Effectiveness of Ambrisentan Therapy in Patients of Protein-Losing Enteropathy after Fontan Operation.

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Aim: The purpose of our study is to investigate the efficacy of ambrisentan in patients with steroid-dependent PLE after Fontan procedure who has been treated by steroids, immuno-suppressant, and pulmonary vasodilators (PDE5-inhibitors, bosentan, and beraprost).

Method: The study subjects were 4 patients with PLE after Fontan (4 Males and age ranged 4-19y, the interval of PLE from Fontan 1.3-8 months). All were steroid-dependent with immuno-suppressant and pulmonary vasodilators(bosentan, and sildenafil) and needed repetitive infusion of albumin and γ-globulin.

Treatment protocol: ambrisentan was given at the dose from 0.09 to 0.19mg/kg per day, replacing bosentan without changing the dose of the rest medications. Then the efficacy of ambrisentan was assessed (1) by the supplement dosage of albumin and γ-globulin and (2) by the dosage of steroids.

Results: No severe adverse effect but transient puffy face (2pts) was found. Ambrisentan could reduce the daily dose of steroid from 15mg (median) to 9mg, and could prolong the supplement cycles of albumin/γ-globulin from 2days to 25days as a median for 3.3 months, comparing to bosentan. The serum albumin also increased from 3.7 to 4.3g/dl.

Conclusion: Ambrisentan is a potent therapeutic option for steroid-dependent PLE after Fontan procedure.