Gastrointestinal bleeding as a symptom of failing Fontan circulation

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Fontan procedure is a standard surgical palliation for single ventricle congenital heart defects. As a result of the procedure, systemic venous return is directly connected to pulmonary circulation without the interposition of the ventricle which leads to chronic venous hypertension. There are many widely known late consequences of Fontan physiology [1] such as the development of plastic bronchitis, protein-losing enteropathy, Fontan-associated liver disease, portal hypertension with esophageal varices, splenomegaly, and hypersplenism. We present a case that demonstrates a different manifestation of failing Fontan circulation.

A 10-year-old girl with a congenital heart disease including a double-outlet right ventricle, hypoplastic aortic arch, subvalvular aortic stenosis, and large ventricular septal defect was admitted to the hospital with symptoms of heart failure and acute lower gastrointestinal bleeding. She had undergone surgical palliation at 6 years of age: nonfenestrated extracardiac Fontan operation (with an additional source of pulmonary blood flow from the ventricle).

Laboratory investigations showed hypoalbuminemia, hypoproteinemia, and elevated stool alpha-1-antitrypsin level. Technetium labeled albumin scintigraphy was performed and confirmed protein-losing enteropathy.

Cardiac imaging was planned to assess the anatomy of Fontan circulation. Computed tomography angiography showed endothelial hyperplasia causing left pulmonary artery in-stent restenosis. The patient underwent successful percutaneous stent redilatation.

Although the signs of active gastrointestinal bleeding diminished, the patient suffered from severe recurrent anemia that required repetitive blood transfusions and during the hospital stay, melena was present. Abdominal ultrasound showed portal hypertension. There were no endoscopic signs of active or recent bleeding, but esophageal varices were found in gastroscopy. Bone marrow biopsy analysis showed normal hematopoietic function. Due to persistent positive fecal occult blood test (FOBT) — technetium labeled erythrocyte scintigraphy was scheduled and localized the source of bleeding.

Both scintigraphy examinations (Figure 1A — with albumin and 1B — with erythrocytes) show leakage in a similar region of the small intestine. After excluding other causes of intestinal bleeding, regarding the same localization of protein and erythrocyte leakage, failing Fontan circulation was identified as the most probable underlying cause of enteric erythrocyte loss [2, 3].

Treatment in our patient involved optimization of Fontan hemodynamics and pharmacotherapy: propranolol, lisinopril, spironolactone, low-fat high-protein diet, and enoxaparin (as data are showing good effects on intestinal epithelial cells of heparin therapy in protein-losing enteropathy [4]). Surgical segmental resection of the abnormal region of the small bowel was considered. One month later, as soon as laboratory assessment showed no anemia with negative FOBT, serum albumin, and protein levels within a normal range; any surgical intervention was postponed. The patient remained stable in a one-year follow-up.

Chronically elevated central venous pressure in Fontan physiology has a significant impact on subdiaphragmatic hemodynamics. Potentially, the same mechanism may play a role in erythrocyte enteric loss causing in-

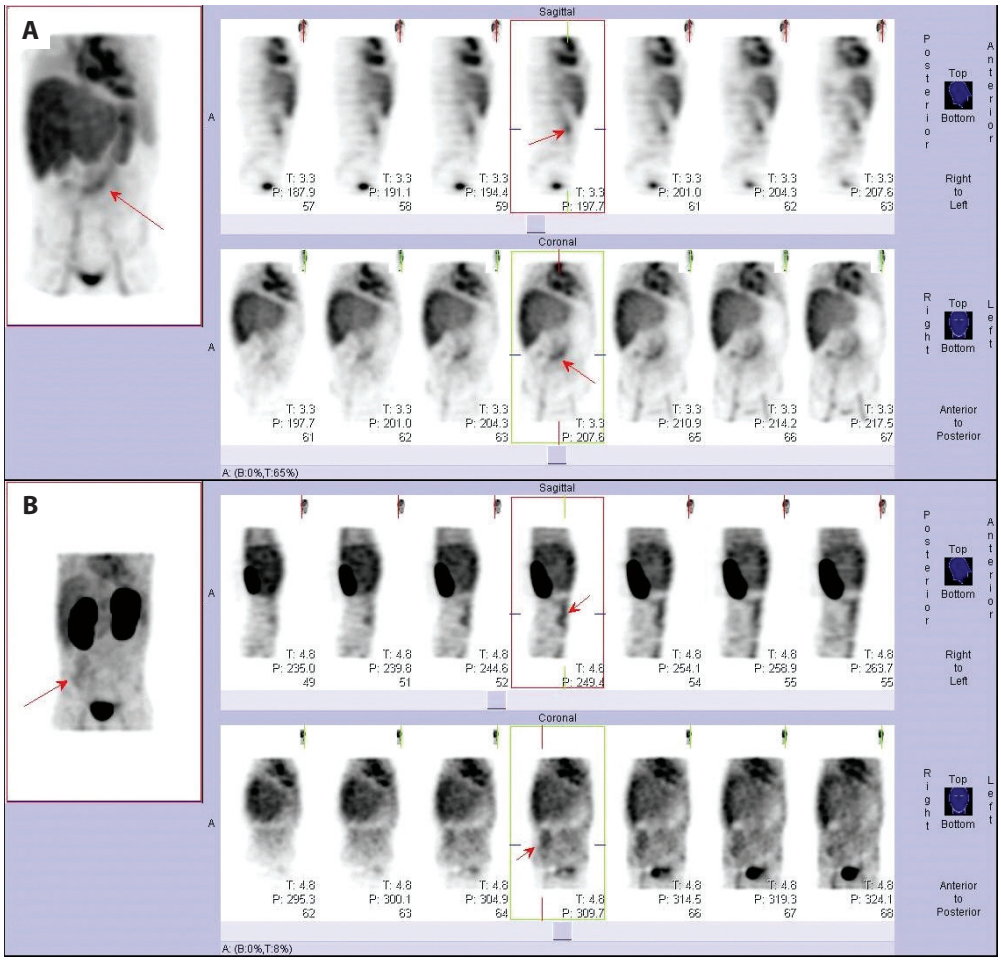

Figure 1. A. Technetium 99m-labeled human serum albumin scintigraphy (the arrows show leakage of albumin in the region of the small intestine). **B.** Technetium 99m-labeled erythrocytes scintigraphy, the arrows show leakage of erythrocyte in the region of the small intestine

creased vascular permeability for larger cells or molecules: lymphocytes, erythrocytes, serum albumin, and protein. Recurrent anemia resulting from enteric erythrocyte loss may be another significant complication related to Fontan palliation. Future studies are needed to confirm that.

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