ORIGINAL ARTICLE

Management and outcome of antenatally diagnosed congenital cystic adenomatoid malformation of the lung

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Objective To review the management and outcome of babies with antenatally diagnosed congenital cystic adenomatoid malformation.

Design Retrospective cohort review.

Setting Tertiary neonatal care unit at Queen Mary Hospital and antenatal diagnostic centre at Tsan Yuk Hospital.

Patients Consecutive patients with antenatally suspected congenital cystic adenomatoid malformation in their concepti among antenatal patients attending Tsan Yuk Hospital from 1994 to 2002. Twenty-four of 33 cases were referred to Queen Mary Hospital for postnatal management and for whom comprehensive records were available for analysis in 23.

Interventions Postnatal interventions in their babies included investigational imaging for congenital cystic adenomatoid malformation and surgery.

Main outcome measures Antenatal and postnatal outcome, as well as pathology of the excised lesions.

Results Antenatal outcome: termination of pregnancy in two cases and spontaneous abortion in one; in-utero regression was documented in nine cases and in one hydropic change was apparent. Postnatal outcome: only eight of 20 babies born alive had symptoms in neonatal period. Two developed serious infective complications in infancy, one with documented in-utero regression. Pulmonary parenchymal abnormalities were detected on computed tomography of the thorax in six of seven cases with normal or non-specific chest radiograph findings. Among nine cases with in-utero regression, congenital cystic adenomatoid malformation was confirmed by operative histology in five and abnormal computed tomography findings in three. Fifteen babies underwent surgical excision, one of whom died because of severe pre-existing pulmonary hypoplasia and nine endured minor postoperative complications. A favourable outcome was documented at a mean follow-up of 22 months (range, 2 months-7 years).

Conclusions In-utero regression of congenital cystic adenomatoid malformation on antenatal ultrasound may not represent genuine resolution. Computed tomographic thorax should be considered in all newborns with antenatally diagnosed congenital cystic adenomatoid malformation, and if confirmed early operation before first hospital discharge is recommended.

Key words
Cystic adenomatoid malformation of lung, congenital; Prenatal diagnosis; Treatment outcome

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Introduction

Congenital cystic adenomatoid malformation (CCAM) is one of the major congenital pulmonary anomalies.[1-4] With the advent of antenatal ultrasonography (USG), it was increasingly diagnosed as was its in-utero regression.[5-7] Antenatal USG provides important information about the natural course of the condition and is pivotal in antenatal counselling and peri-natal management. However, the accuracy of findings from antenatal USG and postnatal chest radiographs (CXRs) have been questioned.[8-12] We review the management and outcome of a cohort of cases with

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目的 檢討對產前已被診斷為肺部先天囊腫性腺瘤樣畸形的嬰兒的治療和結果。

設計 隊列回顧性檢討。

安排 瑪麗醫院的第三級初生嬰兒護理部和贊育醫院的產前診斷中心。

患者 1994至2002年間往贊育醫院求診，孕體懷疑有先天囊腫性腺瘤樣畸形的孕婦。33名病人中有24人被轉介到瑪麗醫院作產後治療，是次研究獲得當中23人的詳細病歷作分析。

治療 對受影響的嬰兒作產後治療，包括掃描檢查是否患有先天囊腫性腺瘤樣畸形和進行手術。

主要結果測量 產前、產後結果，以及切除組織的病理結果。

結果 產前結果：兩人終止懷孕，一人自然流產；9人有子宮內退化的紀錄，一人有明顯水泡病變。產後結果：20個出生後存活的嬰兒中，只有8個在初生時有病徵。兩個在嬰兒期出現嚴重感染併發症，一個有子宮內退化的紀錄。7個接受X光檢查而結果正常或無特別發現的嬰兒中，有6個以電腦X光斷層成像檢查胸膛時發現有不正常肺部擴散。子宮內退化的9個嬰兒中，以手術作組織檢查證實了其中5個有先天囊腫性腺瘤樣畸形，以電腦X光斷層成像檢查則證實了3個。15個嬰兒接受切除手術，其中1人因肺部原本已嚴重發育不全而死亡，另外9人出現輕微的手術後併發症。經過平均22個月的跟進期（介乎2個月至7年）後，結果令人滿意。

結論 即使產前超音波檢查顯示有先天囊腫性腺瘤樣畸形和子宮內退化，這診斷並非一定正確。對於產前診斷到先天囊腫性腺瘤樣畸形的新生嬰兒，應考慮在其出生後作胸膛電腦X光斷層成像檢查，若果確診患上此症，應在首次出院前進行手術治療。

antenatally diagnosed CCAM to verify the true incidence of in-utero regression, ascertain the best modality of postnatal imaging and the optimal timing of surgery.

Methods

The records of 33 consecutive mothers referred to the antenatal diagnostic centre at Tsan Yuk Hospital between 1994 and 2002 with concepti suspected to have CCAM were retrieved. Twenty-four were referred to Queen Mary Hospital for subsequent management, 23 of whom had comprehensive records available for analysis; data pertaining to the nine remaining patients managed in other hospitals were not available. Antenatal USG was performed by experienced obstetricians using Acuson Sequoia or Acuson XP 128 (Mountain View, CA, US). Congenital cystic adenomatoid malformation was classified according to Adzick's classification, based on sonographic features.[13] Antenatal clinical information retrieved included the timing of the first and subsequent USG, the sonographic appearance of the lesions, presence or absence of polyhydramnios, mediastinal shift and hydrops and all antenatal interventions. Clinical data of babies born alive (sex, birth weight, gestational age, mode of delivery, Apgar scores, clinical presentation at birth) as well as postnatal imaging results including CXR and/or computed tomography (CT) of the thorax, and the type of surgery and age at that time were recorded. Pathology of the excised lesions, cases involving termination of pregnancy (TOP), spontaneous abortion, or neonatal death were also recorded if available. Histological types of the lesions were classified according to Stocker's classification.[14] Surviving cases were followed up at Queen Mary Hospital for a mean period of 22 months (range, 2 months-7 years). Growth parameters and respiratory symptoms were documented. The association between categorical variables was assessed by the Chi squared test. Ethical approval was not required as this was a retrospective descriptive review of clinical findings and all patients' identities were kept confidential.

Results

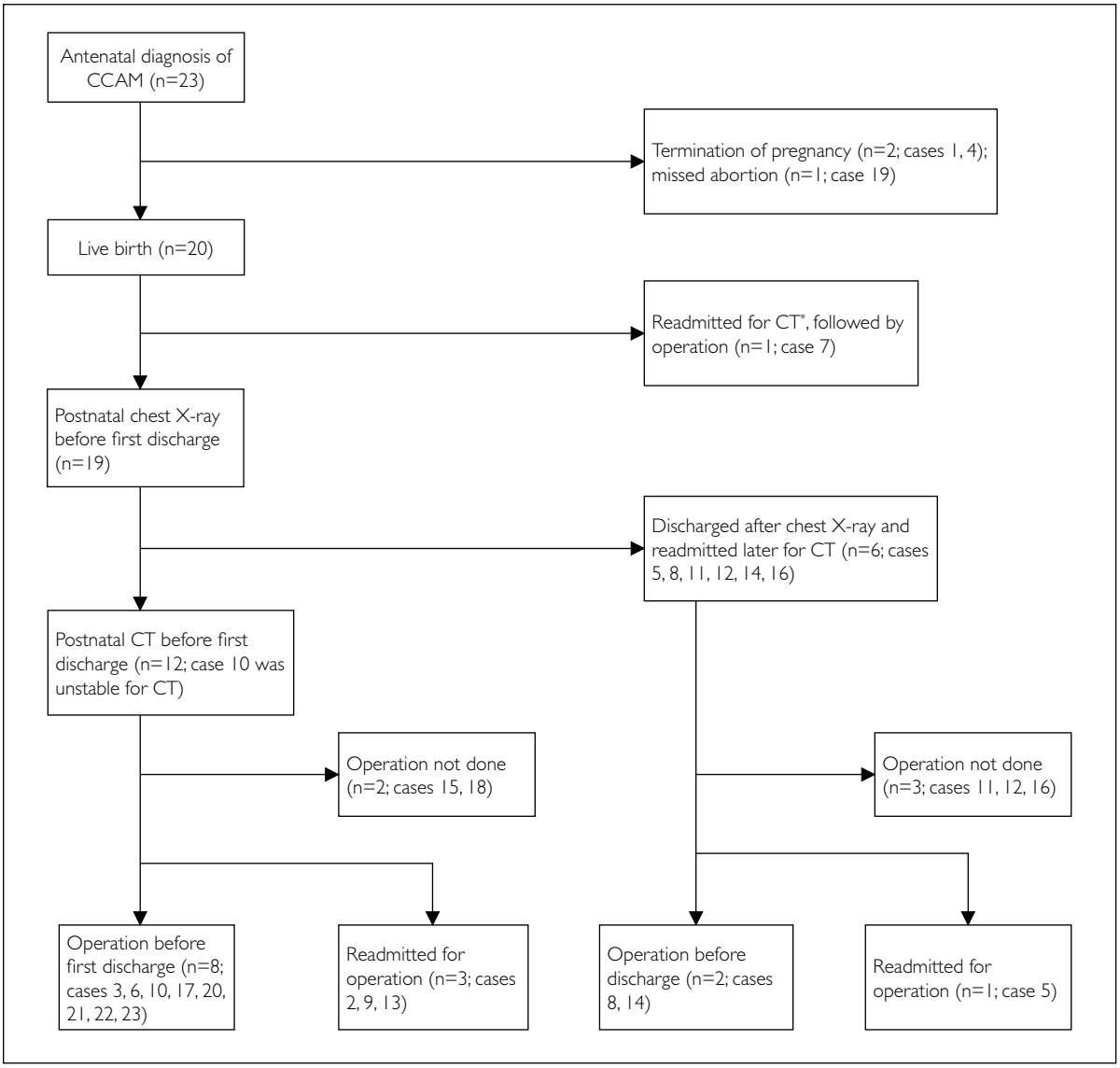
Twenty-three cases with complete data were included in the analysis (Fig).

Antenatal ultrasound and progress

The diagnosis of CCAM was made at a mean of 21 weeks of gestation (range, 15-33 weeks); nine (39%) of 23 were detected during routine scans for anomalies. Lesions were noted in the left lung in 12 (52%) cases and in the right lung in 11 cases. Macrocystic lesions were noted in seven (30%) of the cases, all in the right lung. In contrast, all six (26%) cases with microcystic lesions were found in the left lung, five of which involved the lower lobe. The remaining 10 cases were classified as mixed type. Two mothers had TOP and one a spontaneous abortion. In the remaining 20 mothers, in-utero regression of the lesions was documented in nine (45%) cases; at 27 to 33 weeks of gestation, six appeared to have complete regression and three partial regression. One (case 11) progressed with mediastinal shift and hydropic changes. Repeated amnio-reduction was performed and a cysto-amniotic shunt was inserted at 28 weeks of gestation. The baby was delivered prematurely at 33 weeks of gestation. In-utero regression was noted in seven of 10 cases affecting the left lung and two of eight with lesions in the right lung (P=0.07) but there was no association between the types of lesions and in-utero regression (P=0.2) [Table 1].

Postnatal imaging results

Postnatal CXRs were available in 19 of the 20 live births. Findings were normal or non-specific in seven babies.



* CT denotes computed tomography of the thorax

FIG. Management flowchart of antenatally diagnosed congenital cystic adenomatoid malformation (CCAM)

Abnormal findings included cystic lesions in eight babies (five in the right and three in the left lung), ill-defined opacities in three, and hyperlucency in one. Mediastinal shift was present in five babies. One baby (case 7) did not have a postnatal CXR as complete in-utero regression was documented on antenatal USG and he was asymptomatic at birth. Computed tomography of the thorax was performed in all except one baby (case 10) who underwent emergency surgery. Twelve cases (cases 2, 3, 6, 9, 13, 15, 17, 18, 20, 21, 22, 23) had CT thorax performed before their first hospital discharge. All revealed abnormalities in lung parenchyma, including multicystic lesions in nine babies, focal emphysematous change in one and hyperdense lesions in another. Chest X-ray at birth was normal or non-specific in three of these 12 babies. Computed tomography of the thorax in case 7 presenting as severe pneumonia at 5 months old, showed multicystic lesions with fluid levels suggestive of abscess formation and pleural effusion. In case 14, who had transient neonatal tachypnoea, his CXR at birth showed cystic lesions in the right side, but was discharged after a few days when his respiratory distress resolved. While waiting for an elective CT thorax at home, he developed a serious chest infection and cardiopulmonary arrest on day 12 of life. Computed tomographic thorax revealed multicystic lesions with fluid levels compatible with abscess formation secondary to CCAM. Cases 5, 8, 11, 12, and 16 were asymptomatic at birth. Chest X-ray of case 8 showed cystic lesions in the right lung with no mass effect while the CXRs of the others were unremarkable. Computed tomographic thorax performed subsequently (after discharge) revealed multicystic lesions in two (cases 5, 8), a small subpleural wedge density in the right lower lobe in one (case 12), focal emphysema in

TABLE 1. Antenatal ultrasonography findings

Case No.	Year of birth	Gestational age at diagnosis (weeks)	CCAM location*	CCAM appearance	Mediastinal shift	CCAM progress
1	1994	18†	L	Mixed	Yes	Termination of pregnancy
2	1995	26	L	Mixed	Resolved	Regressed, no shift
3	1995	33	R	Macrocystic	Yes	Static
4	1995	19†	R	Macrocystic	Yes	Termination of pregnancy
5	1996	23	R	Mixed	No	Static
6	1996	16	LWL	Microcysitc (echogenic)	Yes	Static
7	1997	19†	R	Mixed	Resolved	Regressed, no shift at 30 weeks
8	1998	18	R	Mixed	Yes	Static
9	1998	18	R	Macrocystic	Yes	Static
10	1998	24	R	Macrocystic	Yes	Hydropic change and polyhydramnios. Repeated amnio-reduction, cyst-amniotic shunt at 28 weeks
11	1998	32	L	Mixed	No	Regressed
12	1999	15	R	Macrocystic	No	Regressed at 27 weeks
13	1999	22	R	Macrocystic	Yes	Static
14	2000	32	R	Macrocystic	Yes	Static
15	2000	20†	LLL	Microcystic (echogenic)	No	Regressed at 32 weeks
16	2000	21†	LLL	Microcystic (echogenic)	Resolved	Regressed, no shift at 28 weeks
17	2001	18†	LLL	Microcystic (echogenic)	Resolved	Regressed, no shift at 33 weeks
18	2001	20	LLL	Microcystic (echogenic)	Yes	Static
19	2001	18	L	Mixed	Yes	Missed abortion
20	2002	20†	L	Mixed	Resolved	Regressed, no shift at 33 weeks
21	2002	23†	L	Mixed	No	Static
22	2002	22†	LLL	Microcystic (echogenic)	Yes	Regressed
23	2002	18	R	Mixed	Yes	Static

* CCAM denotes congenital cystic adenomatoid malformation, L left lung, R right lung, LWL left whole lung, and LLL left lower lobe
† Abnormality detected during routine anomaly scan

left lower lobe in another (case 16), and a high-seating left diaphragm in the remaining baby (case 11). Real-time ultrasound later confirmed the diagnosis of a left diaphragmatic hump (Table 2).

Postnatal symptoms, clinical course and management

All except three babies were born at term (range 33 to 40 weeks of gestation). Twelve were male and eight were female. Eight babies (cases 3, 6, 10, 13, 14, 20, 21, 22) developed respiratory symptoms at birth but there was no particular association with the type of antenatally diagnosed lesions (P=0.27). The baby with hydropic changes (case 10) required active resuscitation at birth and emergency lobectomy. The postnatal course of case 14 was described above. His lobectomy was performed after CT thorax demonstrated multicystic lesions with abscess formation. The remaining six symptomatic cases underwent surgery before discharge when CT thorax suggested CCAM. Twelve babies were asymptomatic at birth. The diagnosis of CCAM was made at the age of 5 months in case 7, when he presented as a chest infection. Lobectomy was performed at 1 month later. Six asymptomatic babies (cases 2, 5, 7, 8, 9, 17) underwent lobectomy at the age of 22 days to 20 months. Conservative management was adopted in the remaining five (cases 11, 12, 15, 16, 18) in whom CT findings were not suggestive of CCAM. Pathology of the resected specimens showed CCAM in all 15 cases that had an operation; eight (53%) cases had type I and seven had type II. Congenital cystic adenomatoid malformation type I appeared to be more prevalent in right-sided lesions (6/8 cases), while on the left side 4/7 had type II, but this difference did not reach statistical significance (P=0.31). Postoperative complications were noted in nine babies, which included pneumothoraces (6 cases), segmental collapse of the lung (2 cases), chylothorax (1 case) and right phrenic nerve palsy (1 case). All recovered with conservative management. One patent (case 10) died postoperatively due to severe pre-existing pulmonary hypoplasia (Tables 3, 4).

TABLE 2. Postnatal imaging and pathology of congenital cystic adenomatoid malformation (CCAM)

Case No.	Chest X-ray at birth			Computed tomography findings			CCAM pathology	
	Appearance	Location	Shift	Appearance	Location*	Shift	Overall	Location*
2	Cystic	Left	Yes	Multicystic	LLL	Yes	I	LLL
3	Cystic	Right	Yes	Multicystic	RLL	Yes	I	RLL
5	Alveolar shadow	Right	No	Multicystic	RLL	No	II	RLL
6	Cystic	Left	Yes	Multicystic	LUL, LLL	No	II	LUL
7	N/A†	N/A	No	Multicystic with abscess	RLL	Yes	I	RLL
8	Cystic	Right	No	Multicystic	RLL	Yes	II	RLL
9	Ill-defined shadow	-	No	Multicystic	RUL	No	I	RUL
10	Cystic	Right	Yes	N/A	N/A	N/A	I	RUL
11	Non-specific	-	No	Left diaphragmatic hump	N/A	No	N/A	N/A
12	Normal	-	No	Wedge density	RLL	No	N/A	N/A
13	Cystic	Right	Yes	Multicystic	RLL, ML	Yes	I	RLL
14	Cystic	Right	No	Multicystic with abscess	RUL, ML	Yes	I	RUL
15	Hyperlucency	Left	No	Focal emphysema	LLL	No	N/A	N/A
16	Normal	-	No	Focal emphysema	LLL	No	N/A	N/A
17	Cystic	Left	No	Multicystic	LLL	No	II	LLL
18	Normal	-	No	Focal emphysema	LLL	No	N/A	N/A
20	Opacity	Left	No	Multicystic	LLL	No	II	LLL
21	Opacity	Left	No	Tissue density	LLL	No	I	LLL
22	Normal	-	No	Multicystic	LLL	No	II	LLL
23	Opacity	Right	No	Multiple microcystic	RUL, RML	No	II	RUL, RML

* LLL denotes left lower lobe, RLL right lower lobe, LUL left upper lobe, RUL right upper lobe, and ML middle lobe
† N/A denotes not applicable or not available

TABLE 3. Postnatal management and outcome of cases with symptoms at birth

Case No.	Sex	Symptoms at birth*	Operation			Follow-up	
			Type	Age	Postoperative complications	Period	Outcome
3	F	Mild RD at birth	Lobectomy	Day 3	Pneumothorax	7 years	Well
6	M	Mild RD, NCPAP at hour 6	Lobectomy	Day 5	-	2 years	Bell's palsy at year 2
10	F	Respiratory failure, hydrops	Lobectomy	Day 1	Pneumothorax, haemothorax	N/A†	Died
13	M	RD, fever at day 2	Lobectomy	Day 23	Right upper lobe collapse	2 months	Default at month 2
14	M	TTNB resolved at day 2, pneumonia and arrest at day 12	Lobectomy	Day 15	No	3 years	Mild hypotonia
20	M	Pneumonia	Lobectomy	Day 15	Left upper lobe segmental collapse	12 months	Well
21	M	RD, pneumothorax, IMV	Segmental resection	Day 35	Pneumothorax	11 months	Residual cyst
22	F	RD, sepsis, IMV at hour 10	Lobectomy	Day 25	No	10 months	Well

* RD denotes respiratory distress, NCPAP nasal continuous positive airway pressure, TTNB transient tachypnoea of newborn, and IMV intermittent mandatory ventilation
† N/A denotes not applicable

Longer-term outcome

The 19 surviving babies were followed up for 2 months to 7 years (mean, 22 months). One (case 21) showed mild growth retardation. Two (cases 5 and 9) have

TABLE 4. Postnatal management and outcome of cases with no symptom at birth

Case No.	Sex	Subsequent symptoms or complications	Operation			Follow-up	
			Type	Age	Postoperative complications	Period	Outcome
2	F	Well	Lobectomy	Month 20	No	21 months	Well
5	M	Well	Lobectomy	Month 8	No	5.5 years	Mild infrequent episodic asthma
7	M	Right lung abscess at month 5	Lobectomy	Month 6	Pleural effusion, pneumothorax	-	Well
8	M	Well	Lobectomy	Day 29	Pneumothorax, chylothorax	6 months	Well
9	M	Well	Lobectomy	Day 40	Pneumothorax	4.5 years	Infrequent episodic asthma
11	M	Well	No	N/A*	N/A	3 months	Well, closed at month 3
12	F	Well	No	N/A	N/A	2.8 years	Well
15	M	Well	No	N/A	N/A	2.4 years	Well
16	F	Well	No	N/A	N/A	2.3 years	Well
17	F	Heart failure due to patent ductus arteriosus	Lobectomy, patent ductus arteriosus, ligation	Day 22	No	2 years	Slow growth
18	M	Well	No	N/A	N/A	17 months	Well
23	F	Well	Segmental resection	Day 32	Right phrenic nerve palsy	20 months	Well

* N/A denotes not applicable

mild infrequent episodic asthma. One (case 23) had a suspected residual cyst noted on a follow-up CT scan, but clinically the patient was asymptomatic. The baby who was resuscitated from cardiopulmonary arrest on the 12th day of life was found to have mild hypotonia, but otherwise showed normal development. One baby (case 6) suffered a Bell's palsy at 2 years of age (Tables 3, 4).

Discussion

Congenital cystic adenomatoid malformation was first described by Ch'in and Tang in 1949.[2] It is thought to be due to overgrowth of terminal bronchioles with suppression of alveolar development ensuing between 5 and 8 weeks of gestation,[1] with an incidence of one in 25 000 to 35 000 pregnancies.[15,16] With advances in antenatal USG, CCAM has been increasingly diagnosed. During the study period, the antenatal diagnostic centre was the only tertiary referral centre for prenatal diagnosis and management of various congenital malformations in Hong Kong and there were 33 referrals. The incidence during this period was estimated to be 1:31 000 live births in 1996 to 1:16 000 live births in 2002.

The gestational age at diagnosis was similar to that reported in other series. Left-sided lesions and lower lobe involvement were more common in previous studies[2,14] but the two sides were equally involved in the babies born alive in our cohort. In our study, lesions were more often found in the lower lobe on either side (15 of 20) cases. Involvement of more than one lobe was found in four cases. There was a slight male predominance in our cohort, which was similar to a previous report.[14] Eight (40%) of our patients presented with a variable degree of respiratory distress at birth. Symptoms at birth were reported in around 25 to 44% of other reports.[17-19] Hydropic change was associated with more severe symptoms and poor prognosis.[2,15,16,20-25] Earlier reports showed a higher incidence of up to 40%,[2,14,22] but the figure was lower (2-10%) in recent literature.[6,15,16,20,26] This complication was rarely seen in our cohort. Late complications of CCAM include severe or recurrent infection of the lungs and malignant change in the lesion. Two of our babies presented with infective complications, though they were relatively asymptomatic at birth. Overall postnatal mortality was reported to be 4 to 37%,[15-17,20-24,27] and in our series of live births only one of 20 died. There have been many studies on the outcome of antenatally diagnosed CCAM[11,12,15,16,19-27] and various USG features have been associated with a poor prognosis, including the type and size of CCAM, presence of mediastinal shift and hydropic change.[2,6,18,20,22-24,27,28] Identification of these factors calls for antenatal counselling, possible foetal intervention, and preparation of perinatal management at tertiary care facilities. In-utero regression was reported to occur in 6.3 to 55% of cases (Table 5). This was documented in nine (45%) of 20 babies born alive in our series and was apparently associated with left-sided

TABLE 5. Comparison with previous studies on antenatally diagnosed congenital cystic adenomatoid malformation (CCAM)[2,4,11,12,15-24,26,27,30]

Studies	No. of cases	Gestational age at diagnosis	Ultrasonography types of CCAM	Antenatal outcome*	Postnatal imaging†	Postnatal outcome‡
Present study, 2003	23	15-33 weeks	Macrocystic=7, microcystic=6, mixed=10	TOP=2, abortion=1, hydrops=1, regression=9, antenatal shunt=1	7/19 CXR normal, 1/19 no pulmonary lesions on CT	S/m=8/20, death=1/20 (5%), OT=15/20 (type I=8, type II=6)
Sauvat et al,[30] 2003	29 (asymptomatic at birth)	18-33 weeks	N/A	TOP=0, regression=12 (41%), hydrops=0	12 (41%) CXR normal, 4 (14%) CT normal	OT=17, postnatal regression=6 (21%), no death
Duncombe et al,[15] 2002	21	19-22 weeks	Macrocystic=16, microcystic=5	TOP=4, hydrops=2 (9.5%), regression=4 (19%)	Concurred with prenatal diagnosis	Death=1/17 (6%), OT=12/17
Laberge et al,[16] 2001	48	N/A	N/A	TOP=7, SA=1, hydrops=1 (2%), regression=22 (46%)	N/A	Death=4/40 (10%)
Monni et al,[26] 2000	26	19-34 weeks	Macrocystic=13, microcystic=13	TOP=9, hydrops=2 (8%), regression=3 (12%)	Lesions confirmed clinically/CXR in 11	Death=0/17, decreased size of lesion=2/17, OT=9/17
Bunduki et al,[20] 2000	18	18-36 weeks	Macrocystic=11, microcystic=7	TOP=0, IUD=2, hydrops=2 (11%), regression=3 (17%)	N/A	Death=4/16 (25%), OT=13/16
van Leeuwen et al,[12] 1999	16	16-28 weeks	Stocker type I=3, II=9, III=1, N/A=3	TOP=0, IUD=0, hydrops=0, regression=7 (44%)	9/15 CXR negative, CT showed lesions in those asymptomatic and negative CXR	Death=0/16, OT=8/16, type I=4/16, type II=2/16, pulmonary sequestration=1/16, bronchogenic cyst=1/16
Waszak et al,[17] 1999	20 (with antenatal ultrasonography among 21 cases)	21-35 weeks	Macrocystic=11, microcystic=5, N/A=4	TOP=0, hydrops=N/S, regression=N/S	3/21 CXR normal, CT confirmed diagnosis in 15	S/m=5/20, death=0/20, OT=18/20 (type I=7, II=10, III=1)
Bagolan et al,[18] 1999	9	20-36 weeks	Macrocystic=3, microcystic=6	TOP=0, regression=5 (56%)	5 regression: 4/5 CXR normal, 3/3 CT normal; 4 non-regression: all CXR and CT positive	S/m=4/9, death=0/9, OT=5/9, well=9/9
Golaszewski et al,[27] 1998	14	18-23 weeks	Stocker type I=6, II=5, III=3	TOP=4, regression=4 (29%), hydrops=0	N/S	S/m, death=1/10 (10%), OT=5/10
Sapin et al,[19] 1997	18	17-36 weeks	N/A	Abortion=1, regression=5 (28%), static=8, not known=4, hydrops=0	All CXR abnormal, CT N/S	S/m=6/17, OT=14/17, death=0/17, postnatal regression=3/17
Cacciari et al,[4] 1997	16	21-26 weeks	N/A	TOP=3 (type II), hydrops=1 (6%), regression=1 (6%)	N/A	OT=9, type I=10, type II=3, NND=1/13 (8%)
Miller et al,[11] 1996	17	20-34 weeks	Stocker type I=12, II=2, III=3	IUD=2, TOP=3, hydrops=4 (24%), regression=7 (41%)	All had radiological evidence of CCAM	OT=12/12
Bromley et al,[21] 1995	25	16-37 weeks	Stocker type I=10, II=5, III=10	IUD=5, hydrops=1 (4%), regression=8 (32%)	N/A	NND=2/16 (13%), S/m=11/16, OT=10/16
Thorpe-Beeston and Nicolaides,[2] 1994	58	17-39 weeks	Macrocystic=27, microcystic=31	TOP=22/58, IUD=3/58, regression=3 (5%), hydrops=17 (29%), antenatal shunt=5	N/S	NND=3/33 (9%), OT=13/33
Adzick and Harrison,[22] 1993	22	20-36 weeks	Multicystic=12, echogenic=10	TOP=3, hydrops=9 (41%), regression=4 (18%)	N/A	Death=7/19 (37%), OT=12/19
Dumez et al,[23] 1993	18	19-25 weeks	Stocker type I=5, II=9, III=4	TOP=4, hydrops=3 (17%), regression=3 (17%)	N/A	Death=1/14 (7%), OT=7/14
Revillon et al,[24] 1993	32	16-38 weeks	Stocker type I=12, II=15, III=5	TOP=5, IUD=1, hydrops=6 (19%), regression=13 (41%)	N/A	Death=1/26 (4%), OT=18/26

* TOP denotes termination of pregnancy, SA spontaneous abortion, and IUD intrauterine death
† CXR denotes chest X-ray, CT computed tomography, N/A not available, and N/S not specified
‡ S/m denotes symptomatic after birth, OT operation with resection of the lesion, and NND neonatal death

lesions (7/10 cases), microcystic lesions (4/6 cases), and disappearance of mediastinal shift (5 cases). Lack of histological confirmation remains one of the challenges to establishing the incidence and mechanisms of in-utero regression. More importantly, in-utero regression noted on antenatal USG may not represent 'genuine' regression.[8,9] In our series showing such in-utero regression, five were diagnosed to have CCAM after birth. In the remaining

four cases, focal emphysema was noted in the CT scan in two, one showed a small focal opacity at left lower lobe (possibly a remnant of a regressed lesion) and one revealed a left diaphragmatic hump. However, we could only confidently confirm genuine in-utero regression in the baby with the diaphragmatic hump (5%). Thus, our study added further supporting evidence that in-utero regression apparent on USG may not be genuine and that appropriate postnatal evaluation is required.

One interesting antenatal USG findings in our cohort, was the predilection for the left lung, especially the left lower lobe for microcystic lesions (Table 1). To our knowledge, such an association has not been reported before. In the five babies with lesions detected in the left lower lobe (cases 15, 16, 17, 18, 22), four showed varying degrees of 'in-utero regression'. After birth, focal emphysema was identified in three, while two had type II CCAM. This apparently favourable prognosis associated with microcystic lesions of left lower lobe warrants larger studies for verification.

Although CXR was the most commonly used imaging modality, its accuracy was questionable. van Leeuwen et al[12] reported a high false negative rate in their cohort consisting of 15 cases. Kim et al[28] demonstrated that CT appearance of the lesions correlated well with pathological findings. Others also supported the use of CT scan thorax.[29,30] In our study, seven of 20 babies had normal or non-specific findings in the CXR at birth. Subsequent CT thorax demonstrated pulmonary parenchymal abnormalities in six, three of whom had histologically confirmed CCAM. Postnatal CT scan of thorax should therefore be considered in all the babies with antenatally diagnosed pulmonary lesions, irrespective of in-utero regression on antenatal ultrasound, normal postnatal CXR findings, or absence of symptoms at birth.

There has been controversy regarding the postnatal management of CCAM. Bagolan et al[18] proposed that babies with large lesions or those symptomatic at birth should have early evaluation and surgery, while for babies with small asymptomatic lesions, surgical resection could be performed at 6 to 18 months of age.[18,27] Sauvat et al[30] suggested early surgery within the first month of life if the CCAM is voluminous (>3 cm), or liquid-filled on the first CT scan and a conservative approach for those asymptomatic small lesions. Others claimed that early surgery in asymptomatic cases was associated with low morbidity and mortality and suggested that early operation was warranted.[17,31] In our series, significant postoperative morbidity and mortality was low and the long-term outcome was favourable. On the other hand, one of our babies (case 7) with in-utero regression diagnosed by antenatal USG presented with a serious infective complication at 5 months old and another developed severe pneumonia and cardiopulmonary arrest at home on day 12 of life while waiting for CT thorax. Hence, we advocate and have adopted a policy of early (pre hospital discharge) CT scan of the thorax for all the babies with antenatally diagnosed CCAM and early operation once the diagnosis is confirmed.

Conclusion

Our study showed that genuine in-utero regression of CCAM is uncommon and that CXR at birth is unreliable. We advocate early, in-hospital CT thorax for all newborn babies with antenatally diagnosed CCAM, irrespective of subsequent antenatal USG or postnatal CXR findings and clinical presentation at birth. All confirmed cases should undergo early operation before discharge, in view of the significant risk of serious infective complications, relatively low postoperative morbidity, and good long-term outcome.

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