A 4-year old boy with D-transposition of the great arteries, pulmonary atresia with intact ventricular septum, and a hypoplastic left ventricle with mitral atresia underwent a Fontan operation. He had previously been treated with neonatal balloon atrial septostomy followed by a left modified Blalock-Taussig shunt and, at the age of 2 years, a Hemi-Fontan operation was performed. The Fontan operation was completed with an extracardiac conduit (Goretex) from the inferior vena cava to the superior vena cava. High central venous pressures and bilateral chylothoraces complicated the postoperative course. Despite 6 weeks of conservative treatment with fat-free nutrition and subsequent total parenteral nutrition, pleural fluid loss remained excessive, and a pleurodesis was performed bilaterally. A few days postoperatively, acute respiratory failure occurred due to bronchial casts. Cast analysis showed high triglyceride and protein concentrations (cast triglyceride, 0.96 mmol/L; cast protein, 66 g/L; serum triglyceride, 0.36 mmol/L; and serum protein, 47 g/L). Bronchoscoplc attempts to remove the bronchial casts failed, and the child died of cardiorespiratory failure.

At autopsy, the bronchial system was completely plugged by casts resembling bronchial tree anatomy (Figure 1). Thromboses obliterated the left innominate vein partially and reduced the lumen of the extracardiac conduit by 50%. These thromboses occurred despite full anticoagulation and were not detected by previous transthoracic echocardiography. Histopathology demonstrated massively dilated pulmonary lymph vessels within the whole lung (Figure 2). Ruptures of lymphatic vessels and the influx of chyle into the alveoli could clearly be demonstrated (Figure 3). The material in the alveoli was proven to be chyle by lipid staining (Figure 4). The mechanism of lymphatic leakage leading to the formation of bronchial casts has been discussed, but it had not been demonstrated histologically before. This condition must be distinguished from mucus hypersecretion as an unusual response of the pulmonary epithelium to elevated venous pressure (classified as type II or as acellular mucin casts by Seear et al.), where no alveolar flooding with chyle is present.

References


**Figure 1.** Bronchial cast consisting of clotted chyle that was extracted from lung at autopsy.

**Figure 2.** Massively dilated lymphatic vessel with chyle content. The chyle-containing space (*) is lined by lymphatic walls on one side (arrow) and by alveolar walls on the other side (arrowhead). Immunohistochemistry for collagen IV; magnification, 115×.
Figure 3. Lung tissue with dilated lymphatic channel (*) and rupture into alveolar spaces (arrow). Immunohistochemical staining for collagen IV; magnification, 60×.

Figure 4. Fat-containing chyle (*) and fat-laden macrophages (arrow) within lymphatics and adjacent to alveolar spaces. Frozen section with Sudan-red; magnification, 235×.
Chylous Bronchial Casts After Fontan Operation
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