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May infections advantage development of protein losing enteropathy (PLE) after Fontan procedure?

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Objectives: One of the serious complications in patients after Fontan surgery could be the protein losing enteropathy (PLE). PLE may be a rare complication but it is often lethal or at least decreases the quality of life immensely. The aim of this study was to analyse the correlation between pulmonal and gastrointestinal infections and development of PLE after a Fontan procedure.

Methods: This retrospective study reviewed the data of all patients undergoing a Fontan completion on our hospital from 2003-2015. PLE was diagnosed if clinical symptoms such as diarrhea, pleural effusions, ascites, peripheral edema appeared, if the total protein level in Serum was below 6g/dl, serum albumin was below 3.5g/dl and Alpha-1-Antitrypsin level in stool was above 60mg/dl or if a doctor treating the patient documented the diagnosis. Infections were assumed if documented, CRP, IL-6 represented markers for inflammation.

Results: There were 201 children undergoing a Fontan completion at LMU matching the criteria between 2003 and 2015. There were ten patients presenting with PLE after the operation. The average time between Fontan procedure and onset of PLE was 2.26 years (0.07- 4.8 years). An infection associated with the onset was traced in 6 of the ten cases. In one patient an AV - valve insufficiency and a limited ventricular function was documented additionally. In the other four cases dysfunction of the Fontan circulation was noted. Laboratory findings at admission due to PLE were: total protein level 4,26g/dl, Serum-Albumin 2.44g/dl. Alpha-1-Antitrypsin(A1AT) level in stool in n=4 was 425mg/dl. AAT levels in stool before manifestation of PLE were pathological in two cases (n=3). CRP was increased in three patients. IL-6 was increased in two cases.

One patient died of PLE and plastic bronchitis. There were also 2 patients who showed transient elevated A1AT in stool without ever developing any symptoms of PLE.

Conclusion: For better outcome patients who are showing symptoms of infections could be tested for A1AT in stool for an early detection of PLE

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