Outcomes after Protein-Losing Enteropathy in Univentricular Hearts: a Multicenter Study.

Centre Hospitalier Régnional Universitaire de Lille, Lille, France (1); Centre Chirurgical Marie Lannelongue, Le Plessis Robinson, France (2); Centre Hospitalier Universitaire de Lyon, Lyon, France (3); Cabinet de Cardiologie Vendôme-Cardio, Lille, France (4); Hôpital Européen Georges Pompidou, Paris, France (5); Centre Hospitalier Universitaire de Bordeaux, Bordeaux, France (6); Hôpital Necker - Enfants Malades, Paris, France (7)

Background: Protein-losing enteropathy (PLE) is a rare but severe complication after Fontan surgery in patients with univentricular hearts (UVH). Outcomes are compromised since mortality is high and treatment efficiency appears limited.

Methods: This was a retrospective observational study carried out in sixteen Pediatric Cardiology Centres in France. All UVH patients diagnosed with PLE after Fontan type surgery such as atriopulmonary/atrioventricular anastomosis or intracardiac/extracardiac total cavopulmonary connection were included. The study period went from 1988 until 2014.

Results: PLE was diagnosed in thirty-five patients at a median age of 9.7 years. Median delay after Fontan type surgery was 3.6 years. At diagnosis, cardiac catheterization revealed a hemodynamic dysfunction of the Fontan circulation in 63% (n=22/35). Treatment modalities included medical treatment alone (e.g. oral/topic steroids, Calciparine®, sildenafil) in 46% (n=16/25) or combined treatment options (interventional or and surgical therapy associated to medical treatment) in the other 54% (n=19/35). Treatment was considered to be efficient in case of normalization of albumin level (> 30g/l). Medical treatment alone led to complete recovery in 13% (n=2/16), to transient improvement in 31% (n=5/16) and to no improvement in 56% of patients (n=9/16). No patient treated medically died but two (13%) were finally transplanted (one recovery, one transient improvement). Combined treatment modalities led to 21% recoveries (n=4/19) and to 37% transient improvements (n=7/19). No improvement was noted in 42% of patients (n=8/19). 21% (n=4/19) in this treatment group died.16% (n=3/19) finally underwent heart transplantation but all three subsequently died (one early death, two late deaths). Five and 10 years survival of the whole cohort were 89.7% (Cl95%:±11.3%) and 74.9% (Cl95%:±21.3%) respectively. Median follow-up was 4.5 years [0.5-21.7]. At last visit, 75% of survivors (n=21/28) were in functional NYHA class I/II. However, 75% still had patent hypoalbuminemia and 61% received specific medical therapy for PLE.

Conclusions: Despite decreased mortality, PLE remains a significant burden after Fontan surgery since complete recovery is rare and treatment modalities remain unsatisfying. Heart transplantation is an ultimate therapeutic option but carries a high risk. Further studies are needed to develop innovative treatment strategies and improve outcomes.