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Protein Losing Enteropathy after Fontan surgery - the Swedish experience

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Background
Protein-Losing Enteropathy (PLE) after operation with Total Cavo Pulmonary Connection (TCPC) has a prevalence between 5-15%. 5-year survival rate varies between 50-88%. PLE is characterized by enteric loss of proteins due to poorly understood mechanisms and treatment is difficult. This study presents prevalence, treatment and outcome of PLE in the Swedish cohort of TCPC patients.

Materials and methods
All patients with univentricular heart born in Sweden after Jan 1st, 1993 and operated with TCPC before 2017 (n=482) were reviewed and patients with PLE diagnosis before the age of 18 were included.

Results
Twenty-three patients with PLE were identified, corresponding to a prevalence of 4.8% with a median follow-up of 4.3 years (range 11 months – 15.2 years). In all cases PLE was suspected clinically and confirmed with elevated F-α1-antitrypsin. The most common morphological diagnoses were Hypoplastic Left Heart Syndrome (8/23) and tricuspid atresia (3/23). Eight patients had various forms of complex malformations. All patients had been operated in the neonatal period. The most common procedures were the Norwood procedure (9/23), pulmonary banding (6/23) and systemic-to-pulmonary shunts (6/23). Bidirectional Glenn, in some patients combined with other procedures was performed at median 6 months of age (range 3-19). The median age at TCPC was 2.7 years (range 1.2 – 6 years). Four patients had a lateral tunnel and 19 an extracardiac. Five patients had additional surgery concomitant with TCPC. The median interval between TCPC and PLE diagnosis was 11 months (range 3 months – 13.5 years). Pharmacological treatment was used in 19 pts and consisted of combinations of pulmonary vasodilators, heparin and budesonide. Eleven pts underwent 12 catheter interventions. Six patients underwent heart transplant. Nineteen patients are alive, 10 with continued pharmacological treatment, five in partial or complete remission and four after a heart transplant. Four pts have died corresponding to a mortality of 17%.

Conclusion
In this national study survival after the diagnosis of PLE was better compared to other studies. No single specific treatment was identified as successful, but transplantation was fairly common (26%). The study also indicates that remission of PLE occurs.