Pediatr Surg Int (2009) 25:479–485
DOI 10.1007/s00383-009-2371-5

ORIGINAL ARTICLE

Long-term outcome of asymptomatic patients with congenital cystic adenomatoid malformation

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Accepted: 17 April 2009 / Published online: 30 April 2009

Abstract
Purpose Congenital cystic adenomatoid malformation (CCAM) represents a rare congenital anomaly of the lung. It remains controversial whether patients with asymptomatic lesions warrant early surgical intervention. Our aim was to review the outcome of asymptomatic CCAM patients at a paediatric tertiary centre.
Methods The medical case notes of all children with CCAM presenting to our institution between 1986 and 2007 were reviewed. Data on pre- and post-natal investigations, clinical presentation, lesion site, type of surgical procedure, timing, and outcomes of surgery were reviewed.
Results A total of 35 patients were diagnosed with CCAM during the 21-year study period (1986–2007). Sixty percent ($n = 21$) were asymptomatic at birth including eight patients with prenatal ultrasound scan confirming CCAM. In this group, 18 patients (86%) subsequently developed symptoms (median age 2 years, range 1 month–13 years) and required surgery. Symptoms included pneumonia with or without infected CCAM (43%), respiratory distress (14%) and spontaneous pneumothorax (14%). Eight patients underwent multiple hospital presentations with complications related to CCAM. Of the 21 initially asymptomatic patients, 17 (81%) underwent surgical resection. Only one of these patients was completely asymptomatic prior to surgery. There were eight post-operative complications and no mortality. One patient underwent a second thoracotomy for residual CCAM. The median length of hospital stay was 9 days (range 3–32 days).
Conclusion This study suggests patients who present with asymptomatic CCAM will subsequently become symptomatic. Early surgical referral and intervention may be beneficial to avoid the development of complications.

Keywords
Congenital cystic adenomatoid malformation · Asymptomatic · Surgery

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Introduction

Congenital cystic adenomatoid malformation of the lung (CCAM) represents a rare anomaly of the lung, characterised by a multicystic mass of pulmonary tissue from an abnormal proliferation of the terminal respiratory bronchioles, with suppression of alveolar development between the 7th and 15th weeks of gestation [1]. CCAM has been classified histologically by Stocker et al. [2] based on cyst size and by Adzick et al. [3] based on cyst appearance (micro or macrocystic) on antenatal ultrasound (US).

Large congenital lung malformations can give rise to serious complications in utero such as polyhydramnios, cardiovascular compromise, fetal hydrops and death [3–7]. Others have been reported to have 14–56% chance of regression in utero in the third trimester [1, 4, 5, 8–11].

Postnatally, patients with suspected CCAM can be either symptomatic or asymptomatic at birth. Patients may be symptomatic at birth due to mass effect causing mediastinal shift, pneumothorax, pulmonary hypoplasia or subsequently

early infection [3, 5, 8]. These patients present with respiratory distress in the neonatal period and will require emergent surgery.

More commonly, children remain asymptomatic postnatally [8, 9, 12–14]. The management of these asymptomatic CCAM patients remains controversial. In part, this has been due to concerns about operative morbidity, coupled with uncertainty in relation to the natural history of unresected CCAM. Whilst several series have suggested subsequent adverse outcomes in those patients initially asymptomatic with CCAM [13–16], others have documented either regression or long-term dormancy [1, 4, 17]. Such debate indicates a need for further clinical data to support the development of appropriate management guidelines for this group of patients.

The study sought to review outcome of asymptomatic CCAM patients from birth at a paediatric centre.

Materials and methods

The medical case notes of all children with CCAM admitted to our institution, one of two paediatric tertiary referral centres in Sydney, New South Wales, between January 1986 and 2007 (21 years) were reviewed. Data on pre- and post-natal investigations, clinical presentation, lesion site, type of surgical procedure, timing and outcomes of surgery were collected. Patients who had been incorrectly coded were excluded from further analysis.

Asymptomatic patients in our study comprised two separate groups. The first were those that had a prenatally diagnosed CCAM on US and were asymptomatic at birth. The second included those patients that had a delayed presentation, with symptoms at a later age following an unremarkable ante and postnatal course.

Results

Patient cohort

A total of 35 patients were diagnosed with CCAM during the 21-year review period. Of these, 21 patients (60%) were asymptomatic and 14 (40%) were symptomatic at birth. In the asymptomatic group, the median gestational age at delivery was 40 weeks (range 36–41 weeks). The median birth weight was 3,544 g (range 2,655–4,500 g). There were 13 males (62%) and none had any associated congenital anomalies. In five patients, the prenatal data was not available (Table 1). Three patients in the symptomatic group had associated congenital anomalies. These include a patient with Prune Belly Syndrome, a patient with hydrocephalus and a patient with macrocephaly.

Table 1 Comparison between asymptomatic and symptomatic patients

	Asymptomatic[a]	Symptomatic
Patient number	$N = 21$	$N = 14$
Male-to-female ratio	13:8	5:2
Birth weight (grams, range)	3,544 (2,655–4,500)	3,310 (1,120–4,015)
Gestational age at delivery (weeks, range)	40 (36–41)	38.5 (27–41)
Anomalies	–	3
Mechanical ventilation	–	8
Mortality	–	2
Age at presentation (month, range)	24 (1–156)	0 (0–108)
Age for operation (month, range)	24 (1.5–156)	1 (0–108)
Total length of hospital stay (days, range)	9 (3–32)	21 (1–173)

[a] Five patients missing prenatal data

Investigations

Of the 21 initially asymptomatic patients, 8 (38%) patients had a prenatal US suggesting the diagnosis of CCAM (Table 2). Of these eight, seven patients had a cystic lesion suggestive of CCAM and one patient a hyper-echoic left lung. In the symptomatic group, 10 (71%) patients had an abnormal prenatal US. Seven revealed a cystic lesion, one case reduced lung volume, one was initially thought to have a diaphragmatic hernia and in another the data was missing. Six patients had associated mediastinal shift and two polyhydramnios. The low number of prenatal ultrasound scans in the asymptomatic group most likely represents an underestimate of the true detection rate due to a combination of poor documentation in patients with delayed presentation and the retrospective nature of this review.

Of the 20 patients in the asymptomatic group that had a chest radiograph (CXR) performed at presentation with symptoms, 19 (95%) had abnormalities such as cystic lesion with or without air-fluid level, hyperinflation, pneumothorax or opacity (Table 2). Additional investigations used to help ascertain diagnosis included a CT scan of the chest in 11 patients (52%), US of the chest and/or abdomen, a ventilation/perfusion scan, bronchogram, aortogram or upper gastro-intestinal contrast study. In the symptomatic group, 13 (93%) patients had similar CXR abnormalities (Table 2). One patient had bilateral cystic disease, though the left lung was noted to be more severely affected than the right. This patient died prior to any surgical intervention. There were more patients with mediastinal shift in the symptomatic than asymptomatic group.

Table 2 Summary of investigations

	Asymptomatic	Symptomatic
Prenatal USS	8 (38%)	10 (71%)
Chest radiograph	19 (95%)	13 (93%)
Cystic lesion	10	9
Air/fluid level	5	–
Hyperinflation	2	1
Pneumothorax	1	1
Opacity	1	1
Mediastinal shift	4	11
Collapse/consolidation	3	–
Bilateral	–	1
Computer tomography	11 (52%)	6 (43%)
Other	Chest or abdominal ultrasound scan, ventilation/perfusion scan, bronchogram, aortogram, upper gastro-intestinal contrast study	Echocardiogram, Doppler, renal or abdominal ultrasound scan, ventilation/perfusion scan, upper gastro-intestinal contrast study

One patient in the asymptomatic group had an antenatally diagnosed CCAM lesion at 18 weeks gestation with subsequent ultrasound scan showing regression 4 weeks prior to delivery. The CXR for this child was normal at birth and also at 3 months. He developed unexplained fevers and a CT chest scan performed at 3 years of age revealed a multi-loculated cystic lesion in the left lower lobe consistent with a CCAM. This was confirmed histologically following a left lower lobectomy.

Symptoms

In the symptomatic group, majority (86%, $n = 12$) of patients were in respiratory distress at birth (Table 1). Eight patients required mechanical ventilation. Two patients were electively intubated at birth. Five others were subsequently intubated and one placed on continuous positive airway pressure due to persisting respiratory distress. One patient in this group died from overwhelming sepsis and respiratory failure prior to any surgery. He was one of triplets, born with hydrocephalus, subsequently developing intraventricular haemorrhages and recurrent pneumothoraces from bilateral CCAMs.

In the initially asymptomatic group, the majority (86%, $n = 18$) of patients subsequently developed symptoms at a median age of 2 years (range 1 month–13 years, Table 1), with the majority of patients presented with symptoms as an infant (Fig. 1). Symptoms included pneumonia with or without infected CCAM (43%), respiratory distress (14%), spontaneous pneumothorax (14%) and chronic cough (14%, Fig. 2). Two patients had insufficient data regarding symptom presentation. Both patients had prenatal ultrasound scan showing CCAM and one subsequently underwent surgery. One patient had an increase in size of their cystic lesion and was completely asymptomatic prior to elective surgery at 1 month of age.

Eight patients (38%) required more than one hospital admission for treatment of CCAM-related symptoms. The majority ($n = 6$) had recurrent episodes of pneumonia or a single episode that failed to settle despite antibiotic treatment. One patient had recurrent pneumothoraces and another had persistent shortness of breath requiring admission. These patients developed symptoms and were referred for surgery at a median age 4 years (range 6 months–13 years). The median length of total hospital stay for this group was 13.5 days (range 6–32 days).

Ten patients were noted in retrospect to have suffered earlier symptoms. These included previous pneumonia, chronic cough, laboured breathing since birth, frequent

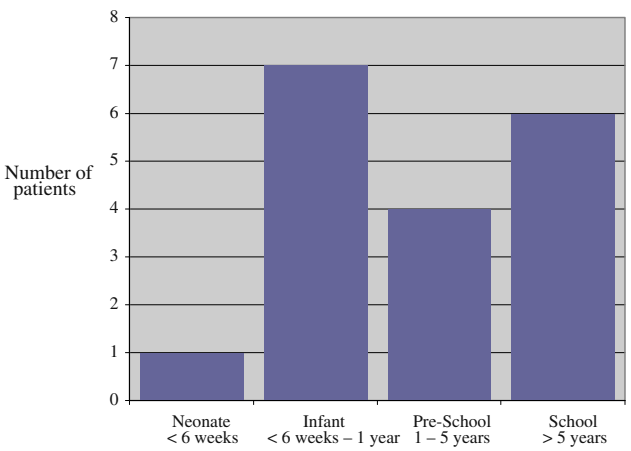

Fig. 1 Age at symptomatic presentation. Missing data two patients

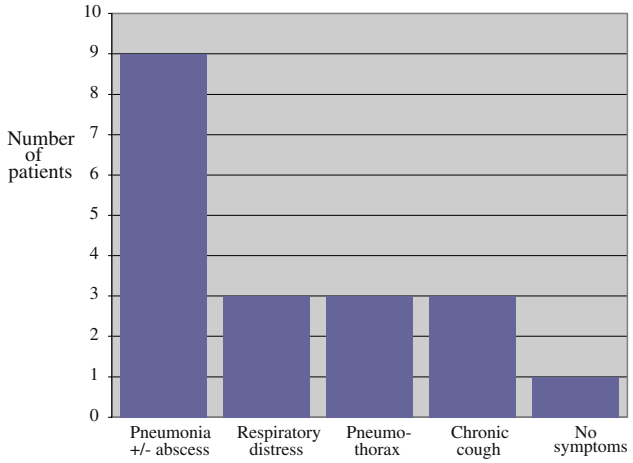

Fig. 2 Symptoms at presentation. Insufficient data in two patients

'asthma' attacks or recurrent severe upper respiratory tract infections. Taking these earlier presentations into account, near half (48%) of asymptomatic CCAM patients at birth developed symptoms during their first year of life.

Surgical treatment

In the asymptomatic group, 19 patients had surgical treatment of which 17 (81%) underwent a lobectomy (Table 3). One patient each had a segmental resection and marsupialization of the right lower lobe lung cyst. This later patient was readmitted to hospital 2 months after discharge with a spontaneous pneumothorax and returned to theatre for a right lower lobe lobectomy. Median age at operation was 2 years (range 6 weeks–13 years). CCAM occurred more frequently on the left (11 cases, 52%) than the right (8 cases, 38%), with the right lower lobe (7 cases, 33%) being most commonly affected. Three patients (14%) had more than one lobe affected. The diagnosis was confirmed in all cases on subsequent histopathology.

Two patients have not yet received surgery. One patient had pneumonic symptoms but had an initial bronchoscopy that was reportedly normal. A follow-up CXR at 6 months revealed a cystic lesion in the right middle lobe. This patient did not have a CT scan and was subsequently lost to follow-up. Another patient had prenatal US diagnosed CCAM of the right lung, which was also noted on CXR at birth. Follow-up CXR at 7 months of age was reported as normal. A CT chest scan was performed to confirm resolution but instead revealed a lobulated cystic structure in the right lower lobe. This patient awaits further imaging.

In the symptomatic group, 13 patients had surgical treatment of which 12 (86%) underwent a lobectomy (Table 3). One patient underwent a marsupialisation of the left lower lobe lung cyst. Unfortunately, this patient was subsequently transferred to another hospital and was lost to follow-up. Median age at operation in this group was at 1 month (0–9 years). CCAM occurred more frequently on the left (10 cases, 71%) than the right (3 cases, 21%), with the left upper lobe (6 cases, 43%) being most commonly affected. One patient presented with a spontaneous right pneumothorax and a lobectomy was performed at 9 years of age. This patient was born overseas and had recently migrated to Australia. A congenital lung cyst in the right lung was known since birth. The patient was reported to be in respiratory distress at birth for 2 weeks treated with supplementary oxygen. He had a number of other anomalies including macrocephaly, convergent strabismus, deafness, agenesis of the corpus callosum and developmental delay. This patient was placed in the symptomatic group based on his initial presentation at birth from CCAM.

Eight (38%) patients developed post-operative complications in the asymptomatic group (Table 2). There was no mortality but three patients returned to theatre. One patient developed immediate post-operative bleeding and returned to theatre on the same day where a bronchial arterial bleed was identified and controlled. Another patient had persistent air leak for 2 weeks. This patient returned to theatre and their bronchopleural fistula was controlled. One patient had residual CCAM. This patient re-presented with spontaneous pneumothorax 2 months post marsupialization of the right lower lobe lung 'cyst' and returned to theatre for a right lower lobe lobectomy. The median length of total hospital stay in the asymptomatic group was 9 days (range 3–32 days). Ten (71%)

Table 3 Surgical procedure and morbidity

	Asymptomatic[a]	Symptomatic
Type of procedure		
Lobectomy	17	12
Segmental resection	1	–
Marsupialisation	1	1
Complications[b]		
Pneumothorax	2	3
Sepsis	2	1
Residual CCAM	1	–
Bleeding	1	–
Bronchopleural fistula	1	3
Pneumonia	1	4
Wound infection	–	2
Empyema	–	1
Transfusion	3	5
Death	–	2
Re-operation	3	3

[a] Two patients missing surgery-related data

[b] Some patients will have more than one concurrent complication

patients developed peri-operative complications in the symptomatic group (Table 3). There were two mortalities. One patient died prior to any surgery due to overwhelming sepsis and respiratory failure. One patient underwent emergency left lower lobe lobectomy but died 21 h later due to respiratory failure from severe pulmonary hypoplasia and persistent pulmonary hypertension. Three patients in this group also returned to theatre. Two patients had persistent air leaks and returned to theatre where their bronchopleural fistula was repaired. One patient represented 12 days post-operatively with pneumonia and empyema. This patient returned to theatre for a decortication of their loculated effusion. The median length of total hospital stay in the symptomatic group was 21 days (range 1–173 days).

Discussion

With the increasing use and sensitivity of antenatal US, more congenital cystic lesions are now detected antenatally. This creates a new spectrum of patients who are asymptomatic at birth with an antenatally suspected CCAM lesion. In our study, 60% of patients were asymptomatic at birth. Published series have shown similar figures of between 55 and 74% of CCAM patients who were asymptomatic at the time of birth [8, 9, 12–14, 18–20]. This group of patients creates a treatment dilemma in the absence of a definitive histological diagnosis as well as the uncertain natural history of CCAM.

Antenatal ultrasound scan may detect a wide spectrum of congenital cystic lung lesions, which include CCAM, bronchopulmonary sequestration, bronchogenic cyst and congenital lobar emphysema. The sensitivity for prenatal detection of CCAM using US has been reported to be 81% [10]. The positive predictive value appears to be only 57% [10, 21]; however, as some congenital lung lesions undergo apparent prenatal resolution.

Whether these lesions truly disappear remains questionable. Studies have shown that lesions not found after delivery using conventional plain CXR were demonstrated on CT [8, 9, 14, 17]. In our study, one patient with an antenatal diagnosis of CCAM had CXR reported as normal at birth and at 3 months but the lesion was subsequently confirmed on CT 3 years later. It would seem appropriate that infants with antenatally suspected CCAM require postnatal investigation irrespective of signs of antenatal resolution [14, 18]. Currently, CT scan has been recommended as the first-line postnatal investigation for asymptomatic babies diagnosed antenatally with suspected CCAM [13]. Recently, one study has shown that CT reports correlated with pathological diagnosis of CCAM with a 100% concordance rate [22].

Studies have suggested a non-operative approach to asymptomatic patients, but with no consensus on the frequency, type and length of clinical or radiological review [8, 16]. This recommendation has been based on an assessment of the relative risks of elective operative morbidity in infancy versus the potential risk of developing complications of CCAM. Aziz et al. [16] noted in their series that only approximately 10% of asymptomatic CCAM patient developed infection, but the follow-up period was only 3 years. Similarly, the follow-up study period by Van Leeuwen et al. [8] was only 4 years, which may not reflect the true natural history of initially asymptomatic CCAMs.

In our study, nearly all of asymptomatic CCAM patients at birth developed symptoms as they progressed into their later pre-school years or even during adolescence. Our findings suggest that with increasing duration of follow-up the majority of patients will develop symptoms. One study reports variability in the age of late onset clinical symptoms attributable to CCAM between 6 months and 21 years [23]. Others also report treating complications of CCAM such as recurrent pneumonias, haemoptysis and upper gastrointestinal bleeding in adults [24, 25]. We also found 48% of our patients presented with symptoms in their first year of life, with some having earlier symptoms such as chronic cough, frequent chest infections and fever that in retrospect may have been related to their CCAM.

Early surgical excision has been recommended by a number of authors [4, 6, 7, 15, 26, 27]. Lobectomy remains the procedure of choice as noted from our study and others [14, 15] to avoid residual disease and subsequent recurrence. Some studies have suggested elective resection for antenatally suspected CCAM at 3–6 months [13] or 12–18 months [28] to prevent complications of CCAM. Several studies has shown that surgery can be performed safely at 1–3 months of age on patients with asymptomatic CCAM and other congenital lung lesions with no mortality and minimal morbidity [12, 14, 15]. More recently, a minimally invasive approach has been advocated, with initial results encouraging [28].

Given the reported low complication rates for elective surgery for asymptomatic CCAM and the capacity for compensation in infants [12, 28] the benefit of early resection would seem appropriate. The median age at surgery was 2 years of age in our study. This may reflect the heterogeneity in management of this group of asymptomatic CCAM patients with delayed referral to a surgical service. Eight of our patients underwent multiple hospital admissions from complications of CCAM prior to their surgical referral and definitive surgery.

The complication rate in this study was higher than anticipated and may reflect referral of more complex cases to our centre. Six of the eight asymptomatic patients and 10

of 13 symptomatic patients developed complications. This may reflect an increased risk when operating on symptomatic patients or those patients, although asymptomatic at their birth, who have subsequently developed CCAM-related complications [15, 29]. A recent study has shown that patients who presented postnatally with symptoms of pneumonia and underwent subsequent surgery had a significantly longer duration of surgery and greater intraoperative blood loss [30]. Perhaps, surgery might be more safely performed before these complications develop.

A further concern has been the risk of developing lung malignancies such as rhabdomyosarcoma or bronchogenic carcinoma from untreated CCAM. Currently, about 40 cases of malignancies thought to have arisen from CCAM have been reported in the literature. These include pulmonary blastoma, rhabdomyosarcoma, bronchogenic carcinoma and mesenchymoma [31–34]. The risk, whilst small, should be taken into consideration when managing asymptomatic CCAM patients.

Although our data would suggest that infants with a suspected CCAM would benefit from early surgical intervention, several limitations of this retrospective review need to be considered. Not all mothers with an antenatal US suggestive of a CCAM would have agreed to review by a paediatric surgeon and/or have elected to continue with their pregnancy. Further, some parents would have either been referred or elected for care at one of the two other paediatric tertiary centres in New South Wales or perhaps even interstate. Clearly, only a prospective, multi-centre study would be likely to capture all of these patients and thus facilitate a complete picture of this abnormality.

This study suggests patients who present with asymptomatic CCAM will subsequently become symptomatic. Although many will present in their first year of life, others will present with their first symptoms much later in childhood. Post-natal imaging with a CT scan appears valuable in confirming a prenatal US diagnosis. We believe early surgical referral, including antenatal counselling, with appropriate and timely operative intervention may be beneficial to avoid the development of subsequent CCAM-related complications.

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