

## MP3-16

### Treatment strategies for protein-losing enteropathy in Fontan patients

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**Objectives:** Protein-losing enteropathy (PLE) is a rare but severe complication occurring after Fontan procedure. Reduced cardiac output, chronic venous congestion and intestinal inflammation are believed to play a role in PLE development. However, given the multifactorial and largely unknown pathogenesis, no single proposed treatment strategy has proven universally successful. We thus evaluated the use of several surgical, interventional and medical therapeutic strategies and outcome in our PLE patients.

#### Methods:

In a retrospective review of the entire cohort of 351 Fontan patients treated in our institution (n=272 originally operated in our institution) we identified 24 patients (6.9%) diagnosed with PLE. Diagnosis was established when clinical criteria as diarrhoea and/or recurring edema, pleural effusions or ascites and abnormal laboratory values suggesting intestinal protein loss as low serum albumin and protein and elevated faecal alpha-1-antitrypsin were present. Data from clinical history, treatment and outcome was extracted and analysed.

#### Results:

Freedom from PLE was 89.9% and 87.9% at 10 and 20 years. PLE developed at a median of 3.0 [0.1-16.5] years after Fontan. Haemodynamic issues as Fontan pathway obstructions, aortopulmonary collaterals, dysrhythmias or phrenic palsy were identified and addressed in 22 patients (91.7%). Treatment with one or more medication was initiated in 14 patients (58.3%) and included Sildenafil in 12 (50%), Bosentan in 4 (16.7%) and Budesonid in 8 patients (33.3%). Surgical and interventional procedures alone yielded stable clinical remission in 4 patients (16.7%). Additional medication achieved stable remission in another 6 patients (25.0%) and relapsing remission in 4 (16.7%). PLE-associated mortality is substantial, 10 patients (41.7%) died during follow-up. In Fontan patients with PLE, freedom from death or transplantation was significantly decreased compared to those without PLE (33.0% vs. 87.1% at 20 years after Fontan,  $p<0001$ ).

#### Conclusions:

Given the small numbers of Fontan patients developing PLE, studies investigating therapeutic strategies are difficult to perform. Treatment has to adjust to the individual patient's findings and may thus include elimination of Fontan pathway obstructions, optimizing ventilatory conditions, reducing pulmonary vascular resistance and anti-inflammatory therapies. Nevertheless, despite individually focussed therapy, reoccurrence is frequent and mortality in persistent PLE remains substantial. Cardiac transplantation should be considered early.