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CYSTIC ADENOMATOID MALFORMATION OF THE LUNG: PRENATAL DIAGNOSIS AND OUTCOME

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SUMMARY

During an 8-year period (1984–1992), we made the ultrasonographic diagnosis of cystic adenomatoid malformation (CAM) of the lung in 58 fetuses at 17–39 weeks' gestation. We reviewed the records of these fetuses and combined the data from 74 cases reported in the literature to determine the incidence of the different types of CAM, associated malformations, and outcome. The lesions were macrocystic in 78 (59 per cent) and microcystic in 54 (41 per cent) of the cases. CAM was left-sided in 51 per cent, right-sided in 35 per cent, and bilateral in 14 per cent of the fetuses. In 15 (11 per cent) of the fetuses there were additional malformations and 57 (43 per cent) were hydropic. The pregnancy was electively terminated in 44 (33 per cent) of the cases, including all those with bilateral CAM. There were six (5 per cent) intrauterine deaths, five in association with hydrops, and one with growth retardation and heart defect. Of the 82 (62 per cent) infants that were liveborn, 21 (26 per cent) died in the neonatal period, 15 before and six after surgery. Of the 61 survivors, 16 (26 per cent) did not require surgery. In the 88 cases where the pregnancy was not terminated, survival was better if the CAM was macrocystic (74 per cent versus 58 per cent for microcystic), if there was no hydrops (92 per cent versus 21 per cent for hydrops), and if the amniotic fluid volume was normal or decreased (82 per cent versus 53 per cent for polyhydramnios).

KEY WORDS—Cystic adenomatoid malformation, ultrasonography, prenatal diagnosis, lung tumour.

INTRODUCTION

Congenital cystic adenomatoid malformation (CAM) of the lungs is a rare developmental abnormality arising from an overgrowth of the terminal respiratory bronchioles. The embryological insult occurs before the seventh week of gestation (Stocker *et al.*, 1977) and was first described by Ch'in and Tang in 1949. The condition may be bilateral involving all lung tissue or it may be confined to a single lobe. Postnatally, there is a wide spectrum of presentation, from neonatal death due to pulmonary hypoplasia to mild respiratory symptoms in childhood (Miller *et al.*, 1980).

CAM can be diagnosed prenatally by ultrasonography, but published data are limited to case reports and small series. Some studies have emphasized the good prognosis of this condition, which may even regress antenatally (Saltzman *et al.*, 1988; Fine *et al.*, 1988; Sonek *et al.*, 1991; Glaves and Baker, 1983), and others reported that the prognosis is often poor and may require prenatal surgery (Kuller *et al.*, 1992).

The aim of this paper is to provide a better understanding of the condition by combining data from 58 fetuses with CAM diagnosed in our centre and 74 cases reported in the literature to determine the incidence of the different types of CAM, associated malformations, and outcome of pregnancies.

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Table I—Ultrasound findings in 44 cases of cystic adenomatoid malformation undergoing termination of pregnancy

Reference	Case No.	GA (weeks)	Type	Site	Hydrops	Mediastinal shift	AFV	Other defects	Outcome
Boulot *et al.* (1991)	1	20	Mac	R	+	+	N		TOP
Chou *et al.* (1992)	2	16	Mic	Bil	+	−	N		TOP
Catanzarite *et al.* (1992)	3	19	Mac	L	−	+	N		TOP
	4	20	Mac	L	−	+	N		TOP
Cave and Adam (1984)	5	21	Mic	L	+	+	P		TOP
Sherer *et al.* (1992)	6	17	Mic	Bil	+	−	R		TOP
Rempen *et al.* (1987)	7	23	Mic	Bil	+	−	R		TOP
	8	23	Mic	Bil	+	−	R	Exomphalos	TOP
Morcos and Lobb (1986)	9	18	Mic	Bil	+	−	N		TOP
Mendoza *et al.* (1986)	10	24	Mic	R	+	−	N		TOP
Deacon *et al.* (1990)	11	18	Mac	R	−	−	P		TOP
Morris *et al.* (1991)	12	23	Mic	R	−	−	N		TOP
	13	23	Mac	L	+	+	N		TOP
	14	21	Mac	L	−	−	N		TOP
	15	20	Mic	Bil	+	−	N	CHD	TOP
	16	26	Mac	L	+	+	P		TOP
Neilson *et al.* (1991)	17	20	Mic		−	−	R	Renal agenesis	TOP
	18	19	Mic		+	−	P	CHD	TOP
Adzick *et al.* (1985)	19	22	Mic	R	+	NA	P		TOP
	20	24	Mic	L	+	NA	P		TOP
Heydanus *et al.* (1993)	21	23	Mic	Bil	+	−	P		TOP
	22	20	Mac	L	−	+	N		TOP

KCH	23	17	Mic	Bil	+	−	N	TOP
	24	23	Mic	Bil	+	−	R	Multicystic kidneys TOP
	25	19	Mac	R	+	+	N	TOP
	26	24	Mic	L	+	+	N	TOP
	27	21	Mic	Bil	−	−	N	Exomphalos TOP
	28	19	Mic	Bil	+	−	N	TOP
	29	20	Mic	L	−	+	N	TOP
	30	27	Mac	R	−	+	R	Multicystic kidneys TOP
	31	21	Mac	R	+	+	N	TOP
	32	18	Mic	Bil	+	−	N	TOP
	33	19	Mic	L	+	+	N	Facial cleft TOP
	34	22	Mic	Bil	+	−	N	Encephalocoele TOP
	35	19	Mac	R	−	+	N	TOP
	36	16	Mic	Bil	+	−	R	TOP
	37	27	Mic	Bil	+	−	P	TOP
	38	21	Mic	Bil	+	−	R	TOP
	39	21	Mac	R	−	+	N	TOP
	40	19	Mic	Bil	+	−	N	TOP
	41	20	Mic	L	−	+	N	Trisomy 18 TOP
	42	19	Mic	Bil	+	−	R	TOP
	43	21	Mic	L	−	+	N	TOP
	44	19	Mac	L	−	+	N	TOP

GA=Gestational age; AFV=amniotic fluid volume (N=normal, P=polyhydramnios, R=reduced); Mic=microcystic; Mac=macrocystic; CHD=congenital heart disease; TOP=termination of pregnancy; Bil=bilateral; L=left; R=right; NA=information not available; KCH=King's College Hospital.

PATIENTS AND METHODS

During an 8-year period (1984–1992), CAM of the lungs was diagnosed in 58 fetuses referred to our unit for ultrasound examination. The antenatal and postnatal records of these patients were examined and the following data were extracted: (i) gestational age at diagnosis; (ii) type of CAM, which was either microcystic (uniformly hyperechogenic) or macrocystic (with translucent areas); (iii) site of the lesion, left, right, or bilateral; (iv) mediastinal shift, which was considered to be significant if in a transverse section of the fetal thorax the heart was found to be entirely in either hemisphere created by an imaginary line between the sternum and the spine; (iv) hydrops fetalis or other malformations; (v) amniotic fluid volume, which was classified as normal, reduced, or polyhydramnios (vertical pool of amniotic fluid >8 cm); and (vi) outcome, including gestation at delivery, neonatal surgery, and survival.

These data were combined by extracting the necessary information from the 74 cases in the published reports which were identified by a search of the literature between 1975 and 1992. We did not include published cases if the data provided were insufficient. One paper (Kuller *et al.*, 1992) summarized the findings and outcome in 22 cases of antenatally diagnosed CAM. However, a subgroup of these cases had previously been reported by Adzick (1991). The outcome of those cases in the study of Kuller *et al.* (1992) that underwent fetal surgery will be discussed separately.

RESULTS

The prenatal findings and outcome of the combined data are shown in Tables I–III. The mean gestational age at diagnosis was 24·5 (range 17–39) weeks. The CAM was unilateral in 86 per cent and macrocystic in 58 per cent of the 132 cases; associated hydrops fetalis was present in 43 per cent and other defects in 11 per cent (Table IV). In cases of unilateral lung disease, hydrops was not more commonly seen if the disease was left- ($n=18$) or right ($n=17$) -sided. Other defects (Table V) were more common if the lesion was bilateral rather than unilateral (28 per cent versus 10 per cent) and microcystic rather than macrocystic (18 per cent versus 8 per cent). Polyhydramnios was present in 46 (35 per cent) of the cases.

Pregnancy outcome in relation to antenatal findings is shown in Table IV. There were 44 (33 per cent) terminations of pregnancy, because the lesions were bilateral or they were associated with other abnormalities (Table V). Of the remaining 88 cases, six (7 per cent) resulted in intrauterine death, 21 (24 per cent) in neonatal death, and 61 (69 per cent) fetuses survived. Of the 21 neonatal deaths, 15 died before and six died after surgery (Table VI). Of the 61 survivors, 45 had surgery and 16 did not because the infants were asymptomatic or the lesion was resolving. Survival was better if the lesion was macrocystic rather than microcystic (74 per cent versus 58 per cent), if there was no hydrops (92 per cent versus 21 per cent for hydropic fetus), and if there was normal or decreased amniotic fluid volume rather than polyhydramnios (82 per cent versus 53 per cent) (Table IV).

In 12 cases there was an apparent antenatal improvement in the disease with a decrease in the size of the tumour, return of a deviated mediastinum to its normal position, or resolution of polyhydramnios or ascites; all these pregnancies resulted in livebirths. Postnatal investigation of the neonates included chest X-ray and computed tomography, and magnetic resonance imaging demonstrated normal lungs with no evidence of CAM in two cases and possible sequestration in one. In nine cases, there was postnatal evidence of residual lung disease, and in six of these, thoracotomy was performed to resect the affected lung.

In 17 cases with either a large unilateral pulmonary cyst causing mediastinal shift and compression of the contralateral lung or non-immune hydrops, thoracocentesis, thoraco-amniotic shunting, or fetal surgery was undertaken. Two of the three cases treated with thoracocentesis resulted in deaths due to pulmonary hypoplasia. In contrast, six of the eight cases that were treated with thoraco-amniotic shunts survived. In six cases, hysterotomy was performed and the affected fetal lung was removed, and four survived (Table VII).

DISCUSSION

The data of this study demonstrate that a high percentage of fetuses with CAM have a poor perinatal outcome, especially if the lesion is bilateral or there are other defects or hydrops fetalis. Nevertheless, the majority (68 per cent) of cases with isolated unilateral lesions survive. In some cases, there is unpredictable apparent antenatal resolution of the tumour with no need for postnatal surgery, and in others, there is progressive

Table II—Ultrasound findings in 27 cases of cystic adenomatoid malformation suffering intrauterine or neonatal deaths

Reference	Case No.	GA (weeks)	Type	Site	Hydrops	Mediastinal shift	AFV	Other defects	Outcome (weeks)
Mayden *et al.* (1984)	1	28	Mic	R	+	−	N		NND 29
Johnson *et al.* (1984)	2	28	Mic	L	+	+	P		NND 30
Diwan *et al.* (1983)	3	32	Mic	L	+	+	P		NND 32
Chao and Monoson (1990)	4	27	Mac	R	+	+	N		NND 35* Thoracotomy
Avni *et al.* (1986)	5	26	Mac	L	+	+	P		NND 29*
Carles *et al.* (1986)	6	24	Mic	R	+	+	N		IUD 26
Donn *et al.* (1981)	7	27	Mac	R	+	−	P		NND 30 Thoracotomy
Garrett *et al.* (1975)	8	32	Mic	R	+	−	P		IUD 34
Oster and Fortune (1978)	9	31	Mac	R	+	+	P		IUD 31
Morris *et al.* (1991)	10	31	Mac	L	+	+	P	CDH	NND 32
Heydanus *et al.* (1993)	11	28	Mac	L	+	+	P		NND 35
	12	25	Mac	L	+	+	P		NND 26
	13	25	Mac	R	+	NA	R		NND 31
Neilson *et al.* (1991)	14	30	Mac	NA	+	+	P		NND 30 Thoracotomy
	15	30	Mac	NA	+	+	P		NND 34* Thoracotomy
Adzick *et al.* (1985)	16	32	Mac	R	+	+	N		NND 33
	17	22	Mac	R	+	NA	P		NND 25
	18	26	Mac	L	+	NA	P	CHD	NND Thoracotomy
	19	33	Mac	L	−	NA	P		NND 36 Thoracotomy
	20	32	Mic	R	+	NA	P		NND 34
	21	30	Mic	L	+	NA	P		NND 36
KCH	22	31	Mic	L	+	+	P		IUD 32
	23	31	Mac	L	−	−	P	IUGR, CHF, TOF	IUD 32
	24	24	Mac	L	+	+	N		IUD 29
	25	23	Mic	R	−	+	N		NND 33
	26	22	Mac	L	−	−	N		NND 26
	27	20	Mic	R	−	+	N		NND 24

GA=Gestational age; AFV=amniotic fluid volume (N=normal, P=polyhydramnios); Mic=microcystic; Mac=macrocystic; CHD=congenital heart disease; IUGR=intrauterine growth retardation; TOF=tracheoesophageal fistula; CDH=congenital diaphragmatic hernia; Bil=bilateral; L=left; R=right; NA=information not available; NND=neonatal death; IUD=intrauterine death; KCH=King's College Hospital.

*??

Table III—Ultrasound findings in 61 surviving cases of cystic adenomatoid malformation

Reference	Case No.	GA (weeks)	Type	Site	Hydrops	Mediastinal shift	AFV	Other defects	Outcome (weeks)
Graham *et al.* (1982)	1	25	Mac	R	−	−	P		LB 40 Thoractomy
Cohen *et al.* (1983)	2	30	Mac	R	−	−	N		LB 40 Thoracotomy
Clairborne *et al.* (1985)	3	30	Mic	R	−	−	P		LB 33 Thoracotomy
Stauffer *et al.* (1984)	4	27	Mac	L	−	−	N		LB 40 Thoracotomy
Deacon *et al.* (1990)	5	25	Mac	R	−	+	N		LB 36
Pezzuti and Isler (1983)	6	36	Mac	R	−	−	N		LB 40 Thoracotomy
Clark *et al.* (1987)	7	20	Mac	R	+	NA	N		LB 37* Thoracotomy
Saltzman *et al.* (1988)	8	32	Mac	R	+	+	P		LB 40†
	9	33	Mac	R	−	+	P		LB 39† Thoracotomy
Sonek *et al.* (1991)	10	24	Mic	L	−	+	P	Spina bifida	LB 37†
Glaves and Baker (1983)	11	32	Mac	L	+	−	P		LB 40† Thoracotomy
Morris *et al.* (1991)	12	19	Mac	L	−	+	P		LB 40 Thoracotomy
Hatjis and Wall (1992)	13	22	Mac	R	−	+	N		LB 36†
Heydanus *et al.* (1993)	14	31	Mac	L	−	+	N		LB 38 Thoracotomy
	15	25	Mac	L	−	+	P		LB 38† Thoracotomy
Neilson *et al.* (1991)	16	26	Mac		+	NA	N		LB 31 Thoracotomy
	17	18	Mac	R	−	+	N		LB 40† Thoracotomy
	18	34	Mac		−	NA	P	Cleft plate	LB 37† Thoracotomy
	19	32	Mac		−	NA	P	Sequestration	LB 41† Thoracotomy
	20	20	Mac		+	NA	P		LB 38 Thoracotomy
	21	36	Mac		−	NA	N		LB 40 Thoracotomy
Adzick *et al.* (1985)	22	33	Mac	R	−	+	N		LB 37* Thoracotomy
	23	26	Mic	L	−	+	P		LB 32 Thoracotomy
	24	24	Mac	R	−	−	P		LB 37 Thoracotomy
	25	24	Mac	L	−	+	P		LB 37 Thoracotomy
	26	32	Mac	R	−	NA	P		LB 39 Thoracotomy
	27	33	Mac	L	−	NA	P		LB 39 Thoracotomy
	28	35	Mac	L	−	NA	P		LB 36 Thoracotomy
	29	20	Mac	L	−	NA	N		LB 40 Thoracotomy
	30	26	Mac	L	−	NA	N		LB 40 Thoracotomy
	31	27	Mac	L	−	NA	N		LB 40 Thoracotomy

KCH	32	23	Mic	L	−	−	N	LB 39
	33	25	Mac	R	−	+	N	LB 39* Thoracotomy
	34	31	Mac	L	−	+	P	LB 37* Thoracotomy
	35	30	Mac	L	+	+	P	LB 30
	36	24	Mac	L	−	−	N	LB 40
	37	20	Mac	L	−	−	N	LB 39 Thoracotomy
	38	28	Mic	L	−	−	N	LB 40†
	39	24	Mac	L	−	+	N	LB 40* Thoracotomy
	40	20	Mic	L	−	−	N	LB 40†
	41	19	Mac	L	−	+	N	LB 36†
	42	21	Mac	L	−	−	N	LB 38
	43	23	Mac	L	−	+	N	LB 41
	44	22	Mac	R	−	+	N	LB 40 Thoracotomy
	45	32	Mac	L	−	+	P	LB 33* Thoracotomy
	46	20	Mac	R	−	−	N	LB 34
	47	19	Mic	L	−	+	N	LB 40 Thoracotomy
	48	21	Mac	R	−	+	N	LB 38 Thoracotomy
	49	20	Mic	L	−	+	N	LB 39 Thoracotomy
	50	28	Mic	R	−	−	N	LB 40
	51	26	Mic	L	−	−	N	LB 40
	52	21	Mic	L	−	+	N	LB 40
	53	39	Mac	L	−	−	O	LB 40 Thoracotomy
	54	19	Mic	L	−	−	N	LB 37
	55	29	Mac	R	−	−	N	LB 43
	56	26	Mac	R	−	+	N	LB 38* Thoracotomy
	57	20	Mac	L	−	−	N	LB 40
	58	24	Mac	L	−	+	N	LB 40
	59	20	Mic	R	−	+	N	LB 37 Thoracotomy
	60	19	Mic	L	−	−	N	LB 39 Thoracotomy
	61	20	Mic	R	−	−	N	LB 41 Thoracotomy

GA=Gestational age; L=left; R=right; Mic=microcystic; Mac=macrocystic; NA=information not available; AFV=amniotic fluid volume (N=normal, P=polyhydramnios, O=oligohydramnios); KCH=King's College Hospital.

*Antenatal surgery.

†Antenatal resolution.

Table IV—Pregnancy outcome and antenatal findings in 132 cases of cystic adenomatoid malformation of the lung

			All cases			
		n	TOP	IUD	NND	Alive
Site	Bilateral	18	18	0	0	0
	Unilateral	114	26	6	21	61
Type	Macrocystic	77	15	3	13	46
	Microcystic	55	29	3	8	15
Hydrops	Present	57	29	5	17	6
	Absent	75	15	1	4	55
Other defects	Present	16	10	1	2	3
	Absent	116	34	5	19	58
Amniotic fluid	Polyhydramnios	46	8	4	14	20
	Normal/reduced	88	36	2	7	41

TOP=Termination of pregnancy; IUD=intrauterine death; NND=neonatal death.

enlargement leading to hydrops and polyhydramnios. In the latter group, there may be a role for prenatal excision of the tumour. Cases with solitary enlarging cysts benefit from thoraco-amniotic shunting.

The striking hyperechogenicity of the fetal lungs allows easy diagnosis of the condition and this is reflected in the gestational age distribution at diagnosis; the majority were diagnosed at 16–22 weeks when presumably routine ultrasound examination was undertaken. Another peak in the gestational age distribution occurs at around 32 weeks and this presumably reflects either a policy of routine third-trimester scanning or the development of clinically recognizable polyhydramnios, leading to a diagnostic scan. There are three possible explanations for the late detection of some of the cases: first, no scan was undertaken previously; second, the lesion escaped detection at an earlier scan; and third, the lesion may not have developed until the second half of pregnancy.

No first-trimester diagnosis of CAM has been reported. Whilst it is conceivable that such lesions may not occur until the second half of pregnancy, this explanation is unlikely because the embryological insult occurs as early as 8 weeks. The hyperechogenicity apparent at later gestation is thought to represent fluid accumulation in small cystic spaces. Presumably this process is not sufficiently advanced in early pregnancy to cause a distinct alteration in the ultrasonic appearance of

Table V—Cystic adenomatoid malformation—associated major malformations

Malformation	*n*
Facial cleft	2
Neural tube defect	2
Heart defect	4
Diaphragmatic hernia	1
Extralobar sequestration	1
Renal agenesis	1
Renal dysplasia	2
Exomphalos	2
Trisomy 18	1
Total	16/132 (12%)

Associated defects present in bilateral disease 28 per cent vs. unilateral 10 per cent. Associated defects present in microcystic disease 18 per cent vs. macrocystic 8 per cent.

Table VI—Summary of outcome of 132 cases of antenatally diagnosed CAM

Termination of pregnancy	44	(33%)
Continuing pregnancy	88	(67%)
Intrauterine death	6	(7%)
Neonatal death	21	(24%)
		15 after surgery
		6 before surgery
Alive	61	(69%)
		45 after surgery
		16 before surgery

Table VII—Cystic adenomatoid malformation—prenatal surgery

Technique	Reference	GA (weeks)	Ascites	Outcome	GA (weeks)
Aspiration	Chao and Monoson (1990)	27, 30	+	NND	35
Aspiration	Neilson *et al.* (1991)	34	+	NND	34
Aspiration	Kuller *et al.* (1992)	29	+	LB	30
Shunt	Avni *et al.* (1986)	26	+	NND	29*
Shunt	Kuller *et al.* (1992)	23	+	NND	?23
Shunt	Clark *et al.* (1987)	20	+	LB	37* Thoracotomy
Shunt	KCH	24	−	LB	40 Thoracotomy
Shunt	KCH	25	−	LB	39 Thoracotomy
Shunt	KCH	26	−	LB	38 Thoracotomy
Shunt	KCH	31	−	LB	37 Thoracotomy
Shunt	KCH	32	−	LB	33 Thoracotomy
Fetal Surgery	Kuller *et al.* (1992)	27	+	NND	28
Fetal Surgery	Kuller *et al.* (1992)	23	+	LB	30
Fetal Surgery	Kuller *et al.* (1992)	26	+	LB	34*
Fetal Surgery	Kuller *et al.* (1992)	25	+	LB	33
Fetal Surgery	Kuller *et al.* (1992)	24	+	LB	26*
Fetal Surgery	Kuller *et al.* (1992)	21	+	IUD	21

LB=Livebirth; NND=neonatal death; IUD=intrauterine death.
*Failed cyst aspiration.

the lungs. This situation may be considered to be analogous to the delayed ultrasonic diagnosis in cases of duodenal atresia, where the classic 'double bubble' appearance is usually not observed until late in the second trimester.

The differential diagnosis of a fetal thoracic mass includes pulmonary sequestration (Dolkart *et al.*, 1992), bronchogenic and enteric cysts (Hobbins *et al.*, 1979), mediastinal cystic teratoma (Golladay and Mollitt, 1984), laryngeal atresia (Watson *et al.*, 1990), and brain heterotopia (Gonzalez-Cruzzi *et al.*, 1980). Laryngeal obstruction may give rise to a prenatal ultrasound and histological appearance similar to CAM (Choong *et al.*, 1992), making differentiation of the conditions difficult. It has been suggested that the association between upper airways obstruction and hyperechogenicity of the lung fields may help to explain the apparent resolution of some cases of CAM (Nicolaides, 1992). The antenatal role of computed tomography or nuclear magnetic resonance scanning in such cases has yet to be validated, but it may provide more information regarding both the extent and the type of tissue involvement.

In the combined series, 12 per cent of the fetuses had additional major abnormalities. Apart from hydrops, which is likely to be a direct consequence of the pulmonary lesion, the other defects were non-specific. As expected, this incidence is considerably higher than that reported in the literature by paediatric surgeons [3 per cent in a combined series of 30 infants diagnosed postnatally (Neilson *et al.*, 1991; Heij *et al.*, 1990)] and considerably lower than the 26 per cent reported in post-mortem studies (Stocker *et al.*, 1977). This finding is analogous to the antenatal findings in congenital diaphragmatic hernia (Thorpe-Beeston *et al.*, 1989).

Differences in the survival rates of fetuses with CAM have previously been ascribed to the histological nature of the lesion (Stocker *et al.*, 1977) or the presence of fetal hydrops (Adzick *et al.*, 1985; Oster and Fortune, 1978). Hydrops may result from venocaval obstruction and cardiac compression from the extreme mediastinal shift. Hypoplasia of the non-affected lung may be due to compression by the expanding tumour or the pleural effusions and ascites. In the present series, the development of hydrops was associated with a worse outcome, but this was not invariably the case.

Mediastinal shift and polyhydramnios may also be markers of more severe disease. Mediastinal shift was reported (where sufficient information

was provided) in 54 per cent of all cases and in 55 per cent of surviving infants, suggesting that mediastinal shift is a poor predictor of pulmonary hypoplasia in CAM; this is also true for congenital diaphragmatic hernia (Thorpe-Beeston *et al.*, 1989). Polyhydramnios may be due to decreased fetal swallowing, the consequence of oesophageal compression by the mass (Donn *et al.*, 1981), or there may be increased fetal lung fluid production by the abnormal tissue (Krous *et al.*, 1980). Increased amniotic fluid volume or polyhydramnios was present in 33 per cent of surviving fetuses and in 67 per cent of those that subsequently died *in utero* or in the postnatal period. Since polyhydramnios is associated with premature delivery, the prognosis could be improved by repeated amniocenteses; such treatment has been shown to be effective in acute polyhydramnios due to twin–twin transfusion syndrome (Saunders *et al.*, 1992).

Thoraco-amniotic shunting, for drainage of large cysts causing mediastinal shift and compression of the contralateral lung, is minimally invasive and is associated with a good outcome. The role of more invasive intervention, such as hysterotomy and excision of solid tumours, remains to be defined. Kuller *et al.* (1992) have recently suggested that the development of non-immune hydrops may be an indication for open fetal surgery, and four of six cases that were treated survived. Nevertheless, the potential risks to the mother both during the pregnancy and in subsequent confinements should not be underestimated. Although rigorous evaluation of such intervention is being undertaken, the difficulties in organizing a trial may prove insurmountable.

Postnatally, management frequently included thoracotomy and lobectomy as soon as the diagnosis was confirmed, even in asymptomatic infants. Such a policy is based on anecdotal evidence of malignant change (Heij *et al.*, 1990; Ueda *et al.*, 1977) and is analogous to prophylactic nephrectomy in cases of multicystic renal disease, which has now been abandoned. In many cases, the pulmonary tumour was observed to regress antenatally and this improvement may continue postnatally. Although symptomatic infants with severe disease are likely to require postnatal surgery, this is now difficult to justify in asymptomatic neonates. Care must be taken to ensure that prenatal diagnosis should not automatically mean that an infant is subjected to unnecessary postnatal intervention.

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