specifically loss of CD3+ T cells. Thus, lymphopenia in a patient with hypoproteinemia indicates increased loss of protein into the GI tract.

Patients with increased protein loss into the GI tract from lymphatic obstruction often have steatorrhea and diarrhea. The steatorrhea is a result of altered lymphatic flow as lipid-containing chylomicrons exit from intestinal epithelial cells via intestinal lymphatics (Table 325-2; Fig. 325-4). In the absence of mechanical or anatomic lymphatic obstruction, intrinsic intestinal lymphatic dysfunction—with or without lymphatic dysfunction in the peripheral extremities—has been designated *intestinal lymphangiectasia*. Similarly, ~50% of individuals with intrinsic peripheral lymphatic disease (Milroy's disease) also have intestinal lymphangiectasia and hypoproteinemia. Other than steatorrhea and enhanced protein loss into the GI tract, all other aspects of intestinal absorptive function are normal in intestinal lymphangiectasia.

Endoscopy and Imaging Endoscopy with biopsy and video capsule endoscopy may be performed to rule out mucosal disease. Magnetic resonance enterography may be helpful in children with lymphangiectasia.

Other Causes Patients who have idiopathic protein-losing enteropathy without evidence of GI disease should be examined for cardiac disease. As more patients with congenital heart disease reach adulthood, Fontan physiology has become a more common cause of protein-losing enteropathy. Other cardiac causes include right-sided valvular disease and chronic pericarditis **(Chaps. 268 and 270)**. Ménétrier's disease (also called *hypertrophic gastropathy*) is an uncommon entity that involves the body and fundus of the stomach and is characterized by large gastric folds, reduced gastric acid secretion, and, at times, enhanced protein loss into the stomach.

TREATMENT

Protein-Losing Enteropathy