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Prenatal diagnosis and outcome of echogenic fetal lung lesions

P. CAVORETTO*, F. MOLINA*, S. POGGI*, M. DAVENPORT† and K. H. NICOLAIDES*

*Departments of *Fetal Medicine and †Pediatric Surgery, King's College Hospital, London, UK*

KEYWORDS: congenital high airway obstruction syndrome; cystic adenomatoid malformation; echogenic lung; fetal surgery; pulmonary sequestration; ultrasound

ABSTRACT

Objective *To describe the antenatal findings and outcome of fetuses with echogenic lung lesions.*

Methods *This was a retrospective study of the prenatal sonographic features, antenatal management and outcome of 193 fetuses with an echogenic lung lesion diagnosed at 18–35 weeks of gestation. There were nine cases of congenital high airway obstruction syndrome (CHAOS), 170 cases of cystic adenomatoid malformation (CAM) and 14 cases of pulmonary sequestration (PS). A literature search was also carried out to compare our data with those of previous series.*

Results *The prognosis in our series of fetuses with CHAOS was invariably poor, but the literature describes a handful of survivors after delivery by Cesarean section and* ex-utero *intrapartum therapy (EXIT). Of the cases in our series with PS and no pleural effusions, more than 95% survived; in half of these cases the lesion resolved antenatally and in the other half sequestrectomy was carried out postnatally. In cases with PS and pleural effusions, successful treatment was provided by the placement of thoracoamniotic shunts or occlusion of the feeding blood vessel by ultrasound-guided laser coagulation or injection of sclerosants. In cases with CAM and no hydrops, there was more than 95% survival and in up to half of the cases there was sonographic evidence of spontaneous antenatal resolution of the hyperechogenic lesion, which was confirmed by postnatal imaging in about 60% of the cases. Of the cases with CAM with hydrops managed expectantly, more than 95% died before or after birth. Of the cases with macrocystic CAM with hydrops, two-thirds survived after placement of a thoracoamniotic shunt. In cases with microcystic CAM with hydrops, there is some evidence that open fetal surgery with lobectomy could improve survival but such treatment is highly invasive for the mother.*

Conclusions *CHAOS is a severe abnormality, whereas CAM and PS are associated with a good prognosis. In a high proportion of fetuses with hyperechogenic lung lesion, there is spontaneous antenatal resolution and the underlying pathology may be transient bronchial obstruction. Copyright © 2008 ISUOG. Published by John Wiley & Sons, Ltd.*

INTRODUCTION

The most common fetal hyperechogenic lung lesions are congenital cystic adenomatoid malformation (CAM), pulmonary sequestration (PS) and congenital high airway obstruction syndrome (CHAOS). Several studies have described the prenatal diagnosis and outcome of affected fetuses. Some studies are from pediatric surgical centers, consequently underreporting cases with spontaneous prenatal resolution and severe cases resulting in termination of pregnancy or intrauterine death. In most of the prenatal studies, the number of cases examined was less than 30 and such small studies cannot provide definite conclusions as to the natural history and prognosis of these conditions. There have been only eight studies examining more than 30 cases, and these reported contradictory results with respect to the prognosis, including both apparent prenatal resolution and a need for postnatal surgery of the lesions[1–8].

In this study, we describe the prenatal sonographic features, antenatal management and outcome of 193 fetuses with echogenic lung lesions examined in our unit and review the literature on these conditions.

Correspondence to: Prof. K. H. Nicolaides, Harris Birthright Research Centre for Fetal Medicine, King's College Hospital, Denmark Hill, London SE5 8RX, UK (e-mail: fmf@fetalmedicine.com)

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METHODS

This was a retrospective study of the prenatal sonographic features, antenatal management and outcome of fetuses with an echogenic lung lesion diagnosed in local hospitals and referred to our specialist fetal medicine center for further assessment and management. In the fetal medicine center, a detailed ultrasound examination was carried out and the findings were entered prospectively into a database. Subsequent management was based on these findings and included expectant management, antenatal fetal surgical intervention or termination of pregnancy at the request of the parents. For those choosing to continue the pregnancy, follow-up ultrasound examinations were carried out and planned delivery was generally undertaken in hospitals with facilities for neonatal intensive care and pediatric surgery. Postnatal management included confirmation of the antenatal findings by chest X-ray and contrast computerized tomography or magnetic resonance imaging. The need for surgery was determined by the individual surgeons in consultation with the parents.

We searched the fetal medicine center's database to identify all cases with echogenic lung lesions diagnosed between 1994 and April 2006 and obtained data on their antenatal findings, management and outcome. In cases delivering liveborn infants, we obtained data on postnatal findings and management from individual hospital records or reports from their family doctors. We also searched web-based bibliographic databases between 1985 and 2007 to identify all relevant English-language literature. We used a combination of the following keywords: 'CAM', 'cystic adenomatoid malformation', 'pulmonary sequestration', 'CHAOS', 'laryngeal atresia', 'tracheal atresia', 'fetal surgery', 'prenatal diagnosis', 'ultrasound' and 'fetus'.

RESULTS

The fetal medicine database search identified 193 fetuses with an echogenic lung lesion diagnosed at 18–35 weeks of gestation, which included nine cases of CHAOS, 170 of CAM and 14 of PS.

Congenital high airway obstruction syndrome (CHAOS)

In the nine cases of CHAOS, the diagnosis was made at 16–22 (median, 19) weeks. In all cases, there was massive bilateral enlargement with uniform hyperechogenicity of the lungs, compression of the fetal heart and inverted diaphragm, dilation of the trachea and main bronchi (Figure 1), ascites and placentomegaly. In two cases, there was bilateral renal agenesis, raising the possibility of Fraser syndrome. None of the mothers had features suggestive of mirror syndrome. In eight cases, the pregnancies were terminated at the request of the parents and in one case diagnosed at 16 weeks the parents chose to receive expectant management; this fetus died at 20 weeks. In seven of the nine cases, the parents gave permission for postmortem examination, which confirmed the presence of tracheal or laryngeal atresia.

There were 10 fetuses with CHAOS diagnosed at 16–33 weeks and managed expectantly, including one of our cases and nine identified from the literature review[9–16]. Two of the mothers developed mirror syndrome. There were two fetal deaths due to progressive hydrops and seven neonatal deaths due to respiratory insufficiency. In one case, there was spontaneous prenatal regression of the hydrops and the baby survived after delivery at 38 weeks[16]. In this case, as well as in five of the nine deaths, there was a tracheoesophageal fistula.

The literature review identified 11 fetuses with CHAOS, diagnosed at 16–26 weeks, managed expectantly and delivered vaginally or by Cesarean section, and

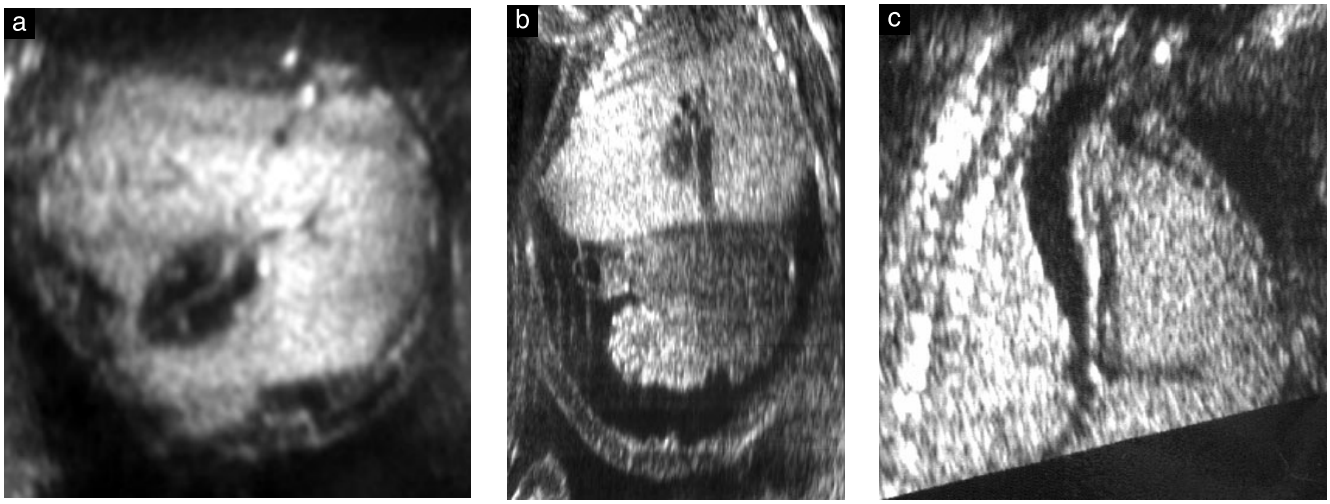

Figure 1 Laryngeal atresia in a fetus at 23 weeks' gestation showing massive bilateral enlargement and hyperechogenicity of the lungs with compression of the heart (transverse view) (a), inverted diaphragm and ascites (coronal view) (b) and dilation of the trachea and main bronchi (longitudinal view) (c).

Table 1 Outcome of 11 fetuses with congenital high airway obstruction syndrome (CHAOS) treated by EXIT procedure (*ex-utero* intrapartum therapy)

Reference	*GA at delivery (weeks)*	*Diagnosis*	*Treatment/Outcome*
Richards *et al.*[17] (1992)*	37	Laryngeal stenosis	Laryngotracheoplasty and stent in the neonatal period. Stent removal planned at 4 months. Good ventilation and laryngeal function.
De Cou *et al.*[18] (1998)	35	Laryngeal atresia	Died at 14 weeks from respiratory arrest due to a tracheostomy-related accident.
Bui *et al.*[19] (2000)	35	Laryngeal atresia	Discharged from hospital at 2 months. Laryngotracheoplasty planned at 24 months.
Lim *et al.*[14] (2003)	31	Tracheal atresia	Laryngotracheoplasty at 17 months. Normal development and speech at 5 years.
	37	Laryngeal atresia	Discharged from hospital at day 19. Laryngotracheoplasty planned at 18 months.
	32	Laryngeal atresia	Needing assisted ventilation at 6 months.
Oepkes *et al.*[20] (2003)	37	Tracheal atresia	Discharged from hospital at 7 weeks. Laryngotracheoplasty planned at 8 months.
Kanamori *et al.*[21] (2004)	39	Laryngeal atresia	Microcephaly due to 5p deletion diagnosed in the neonatal period.
Hirose *et al.*[22] (2004)	32	Tracheal atresia	Breathing with minimal ventilatory support. Awaiting laryngotracheoplasty.
Shimabukuro *et al.*[23] (2007)	36	Laryngeal atresia	Laryngotracheoplasty at 20 months. Awaiting reversal of tracheostomy. Normal physical and mental development but unable to speak.
Colnaghi *et al.*[24] (2007)	29	Laryngeal atresia	Laryngotracheoplasty performed at 22 months. Normal ventilation and speech at 33 months.

*In this case delivery was vaginal. In all other cases it was by Cesarean section. GA, gestational age.

which underwent *ex-utero* intrapartum therapy (EXIT) (Table 1)[14,17–24]. At EXIT, tracheostomy was carried out and positive pressure ventilation was initiated before clamping of the umbilical cord and complete delivery of the neonate. There were 10 survivors and one neonatal death.

The literature review identified three published reports on prenatal fetal intervention in CHAOS[25–27]. In the first case, ultrasound-guided percutaneous fetal tracheostomy was attempted at 18 weeks of gestation but the fetus died a few hours later[25]. In the second case, the fetus presented with classical signs of laryngeal atresia at 24 weeks. A transverse laparotomy was performed at 24 weeks and the uterus was exteriorized, exposing the posterior wall to avoid the anterior placenta[26]. Three 5-mm trocars were inserted into the uterine cavity, one for the fetoscope and two working ports. A transuterine stitch was placed through the fetal chin in order to extend the neck and immobilize the head. During tracheal dissection, the fetus developed severe bradycardia and hysterotomy was performed to expose the chest and apply external thoracic compression. The resuscitation failed and the fetus was delivered by EXIT procedure. The baby was discharged from hospital at 6 months on assisted ventilation with permanent tracheostomy. At 42 months, the child had no speech and received all feeds by gastrostomy. The infant required assisted ventilation at night for 3 years and at 4 years of age was mildly developmentally delayed. In the third case, general anesthesia was administered and three 5-mm trocars were inserted into the uterine cavity percutaneously at 19 weeks of gestation[27]. Under fetoscopic and ultrasonographic guidance, a wire was passed from the pharynx through the atretic region into the trachea. The atretic region was dilated subsequently using a balloon angioplasty catheter and by the placement of a 2.5/8-mm coronary stent. Successful decompression of the trachea into the pharynx became immediately apparent by a sudden decrease in tracheal diameter and within the next few days there was a decrease in the echogenicity of the lungs and the ascites subsequently resolved. At 28 weeks, after a spontaneous rupture of membranes and premature labor, the fetus was delivered by Cesarean section with EXIT procedure and tracheostomy. The baby had Fraser syndrome. However, pulmonary function was good and the baby was weaned off ventilation after 18 days and discharged from hospital after 6 months.

Pulmonary sequestration

In the 14 cases of PS, the diagnosis was made at 19–35 (median, 21) weeks. In 10 cases, the PS was on the left side and in four it was on the right. In all cases, there was a uniformly echogenic lesion, with color Doppler evidence of systemic arterial blood supply arising from the aorta (Figure 2).

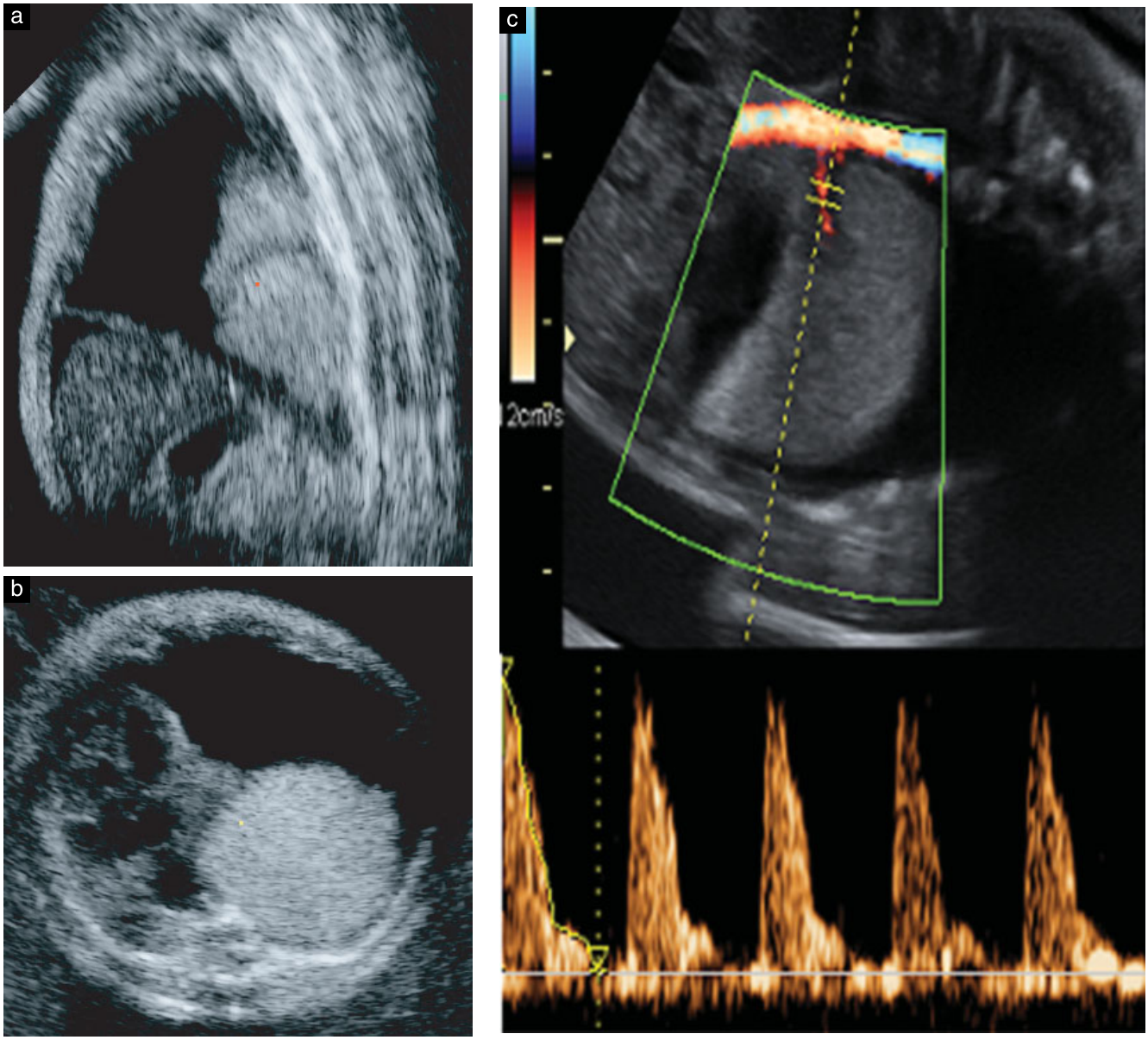

Figure 2 Longitudinal (a) and transverse (b) sections of the fetal thorax at 22 weeks demonstrating the echogenic mass of pulmonary sequestration with pleural effusion, and pulse Doppler study of the feeding artery arising from the fetal aorta (c).

Pulmonary sequestration with no pleural effusions

In six of our cases of PS, there was no mediastinal shift and the pregnancies were managed expectantly. All infants were liveborn and five had sequestrectomy because of postnatal persistence of the PS.

Table 2 summarizes the outcome of 95 fetuses, including our six cases, with PS diagnosed at 18–36 weeks and managed expectantly[3,7,28–46]. There were three neonatal deaths due to hydrops and pulmonary hypoplasia and one neonatal death due to a surgical complication. All other infants survived. In 38 (40%) of the cases, the lesion regressed antenatally, the neonates were asymptomatic and no postnatal surgery was carried out. In the other cases, the lesion persisted, the neonates were usually symptomatic and sequestrectomy was performed.

Pulmonary sequestration with pleural effusions

In eight of our cases of PS there was a large pleural effusion surrounding the PS and mediastinal shift (Table 3). In these cases, local anesthetic (1% lignocaine) was injected into the maternal abdomen down to the myometrium, and an 18-gauge needle was inserted under ultrasound and color Doppler guidance, through the maternal abdomen and into the fetal thorax and then the PS. A Nd : YAG laser fiber (Dornier, Munich, Germany) 400-μm in diameter was then passed through and to 5 mm beyond the tip of the needle and the feeding vessel was coagulated using an output of 30–50 Watts for 5–10 s. Color Doppler demonstrated immediate cessation of blood flow within the tumor. Follow-up ultrasound examinations demonstrated that the effusions resolved and the PS decreased in size, with complete resolution of the PS

Table 2 Outcome of 95 fetuses with thoracic pulmonary sequestration managed expectantly. In total, 91 of the 95 cases survived.

Reference	n	*GA (weeks)*		*Survival* (n)	*Sequest-rectomy* (n)
		Diagnosis	*Delivery*		
Meizner *et al.*[28](1990)	1	23	39	1	1
Langer *et al.*[29] (1995)	2	25–26	37–38	2	0‡
Abuhamad *et al.*[30] (1996)	2	18	39–40	2	0
da Silva *et al.*[31] (1996)	3	25–34	30–35	3	3
Evans[32] (1996)	3	25–34	30–36	3	3
Adzick *et al.*[3] (1998)	37	18–36	—	36*	7
Becmeur *et al.*[33] (1998)	9	20–33	37–40	9	9
Bratu *et al.*[34] (2001)	13	24	—	11*	11§
Wax *et al.*[35] (2002)	1	29	39	1	1
Chen *et al.*[36] (2003)	2	19–20	38–40	2	1
Cuillier[37] (2003)	1	23	> 32	1	1
Jeanty *et al.*[38] (2004)	1	30	—	1	1
Illanes *et al.*[7] (2005)	4	19–29	—	4	2
Ruano *et al.*[39] (2005)	3	21–33	—	2†	3¶
Chen *et al.*[40] (2005)	1	21	41	1	1
Chen *et al.*[41] (2006)	1	30	39	1	—
Kuo *et al.*[42] (2006)	1	28	37	1	1
York *et al.*[43] (2006)	1	21	37	1	1
Stern *et al.*[44] (2007)	1	20	41	1	1
Manson[45] (2007)	1	22	39	1	1**
Hung *et al.*[46] (2008)	1	22	38	1	—
Present series	6	19–35	37–42	6	5
Total	95	18–36	30–42	91	53

*Neonatal death due to hydrops and pulmonary hypoplasia. †Neonatal death after thoracotomy (surgical complication). ‡One lost to follow-up and the other did not require surgery. §In one of the 11 cases there was percutaneous embolization rather than sequestrectomy. ¶One operative thoracoscopy, one open chest sequestrectomy and one percutaneous arterial embolization. **Preoperative arterial embolization. GA, gestational age.

in three. Sequestrectomy was carried out in the five cases with postnatal persistence of the lesion.

The literature review identified several case reports suggesting that PS with pleural effusions with expectant antenatal management is associated with a poor neonatal outcome due to pulmonary hypoplasia[47–51].

The literature review identified 31 fetuses, in addition to our eight cases, with PS that were treated prenatally because of associated pleural effusions (Table 3)[3,33,52–70]. In one case, open fetal surgery, involving laparotomy, hysterotomy and fetal left lower lobectomy, was carried out at 22 weeks[68]; the baby was delivered by Cesarean section at 35 weeks after spontaneous rupture of the membranes and was reported as doing well. In two cases, percutaneous ultrasound-guided laser coagulation of the feeding artery was performed at 23 and 29 weeks and the fetuses were delivered at 39 and 38 weeks, respectively. Both had normal ventilation and were in excellent condition at delivery and one of them did not require postnatal surgical treatment[69,70]. In four cases, ultrasound guidance was used to inject a sclerosant into the feeding vessel at the hilus of the tumor[66,67]. This resulted in immediate cessation of blood flow and subsequent ultrasound examinations demonstrated prenatal resolution of the tumor. All four infants survived and two did not require postnatal sequestrectomy.

In 24 of the 31 cases, treatment was aimed essentially at drainage of the effusions rather than surgery of the tumor. In one case the effusions were drained by thoracentesis and intraperitoneal injection of digoxin and furosemide. The effusions reaccumulated and the procedure was repeated daily between 28 and 32 weeks. The infant was delivered in good condition at 32 weeks after spontaneous labor and was awaiting sequestrectomy within the first year of postnatal life[54]. In 18 cases there was placement of thoracoamniotic shunts with consequent resolution of the effusions, but in two of these the effusions subsequently reaccumulated. In five cases, treatment was by thoracentesis but in all cases there was subsequent reaccumulation of the effusions. In six fetuses undergoing prenatal shunt with resolution of the effusions, a previous thoracentesis had been performed with rapid reaccumulation of hydrothorax. In total, 21/23 survived and two died in the neonatal period due to pulmonary hypoplasia and/or pulmonary hypertension.

Postnatal surgery was carried out in 26 (72.2%) of the 36 cases with available data.

Cystic adenomatoid malformation of the lung

In our 170 cases of CAM, the diagnosis was made at 18–34 (median, 21) weeks. The lesion was left-sided in 88 (51.8%) cases and right-sided in 82 (48.2%), and it was microcystic in 90 (52.9%) cases, macrocystic in 38 (22.4%) and mixed in 42 (24.7%) (Figure 3). Nine fetuses had hydrops and 161 did not. Other major

Table 3 Outcome of 39 fetuses with thoracic pulmonary sequestration treated prenatally

	Fetal therapy				
Reference	*Technique*	*GA at procedure (weeks)*	*Effusion*	*GA at delivery (weeks)*	*Postnatal surgery*
Hernanz-Schulman *et al.*[52] (1991)	Thoracentesis	—	Reaccumulated	31	Sequestrectomy
Jones *et al.*[53] (1992)	Thoracentesis	24	Reaccumulated	29	None; NND†
Adzick *et al.*[3] (1998)	Thoracentesis	27	Reaccumulated	33–35	Sequestrectomy
Anandakumar *et al.*[54] (1999)	Thoracenteses + digoxin, furosemide	28	Reaccumulated	32	Surgery planned
Morville *et al.*[55] (2003)	Thoracentesis	27	Reaccumulated	32	Arterial embolization
Pumberger *et al.*[56] (2003)	Thoracenteses	—	Reaccumulated	22–27	Sequestrectomy
Kitano *et al.*[57] (2006)	Thoracenteses* + thoracoamniotic shunt	28–31	Resolved	35	Sequestrectomy
Kitano *et al.*[57] (2006)	Thoracenteses* + thoracoamniotic shunt	27–28	Resolved	33	Sequestrectomy
Kitano *et al.*[57] (2006)	Thoracenteses* + thoracoamniotic shunt	30	Resolved	35	Sequestrectomy
Hayashi *et al.*[58] (2006)	Thoracenteses* + thoracoamniotic shunt	30	Resolved	35	Sequestrectomy
Hayashi *et al.*[58] (2006)	Thoracenteses* + thoracoamniotic shunt	28	Resolved	33	Sequestrectomy
Hayashi *et al.*[58] (2006)	Thoracenteses* + thoracoamniotic shunt	30	Resolved	35	Sequestrectomy
Weiner *et al.*[59] (1986)	Thoracoamniotic shunt	24	Reaccumulated	29	Sequestrectomy; NND†
Slotnick *et al.*[60] (1990)	Thoracoamniotic shunt	32	Resolved	34	Sequestrectomy
Hernanz-Schulman *et al.*[52] (1991)	Thoracoamniotic shunt	27	Resolved	—	Sequestrectomy
Favre *et al.*[61] (1994)	Thoracoamniotic shunt	30	Resolved	38	Sequestrectomy
Adzick *et al.*[3] (1998)	Thoracoamniotic shunt	29	Resolved	33–35	Sequestrectomy
Adzick *et al.*[3] (1998)	Thoracoamniotic shunt	30	Resolved	33–35	Sequestrectomy
Becmeur *et al.*[33] (1998)	Thoracoamniotic shunt	30	Resolved	38	Sequestrectomy
Lopoo *et al.*[62] (1999)	Thoracoamniotic shunt	23	Resolved	33	—
Lopoo *et al.*[62] (1999)	Thoracoamniotic shunt	30	Resolved	33	—
Salomon *et al.*[63] (2003)	Thoracoamniotic shunt	34	Resolved	36	None
Picone *et al.*[64] (2004)	Thoracoamniotic shunt	19–36	—	28–40	—
Odaka *et al.*[65] (2006)	Thoracoamniotic shunt	28	Reaccumulated	37	Sequestrectomy
Nicolini *et al.*[66] (2000)	Alcohol injection + thoracoamniotic shunt	27	Resolved	40	None
Bermudez *et al.*[67] (2007)	Polidocanol injection	26	Resolved	38	Sequestrectomy
Bermudez *et al.*[67] (2007)	Polidocanol injection	26	Resolved	38	None
Bermudez *et al.*[67] (2007)	Polidocanol injection	24	Resolved	38	Sequestrectomy
Cass *et al.*[68] (1997)	Fetal lobectomy	22	Resolved	35	None
Oepkes *et al.*[69] (2007)	Laser coagulation	23	Resolved	39	None
Ruano *et al.*[70] (2007)	Laser coagulation	29	Resolved	38	Sequestrectomy
Present series	Laser coagulation	31	Resolved	38	Sequestrectomy
Present series	Laser coagulation	30	Resolved	38	Sequestrectomy
Present series	Laser coagulation	32	Resolved	34	None
Present series	Laser coagulation	27	Resolved	41	None
Present series	Laser coagulation	24	Resolved	40	None
Present series	Laser coagulation	31	Resolved	34	Sequestrectomy
Present series	Laser coagulation	23	Resolved	35	Sequestrectomy
Present series	Laser coagulation	28	Resolved	39	Sequestrectomy
Total ($n = 39$)	Effusion drainage, $n = 24$; laser coagulation, $n = 10$; sclerosant, $n = 4$; fetal lobectomy, $n = 1$	Mean, 27.8	Resolution, 30/37 (81.1%)	Mean, 35.1	Alive, 37/39 (94.9%) Surgery, 26/36 (72.2%)

In 37 of the 39 cases, the infant survived. *Thoracenteses followed by rapid reaccumulation of hydrothorax. †Neonatal death (NND) due to pulmonary hypoplasia. GA, gestational age.

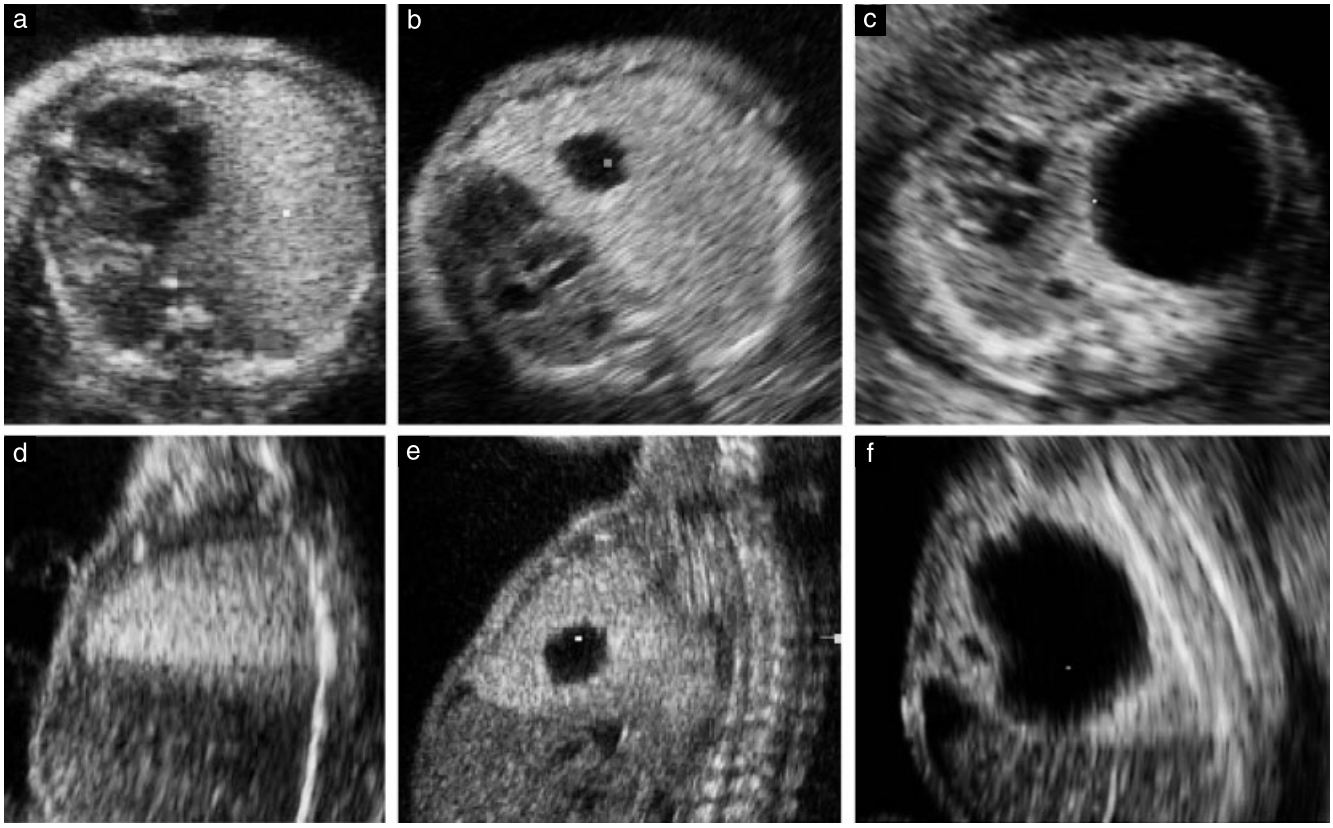

Figure 3 Transverse (a–c) and longitudinal (d–f) sections of the fetal thorax at 20–22 weeks' gestation demonstrating cystic adenomatoid malformation of the lungs of the microcystic (a, d), mixed (b, e) and macrocystic (c, f) types.

defects were observed in four cases: one case each of bilateral multicystic renal dysplasia, esophageal atresia, coarctation of the aorta and sacrococcygeal teratoma.

Cystic adenomatoid malformation with no hydrops

In the non-hydropic group ($n = 161$) there were 154 infants that were delivered at 29–42 (median, 39) weeks and survived, two terminations of pregnancy at the request of the parents, three unexplained fetal deaths at 34–37 weeks, one fetal death due to placental abruption at 39 weeks and one neonatal death in a case known to have bilateral multicystic kidneys but the parents had chosen to continue with the pregnancy. In two of the four fetal deaths there was spontaneous resolution of the CAM (Table 4).

In 76 (49.4%) of the 154 cases that survived, there was sonographic evidence of antenatal resolution of the CAM by 28–37 (median, 32) weeks. Of these 76 cases, postnatal chest X-ray showed no lesion in 54 (71.1%) and this was confirmed by contrast computerized tomography or magnetic resonance imaging carried out in 34 of the 54 cases. None of these 54 cases required surgery. In 22 cases with apparent antenatal resolution of the CAM, there was postnatal evidence of a persisting lesion (chest X-ray positive in 17 and negative in five; computerized tomography positive in 18, negative in two and not done in two) and 16 (72.7%) of these cases had surgery. Therefore, postnatal computerized tomography or magnetic resonance imaging was carried out in 54 of the cases with antenatal sonographic evidence of resolution of the lesion and this was negative in 34 (62.9%) and positive in 20 (37.1%) of the 54 cases.

In the 78 cases with evidence of prenatal persistence of the CAM, the lesion was detected postnatally in 75 (96.2%) cases and in 55 (73.3%) of these surgery was performed. There were three cases with prenatal but not postnatal persistence of the CAM and none of these had surgery. The chest X-ray was positive in 71 and negative in seven cases and computerized tomography was positive in 71, negative in two and not done in five cases.

The literature review identified 486 non-hydropic fetuses with CAM diagnosed prenatally in pregnancies which the parents chose to continue (Table 4)[1–4,6–8,39,56,71–84]. Of the total of 645 fetuses (including our 159 cases), there were 18 (2.8%) intrauterine or neonatal deaths and 627 (97.2%) survived. Data on postnatal imaging was not available in all reports and it was therefore not possible to substantiate the true extent of prenatal resolution of the lesion. However, of the survivors with data available, apparent prenatal resolution of the CAM was observed in 29.6% and postnatal surgery was carried out in 62.7%.

Thoracoamniotic shunting. In six of our 161 non-hydropic fetuses, there was a large cyst causing a

Table 4 Outcome of 645 non-hydropic fetuses with cystic adenomatoid malformation in which the parents chose to continue with the pregnancy

		GA (weeks)				
Reference	n	*Diagnosis*	*Delivery*	*Survival* (n (%))	*Apparent prenatal resolution* (n (%))	*Postnatal surgery* (n (%))
Neilson *et al.*[71] (1991)	6	18–36	37–41	6 (100)	1/6 (16.7)	5/6 (83.3)
Thorpe-Beeston & Nicolaides[1] (1994)	38	19–39	26–41	34 (89.5)	3/34 (8.8)	19/34 (55.9)
Barret *et al.*[72] (1995)	8	17–32	40	8 (100)	3/8 (37.5)	2/8 (25.0)
Bromley *et al.*[73] (1995)	17	17–37	27–40	16 (94.1)	0/16 (0)	9/16 (56.3)
Miller *et al.*[74] (1996)	12	20–34	31–41	12 (100)	—	12/12 (100)
Dommergues *et al.*[2] (1997)	20	20–27	31–41	18 (90.0)	2/18 (11.1)	12/18 (66.7)
Adzick *et al.*[3] (1998)	79	17–38	—	79 (100)	0/79 (0)	79/79 (100)
Van Leeuwen *et al.*[75] (1999)	16	18–28	—	16 (100)	6/16 (38)	8/16 (50)
Lacy *et al.*[76] (1999)	16	18–22	—	16 (100)	9/16 (56.2)	3/16 (18.7)
Bunduki *et al.*[77] (2000)	11	18–36	36–40	11 (100)	0/11 (0)	11/11 (100)
De Santis *et al.*[78] (2000)	13	19–37	26–41	13 (100)	3/13 (23.1)	4/13 (30.8)
Monni *et al.*[79] (2000)	17	21–34	33–40	17 (100)	3/17 (17.6)	9/17 (52.9)
Laberge *et al.*[4] (2001)	37	16–40	—	36 (97.3)	0/36 (0)	—
Crombleholme *et al.*[8] (2002)	39	—	27–40	38 (97.4)	—	—
Duncombe *et al.*[80] (2002)	15	19–22	36–40	15 (100)	0/15 (0)	12/15 (80.0)
Pumberger *et al.*[56] (2003)	23	16–35	—	22 (95.6)	11/22 (50)	14/22 (63.6)
Hsieh *et al.*[81] (2005)	8	28–39	28–39	7 (87.5)	0/7 (0)	0/7 (0)
Ruano *et al.*[39] (2005)	4	21–28	—	4 (100)	—	4/4 (100)
Ierullo *et al.*[6] (2005)	28	>18	35–40	27 (96.4)	15/27 (55.6)	18/27 (66.7)
Illanes *et al.*[7] (2005)	32	19–29	—	32 (100)	21 of 32 (65.6)	19/32 (59.4)
Calvert *et al.*[82] (2006)	21	—	36–42	21 (100)	4/21 (19.0)	16/21 (76.2)
Kunisaki *et al.*[83] (2007)	6	17–21	31–40	6 (100)	3/6 (50.0)	6/6 (100)
Chow *et al.*[84] (2007)	20	16–32	33–40	19 (95)	9/19 (47.4)	14/19 (73.7)
Present series*	159	18–34	29–42	154 (96.9)	76/154 (49.4)	71/154 (46.1)
Total	645	16–40	26–42	627 (97.2)	169/573 (29.5)	347/553 (62.7)

*Davenport *et al.*[5] (2004) reported 57 cases that are included in the present series. GA, gestational age.

major mediastinal shift and a thoracoamniotic shunt was inserted at 22–27 weeks (Table 5); all infants survived after delivery at 36–39 weeks. In five infants, lobectomy was performed and one was being managed expectantly with persistence but diminution of the lesion by the age of 3 years.

The literature review identified another 18 fetuses with a large cyst causing major mediastinal shift that were treated by placement of a thoracoamniotic shunt (Table 5)[1–3,6,74,85–90]. In five of these fetuses, thoracentesis was first performed, with subsequent rapid reaccumulation of fluid within the cyst. All infants were liveborn but three died in the neonatal period due to pulmonary hypoplasia.

In one of our fetuses with major mediastinal shift but microcystic disease, ultrasound-guided laser coagulation of the major vessels within the substance of the tumor was performed at 24 weeks. Follow-up ultrasound examinations demonstrated diminution of the tumor with return of the mediastinum to its normal position. The infant was delivered at 38 weeks and was asymptomatic at birth, but underwent lobectomy at 14 months because of persistence of the tumor.

Congenital cystic adenomatoid malformation with hydrops

In the hydropic group ($n = 9$), the lesion was macrocystic in four cases, microcystic in three and mixed in two. In the macrocystic group, one pregnancy was terminated at the request of the parents and the other three fetuses were treated by placement of a thoracoamniotic shunt; these infants survived (Table 6). The two cases with a mixed lesion were also treated by placement of a thoracoamniotic shunt; one fetus died *in utero* and the other infant survived. In the microcystic group, two cases were managed expectantly and the infants died in the neonatal period. In the third fetus with microcystic disease, ultrasound-guided laser coagulation of the major vessels within the substance of the tumor was performed at 19 weeks. Follow-up scans demonstrated decreases in the size of the tumor and in the hydrops by 23 weeks. However, the occlusion of the vascular supply to the tumor may have been incomplete, because there was subsequent expansion of the mass and recurrence of major mediastinal shift. Ultrasound-guided laser coagulation was repeated at 31 weeks. The infant was delivered at 37 weeks and died in the neonatal period due to pulmonary hypoplasia.

The literature review identified a few papers reporting on one to four hydropic fetuses with CAM, the majority of which died either prenatally or in the neonatal period. In two papers from the same research group on a total of 45 hydropic fetuses with CAM that were managed expectantly, all but one died either before or after delivery at 25–36 weeks[3,91].

Attempts at fetal therapy in hydropic fetuses with CAM are summarized in Table 6[2–4,6–8,39,56,71,77,84–87,92–105].

Table 5 Outcome of 24 non-hydropic fetuses with cystic adenomatoid malformation and a large cyst treated by placement of a thoracoamniotic shunt

	GA (weeks)		
Reference	*Thoracic shunt*	*Delivery*	*Outcome*
Thorpe-Beeston & Nicolaides[1] (1994)*	24	40	Alive, lobectomy
Thorpe-Beeston & Nicolaides[1] (1994)*	25	39	Alive, lobectomy
Thorpe-Beeston & Nicolaides[1] (1994)*	26	38	Alive, lobectomy
Thorpe-Beeston & Nicolaides[1] (1994)*	31	37	Alive, lobectomy
Thorpe-Beeston & Nicolaides[1] (1994)*	32	33	Alive, lobectomy
Bernaschek *et al.*[85] (1994)	24	36	Alive, lobectomy
Bernaschek *et al.*[85] (1994)†	35	40	Alive, lobectomy
Miller *et al.*[74] (1996)*	25	—	Alive, lobectomy
Dommergues *et al.*[2] (1997)*	27	36	Alive, lobectomy
Dommergues *et al.*[2] (1997)*	30	31	Neonatal death§
Dommergues *et al.*[2] (1997)*	23	36	Neonatal death§
Adzick *et al.*[3] (1998)*†	25	36	Alive, surgery?
Adzick *et al.*[3] (1998)*†	28	38	Alive, surgery?
Adzick *et al.*[3] (1998)*†	30	32	Alive, surgery?
Morikawa *et al.*[86] (2003)†	29	37	Alive, lobectomy
Wilson *et al.*[87] (2004)‡	24–29	27–30	Neonatal death§
Ierullo *et al.*[6] (2005)*	27–30	40	Alive, surgery?
Viggiano *et al.*[90] (2006)†	28	38	Alive, lobectomy
Present series*	22	36	Alive, lobectomy
Present series*	24	39	Alive, lobectomy
Present series*	24	37	Alive, lobectomy
Present series*	25	39	Alive, no surgery
Present series*	25	38	Alive, lobectomy
Present series*	27	37	Alive, lobectomy
Total ($n = 24$)	Mean, 26.8	Mean, 36.6	Alive, 21/24 (87.5%) Dead, 3/24 (12.5%)

*Included in Table 4. †Thoracentesis first performed with subsequent rapid reaccumulation of fluid within the cyst. ‡Case obtained by integrating information from four different papers of the same research group[3,87–89]. §Neonatal death due to pulmonary hypoplasia. GA, gestational age.

Table 6 Outcome of 85 hydropic fetuses with cystic adenomatoid malformation treated prenatally

				GA (weeks)		
Reference	*Lesion*	*Fetal therapy*	*Indication*	*Therapy*	*Delivery*	*Outcome*
Chao & Monoson[92] (1990)	Macrocystic	Thoracenteses ×3	A, M, E	27	35	Neonatal death*
Neilson *et al.*[71] (1991)	Macrocystic	Thoracentesis ×1	A, P	30	34	Neonatal death*
Brown *et al.*[93] (1995)	Macrocystic	Thoracenteses ×6	M, P	28	34	Alive, lobectomy
Sugiyama *et al.*[94] (1999)	Macrocystic	Thoracentesis ×1	M, P	29	29	Neonatal death†
Crombleholme *et al.*[8] (2002)	Macrocystic	Thoracentesis ×1	—	—	—	Alive, surgery?
Crombleholme *et al.*[8] (2002)	Macrocystic	Thoracentesis ×1	—	—	—	Alive, surgery?
Crombleholme *et al.*[8] (2002)	Macrocystic	Thoracenteses (several)	TPTL	—	—	Alive, surgery?
Crombleholme *et al.*[8] (2002)	Macrocystic	Thoracenteses (several)	TPTL	—	—	Alive, surgery?
Pumberger *et al.*[56] (2003)	Macrocystic	Thoracenteses ×3	A, M, P	—	—	Alive, lobectomy
Bunduki *et al.*[77] (2000)	Macrocystic	Thoracentesis ×1	—	25	38	Neonatal death‡
Clark *et al.*[95] (1987)	Macrocystic	Thoracoamniotic shunt	FT, A, E	20	37	Alive, lobectomy
Bernaschek *et al.*[85] (1994)	Macrocystic	Thoracoamniotic shunt	FT	22	33	Neonatal death*
Bernaschek *et al.*[85] (1994)	Macrocystic	Thoracoamniotic shunt	FT	29	39	Alive, lobectomy
Dommergues *et al.*[2] (1997)	Macrocystic	Thoracoamniotic shunt	A, PE	26	36	Alive, lobectomy
Dommergues *et al.*[2] (1997)	Macrocystic	Thoracoamniotic shunt	A, E, H	26	37	Alive, lobectomy
Dommergues *et al.*[2] (1997)	Macrocystic	Thoracoamniotic shunt	A, PE	20	35	Alive, lobectomy
Dommergues *et al.*[2] (1997)	Macrocystic	Thoracoamniotic shunt	A, E	25	34	Neonatal death*
Dommergues *et al.*[2] (1997)	Macrocystic	Thoracoamniotic shunt	A, E	18	39	Neonatal death*
Ryo *et al.*[96] (1997)	Macrocystic	Thoracoamniotic shunt	A, M, P	27	37	Alive, lobectomy
Adzick *et al.*[3] (1998)	Macrocystic	Thoracoamniotic shunt	FT, M	24	34	Alive, surgery?
Adzick *et al.*[3] (1998)	Macrocystic	Thoracoamniotic shunt	FT, M	30	34	Alive, surgery?
Adzick *et al.*[3] (1998)	Macrocystic	Thoracoamniotic shunt	FT, M	22	22	Fetal death§
Golaszewski *et al.*[97] (1998)	Macrocystic	Thoracoamniotic shunt	A, E, FT	25	36	Alive, lobectomy
Sugiyama *et al.*[94] (1999)	Macrocystic	Thoracoamniotic shunt	A, M, P	27	37	Alive, lobectomy
Bunduki *et al.*[77] (2000)	Macrocystic	Thoracoamniotic shunt	A, M	22	33	Alive, lobectomy
Laberge *et al.*[4] (2001)	Macrocystic	Thoracoamniotic shunt	A, H, M, P	26	36	Neonatal death*
Crombleholme *et al.*[8] (2002)	Macrocystic	Thoracoamniotic shunt	FT	—	27	Alive, surgery?
Crombleholme *et al.*[8] (2002)	Macrocystic	Thoracoamniotic shunt	FT	—	29–38	Alive, surgery?

Table 6 (*Continued*)

Reference	Lesion	Fetal therapy	Indication	GA (weeks)		Outcome
				Therapy	Delivery	
Crombleholme *et al.*[8] (2002)	Macrocystic	Thoracoamniotic shunt	FT	—	29–38	Alive, surgery?
Crombleholme *et al.*[8] (2002)	Macrocystic	Thoracoamniotic shunt	FT	—	29–38	Alive, surgery?
Crombleholme *et al.*[8] (2002)	Macrocystic	Thoracoamniotic shunt	FT	—	29–38	Alive, surgery?
Crombleholme *et al.*[8] (2002)	Macrocystic	Thoracoamniotic shunt	FT	—	29–38	Alive, surgery?
Adzick *et al.*[98] (2003)	Macrocystic	Thoracoamniotic shunt	—	—	—	Alive, surgery?
Adzick *et al.*[98] (2003)	Macrocystic	Thoracoamniotic shunt	—	—	—	Alive, surgery?
Adzick *et al.*[98] (2003)	Macrocystic	Thoracoamniotic shunt	—	—	—	Death¶
Gornall *et al.*[99] (2003)	Macrocystic	Thoracentesis	—	20	38	Alive, lobectomy
Gornall *et al.*[99] (2003)	Macrocystic	Thoracentesis	—	22	37	Alive, lobectomy
Gornall *et al.*[99] (2003)	Macrocystic	Thoracoamniotic shunt	—	—	40	Alive, lobectomy
Morikawa *et al.*[86] (2003)	Macrocystic	Thoracoamniotic shunt	FT	21	40	Alive, lobectomy
Wilson *et al.*[87] (2004)§	Macrocystic	Thoracoamniotic shunt	A, E, P	—	—	Alive, surgery?
Wilson *et al.*[87] (2004)§	Macrocystic	Thoracoamniotic shunt	A, E, P	—	—	Alive, surgery?
Wilson *et al.*[87] (2004)§	Macrocystic	Thoracoamniotic shunt	A, E, P	24–29	27–30	Neonatal death**
Illanes *et al.*[7] (2005)	Macrocystic	Thoracoamniotic shunt	FT	26	26	Fetal death††
Illanes *et al.*[7] (2005)	Macrocystic	Thoracoamniotic shunt	—	27	30	Neonatal death*
Asabe *et al.*[100] (2005)	Macrocystic	Thoracoamniotic shunt	M, P	29	37	Neonatal death††
Ierullo *et al.*[6] (2005)	Macrocystic	Thoracoamniotic shunt	M	27	40	Alive, lobectomy
Ruano *et al.*[39] (2005)	Microcystic	Thoracoamniotic shunt	H	22	23	Fetal death‡‡
Isnard *et al.*[101] (2007)	Macrocystic	Thoracoamniotic shunt	A, M, P	26	37	Alive, lobectomy
Chow *et al.*[84] (2007)	Macrocystic	Thoracoamniotic shunt	M, P	28	33	Neonatal death†
Vu *et al.*[102] (2007)	Macrocystic	Thoracoamniotic shunt	—	25	34	Neonatal death†
Present series	Macrocystic	Thoracoamniotic shunt	A, E, M, P	21	38	Alive, lobectomy
Present series	Macrocystic	Thoracoamniotic shunt	A, E, M	24	41	Alive, lobectomy
Present series	Macrocystic	Thoracoamniotic shunt	A, E, M, P	26	38	Alive, lobectomy
Present series	Mixed	Thoracoamniotic shunt	A, E, H, M, P	26	38	Alive, lobectomy
Present series	Mixed	Thoracoamniotic shunt	A, E, M, P	28	35	Fetal death‡‡
Grethel *et al.*[91] (2007)	—	EXIT	—	36	36	Alive, lobectomy
Grethel *et al.*[91] (2007)	—	EXIT	—	28	28	Neonatal death**
Grethel *et al.*[91] (2007)	—	EXIT	—	30	30	Neonatal death**
Adzick *et al.*[3] (1998)	Macrocystic	Fetal lobectomy	FS	26	34	Alive
Adzick *et al.*[3] (1998)	Microcystic/mixed	Fetal lobectomy	ST	21	21	Fetal death††
Adzick *et al.*[3] (1998)	Microcystic/mixed	Fetal lobectomy	ST	25	25	Fetal death††
Adzick *et al.*[3] (1998)	Microcystic/mixed	Fetal lobectomy	ST	24	24	Fetal death††
Adzick *et al.*[3] (1998)	Microcystic/mixed	Fetal lobectomy	ST	21	21	Fetal death††
Adzick *et al.*[3] (1998)	Microcystic/mixed	Fetal lobectomy	ST	27	28	Neonatal death*
Adzick *et al.*[3] (1998)	Microcystic/mixed	Fetal lobectomy	ST	23	30	Alive
Adzick *et al.*[3] (1998)	Microcystic/mixed	Fetal lobectomy	ST	26	33	Alive
Adzick *et al.*[3] (1998)	Microcystic/mixed	Fetal lobectomy	ST	24	26	Alive
Adzick *et al.*[3] (1998)	Microcystic/mixed	Fetal lobectomy	ST	24	30	Alive
Adzick *et al.*[3] (1998)	Microcystic/mixed	Fetal lobectomy	ST	22	35	Alive
Adzick *et al.*[3] (1998)	Microcystic/mixed	Fetal lobectomy	ST	22	35	Alive
Adzick *et al.*[3] (1998)	Microcystic/mixed	Fetal lobectomy	ST	29	37	Alive
Crombleholme *et al.*[8] (2002)	Microcystic	Fetal lobectomy	ST	—	35	Alive
Crombleholme *et al.*[8] (2002)	Microcystic	Fetal lobectomy	ST	—	36	Alive
Crombleholme *et al.*[8] (2002)	Microcystic	Fetal lobectomy	ST	—	—	Fetal death††
Crombleholme *et al.*[8] (2002)	Microcystic	Fetal lobectomy	ST	—	—	Fetal death††
Crombleholme *et al.*[8] (2002)	Microcystic	Fetal lobectomy	ST	24	24	Neonatal death**
Crombleholme *et al.*[8] (2002)	Microcystic	Fetal lobectomy	ST	—	—	Fetal death‡‡
Crombleholme *et al.*[8] (2002)	Microcystic	Fetal lobectomy	ST	—	32	Neonatal death§§
Adzick *et al.*[98] (2003)	Microcystic/mixed	Fetal lobectomy	ST	21–31	—	Alive
Adzick *et al.*[98] (2003)	Microcystic/mixed	Fetal lobectomy	ST	21	21	Fetal death††
Vu *et al.*[102] (2007)	—	Radiofrequency ablation	ST	26	26	Fetal death††
Fortunato *et al.*[103] (1997)	Microcystic	Laser coagulation	M	21 and 23	—	Alive, surgery?
Bruner *et al.*[104] (2000)	Microcystic	Laser coagulation	M, P, ST	22	24	Fetal death‡‡
Ong *et al.*[105] (2006)	Microcystic	Laser coagulation	A, M, P, ST	21	38	Alive, lobectomy
Present series	Microcystic	Laser coagulation	A, M, H, ST	19 and 31	37	Neonatal death*
Total ($n = 85$)				Mean, 25.0	Mean, 32.2	Alive, 51/85 (60.0%)

*Lung hypoplasia. †Neonatal surgery-related complication. ‡Sepsis after lobectomy. §Preterm premature rupture of membranes after procedure. ¶No information available. **Prematurity. ††Fetal surgery-related complication. ‡‡Progression of hydrops. §§Chromosomal abnormality. A, ascites; E, skin edema; EXIT, *ex-utero* intrapartum therapy; FT, failed thoracentesis (thoracentesis attempted in cases with large cysts and hydrops, with thoracoamniotic shunt placement only when reaccumulation of fluid occurred); FS, failed thoracoamniotic shunt; GA, gestational age; H, hydrothorax; M, mediastinal shift; P, polyhydramnios; PE, pericardial effusions; TPTL; threatened preterm labor reason for repeated thoracenteses rather than shunt placement; ST, predominantly solid tumor.

Essentially, there were 50 cases treated by placement of a thoracoamniotic shunt or thoracentesis; among these, there were 17 (34%) intrauterine or neonatal deaths and 33 (66%) survivors. Prenatal surgery by hysterotomy and lobectomy was carried out in 22 cases with predominantly microcystic or mixed lesions, and 11 (50%) of the infants survived. In another three cases with microcystic disease, ultrasound-guided laser coagulation was carried out: two survived and one died *in utero* due to progression of hydrops. In one case, percutaneous radiofrequency ablation was performed but the fetus died due to a procedure-related accident. In another case of microcystic CAM with associated pleural effusion, a thoracoamniotic shunt was placed at 22 weeks but the fetus died due to progression of hydrops. Three cases with late-onset hydrops were treated by EXIT delivery and one of these survived[91].

DISCUSSION

The data from our study and previous reports indicate that CHAOS is a serious abnormality, whereas CAM and PS in the absence of hydrops are associated with a good prognosis.

In CHAOS, due to laryngeal and/or tracheal atresia, the massively enlarged lungs result in cardiac and superior mediastinal compression with secondary progressive hydrops and fetal or neonatal death. In the majority of cases diagnosed antenatally, the parents choose the option of pregnancy termination. In the few cases in which therapeutic intervention was undertaken either prenatally or during delivery by EXIT, some of the infants survived. A recent study highlighted the existence of a subtype of CHAOS in which the tracheal obstruction is incomplete due to the presence of a pharyngotracheal or laryngotracheal fistula and in these cases the prognosis may be good[106]. The sonographic features at 16–22 weeks are similar to those in complete obstruction, but with advancing gestation the fetal condition improves and by 32 weeks there is regression of hyperechogenicity of the lungs, eversion of the diaphragm, ascites and polyhydramnios.

In PS, a portion of lung parenchyma is supplied directly by an aberrant branch of the aorta rather than by a branch of the pulmonary artery. In the vast majority of cases, there is no obvious connection with the tracheobronchial tree. In a few cases, there is histological evidence of a mixed CAM-PS lesion[68]. We observed such lesions in nine of our fetuses, including seven in which the prenatal diagnosis was CAM and two in which it was PS. There are also cases with concomitance of CAM and PS, suggesting that these conditions may have a common embryological origin[107]. We had two such cases in which the prenatal diagnosis was CAM, but after postnatal surgery there was histological evidence for the presence of both CAM and PS[108,109].

In PS, ultrasound examination demonstrates a uniformly echogenic lesion with or without an associated pleural effusion and with color Doppler it is possible to visualize the systemic arterial blood supply arising from the aorta. In PS with no pleural effusions, expectant management is associated with survival in all cases and in about half of fetuses the lesion regresses antenatally with no need for postnatal surgery. In PS with pleural effusions the condition may progress to hydrops and perinatal death. Effective antenatal intervention is provided either by placement of thoracoamniotic shunts and consequent resolution of the effusions or by occlusion of the feeding vessel at the hilus of the tumor by ultrasound-guided laser coagulation or injection of a sclerosant agent. In the case of drainage of the effusions, postnatal surgery is usually necessary to remove the tumor, whereas in those treated by antenatal occlusion of the feeding vessel, postnatal surgery was necessary only in half of our cases because in the other half the tumor resolved antenatally. This issue merits further investigation.

In CAM, prenatal diagnosis is based on the demonstration of a uniformly hyperechogenic mass (microcystic), echo-free cysts (macrocystic) or a multicystic tumor with echogenic stroma (mixed type), usually involving one lobe of the lungs. The macrocystic and mixed types usually persist throughout pregnancy and necessitate postnatal thoracotomy and lobectomy. Cases with a large cyst causing a major mediastinal shift can be treated successfully by placement of a thoracoamniotic shunt. A previous study found a 74% survival rate in 23 fetuses with a large cyst, including 18 with hydrops, that were treated by thoracoamniotic shunt[110].

In microcystic CAM with no hydrops, the survival rate is more than 95% without the need for antenatal intervention. In half of cases there is apparent antenatal resolution of the hyperechogenic lesion, usually at around 32 weeks of gestation. In about 60% of cases with apparent antenatal resolution no lesion can be demonstrated by postnatal imaging and it is possible that at least in some of these cases the underlying cause may be not CAM but rather a transient bronchial tree obstruction with retention of mucoid fluid distal to the obstruction[82,111–115]. In contrast, in more than 95% of cases with prenatal persistence of the hyperechogenic lesion postnatal imaging confirms the presence of CAM. Postnatal surgery is carried out in about 75% of cases in which postnatal imaging demonstrates presence of a lesion.

In CAM with hydrops managed expectantly, the infants usually die before or after birth. In macrocystic disease, two-thirds of cases survive after placement of a thoracoamniotic shunt. In those with microcystic disease, open fetal surgery with lobectomy could improve survival but such treatment has not been accepted widely because it is highly invasive for the mother. The extent to which the less invasive approach of ultrasound-guided laser coagulation of the vascular supply to the tumor could improve survival merits further investigation.

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