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

J K Calvert, P A Boyd, P C Chamberlain, S Said, K Lakhoo

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 asymptomatic 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.10,14

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 and had 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 investigations.
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

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Number (n)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gestation (weeks)</td>
<td>39 (35–42)</td>
</tr>
<tr>
<td>Birth weight (g)</td>
<td>3583 (2640–4674)</td>
</tr>
<tr>
<td>Sex</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>13 (57%)</td>
</tr>
<tr>
<td>Female</td>
<td>10 (43%)</td>
</tr>
<tr>
<td>Unilateral</td>
<td></td>
</tr>
<tr>
<td>Right</td>
<td>9 (39%)</td>
</tr>
<tr>
<td>Left</td>
<td>14 (61%)</td>
</tr>
<tr>
<td>Bilateral</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Hydrops</td>
<td>2 (8.7%)</td>
</tr>
<tr>
<td>Mediastinal shift</td>
<td>16 (70%)</td>
</tr>
<tr>
<td>Antenatal regression</td>
<td>11 (48%)</td>
</tr>
<tr>
<td>Complete antenatal resolution</td>
<td>2 (8.7%)</td>
</tr>
</tbody>
</table>

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

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

ACKNOWLEDGEMENTS

We acknowledge Dr Mary Anthony for her support and guidance for this study.

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

What this study adds

- Most cases persist and are present at 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

REFERENCES