PAEDIATRIC RESPIRATORY REVIEWS (2007) **8**, 67–76

Paediatric Respiratory Reviews

CME ARTICLE

Congenital cyst adenomatoid malformations: resect some and observe all?

Dominic A. Fitzgerald

Department of Respiratory Medicine, The Children's Hospital at Westmead, Sydney, Locked Bag 4001, Westmead, NSW, Australia 2145

EDUCATIONAL AIMS

- To discuss the clinical course of congenital cyst adenomatoid malformations (CCAM) from the time of diagnosis to resection or spontaneous resolution.
- To illustrate the merits of surgical resection of large symptomatic CCAMs.
- To discuss the limited evidence for the universal surgical resection of all CCAMs based on the risk of malignancy.
- To help the reader to appreciate the rationale for conservative management of infants with an asymptomatic CCAM.
- To emphasise the importance of longitudinal clinical follow-up of all patients with CCAMs, whether a conservative or a surgical approach has been undertaken.

KEYWORDS
congenital cyst adenomatoid malformation (CCAM);
antenatal diagnosis;
radiologic imaging;
surgery;
cancer risk;
prognosis;
conservative management

Summary With the advent of improved antenatal imaging over the past 10 years, the diagnosis, assessment and management of congenital cystic lung abnormalities have changed. These were once considered the exclusive domain of the surgeon, who had the authority to operate on all congenital cystic lung abnormalities regardless of size or clinical signs in order to avoid the risk of cancer and improve lung growth in even asymptomatic infants. Clinicians are reconsidering this approach in the light of an appreciation of the spontaneous improvement and possible resolution that occurs over months to years with many of these lesions. The risks of subsequent cancer are poorly understood and probably overstated, whereas the magnitude of compensatory lung growth is poorly defined in the majority of children with small unilateral congenital cystic abnormalities. The evolution of regional, national and ideally international databases will provide much-needed longitudinal data to better inform clinicians of the optimal way in which to manage these children.

The optimal management of cystic lung lesions in young children has been based upon the surgical aphorism 'When in doubt, cut it out!' With regard to congenital cyst adenomatoid malformations (CCAMs), the most common of congenital cystic lung lesions, this view appears to be changing in the light of a growing understanding of the likelihood that antenatal cystic lung lesions tend to shrink in the last trimester of pregnancy and in the early years of life. The evolution of better antenatal imaging has been crucial in changing thinking to the point at which it is no longer considered heresy to observe smaller lesions in otherwise well, asymptomatic infants. The central issues of later cancer risk and optimising lung growth are being re-evaluated. This has been prompted

E-mail address: dominif2@chw.edu.au

1526-0542/$ – see front matter © 2007 Elsevier Ltd. All rights reserved.
doi:10.1016/j.prrv.2006.10.001

KEY POINTS

- The improving resolution of fetal imaging has increased the likelihood of a congenital cystic lung lesion being recognised with routine antenatal ultrasound.
- Congenital cyst adenomatoid malformations (CCAMs) are the most common form of congenital cystic lung lesion.
- The natural tendency for many cystic lung lesions to shrink in the third trimester of pregnancy has become apparent.
- Infants with large (including bilateral) cystic lung lesions may develop hydrops in utero or severe respiratory distress after birth and require urgent resection of the cystic lesion.
- For a child with a congenital cystic lung lesion, a 'normal' early postnatal chest radiograph does not exclude a residual cystic lesion so that a computed tomography scan is recommended to delineate the residual lesion in early infancy.
- In infants with asymptomatic, unilateral, small CCAMs, the opportunity for clinically significant lung growth and the true risk of subsequent cancer is probably overstated as an indication for surgery.
- The majority of children with CCAMs diagnosed antenatally or in the early postnatal period have traditionally undergone surgery in infancy. There is, however, an increasingly held view that a more conservative approach with regular review is warranted, particularly in those infants who are minimally symptomatic or asymptomatic in early infancy.

Table 1 Classification of congenital lung malformations

Bronchopulmonary malformations (more common)
1. Bronchogenic cyst
2. Bronchial atresia
– Isolated
– With vascular connection (intralobar sequestration)
– With connection to the gut
– With a systemic artery connection to normal lung
3. Cystic adenomatoid malformation, large cyst type/ macrocystic (Stocker type I)
– Isolated
– Systemic vascular connection
4. Cystic adenomatoid malformation, small cyst type/ microcystic (Stocker type II)
– Isolated
– Systemic vascular connection
5. Extralobar sequestration
– With connection to the gut
– Without connection to the gut
Pulmonary hyperplasia and related lesions (uncommon)
1. Laryngeal atresia
2. Solid cystic adenomatoid malformation (Stocker type III)
3. Polyalveolar lobe
Congenital lobar overinflation/congenital lobar emphysema (more common)
Other cystic lesions (rare)
1. Lymphatic cystic malformations
2. Enteric cysts
3. Mesothelial cysts
4. Parenchymal cysts
5. Low-grade cystic pleuropulmonary blastoma (Stocker type IV)

Derived from.[2–5]

by an improved understanding of the classification of CCAMs and their natural course, and a recent move toward the generation of large-scale databases to more accurately define the natural history of CCAMs and their association, probably exaggerated, with pleuropulmonary blastomas (PPBs) and bronchoalveolar carcinoma. This article will review current knowledge on CCAMs and discuss management options for both symptomatic and asymptomatic young children.

NOMENCLATURE

The descriptive terms for congenital cystic lung lesions are not straightforward. Although the first report of a congenital cyst adenomatoid malformation occurred in 1949,[1] the nomenclature of congenital cystic lung lesions has evolved over time, largely based on histopathological descriptions from 30 years ago.[2] There have been more recent attempts[3] to update the classification, these primarily including the following bronchopulmonary malformations: congenital cyst adenomatoid malformation (CCAM), bronchogenic cysts, extralobar pulmonary sequestration and bronchial atresia. These are derived from Langston (2003),[3] as seen in Table 1. This includes the terms popularised by Stocker *et al.*[2] and expanded subsequently to include types 0 and IV.[4]

There is disagreement between the views of leading authors, Stocker and Langston, over the aetiology of the congenital cystic abnormalities, Langston favouring a malformation sequence based primarily upon airway obstruction during development while suggesting that Stocker's classification has a more diverse aetiology.[3,5] This may include a disruption of humoral factors from the mesenchyme at a critical stage of distal airway development leading to an abnormal differentiation of lung tissue, analogous to a hamartomatous malformation.

The most noteworthy aspect of the controversy relates to the relationship between a low-grade PPB and a Stocker type IV CCAM and whether the classifications are identical. The advent of better antenatal imaging has facilitated a deeper appreciation of the dynamic nature of cystic lung lesions in utero[6–8] and coincided with a laudable suggestion for more clinically descriptive terms to be applied to all