

Long Term Outcome with Congenital Pulmonary Airway Malformation (CPAM). Spirometry Following Conservative or Surgical Management.

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Children with a Congenital Pulmonary Airway Malformation (CPAM) may require emergency surgery shortly after birth. Many, however, remain asymptomatic and are offered either elective surgery or a conservative approach. All children should be followed up long term to identify potential complications. Since 2004 we have ongoing follow up of 63/88 children aged over 5 years (from a cohort of 166 children & infants) with an antenatal diagnosis of CPAM.

Methods:

60 children had spirometry as part of their most recent annual follow up. 3 others were unable to perform the technique. 25 have been lost to follow up.

Results:

42 had surgery, 6 as an emergency for early respiratory distress, and 18 managed conservatively.

Spirometry was performed at a mean of 8.37 years (5.4-16.0 years): Mean FEV₁ 1.48L (93% predicted), FVC 1.79L (97% predicted).

There were no significant differences between children with conservative and surgical management plans. There were no significant differences in spirometry in children with an histological diagnosis of type1 CPAM, type 2 CPAM or pulmonary sequestration.

There was a trend to higher lung function in children managed with thoroscopic surgery (n=15) compared to thoracotomy (n=26).

Spirometry from children (n=6) requiring emergency surgery was reduced; Mean FEV₁ 1.27L (77% predicted), FVC 1.65L (88% predicted).

Discussion:

There is ongoing debate about the optimal management for children with asymptomatic CPAM. Rates of elective surgery have probably declined and surgical techniques also modified from thoracotomy/lobectomy towards thoracoscopy/segmentectomy or sequestrectomy. The pulmonary outcome may also be affected by co-existent pathology.

Conclusion:

This data supports the evidence that the outcome from CPAM is good whether surgically or conservatively managed. Further information looking at static lung volumes and exercise tolerance in a larger cohort would be useful to inform management decisions further and, if appropriate, the surgical approach.

