

### Protein losing enteropathy and cardiac disease in children and young adults

*Perouse de Monclos T., Martin-Bonnet C., Bakloul M., Ducreux C., Veyrier M., Walton C., Ninet J., Henaine R., Metton O., Mitchell J., Di Filippo S.  
Cardiovascular Hospital Louis Pradel, University Claude Bernard Lyon1, Hospices Civils de Lyon,  
Lyon France*

The objective of this study was to assess the severity and the outcome of PLE in patients with congenital heart disease (CHD).

Material and methods: Single-center retrospective chart-analysis of patients with CHD and diagnosed with PLE during follow-up. Cardiac disease past history, demographics, clinical data, therapeutic management and outcomes were assessed.

Results: 15 patients were included in the study (8 males). Underlying cardiac disease was: tricuspid atresia (4), double inlet ventricle (2), complex CHD (2), TGA with VSD and pulmonary stenosis (1), VSD and pulmonary hypertension (1), AVSD (1), pulmonary valve stenosis (1), TGA (1), total abnormal pulmonary venous return (1) and restrictive cardiomyopathy (1). Cardiac surgery was performed in 14 patients at the mean age of 5.4years (median 5y). Postoperative course was uncomplicated in 4 cases and 9 (60%) presented prolonged pleural and/or pericardial effusion after surgery, 2 of them had a phrenic paralysis. Long-term cardiac events included arrhythmias in 3 cases, edemas in 2.

PLE was diagnosed at mean age of 8.5years (mean 7.5y). Mean time from surgery to PLE onset was 3.75years (median 0.8y). PLE was mild in 6 patients (40%), moderate in 4 (27%) and severe in 5 (33%). No prognosis factors for severity of PLE was found. All patients had myocardial function within normal range. Treatment for PLE included: diuretics and albumin supplementation in all cases, steroids in 4, fenestration of Fontan in 3. Four cases (27%) had poor outcome: of them, 2 patients died at 1.4 and 2.5y after onset of PLE because of uncontrolled compromised hemodynamics, and 2 were transplanted (1 PLE resolved within 6 months after transplantation and 1 died of uncontrolled graft failure). Eleven non-transplanted survivors (73%) clinical status ranged in: NYHA I (5cases), NYHA II (4), or NYHA III (2). No PLE resolved in non-transplanted survivors.

Conclusion: PLE is a severe complication of CHD with high rates of mortality or transplantation. No significant predictive prognosis factor was found. No case resolved over time but disease was controlled in 73% of the cases.