Foetal Intralobar Lung Sequestration: Antenatal Diagnosis and Management

J S C Chen, N Walford, Y L Yan, C L Ong, G S H Yeo

ABSTRACT

Objectives: The objectives of this study are to discuss the use of ultrasonography for the diagnosis of foetal intralobar sequestration (FILS) antenatally and the management options available for these pregnancies.

Methods: This is a retrospective review of six cases of FILS diagnosed antenatally by two dimensional (2D) and colour Doppler ultrasonography out of a total of 31,508 deliveries over a two-year period at the KK Women's and Children's Hospital.

Results: The incidence of FILS in this hospital was 1 in 5,251 deliveries. 2D ultrasonography showed an echogenic lung in all cases. FILS was confirmed by the demonstration of a systemic vessel leading to the affected lung on colour Doppler examination. After counselling, four terminated their pregnancies during mid-trimester, while two continued their pregnancies to term. Confirmation of the terminated cases was by post-mortem. In the two pregnancies that continued, regular growth scans were done to monitor the progression of the condition. Computed tomography confirmed the diagnosis post-delivery. Both were well but one had a resection of the sequestrated lung although he was asymptomatic. Histology also confirmed the diagnosis.

Conclusion: FILS is a rare anomaly. 2D and colour Doppler ultrasonography are used to diagnose the condition antenatally. Termination of the pregnancy is not always indicated, as there are favourable outcomes from FILS.

Keywords: echogenic lungs, intralobar sequestration, Colour Doppler Ultrasonography, foetal anomalies

INTRODUCTION

Intralobar lung sequestration (ILS), by definition, consists of a portion of lung that is isolated (sequestered) from the tracheobronchial tree and is supplied by a systemic artery. It develops from an accessory lung bud that arises before the formation of the pleura and becomes surrounded by the same pleura as the normal lung. The vascular supply of the sequestrated lung is from the aorta. ILS is a rare anomaly and genetic predisposition has been suggested. It accounts for 75% of pulmonary sequestrations involving mainly the lower lobe of the lung. This condition affects either side equally though bilateral involvement is rare. The majority of the arteries originate from the thoracic (93%) or abdominal aorta and the venous drainage terminates in the pulmonary veins (96%). Macroscopically, the section areas may show either a cystic appearance to the less frequent pseudotumorous form.

The origin of ILS has been a source of debate in the past. There are two schools of thought: one that believes that it is congenital or foetal in origin and the other, an acquired lesion. More papers have been published recently supporting the congenital origin of ILS. The main reason for this is that more cases of ILS are now detected antenatally with advances in ultrasonography, especially with the use of colour Doppler scans to detect the systemic vessel leading to the sequestrated lung. In the past, as these tools of diagnosis were not available, cases were detected only later in life when complications from the condition occurred. This could account for ILS being thought to be an acquired lesion. The current medical literature has limited documentation of foetal intralobar lung sequestration (FILS). There are very few papers written on the antenatal diagnosis of this condition by ultrasonography, and the current series is the first in Singapore.

METHODS

This is a retrospective review of six cases of FILS diagnosed during the mid-trimester screening scans over a two-year period, between 9 May 2000 and 21 May 2002, in the KK Women’s and Children’s Hospital in Singapore. There were a total of 31,508 deliveries during this period of time. Routine screening ultrasonography scans were usually done for all
antenatal patients except for an approximate 3% of patients who may not have been screened because they had booked late in their pregnancies. However, all babies were screened after birth by the neonatologists. All cases with abnormalities diagnosed antenatally on screening ultrasonography scans or postnatally after screening by the neonatologists were reported to the hospital’s birth defect registry. This is where discussion, verification and classification of the fetal abnormalities are done.

Cases of FILS were identified from the birth defect registry. The case notes for these pregnancies were then obtained from the hospital’s medical records and analysed.

The following data were analysed:
1. Age and race of the patients
2. Screening scan findings
3. Counselling and treatment options given
4. Decision made about the pregnancy
5. Subsequent scans and complications that occurred
6. Postnatal outcome of the baby
7. Post-mortem findings
8. Computed tomography (CT) findings
9. Histology results

RESULTS
The incidence of FILS in this study was 1 in 5,251 deliveries. The mean age of the patients was 28.33 years of age (SD ± 2.42). Five (83.3%) were Chinese and one (16.7%) was a Malay. There was an equal distribution of primigravidas and multiparous patients. The mean gestation period at screening was 20.72 weeks (SD ± 0.71) (see Table I for a summary of the six cases). 2D and colour Doppler ultrasonography showed echogenic lungs and

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**Table I. A summary of the six cases of Foetal Intralobar Lung Sequestration (FILS).**

<table>
<thead>
<tr>
<th>Race</th>
<th>Age (years)</th>
<th>Screening Scan findings</th>
<th>Decision</th>
<th>Subsequent scans</th>
<th>Postnatal outcome</th>
<th>Post-mortem Findings</th>
<th>CT scan findings</th>
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<tr>
<td>Chinese</td>
<td>28</td>
<td>Echogenic right lung with large vessel from aorta at 20.9 weeks</td>
<td>Termination of pregnancy Male fetus</td>
<td>–</td>
<td>–</td>
<td>Right FILS with blood supply from infradiaphragmatic aorta</td>
<td>–</td>
</tr>
<tr>
<td>Chinese</td>
<td>28</td>
<td>Echogenic left lung with vessel from thoracic aorta at 19.7 weeks Heart pushed to the right</td>
<td>Termination of pregnancy Male fetus</td>
<td>–</td>
<td>–</td>
<td>Left FILS with arterial supply from thoracic aorta</td>
<td>–</td>
</tr>
<tr>
<td>Chinese</td>
<td>30</td>
<td>Echogenic left lung with vessel from the aorta at 21.9 weeks Heart pushed to the right</td>
<td>Termination of pregnancy Female fetus</td>
<td>–</td>
<td>–</td>
<td>Left FILS with arterial supply from descending aorta</td>
<td>–</td>
</tr>
<tr>
<td>Chinese</td>
<td>27</td>
<td>Echogenic left lung with vessel from descending aorta at 20.5 weeks Mediastinum pushed to the right</td>
<td>Termination of pregnancy Female fetus</td>
<td>–</td>
<td>–</td>
<td>Left FILS with blood supply from descending thoracic aorta</td>
<td>–</td>
</tr>
<tr>
<td>Malay</td>
<td>25</td>
<td>Echogenic left lung with vessel from aorta at 20.7 weeks Heart pushed to the right</td>
<td>Continuation of pregnancy</td>
<td>–</td>
<td>–</td>
<td>Left intralobar sequestration with blood supply from distal thoracic aorta</td>
<td>–</td>
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<tr>
<td>Chinese</td>
<td>32</td>
<td>Echogenic left lung with arterial supply from aorta at 20.6 weeks Heart displaced to the right</td>
<td>Continuation of pregnancy</td>
<td>–</td>
<td>–</td>
<td>Left intralobar sequestration with blood supply from distal thoracic aorta</td>
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demonstrated the presence of a systemic blood supply to the sequestrated lung in all cases (Figs. 1a, 1b).

Of the six cases, four were male (66.7%) and two were female (33.3%) fetuses. Five left lungs (83.3%) and one right lung (16.7%) were affected. All had arterial supply from the aorta. The displacement of the heart to the right side of the thorax was present in five cases (83.3%). Four out of the six patients (66.7%) opted for mid-trimester termination of pregnancy.
while the other two patients (33.3%) continued their pregnancies to term. The diagnosis for the terminated cases were confirmed at post-mortem.

In the two cases that continued to term, ultrasonography showed apparent resolution of the lung masses later in the pregnancies (Figs. 2a, 2b). There were no antenatal complications in these two cases. Both delivered vaginally at term male infants that were well and did not require any respiratory support. CT scans performed after birth revealed lung lesions that confirmed the diagnosis. One of the cases (Case 6), though asymptomatic at birth, had a surgical resection of the affected lung to prevent complications later in life. He made a good post-operative recovery. The diagnosis was confirmed on histology. The other case (Case 5) did not have a surgical resection of the affected lung and remained well at one year of age (Figs. 2c, 2d).

**DISCUSSION**

FILS appears on 2D ultrasonography (transverse scan) as an echogenic lung mass, with or without cysts, usually within the lower lobe of the lung\(^\text{11}\). Differential diagnosis for solid echogenic lung lesions include FILS, mediastinal teratomas, an obstruction of a major airway or bronchial cysts. Possible diagnosis for cystic lesions in the echogenic lung include congenital cystic adenomatoid malformation (CCAML) of the lung, types 1 and 2. Colour Doppler ultrasonography is used to differentiate FILS from other echogenic solid lung lesions by the identification of a systemic vessel leading to the sequestrated lung\(^\text{6-8}\). The vessel is detected by 2D scan taken in the coronal or transverse planes. In order to make a diagnosis of FILS, both the presence of an echogenic lung and the systemic vessel must be detected on scanning. Magnetic resonance imaging has been found to be useful in diagnosing this condition when ultrasonographic findings are equivocal\(^\text{9}\).

The accuracy rate for the antenatal detection of FILS by ultrasonography is not known due to limited feasible data in the current literature. Some cases of FILS are not diagnosed as colour Doppler ultrasonography may not be available during the screening scan. In such cases, the diagnosis is only made postnatally when a problem, such as recurrent chest infections, develops. The accuracy of diagnosing FILS is greater in the second trimester compared to the third trimester of pregnancy. In all cases, confirmation of the diagnosis of FILS is made only at post-mortem (Fig. 3), CT scan after birth, or by histology of the resected lung. About 50% of sequestrations are atypical in presentation or associated with other anomalies\(^\text{10}\). When FILS is discovered antenatally, patients should be counselled about the varied prognosis. Pulmonary sequestrations are a subgroup of congenital lung lesions with a favourable outlook\(^\text{11}\). Many regress prenatally while the persistent ones can be resected postnatally if symptomatic, or left alone. Termination of pregnancy is not the only option available.

In our series of cases, two doctors from the maternal-foetal department of the hospital counselled patients with an affected pregnancy. These doctors attend the birth defect clinic held weekly in the hospital where the management of this condition is discussed. The patients are counselled about the rarity and possible prognosis of FILS. During the counselling, it was explained that the condition could regress, remain stable or deteriorate, leading to complications like hydrops or heart failure. Surgical resection of the lesion postnatally to improve outcome was also discussed. Though all patients were counselled in the same way and told about good prognosis reported in the literature, the majority of our cases decided to undergo termination of pregnancy. The decision of whether to terminate or to continue with an affected pregnancy may be influenced by various factors. This would include taking into consideration an individual’s character and personality. Some patients may not be willing to continue with an affected pregnancy.
pregnancy no matter how favourable the prognosis may be. Also the different cultural and religious beliefs of the individual may play a major part in influencing the decision made. The two cases that decided to continue with their pregnancies were from Muslim backgrounds.

In cases where the pregnancy continues, there is a risk of the fetus developing hydrops\(^\text{(12)}\), congestive heart failure and polyhydramnios. These complications result from the effects caused by the sequestrated lung. In some cases, the lung lesion increases in size and causes a mediastinal shift. This may cause vascular compression and eventually lead to heart failure and hydrops. Premature labour or intrauterine death may also occur. Regular growth scans should be performed to detect such complications. In some cases, thoracocentesis have been used, with the resolution of the hydrops\(^\text{(13)}\). In the absence of non-immune hydrops fetalis or other anomalies, the outcome of the fetuses is excellent, with over 90% survival\(^\text{(14)}\). Our two cases that continued to term deliveries showed apparent regression of the affected lesion on antenatal ultrasonography with no complications occurring.

Delivery should be carried out in a tertiary centre where there is rapid access to neonatal care and paediatric surgery, as the presence of this condition in the newborn can cause respiratory distress\(^\text{(15)}\). Immediate respiratory support may be needed. The placement of a thoracoamniotic shunt for drainage of hydrothorax can be done, or eventual post-natal resection may be needed later in life if recurrent pulmonary infection, haemorrhage, gastrointestinal symptoms or heart failure occur.

**CONCLUSION**

Our series of FILS support existing medical literature that it is a congenital and rare anomaly that can be diagnosed by 2D and colour Doppler ultrasonography antenatally. Identification of FILS by ultrasonography has enabled clinicians to counsel patients about the prognosis. As the outcome for this condition is variable, termination of pregnancy is not the only option. In pregnancies that continue, regular growth scans should be done to exclude complications that can occur. After delivery, surgical resection of the sequestrated lung is not always needed though long-term follow-up of the patient is recommended. Our series have shown good outcomes for this condition. We recommend on-going collection of data for this condition and longer-term follow-up of affected cases.

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**REFERENCES**