Intestinal pneumatosis and gas in the portomesenteric vein are most often associated with intestinal infarction, necrotic small bowel, or gastric volvulus [1,2]. Surgical treatment should be considered in symptomatic patients [3].

We report here the case of a 21-year-old man with Niemann–Pick disease who presented with acute abdominal pain. Due to neuromuscular malfunctions, he had received a percutaneous endoscopic gastrostomy (PEG) with a jejunal extension 2 years previously, as well as a permanent tracheostoma. Initial laboratory findings showed an elevated lipase level, and ultrasonography revealed edematous pancreatitis. Liver enzymes, inflammatory parameters, and serum lactate were only slightly elevated.

When the abdominal symptoms became progressive, a second ultrasound examination showed a nearly completely extinguished liver signal, and ultrasonography revealed edematous pancreatitis. Liver enzymes, inflammatory parameters, and serum lactate were only slightly elevated.

Figure 1  Contrast-enhanced computed tomography (CT) of the abdomen, showing marked air accumulations within the portal venous system (white arrows). The inhomogeneous imaging of the spleen, caused by lipid accumulation (empty arrow) due to the patient’s Niemann–Pick disease. An unenhanced CT 12 h after extraction of the defective jejunal feeding tube showed distinct regression of the intrahepatic pneumatosis (not shown).

Figure 2  The extracted defective jejunal feeding tube. The arrow shows the bare metallic end where the protective cap is missing.

With the absence of intestinal ischemia and the marked regression of the portal gas accumulation after extraction of the defective jejunal tube, the most likely explanation is that it was intramural invasion by the jejunal tube that caused the intestinal and portosystemic pneumatosis.

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