

UNILATERAL LUNG AGENESIS: IMPLICATIONS FOR THE PAEDIATRIC SURGEON

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Background: Unilateral lung agenesis is a rare anomaly, with a paucity of published literature, usually restricted to case reports. We aimed to review our experience to better inform antenatal and postnatal counselling, in terms of respiratory outcome and associated anomalies.

Methods: Retrospective case-notes review of all children at our institution with unilateral lung agenesis 1995-2015.

Results: We identified four patients with unilateral lung agenesis (1-18y, 3 female, 3 right sided). Two had unilateral lung agenesis detected antenatally. One other was initially misdiagnosed as a diaphragmatic hernia, confirmed as isolated lung agenesis with intact diaphragm postnatally.

Patient#1 has right lung agenesis, associated oesophageal atresia (OA, type-C) and underwent repair by right thoracotomy with extra-pericardial approach. She has subsequently required fundoplication and aortopexy (tracheobronchomalacia).

Patient#2 has right lung agenesis, tracheobronchomalacia and reflux (conservatively managed), and a repaired pulmonary artery sling.

Patient#3 has left lung agenesis, left MCDK, and tracheobronchomalacia (conservatively managed).

Patient#4 has right lung agenesis, repaired ASD, right radial club hand/syndactyly, tracheobronchomalacia and reflux (conservatively managed).

Despite frequent admissions during the first few years, the medium term outcome is good without significant respiratory morbidity in the older two patients. All patients are below the 2nd centile for growth and have impaired lung function tests. However one patient's exercise tolerance is good enough to allow him to play badminton and rugby.

Conclusions: Unilateral lung agenesis is rare and may present antenatally. Associated OA can be repaired safely via right thoracotomy in the presence of right lung agenesis. The association of ipsilateral radial deformity and lung agenesis adds weight to this association shown in other reports. Tracheobronchomalacia is universal and reflux is common, both can usually be managed conservatively. Despite impaired lung function tests reasonable exercise tolerance can be anticipated in older children.