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PULMONARY LYMPHANGIECTASIA

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ABSTRACT

Congenital pulmonary lymphangiectasia (PL) is a rare developmental disorder involving the lung and is characterized by pulmonary subpleural, interlobar, perivascular, and peribronchial lymphatic dilatation. Both frequency and etiology are unknown. PL presents at birth with severe respiratory distress, tachypnea, and cyanosis, with a very high mortality rate at or within a few hours of birth.

At birth, mechanical ventilation and pleural drainage are nearly always necessary to obtain a favorable outcome of respiratory distress. Home supplemental oxygen therapy and symptomatic treatment of recurrent cough and wheeze are often necessary during childhood, sometimes associated to prolonged pleural drainage. Recent advances in intensive neonatal care have changed the previously nearly fatal outcome of PL at birth.

Patients affected by PL who survive infancy present medical problems which are characteristic of chronic lung disease.