 months). Data from echocardiographic evaluation shows that patients with functionally single right ventricle have worse diastolic function ($p < 0.05$); AV valve insufficiency is associated with low VO2 max at cardiopulmonary testing ($p = 0.035$, figure 1).

Nephrological

W23% patients have a mildly reduced glomerular filtration rate and 3% moderately reduced. Renal evaluation shows glomerular dysfunction (associated with systolic dysfunction $p < 0.05$) and chronic tubular alterations (associated with diastolic dysfunction $p < 0.05$, figure 2). Renal dysfunction doesn't appear to be worse in patients with a past history of dialysis.

Hepatic

Hepatic tests show that GGT alterations are more frequent than AST elevation (66% vs 9%). Hepatic stiffness higher than reference value is present in 32% of the population, with a lower VO2 max at cardiopulmonary testing ($p < 0.05$).

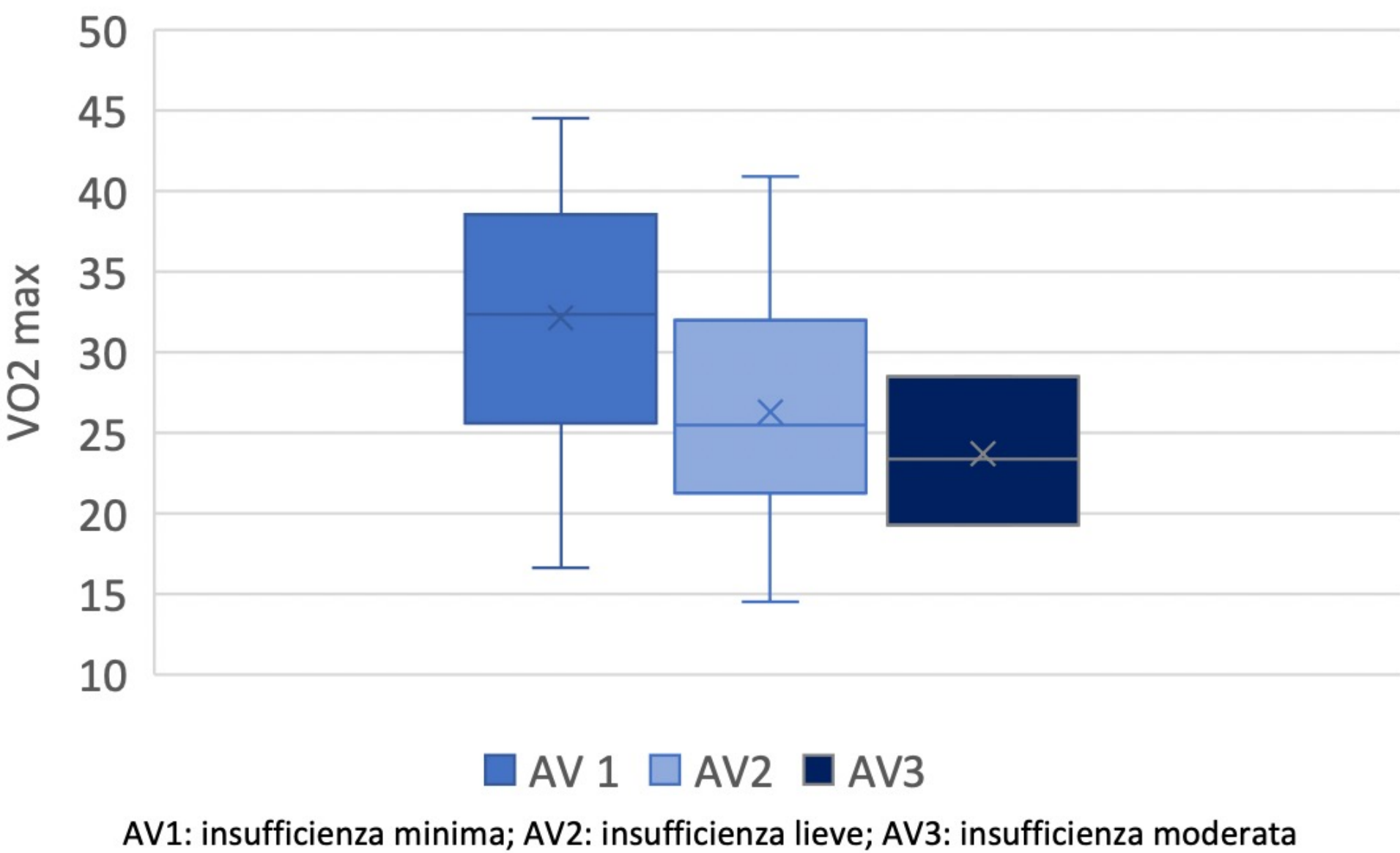


Figure 1: AV valve insufficiency is associated with low VO2 max ($p = 0.035$)

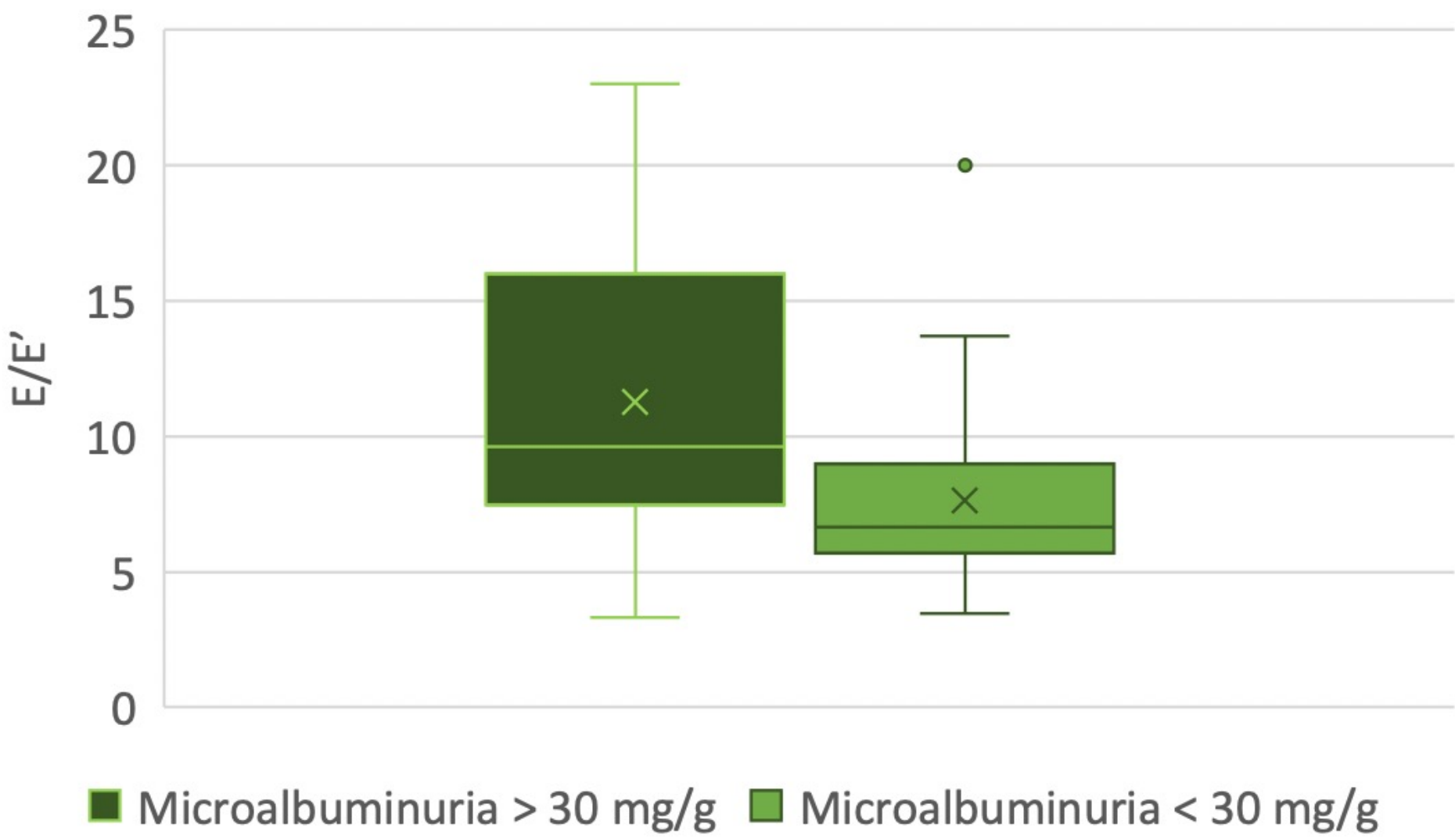


Figure 2: chronic tubular alterations is associated with diastolic dysfunction ($p = 0.02$)

	Valvola AV normale o con grado di insufficienza minima o lieve	Valvola AV con grado di insufficienza almeno moderato	Totale pazienti
Stiffness epatica > 22kPa	14 (20%)	4 (6%)	18
Stiffness epatica < 22kPa	50 (71%)	2 (3%)	52

Figure 3: AV valve insufficiency is associated with hepatic stiffness ($p = 0.015$)

CONCLUSIONS:

Fontan patients show hepatic and renal dysfunctions, that worsen with time and appear to be related with functional ad cardiological tests. This suggests that Fontan patients need a multidisciplinary follow-up.