LETTER

Protein-losing enteropathy after Fontan operation: enteric capsule findings and management with atrial pacing

Dear Editor,

Protein-losing enteropathy (PLE) is associated with multiple conditions, including cardiovascular abnormalities with elevated central venous pressure such as Fontan operation. Apart from PLE, another complication in these patients is sinus node dysfunction, which may limit cardiac output, contributing to PLE.

A girl with a complex congenital heart disease who underwent a Fontan operation, presented at the age of 11 years with peripheral edema without signs of malnutrition. Blood tests revealed hypoalbuminemia (serum albumin 2.5 g/dl), lymphopenia and hypogammaglobulinemia (IgG 190, IgA: 53.4, and IgM: 23.4 mg/dl). Renal and liver function were normal, but a_1-antitrypsin excretion in stool was indicative for PLE (448 mg/dl, normal values <54 mg/dl). Video capsule endoscopy (VCE) was performed to investigate further the cause of PLE which revealed white villi and extensive mucosal edema in the duodenum and jejunum. A high protein (3-4 gr/kg) and low-fat diet (20-22 % of the daily energy intake) supplemented with medium-chain triglyceride (12-15 %) was initiated. The patient was also taking essential fatty acids and lipid soluble vitamins. Transient clinical improvement occurred after the initiation of the diet with recession of peripheral edema, but her albumin levels continued to be low (2.9 g/dl) for the next two years. Because of sinus node dysfunction at the age of 13, a permanent epicardial pacemaker was implanted. After this, there was a significant clinical and laboratory improvement with increased serum albumin levels (3.9 g/dl) and decreased a_1-antitrypsin excretion in stool.

The presented case is rare and there is limited published data regarding the use of VCE for diagnosis PLE (additionally to a_1-antitrypsin) as well as the clinical improvement after implantation of a pacemaker[1,2].

There are two theories about the pathophysiologic mechanism of PLE after Fontan procedure: one theory suggests that elevation in the systemic venous pressure leads to dilation of the lymphatics within the gastrointestinal tract, while another one suggests that the low cardiac output leads to decreased intestinal perfusion and increased epithelial permeability, leading to protein leakage into the gut lumen. There is also an element of chronic inflammation in PLE secondary to low cardiac output[3].

In our patient, extensive edema revealed by VCE could be indicative of chronic inflammation secondary to low cardiac output. The restriction of long-chain triglycerides in the diet (which decreases the lymphatic circulation) had only partial and transient clinical improvement. Hence the effective treatment was the implantation of the pacemaker, which increased the cardiac output and improved the epithelial permeability of the intestine, leading to an increase of serum albumin levels. We did not consider it necessary to undergo a new VCE after applying the pacemaker, as the symptoms had subsided completely and the cost of the examination is substantial.

In conclusion, PLE should be suspected in any patient undergoing Fontan operation. VCE might be useful in the differential diagnosis of the underlying mechanism of PLE and the follow-up evaluation of the treatment effectiveness. Atrial pacing should be considered as a possible intervention to treat PLE especially for patients with signs and symptoms of sinus node dysfunction.

Keywords: Protein losing enteropathy, Fontan operation, pacemaker, video capsule endoscopy

References

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Conflict of interest

Authors declare no conflict of interest.

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