# Successful Resection of Localized Intestinal Lymphangiectasia Post-Fontan: Role of $^{99m}$Technetium-Dextran Scintigraphy

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**ABSTRACT. Intestinal lymphangiectasia is a well-recognized complication of the Fontan procedure, occurring in up to 24% of patients. Because of the loss of chylous fluid into the gut lumen, protein-losing enteropathy results as well as lymphopenia and hypogammaglobulinaemia. In some cases, dilated lymphatics in the intestinal serosa or mesentery also rupture, causing chylous ascites. Standard medical and cardiac surgical interventions are generally ineffective and the condition is frequently lethal. We report a case of intractable and life-threatening chylous ascites and chylothorax in a 14-year-old girl, associated with intestinal lymphangiectasia and protein-losing enteropathy after a Fontan procedure for tricuspid atresia. The condition was refractory to all standard medical therapies, including dietary modifications, diuretics, corticosteroid therapy, albumin infusions, octreotide, heparin, bowel rest, and parenteral nutrition. Cardiac surgery to optimize her hemodynamic status was also ineffective and large volume pleural and ascitic fluid losses continued. Having exhausted all other therapeutic modalities, $^{99m}$technetium-dextran scintigraphy was performed to assess the extent of intestinal protein loss and the potential for surgical intervention. Scintigraphy suggested localized protein loss from the proximal jejunum and subsequent segmental resection was effective. Postoperatively, ascites and pleural effusions resolved, and there was no evidence of short bowel syndrome. Growth has accelerated and the patient has entered puberty. There is mild persistent intestinal protein loss requiring diuretic therapy. Ascites or pleural effusions are absent, and the patient remains well >2 years after surgery. Intestinal lymphangiectasia post-Fontan procedures has traditionally been ascribed to hemodynamic factors such as raised systemic venous pressure, which would predispose to a generalized intestinal lesion. However, in this case, scintigraphy demonstrated a localized, surgically correctible lesion. To our knowledge, this is the first reported case of the use of $^{99m}$technetium-dextran scintigraphy for this indication and of successful partial small bowel resection in such a case.** *Pediatrics* **2003;112:e242–e247. URL: http://www.pediatrics.org/cgi/content/full/112/3/e242;** ***intestinal lymphangiectasia, resection, Fontan procedure, $^{99m}$technetium-dextran, scintigraphy.***

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ABBREVIATIONS. PLE, protein-losing enteropathy; $^{51}$Cr, chromium-51; $^{125}$I, iodine-125; $^{111}$In, indium-111; $^{99m}$Tc, technetium-99m; $^{131}$I, iodine-131; $^{67}$Cu, copper-67.

Intestinal lymphangiectasia and protein-losing enteropathy (PLE) are recognized complications of the Fontan procedure, occurring in 4% to 24% of cases.[1–5] Results of medical and cardiac surgical interventions are generally disappointing. Mortality is ~50% over 5 years[4,5] and continuing morbidity is significant in survivors. Standard endoscopic small bowel biopsy, although confirming the diagnosis of lymphangiectasia, does not provide information on extent and sites of actual protein loss. Even enteroscopy with serial small bowel biopsies may be inaccurate, as the disease may be localized or nonuniform in its distribution.[5,6] Hypoproteinemia, hypogammaglobulinemia, hypoalbuminemia, elevated $\alpha$-1-antitrypsin clearance, hypocalcemia, and lymphocytopenia are useful markers of the severity of the disease process, but reveal little of the location or extent of bowel involvement.[4] Similarly, techniques involving the use of radiolabelled macromolecules such as chromium-51 ($^{51}$Cr) and iodine-125 ($^{125}$I)-conjugated to serum proteins such as albumin, globulin, and transferrin as well as being cumbersome and not readily available, require fecal handling over several days, and are not suitable for imaging purposes to determine the site of leakage.[7]

Although characteristic radiologic features of this condition have been described,[8–10] in practice these features are usually absent.[11] Nuclear scintigraphy is the most sensitive and reliable noninvasive imaging modality for the detection of PLE. Several radiopharmaceuticals have been used, including indium-111 ($^{111}$In) -transferrin[12] and technetium-99m ($^{99m}$Tc)-human serum albumin.[10,13–15] However $^{99m}$Tc-dextran appears to be superior as it is a more chemically stable compound, provides a higher target to-background activity ratio, and false-positive studies are less likely.[16]

It has been reported that intestinal lymphangiectasia and PLE may respond to small bowel resection where the disease process is confined to a relatively small segment of the bowel.[11,17,18] The use of nuclear

scintigraphy to assess the feasibility of partial small bowel resection in such a clinical setting has not been previously described. A case report of a 14-year old female who underwent $^{99m}$Tc-dextran scintigraphy to assess extent of bowel involvement, and who experienced marked and sustained improvement following small bowel resection is presented.

## CASE REPORT

The patient was born with tricuspid atresia, right ventricular hypoplasia, atrial septal defect and ventricular septal defect. Fontan procedure (right atrium to pulmonary artery anastomosis) and atrial septal defect closure were performed at 3 years of age. The duration of cardiopulmonary bypass was 105 minutes. Postoperatively her recovery was uneventful, apart from renal impairment requiring peritoneal dialysis for 36 hours. The patient was discharged from the hospital after 11 days. From 12 months postsurgery, ascites and peripheral edema required diuretic therapy, but ascites gradually increased. Despite this, there was no significant limitation of exercise tolerance.

Investigations included ultrasound examination of liver and portal vessels, which were normal. Echocardiogram showed favorable hemodynamics, with no evidence of right heart failure. Left atrial pressure was 7 mm Hg. Hypoalbuminemia (serum albumin: 21–30 g/L; normal range: 35–50 g/L), diarrhea and elevated stool $\alpha$-1-antitrypsin losses (up to 51.9 g/kg; NR: 0–1.5) suggested PLE. Transferrin isoform electrophoresis was normal, excluding carbohydrate-deficient glycoprotein syndrome. Lymphopenia ($0.38–0.9 \times 10^9$/L; NR: 1–5) and hypogammaglobulinemia (immunoglobulin G: 2.79–3.08 g/L; NR: 6.12–12.93) suggested intestinal lymphangiectasia as the likely cause. Small intestinal lymphangiectasia was confirmed on small bowel biopsy (Fig 1A). Diuretic therapy was combined with a high-protein, low long-chain triglyceride fat diet with medium-chain triglyceride fat supplementation, but without success.

Antigliadin antibodies (AGA) were subsequently found to be elevated, with AGA immunoglobulin A at 141 U/L (NR <25), AGA immunoglobulin G at 139 U/L (<46) and positive antiendomysial antibodies. Review of the biopsy confirmed partial villous atrophy with mixed inflammatory cell infiltrate consistent with celiac disease (Fig 1B). Gluten was now excluded from the diet in addition to previous modifications, but this did not result in any clinical improvement despite normalization of celiac serology over a 6-month period. Prednisone (1 mg/kg/d) led to partial remission of fluid retention, but she remained steroid-dependent and steroid reduction was not tolerated. Trials of subcutaneous octreotide (30 $\mu$g bid)[19–21] and subcutaneous heparin (3000 U bid)[22–25] were similarly unsuccessful. Azathioprine 1.5 mg/kg/d led to initial steroid-sparing; however, symptoms gradually recurred.

At 13 years of age she presented with severe chylous ascites, bilateral chylous pleural effusions, and respiratory compromise. After initial drainage of 9 L of sterile, turbid fluid by bilateral intercostal catheters, which clinically included both pleural and ascitic fluid, losses continued at approximately 2 L per day. Bowel rest, parenteral nutrition, and increased steroid dose were all unhelpful in slowing the losses. Therefore, attempts were made to optimize her cardiovascular hemodynamics by surgical revision to

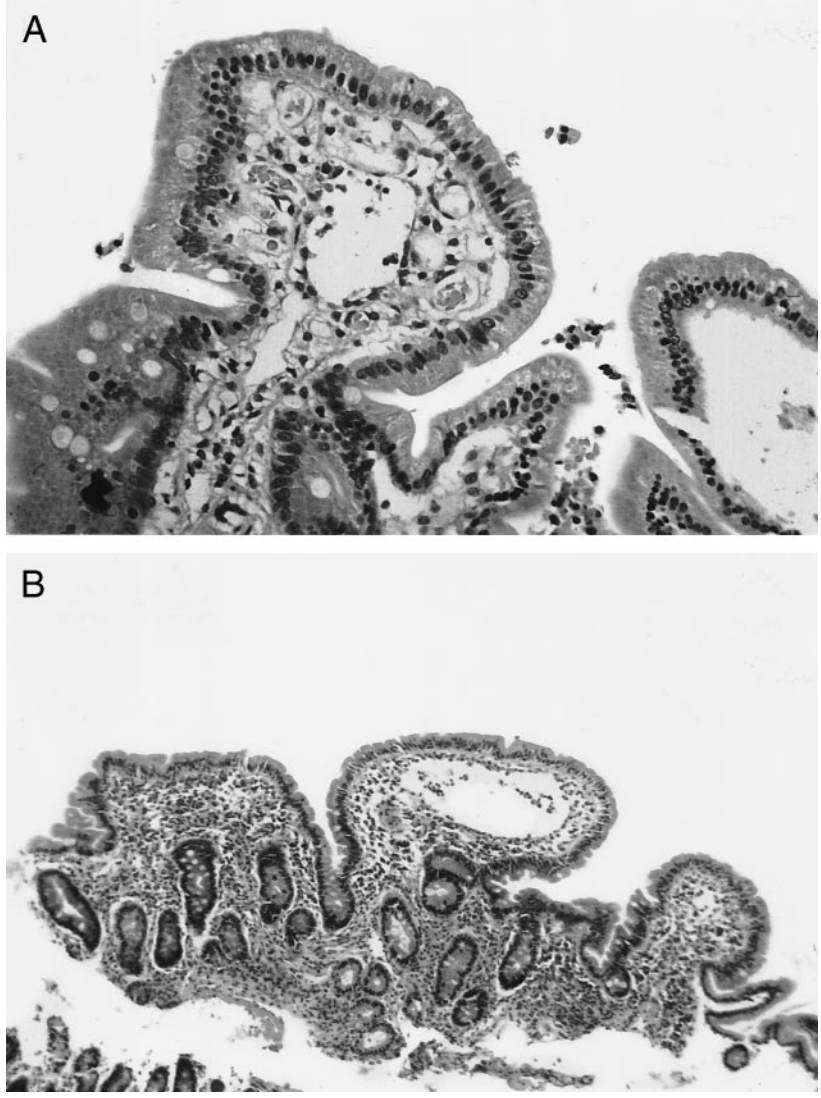


**Fig 1.** Duodenal biopsy from patient with PLE post-Fontan procedure. A, Dilated lymphatics in the lamina propria consistent with intestinal lymphangiectasia. (H&E, original magnification ×66). B, Partial villous atrophy and mild crypt hyperplasia are evident. Increased numbers of plasma cells, eosinophils, and lymphocytes are present in the lamina propria and there is a moderate increase in intraepithelial lymphocytes. The features are consistent with a diagnosis of celiac disease. (H&E, original magnification ×26).

total cavopulmonary connection. However, this also did not lead to improvement in ascites or pleural losses. In the immediate postoperative period she developed a junctional cardiac rhythm for which a permanent pacemaker was inserted. Eight weeks later she developed left hemiplegia and acute dystonia. Computerized tomography of the brain revealed an infarct involving the right parietotemporal lobe, consistent with an embolic etiology. Recovery was prompt with minimal residual left-sided weakness apparent 2 weeks later.

As all other therapeutic options had been exhausted, a $^{99m}$Tc-dextran study was performed to assess the extent of the PLE and the possibility of resecting affected bowel to alleviate symptoms. Dextran 77 kDa (Sigma, St Louis, MO) was labeled with Tc-99m as previously described.[26] Radiopurity was determined at >95%. Commencing immediately after the intravenous administration of the radiotracer (260 MBq), planar scans (anterior and posterior) of the lower thorax, abdomen and pelvis were obtained continuously every 60 seconds for 2 hours with the patient supine. High-resolution collimators and a dual-head gamma camera (AXIS; Philips Medical Systems, Best, the Netherlands) were used.

The scans demonstrated focal abnormal accumulation of radiotracer in the left flank below the spleen from the 10th minute of the study. Thereafter there was passage of tracer down the abdomen and across to the region of the right iliac fossa in a pattern suggestive of transit through the small bowel (Fig 2A-C). No other abnormal uptake of tracer was identified in the lower chest, abdomen, or pelvis. Normal blood pooling of tracer and biodistribution to the liver, kidneys, and bladder were demonstrated. No significant gastric uptake of tracer was evident to suggest the presence of free pertechnetate. Thus, scintigraphy findings were interpreted as consistent with localized PLE.

At subsequent laparotomy, 135 cm of macroscopically affected jejunum, with serosal and mucosal lymphangiectasia and vascular congestion, was excised. Ascites and pleural losses ceased over the following week. After surgery she has remained well, with normal bowel habit and no evidence of short bowel syndrome or of reaccumulation of ascites and pleural fluid. There was mild persistent PLE (stool $\alpha$-1-antitrypsin: 2.7 g/kg; NR: 0–1.5), controlled with diuretic therapy. At 5 months postoperatively, she was readmitted with a pleural effusion after noncompliance with diuretics. This responded to reinstitution of diuretic therapy and albumin infusion. There was no evidence of recurrent ascites. A repeat $^{99m}$Tc-dextran scintiscan at that time documented some ongoing intestinal protein loss (Fig 3); however, since then she has remained well, with no further requirement for supplemental albumin infusion. Now, >2 years postoperatively, her serum albumin and immunoglobulin remain normal, she is showing significant catch-up growth, she has entered puberty, and appears a healthy, happy teenager, which is a far cry form the stunted, bloated, chronically unwell child she had been for many years previously.

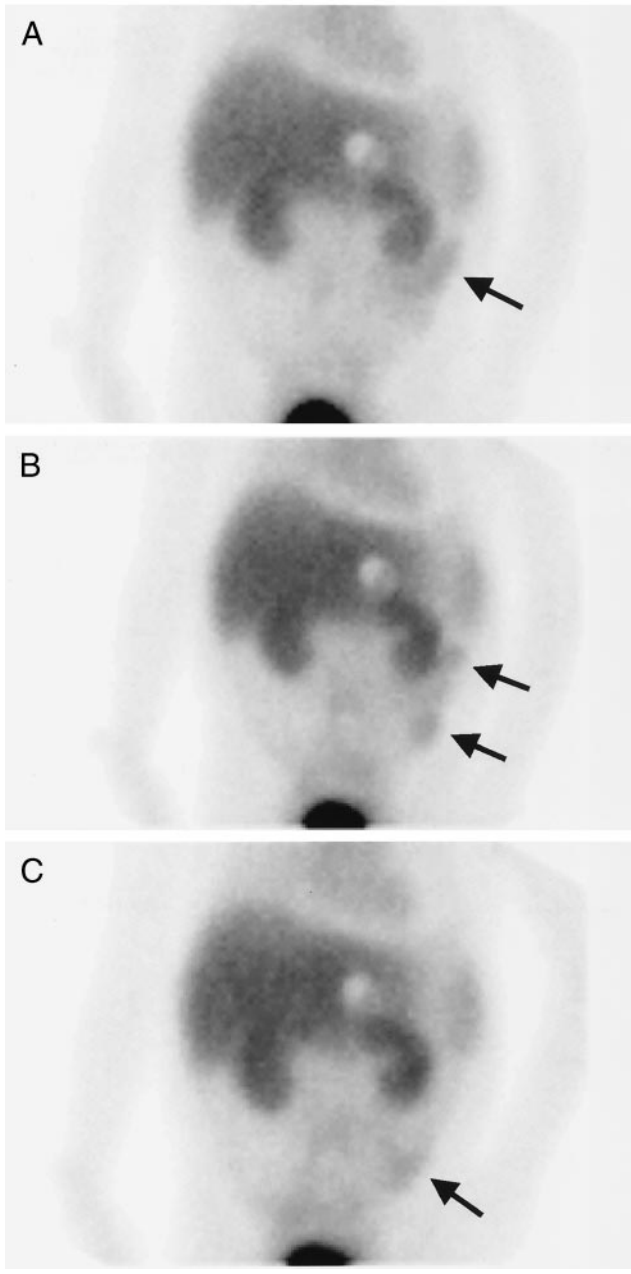


**Fig 2.** $^{99m}$Technetium-labeled dextran scintiscan of the abdomen in patient with intestinal lymphangiectasia post-Fontan procedure demonstrating localized enteric protein loss. Images A-C taken 66, 72, and 78 minutes after injection show abnormal focal accumulation of radiotracer in left upper quadrant below the spleen and left kidney, with subsequent passage of tracer down the abdomen consistent with transit through the small bowel.

## DISCUSSION

PLE associated with the Fontan procedure was initially reported during the early 1980s,[1,2] and occurs in up to 24% of patients undergoing this operation.[1–5] The median time interval between surgery and diagnosis is 2.7 years (range: 0.1–16.4 years), but up to one third of cases develop at least 5 years after surgery.[4]

Although PLE post-Fontan operation is generally thought to result from raised systemic venous pressure leading to increased lymph production and decreased lymph drainage resulting in lymphangiectasia, 23% of cases of PLE have favorable hemodynamics,[4] as in the current case, and cardiac surgery is beneficial in only 50% of survivors.[4] Poorly understood inflammatory and immunologic factors may also contribute to the pathologic process.[4,27]

Patient risk factors for the development of PLE post-Fontan include heterotaxia, polysplenia, anomalies of systemic venous drainage, ventricular anatomic variants (other than dominant left ventricle), increased pulmonary arteriolar resistance, and increased left ventricular end-diastolic pressure.[5,28] Interestingly, patients with tricuspid atresia were less likely to develop PLE than other groups.[5] Perioperative risk factors included longer cardiopulmonary bypass time, increased left atrial pressure after the operation, longer length of hospital stay, and presence of postoperative renal failure.[2,5] The current case had none of these risk factors, apart from transient renal impairment in the postoperative period, requiring dialysis for 36 hours.

Complications of post-Fontan PLE are secondary

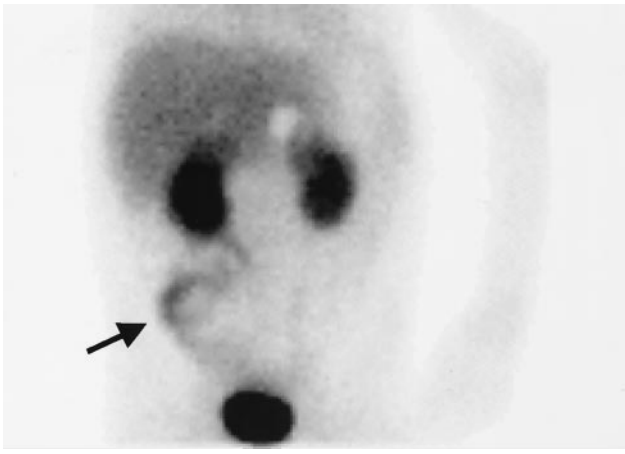

**Fig 3.** Repeat $^{99m}$technetium-labeled dextran scintiscan 6 months postresection. Image taken 140 minutes after injection demonstrating minimal persistent enteric protein loss, with abnormal accumulation of tracer in the right iliac fossa.

to enteric loss of serum proteins (including immunoglobulins), fat, and lymphocytes. These losses lead to peripheral edema (in 79%), ascites (in 53%), pleural effusions (in 22%), respiratory compromise, weight loss, chronic diarrhea (in 11%), hypocalcemia (in 33%), infections (in 16%) and thromboembolism (in 18%).[4] Chylous ascites and pleural effusions may supervene if there is rupture of dilated lymphatics in the intestinal serosa or mesentery. There is often associated growth and pubertal delay.[7,11,29]

Traditionally, treatment for PLE post-Fontan involves both medical and cardiac surgical options. A noninvasive approach consisting of various combinations of drug treatments (digitalis, diuretics, inotropes and afterload reduction agents), albumin infusions, dietary manipulation (salt restriction, high protein/low fat), and possibly steroids is often used initially. The mechanism of action of steroids in this condition is unknown, but may relate to favorable effects on intestinal epithelial permeability and/or local inflammation.[30–32] Unfortunately, many patients are unresponsive to steroid treatment, and responders, including our patient, tend to remain steroid-dependent.[4,32] Other treatments reported to lead to improvement include high molecular weight heparin[22–25] and moving from a high altitude to sea level.[5] Octreotide has been shown to be efficacious in cases of intestinal lymphangiectasia in other clinical contexts.[19–21] In cases of PLE post-Fontan procedure resistant to medical therapy, cardiac surgery (open or transvascular) or pacing may be used.[4] Even cardiac transplantation has been advocated[4,5]; however, response to current treatments is generally disappointing.[4,5] In a recent retrospective review,[4] noninvasive medical treatment was associated with complete resolution of symptoms in 25%, while cardiac surgery, including some cases of transplantation, was associated with relief of symptoms in 19% but with a 60% mortality. Our patient received all of the available medical therapies, but her condition was medically refractory. Although she had initially responded to steroids, she became steroid-dependent and later, steroid-resistant. Steroid toxicity was significant, with restriction of linear growth, osteoporosis, and spinal compression fractures.

Lymphangiectasia observed post-Fontan procedure leads to ongoing gastrointestinal protein loss, which may be detected and quantitated using several techniques. A nonisotopic method involves measuring levels of $\alpha$-1-antitrypsin in stool samples. This serum enzyme has a similar molecular weight to albumin (50 kDa), is resistant to proteolysis, and is not degraded after leaking into the intestinal lumen.[33,34] Measurement of $\alpha$-1-antitrypsin loss has been demonstrated to be reliable as a screening test for the presence of enteric protein loss, and correlates well with disease activity and response to therapy.[7]

The use of radioisotopes is an alternative method for determining gastrointestinal protein loss. After the intravenous administration of radioisotopes, the rate of decline in radioactivity in serum and/or the level of radioactivity in stool samples is determined over several days. Radiolabelled macromolecules such as $^{125}$I-labeled serum albumin, $^{131}$I-labeled serum albumin, $^{51}$CrCl3, $^{51}$Cr-labeled albumin, copper-67 ($^{67}$Cu)-ceruloplasmin[7] and $^{111}$In-labeled transferrin[12] have been used. However, the limited availability of such tracers, difficulties with chemical stability, the need for sequential measurements, and the prolonged collection of stool samples have restricted their clinical use. Furthermore, they provide no information regarding the site of protein loss, nor the extent of bowel involvement. In addition, these radioisotopic methods are no more specific, nor sensitive as to enteric protein loss than $\alpha$-1-antitrypsin measurement.[7]

Scintigraphic assessment of PLE overcomes the limitations of other radioisotopic studies as scintigraphy does not require any prior patient preparation or stool collection. $^{99m}$Tc human serum albumin has been used successfully to image PLE.[10,13–15] However, it is not an ideal radiotracer. Problems have been reported with in vivo chemical stability, with marked dissociation documented over 24 hours,[35] with variable radiolabelling efficiency even within the one batch, with reduced specificity as up to 30% of the activity is in the free or reduced form, and with significant radiation burden on the liver.[35] Furthermore, questions have arisen regarding its sensitivity for detecting PLE.[15] Case reports of other agents used to evaluate PLE include $^{99m}$Tc human immunoglobulin G[36], $^{111}$In- and $^{99m}$Tc-transferrin.[12]

More recently $^{99m}$Tc-dextran has been used as an agent in imaging PLE.[16,37] Dextran is a polysaccharide, commercially available in a large range of molecular weights, and is used clinically as a plasma expander. Higher molecular weight forms are used for imaging purposes as these have long intravascular half-lives and are degraded by the liver slowly over weeks, with smaller end-product molecules excreted renally.[16] Dextran does not leak through normal intestinal mucosa,[16,26,38] and radiolabeling efficiency and in vivo stability are high.[16] Over the initial 2 hours after intravenous administration there is little hepatic or splenic uptake evident. The high sensitivity of $^{99m}$Tc-dextran in imaging PLE is attrib-

uted to a high target to background ratio, predominantly resulting from the rapid clearance of this compound during the initial hours after intravenous administration. Adverse effects on coagulation, renal function, and the cardiovascular system seem negligible because of the minimal amount used.[16] $^{99m}$Tc-dextran scintigraphy was chosen in the current case as the optimal agent for identification of localized enteric protein loss.

Previous barium studies had shown no evidence of intestinal malrotation; therefore, the appearance of the scintigraphic tracer in the left upper quadrant and its progression across the abdomen to the right iliac fossa was consistent with leakage of chylous fluid into the proximal small intestine.

Various techniques have been used to visualize dilated lymphatics at laparotomy and to attempt to localize the site of intraperitoneal chylous fluid leakage. These involve preoperative ingestion of lipophilic dyes by patients, or intraoperative lymphangiography. However, affected segments may be identified at laparotomy without the use of dyes[18] and this approach was favored in the current case.

Resection of affected segments in lymphangiectasia may control symptoms, even in the presence of ongoing protein loss.[11,17,18] Given one of the proposed mechanisms for the development of PLE post-Fontan is raised systemic venous pressure, predisposing to lymphangiectasia, a generalized abnormality of intestinal lymphatics might be expected. However, circumscribed intestinal protein loss has been described in this context.[39] Interestingly, associated factors such as heterotaxia, polysplenia, anomalies of systemic venous drainage, and ventricular anatomy, none of which are present in the current case, have been found to increase the risk of developing PLE post-Fontan procedure.[28] Also, lymphangiectasia and cardiac lesions are associated in several dysmorphic syndromes, including Turner,[40,41] Down,[42] and Noonan[43–45] and with 22q11 microdeletions.[46] Perhaps those cases developing PLE post-Fontan have an underlying localized lymphatic malformation reflecting a defect in angiogenesis that affects both cardiac and lymphatic development.

Any decision to surgically resect areas of intestinal lymphangiectasia must weigh the potential benefits of surgery against the risks of complications and the possibility of persistence or recurrence of PLE. Resection may be complicated by the development of short bowel syndrome, with associated diarrhea, weight loss, and specific nutritional deficiencies. The resection of a segment of jejunum in the current case was not associated with any features of short gut syndrome, in fact nutrition and growth improved dramatically. Minimal ongoing protein loss did persist postoperatively, requiring maintenance diuretic therapy, however there has been no recurrence of chylous ascites or pleural effusions. There is a possibility that symptomatic protein loss may recur in time; however, compared with the known 50% 5-year mortality of her preoperative condition and the multiple life-threatening complications she had already experienced, the 2 years of good health she has enjoyed since the surgery demonstrate the benefits of segmental resection in this case.

In intractable cases of PLE post-Fontan procedure, a careful search for localized intestinal lymphangiectasia is warranted. $^{99m}$Technetium-dextran scintigraphy appears to be a sensitive and well-tolerated investigation to define the site(s) and extent of intestinal protein loss in these circumstances. This technique offers new hope for children with this devastating complication of cardiac surgery.

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