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Pulmonary agenesis

INSERM

Source

INSERM. (1999). *Orphanet: an online rare disease and orphan drug data base. Pulmonary agenesis. ORPHA:984*

A rare, non-syndromic respiratory or mediastinal malformation characterized by unilateral complete absence of lung tissue, bronchi, and pulmonary vessels. It may be isolated or associated with congenital malformations, most commonly with heart anomalies. Presentation is highly variable including airway narrowing, stridor, respiratory distress, recurrent respiratory tract infections, and pulmonary hypertension.