Heterotopic brain tissue in the lung causing acute respiratory distress in an infant

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ABSTRACT A three month male infant with a 24 hour history of cyanotic attacks was found to have heterotopic brain tissue at thoracotomy. He has remained well over 20 years of follow up. The pathogenesis of the condition is discussed.

We report a case of heterotopic brain tissue in the lung of a three month old male infant presenting with cyanotic attacks. Only one previous case of brain heterotopia in the lung causing neonatal respiratory distress has been described.1

Case report

A 3 month old male infant presented with a 24 hour history of repeated cyanotic attacks. The child was born after an uneventful pregnancy and spontaneous vaginal delivery. At birth he was found to have a cleft lip and cleft hard and soft palate, but no other congenital abnormalities. The child was fed by spoon and was gaining weight normally. No corrective surgery had been undertaken. A chest radiograph on admission showed a large tension cyst in the right chest, displacement of the heart to the left, and anterior herniation of the mediastinum. The infant was taken to the operating theatre and a needle was inserted into the right chest cavity with the release of some air, but little clinical improvement occurred. A thoracotomy was hurriedly performed. A large thin walled cyst in the upper lobe and two large cysts in the lower lobe of the right lung were resected. The child made an uneventful recovery and subsequently had surgery to correct his cleft lip and palate. He was followed as an outpatient for 20 years and showed normal mental and physical development.

PATHOLOGICAL FINDINGS

Three collapsed cysts were received, measuring 7, 2.5, and 2 cm in diameter. Microscopic examination showed that the cyst walls were composed of mature neural tissue with neurones, glial cells, and glial fibres (the glial fibres stained with phosphotungstic acid haemotoxylin). Within the neural tissue were irregular ductal structures lined by ciliated columnar epithelium similar in appearance to the bronchiolar epithelium in the surrounding rim of haemorrhagic

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A: Neuroglial tissue (N) containing tubules adjacent to collapsed, haemorrhagic lung (L); a cyst cavity is seen top right.

B: Tubules lined by ciliated epithelium lying in a background of neuroglial tissue.
This within the brain and fail lung systemic theories of brain tissue. The child presented distress, mediastinum showed radiograph of brain tissue. The infant was the brain tissue. The finding of necropsy brain tissue. The infant was the brain tissue, anencephaly and of brain tissue, anencephaly, cleft palate, talipes equinovarus development growth. With the subsequent development of the lung the bronchi grow into the neural tissue but alveoli, muscle, and cartilage fail to develop owing to the effect of the abnormal stroma. This theory explains the finding that the duct structures within the brain tissue are in continuity with the bronchial tree and that there are usually multiple peripheral and subpleural areas of neural tissue. Against the second theory is the fact that intraterine respiratory movements are shallow and insufficient to carry brain tissue out to the lung periphery. Only during birth hypoxia are respiratory movements sufficient to carry amniotic fluid into the peripheral alveoli.

In summary, we present a case of heterotopic brain tissue within the lungs causing tension cysts and respiratory distress in a 3 month old infant with cleft lip and palate. This case is unusual in that the child was without anencephaly and of normal intelligence.

### References