ORIGINAL ARTICLE

Outcome of antenatally suspected congenital cystic adenomatoid malformation of the lung: 10 years' experience 1991–2001

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Objective: To determine the outcome of antenatally suspected congenital cystic adenomatoid malformation of the lung (CCAM) over a 10 year period.

Methods: This is a retrospective study of all babies diagnosed antenatally in the Prenatal Diagnosis Unit and delivered in Oxford between 1991 and 2001. Data were obtained from the Oxford Congenital Anomaly Register, theatre records, and histopathology reports.

Results: Twenty eight cases of CCAM were diagnosed antenatally. Five pregnancies were terminated. Data are available on all 23 of the pregnancies that continued and resulted in two neonatal deaths and 21 surviving babies. Eleven of the 23 cases (48%) showed some regression of the lesion antenatally, and four of these cases appeared to resolve completely on prenatal ultrasound. Three of the 23 babies (13%) were symptomatic in the early neonatal period, and three developed symptoms shortly afterwards. Seventeen of the 23 babies (74%) were asymptomatic, of whom 12 had abnormalities on chest radiograph or computed tomography scan and had elective surgery. Two babies (8%) had completely normal postnatal imaging, and three had abnormalities which resolved in the first year of life. Seventeen of the 23 babies (74%) had surgery. Histology at surgery was heterogeneous. Of the 23 live births, all 21 survivors (91%) are well at follow up or have been discharged.

Conclusions: All babies diagnosed antenatally with CCAM require postnatal imaging with computed tomography irrespective of signs of antenatal resolution. In asymptomatic infants, the recommendations are close follow up and elective surgery for persistent lesions within the first year of life. Histology at surgery was heterogeneous, and this should be considered when counselling parents.

Congenital cystic adenomatoid malformation of the lung (CCAM) is a form of congenital cystic lung disease believed to result from an arrest in fetal lung development.^{1–3} There has been an increase in cases suspected prenatally over the last decade. Antenatal ultrasound findings suggestive of CCAM include an increase in lung echodensity with or without associated cyst formation. Hydrops may occur in the most severe cases. The differential diagnosis includes lobar sequestration or congenital diaphragmatic hernia.^{4–6}

CCAM is usually restricted to a single lobe and occurs in isolation.⁷ Associated anomalies are rare. An incidence of 1 in 25 000 live births has been suggested,⁸ with males and females equally affected. Potential clinical outcomes associated with CCAM are as follows. Antenatally it can lead to hydrops, polyhydramnios, or regress.⁹ Postnatally the baby may have severe respiratory distress, or more commonly be asymptomatic.^{8 10–13} Traditionally, surgery is recommended to remove the CCAM because of the long term risks of morbidity from infection, pneumothorax, or, more rarely, malignancy.^{14–18}

The aims of this study were to document the outcome of all babies diagnosed antenatally with suspected CCAM over a 10 year period from 1991 to 2001 in order to facilitate counselling of parents during pregnancy and to allow optimal timing of postnatal management.

METHODS

A retrospective audit was carried out of all babies diagnosed in the Prenatal Diagnosis Unit and delivered at the John Radcliffe Hospital, Oxford between 1991 and 2001. Data were obtained from the Oxford Congenital Anomaly Register,

theatre records, and histopathology reports, to identify all babies with a diagnosis of CCAM. Information on gestation at diagnosis, progression of the lesion during pregnancy, the presence of other anomalies, postnatal features, results of postnatal investigations, and outcome were recorded. Ethical permission was not required for this study.

RESULTS

Figure 1 illustrates the overall results of the study. Of the pregnancies that continued, data are available on all 23 babies, two of whom died in the neonatal period and 21 are alive and well. Table 1 shows the characteristics of these 23 babies. All 21 babies who survived had postnatal imaging with chest radiograph and computed tomography (CT) scan.

After delivery, three babies were symptomatic, and three developed symptoms later in the first year of life at 2 weeks, 8 weeks, and 5 months. There were two neonatal deaths: one baby previously noted to have massive ascites which resolved, born at 36 weeks gestation with a type 3 CCAM, on the right, who died at 2 hours of age and was found at post mortem examination to also have right bronchial atresia and a hypoplastic left lung; the second baby was born at 35 weeks gestation and had a large CCAM with hydrops. Despite surgery, he died on day 9 with complications from multiple air leaks.

Seventeen babies (74%) were asymptomatic after birth. Of these, 12 (71%) had abnormalities which were detectable on postnatal imaging and went on to have elective surgery. Two babies had their diagnosis changed on the basis of postnatal

Abbreviations: CCAM, congenital cystic adenomatoid malformation of the lung; CT, computed tomography

Figure 1 Outcome of antenatal diagnosis of congenital cystic adenomatoid malformation of the lung (CCAM). TOP, Termination of pregnancy.

imaging to lobar sequestration and a bronchogenic cyst respectively. Five of the 17 asymptomatic babies (29%) did not require surgery. Of these five babies, two had chest

radiograph and CT scans reported as normal in the postnatal period, one baby had a normal chest radiograph and an area of volume loss on the CT scan, but no identifiable abnormality, and two babies had normal chest radiograph and changes on CT scan, which resolved by 9 months of age. Four of these five babies were followed up for 12–17 months and were well at discharge.

Overall, 17 babies had surgery. Sixteen of these survived. Timing of surgery ranged from two days to 23 months with a median of eight and a half months. Histology from surgery was heterogeneous. There were nine cases of isolated CCAM, two of adenomatoid changes within a sequestered lobe, one bronchogenic cyst which had been diagnosed on postnatal magnetic resonance imaging scan at 5 months of age, three cases of isolated lobar sequestration, one of which was diagnosed on postnatal CT scan, and two cases where the histology was of bronchial malformation, not typical of either CCAM or sequestration, with areas of collapse and air trapping.

All 21 survivors (91%) are well at follow up or have been discharged from hospital follow up at between 1 and 3 years of age.

Table 1 Characteristics of babies born after antenatal diagnosis of congenital cystic adenomatoid malformation of the lung	
Gestation (weeks)	39 (35–42)
Birth weight (g)	3583 (2640–4674)
Sex	
Male	13 (57%)
Female	10 (43%)
Unilateral	
Right	9 (39%)
Left	14 (61%)
Bilateral	0 (0%)
Hydrops	2 (8.7%)
Mediastinal shift	16 (70%)
Antenatal regression	11 (48%)
Complete antenatal resolution	2 (8.7%)

Values are median (range) or number (%) (n = 23).

What is already known on this topic

- Congenital cystic adenomatoid malformation is a congenital abnormality of the lung believed to result from an arrest in fetal lung development
- There is a spectrum of outcomes associated with this condition

DISCUSSION

CCAM is an important diagnosis that can be suspected on routine antenatal ultrasound. It has implications for both the ongoing pregnancy and the baby, at delivery and later in life. Although the prognosis is generally good, this study illustrates the spectrum of potential outcome for babies with this diagnosis, ranging from hydrops and severe respiratory distress with pulmonary hypoplasia, to resolution of the lesion either antenatally or postnatally. In this study, as noted by others,^{13–19} a poor outcome was associated with the presence of hydrops and prematurity. It is important that, when this diagnosis is made antenatally, parents are made aware of all potential outcomes and the need for ongoing surveillance in the pregnancy. Ultrasound scanning is subjective, and the findings often non-specific and therefore follow up scanning is important both to confirm a diagnosis and to document any changes in appearances of the lesion.

Although some lesions do resolve antenatally and most babies are asymptomatic at birth, we would recommend that all babies are investigated in the neonatal period with CT scan, even if the lesion appeared to resolve completely antenatally. We document two cases of complete antenatal resolution with normal postnatal investigations, and two cases in which, although the CCAM was not seen late in pregnancy, postnatal investigations confirmed the initial diagnosis. It is also important to distinguish where possible CCAM from lobar sequestration on postnatal imaging to ensure optimal management.

Most lesions do persist in the first year of life and therefore surgery is recommended. However, we have documented three cases in which abnormalities on postnatal imaging resolved by 9 months of age. We would therefore advocate a close follow up of asymptomatic babies, with repeat imaging and surgery for persistent lesions within the first year of life. If symptoms develop at any stage, surgery would need to be considered earlier.

The histology at surgery was variable, with some babies having their diagnosis changed to lobar sequestration. This should be considered when counselling parents antenatally. The non-specific term congenital lung malformation may be more useful when discussing the diagnosis with parents antenatally, as has been suggested by Bush.²⁰

In conclusion, this paper provides valuable information for parents and recommendations for timing of surgery in asymptomatic patients diagnosed antenatally with CCAM.

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What this study adds

- Most cases persist and are present after birth, although most babies are asymptomatic
- In those cases where the lesion persists, surgery is required to remove the abnormality because of the long term risks of infection, pneumothorax, and, more rarely, malignancy

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vein PSCs placed over a 13 month period in one neonatal intensive care unit. All had been inserted in the left leg, and the inserting practitioner failed to identify malpositioning of the PSC tip on a frontal abdominal view obtained with contrast (fig 1A). Radiologists later identified the probable abnormal position based on the course of the PSC, and lateral views of the abdomen on the same day of insertion showed the superficial position of the catheter tip in the anterior abdominal wall (fig 1B).

A literature review showed that most of the reported serious complications secondary to malpositioning of saphenous lines occurred with a PSC inserted via the left leg.¹⁻⁶ Chen *et al*¹ reported one new and three previously reported cases of paraplegia secondary to tracking of a PSC into the lumbar venous plexus from a left saphenous vein. Odaibo *et al*² and Kelly *et al*³ reported serious neurological and respiratory complications in pre-term infants from parenteral nutrition fluid in the cerebrospinal fluid via left leg PSCs. Baker and Imong⁴ reported abdominal wall necrosis as a complication of PSC via a left saphenous vein with the tip in a superficial abdominal vessel, and Coit and Kamitsuka⁵ reported similar findings, but had one case from a right leg, as did Cartwright.⁶

Healthcare providers need to be aware of the expected projection of a normally placed saphenous PSC. The appropriate course of the left leg PSC line passes from the left femoral and iliac veins, gradually ascending and becoming more medial until joining the

inferior vena cava over the right to mid spine at the L5 or L4 level. It then ascends over the right side of the spine or slightly to the right of the spine (fig 1C). Lines that end below the L5 level are not likely to be in the inferior vena cava. Lines that ascend to the left of the spine may be in an anomalous inferior vena cava but may be in a vertebral vein, in an anterior abdominal wall vessel, in the aorta, or extravascular.

We believe that tracking of the PSC line into an anterior abdominal wall vein can be subtle on the frontal abdominal radiograph, and serious catheter malposition related complications can be prevented by careful comparison with the expected course on the anterior-posterior projection (fig 1C). If uncertain, a lateral abdominal radiograph to verify the catheter course and tip location is needed.

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CORRECTION

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J K Calvert, P A Boyd, P C Chamberlain, *et al*. Outcome of antenatally suspected congenital cystic adenomatoid malformation of the lung: 10 years' experience 1991-2001 (*Arch Dis Child Fetal Neonatal Ed* 2006;**91**:F26-8). The surname of the fourth author of this paper was spelt incorrectly; the correct name is S Syed.

Figure 1 (A) Frontal abdominal view of malpositioned saphenous vein percutaneous silastic catheter inserted in the left leg. (B) Lateral view of the abdomen on the day of insertion showing the superficial position of the tip in the anterior abdominal wall of the catheter shown in (A). (C) Frontal abdominal view of an appropriately positioned catheter.

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