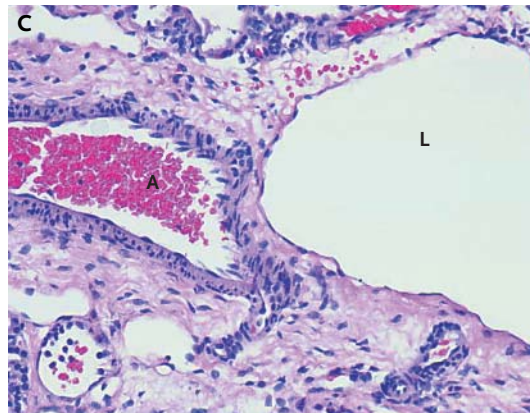
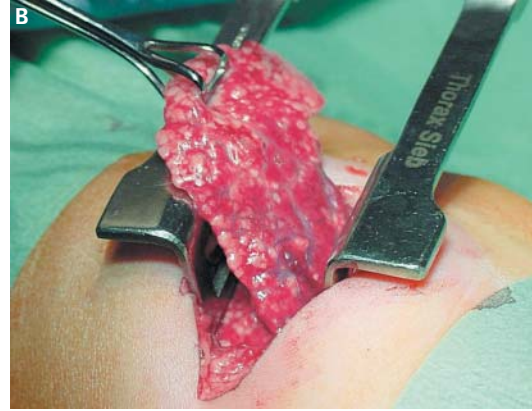


## IMAGES IN CLINICAL MEDICINE

## Congenital Pulmonary Lymphangiectasia



**A** FEMALE INFANT WAS BORN AT 40 WEEKS AND 3 DAYS OF GESTATION, weighing 2970 g. The Apgar scores were 8, 10, and 10 at 1, 5, and 10 minutes, respectively. Within one hour after birth, severe respiratory distress with profound cyanosis developed while the infant was breathing room air, and intubation and mechanical ventilation were required. An initial chest radiograph showed normal lung volumes with diffuse, bilateral nodular changes, and a subsequent chest radiograph obtained on the 17th day of life showed that these changes had become more prominent (Panel A). Surfactant and antibiotics were administered. Lung compliance remained poor, and high-frequency oscillatory ventilation was begun. Congenital heart disease was ruled out, and cultures of blood and tracheal aspirates were unrevealing. After surfactant protein B deficiency was ruled out, an open-lung biopsy was performed, on the 18th day of life. On gross examination, the lung had an irregular surface with scattered nodular changes (Panel B). On microscopical examination, there was subpleural and septal cystic lymphangiectasia (L, Panel C; hematoxylin and eosin,  $\times 150$ ). A denotes artery. These findings are consistent with a diagnosis of congenital pulmonary lymphangiectasia, which is a uniformly fatal disease when it manifests in the newborn period. The infant died on the 20th day of life.

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