

Congenital pulmonary lymphangiectasia (CPL) is a rare developmental disorder that is present at birth (congenital). Affected infants have abnormally widened (dilated) lymphatic vessels within the lungs. The lymphatic system helps the immune system in protecting the body against infection and disease. It consists of a network of tubular channels (lymph vessels) that drain a thin watery fluid known as lymph from different areas of the body into the bloodstream. Lymph accumulates in the tiny spaces between tissue cells and contains proteins, fats, and certain white blood cells known as lymphocytes. According to some reports CPL occurs more often in males than females. However, the data is not conclusive and CPL may occur in males and females in equal numbers. The exact number of cases of CPL is unknown. In the past, the disorder was associated with high mortality in the newborn period. CPL was first described in the medical literature in 1856.