

The Postnatal Management of Congenital Cystic Adenomatoid Malformation

Sergey Keidar MD¹, Liat Ben-Sira MD², Mark Weinberg MD¹, Ariel J. Jaffa MD³, Aviel Silbiger MD⁴ and Itzhak Vinograd MD¹

Departments of ¹Pediatric Surgery, ²Radiology, ³Gynecology and ⁴Anesthesiology, Dana Children's Hospital, Tel Aviv Sourasky Medical Center and Sackler Faculty of Medicine, Tel Aviv University, Israel

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Abstract

Background: Routine prenatal ultrasound has increased the frequency of prenatal diagnosis of congenital cystic lung malformation, such as cystic adenomatoid malformation, pulmonary sequestration, congenital lobar emphysema, and bronchogenic cyst.

Objectives: To evaluate the methods of postnatal diagnosis, the optimal age for operation since surgery is always required, and the optimal extent of lung resection.

Methods: The clinical courses of 11 patients with congenital lung cysts who underwent surgical lung resection (8 lobectomies and 3 segmentectomies) were reviewed.

Results: The diagnosis was confirmed by computed tomography scan in all. In nine patients the diagnosis was made prenatally. Chest X-ray was normal postnatally in all patients except for two who had recurrent pneumonia. Post-operative follow-up showed excellent recovery in all operated children. One patient who underwent surgery for CCAM following episodes of severe pneumonia died from another cause 5 months later. Postoperative chest CT scan showed no residual disease in eight patients. In two who had undergone limited resection, tomography showed a small segment of residual disease in one and a suspected residual lesion in the other.

Conclusion: With prenatal ultrasound the true frequency of congenital cystic lung anomaly appears to be higher than previously reported. Postnatal CT is mandatory to confirm or to rule out the diagnosis. The mere presence of cystic lung malformation is an indication for surgery. Complete removal of the affected lung lobe is recommended. Segmental resection may be inadequate. Early operation is tolerated well by infants and small children and we recommend that surgery be performed in children between 6 and 12 months of age.

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Most cystic anomalies of the lung in infants and children are of congenital origin. During the last decade prenatal sonographic diagnosis of such lesions has become possible from as early as 16–18 weeks of gestation, and has become a valuable and a routine diagnostic method [1–3]. It enables the treating physician to identify more cystic lung lesions, but the distinct type of cystic anomaly is usually indistinguishable. While postnatal chest X-ray assessment is usually negative, computerized tomography provides important information about the size, location and character of the cystic lesion, confirming the diagnosis [4,5].

There are four main types of congenital cystic anomalies of the lung: pulmonary sequestration, cystic adenomatoid malformation, congenital lobar emphysema, and bronchogenic cyst. All have similar embryologic origins and all require surgical management. Pulmonary sequestration is subdivided into intralobar, which carries the risk of infection and therefore requires surgical removal; and extralobar, which is usually asymptomatic, but if diagnosed needs removal for histological confirmation as does any pulmonary mass [6]. Congenital lobar emphysema necessitates surgery due to the possibility of respiratory distress secondary to lung compression by the enlarging cyst or from pneumothorax. Bronchogenic cyst also requires surgery due to possible tracheal obstruction with subsequent pneumonia, and because of its rare association with malignancy [4].

There is a wide range of possible presentations of CAM anomaly, extending from lung hypoplasia with fetal demise, severe fetal hydrops [7], severe respiratory compromise [8], to rare complete *in utero* regression. These lesions may require a variety of different interventions, ranging from fetal drainage *in utero* [2,7] to elective lobectomy or segmentectomy, or merely observation [6]. Stocker et al. [9] classified CAM into three main histological types: Type I, with a good prognosis, is characterized by large cystic cavities greater than 2 cm in diameter; type II, the most common type, consists of multiple smaller cysts; and type III, which is predominantly a solid lesion producing mediastinal shift, carries the worst prognosis due to fetal hydrops.

Traditionally, a vague prognosis was assigned to CAM, possibly because of the delayed manifestation of complications. However, recent series supported by contemporary imaging

CCAM = congenital cystic adenomatoid malformation

modalities have updated our understanding of the natural history of the disease and demonstrated favorable outcomes after surgical treatment in most cases [3,6,10–16]. Postnatally, patients may be divided into those who are symptomatic and require early surgical intervention but improve once the lesion is resected, and those who are asymptomatic. The previously accepted view that asymptomatic lesions need only to be followed has changed following reports of malignant degeneration of CAM lesions [12–16]. State-of-the-art management now calls for surgical removal within normal anatomic margins.

Nonetheless, despite the increasing number of published series of CCAM [1–4,11,14–16], two questions remain to be resolved: the optimal age for surgery in asymptomatic patients, and whether every resection of the viscus should include lobectomy rather than segmental resection.

Materials and Methods

We prospectively analyzed 11 patients with a congenital cystic lung anomaly who underwent surgery in our department during a one year period. Most of them (9 of 11) were sonographically diagnosed and were followed prenatally. Chest X-ray was performed routinely in all patients, and CT of the chest was the image modality of choice in all patients regardless of their symptoms. The routine procedure, after discussion with the parents, was to operate urgently in a symptomatic patient, while in asymptomatic patients with a positive CT scan we recommended a planned resection. One preterm infant was

symptomatic at birth and was ventilated until surgery was performed in early infancy. Chest tomography revealed cystic masses in three children with recurrent pneumonias, and they were referred for surgical resection without delay. The diagnosis was confirmed histologically in all cases. In one child who underwent a segmentectomy a frozen study was undertaken.

Results

We treated 11 patients, 6 girls and 5 boys. The ages of the patients at the time of surgery ranged from 1 week to 23 months. The mean ages of asymptomatic and symptomatic patients were 9.8 and 7.3 months respectively. Table 1 reviews the clinical course of these patients. Nine underwent prenatal sonographic evaluation from the 16th to 24th week of pregnancy with follow-up scans that were consistent with cystic lung malformation. In three cases the cysts had completely disappeared sonographically during the last quarter of pregnancy and in one case there was regression of the original finding. One, with extensive lung involvement, had polyhydramnion. In the two children who did not undergo prenatal sonographic studies the cystic pulmonary lesions were found during investigation for intractable recurrent pneumonia.

The chest film was normal in seven patients, six of whom were asymptomatic. All four symptomatic patients had abnormal chest radiograms. One preterm infant with prenatally known disease and polyhydramnion was ventilated due to respiratory compromise at birth, and right bilobectomy in early

Table 1. Clinical features of 9 cases of CAM and 1 of pulmonary sequestration

Patient no.	Gestational age at diagnosis (wk)	Change on US	Symptoms	Chest X-ray	CT scan	Operation	Pathology	Postop complications	Postop hosp. stay (days)
1	21	Disappeared	None	Normal	Small cysts	RLL segmentectomy	Type II	Susp. residual CCAM	4
2	20	Disappeared	Mild	Normal	Cysts	RLL lobectomy	Type I	–	6
3	24	None	None	Normal	Cysts	RUL lobectomy	Type I	–	4
4	16	Disappeared	None	Normal	Cysts	LLL lobectomy	Intralobar sequestration	–	4
5	20	None	None	Normal	Cysts	RML lobectomy	Type I	–	7
6	22	None	Severe distress	Abnormal	Huge bilobar cysts	RML + RUL bilobectomy	Type II	–	26
7	–	–	Pneumonia	Abnormal	Cysts	RUL lobectomy	Type II, pneumonia	Late death	4
8	24	None	None	Cyst	Cysts	RLL lobectomy	Type II within sequestration	–	4
9	21	Smaller	None	Normal	Small Cysts	RLL segmentectomy	Type II	–	4
10	20	None	None	Normal	Cysts	RML segmentectomy	Type II	Residual CCAM	5
11	–	–	Pneumonia	Abnormal	Cysts	RUL lobectomy	Type II	Pneumonia	42

infancy was tolerated successfully. Two of the symptomatic patients had pneumonia and one had symptoms of respiratory infection. Routine CT assessment was done in all patients. CT follow-up in six asymptomatic patients did not show further regression in the cyst size. Sublobar or subsegmental tomographic lesions were consistent with CAM.

The procedures carried out included lobectomy in seven children, bilobectomy in one, and segmentectomy in three asymptomatic cases. Histological studies in 10 of the children confirmed the diagnosis of CAM, which was located within an extralobar pulmonary sequestration in one, and in another patient intralobar sequestration was confirmed. Using the CAM classification of Stocker et al. [9], three patients had type I cysts and seven had type II cysts.

There were no postoperative complications in the asymptomatic patients. The one (and only) complication, severe long-standing pneumonia, occurred in a child who was severely ill preoperatively. She died 5 months later from congenital heart disease. The mean length of hospital stay in asymptomatic patients was 4.6 days (range 4–7 days). Postoperative follow-up included clinical observation and a CT scan performed within 6 months. Follow-up CT scan in 10 patients (one has died) showed that the cystic anomaly was totally eradicated in 8 patients, and that residual disease and a suspected residual lesion was present in one each of two patients who had a limited resection (segmentectomy). The former of the two patients underwent planned lobectomy and the other patient is under observation.

Discussion

The improvement in prenatal diagnostic techniques has made CAM a more commonly encountered entity within the scope of pediatric surgical practice. Some of these lesions undergo partial or complete regression *in utero* by an unknown mechanism. Despite the finding of this anomaly on ultrasonography, most newborns have normal chest radiographs. While prenatal sonography may show malformation regression, subsequent CT scan usually shows a significant lesion, making CT the postnatal diagnostic method of choice.

The CCAMs, which represent about 25% of the congenital lung malformations, present with a broad and dynamic spectrum of clinical severity – ranging from an asymptomatic lesion, through childhood infection, to neonatal respiratory distress, fetal hydrops and death. This is almost always a unilateral lesion. Respiratory distress at birth is uncommon. The specific cause of the anomaly is unknown. One of the theories explains it as a disturbance in bronchiolar differentiation during the embryonic period, with subsequent mesenchymal overgrowth of the terminal bronchiolar structures. Others suggest that local bronchial collapse followed by consecutive obstruction is responsible for the development of this anomaly. The cystic lesions tend to be unilateral and usually sublobar or subsegmental. The original indications for surgery – respiratory distress and pulmonary infection – have recently been extended

to include an asymptomatic lung cyst or mass. In asymptomatic patients, malignancy and infection are good reasons for removing these congenital lesions, hence the mere presence of CAM is an indication for surgery.

Obviously the decision to operate on an asymptomatic patient must take into account the parents' decision following detailed explanation of the various possibilities. It is known that about 10% of all malignant pulmonary tumors arise in cystic lung malformations; of those, 35% are rhabdomyosarcomas and 20% are pulmonary blastomas. There are some reports of bronchioalveolar carcinoma arising in cystic malformations of the lung. Because pulmonary malignancy arising in CCAM has been reported, surgical resection is of utmost importance and advisable even in asymptomatic lesions. Infants and young children tolerate thoracotomy and resection extremely well, with vigorous growth and expansion of remaining lung tissue.

When to operate – early or late? Since there were no age-related complications and given that infants tolerate the procedure better, we can conclude that the optimal age for surgical intervention is between 6 and 12 months. Other studies [1,4] have confirmed that in limited resection, i.e., segmentectomy, determining the border of resection between affected segment and residual normal parenchyma is difficult. Consequently, this approach carries a high complication rate and frequently requires reoperation.

The outcome of our study showed that total eradication of disease is better achieved by complete lobectomy, since segmental resection was inadequate in two of three cases. Finally, the pediatric surgeon should be closely involved in the prenatal counseling of patients with cystic lung malformation in order to relieve anxiety and ensure a good prognosis after birth.

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- Correspondence: Dr. I. Vinograd, Head, Dept. of Pediatric Surgery, Dana Children's Hospital, 6 Weizmann St., Tel Aviv 69423, Israel. Phone: (972-3) 697-3952, 697-4409, Fax: (972-3) 697-4409, email: vinopesu@tasmc.health.gov.il