Congenital lung malformations: correlation between prenatal and postnatal imaging and pathological findings

Jean-François Lecompte[a], Bastien Gondran-Tellier[b], Geraldine Hery[b], Daniel Sikavi[b], Nathalie Colavolpe[c], Jean-Michel Guys[b] and Pascal de Lagausie[b]

Aim **Congenital lung malformations are a common finding during prenatal ultrasonography (US). Investigations were completed by means of prenatal MRI and postnatal computed tomographic (CT) scan. The purpose of this study was to compare these prenatal findings with postnatal findings and pathological findings after surgical resection.**

Materials and methods **Prenatal examinations and postnatal CT scan results of congenital malformations were compared with pathological findings.**

Results **From 2007 to 2013, 39 prenatally diagnosed congenital lung malformations were resected: 18 congenital cystic adenomatoid malformation, eight pulmonary sequestration, five bronchogenic cyst, one bronchial atresia and six complex lesions. Correlation between imaging and diagnosis was as follows: congenital cystic adenomatoid malformation was seen in 17/17 patients using postnatal CT, in 10/15 patients using prenatal MRI and in 17/18 patients using prenatal US. Correlation between imaging and diagnosis was as follows: bronchogenic cyst was seen in 3/5, 3/5 and 3/5 patients, pulmonary sequestration was seen in 7/9, 5/9 and 4/9 patients, and complex lesion was seen in 4/5, 3/6, and 2/6 patients using postnatal CT, prenatal MRI, and prenatal US, respectively. Overall, 32/37 cases were diagnosed by means of postnatal CT, 21/36 cases were diagnosed by means of prenatal MRI and 26/39 cases were diagnosed by means of prenatal US.**

Conclusion **Discordance between imaging data and definitive diagnosis is not rare. Our results suggest that postnatal CT scan is the most sensible and specific examination. Before birth, US seems better compared with MRI for description of the lesion. MRI seems to be useful in case of complex lesions and pulmonary sequestration.** ***Ann Pediatr Surg*** **12:82–85 © 2016 Annals of Pediatric Surgery.**

Annals of Pediatric Surgery 2016, **12**:82–85

Keywords: congenital lung malformation, imaging, prenatal

[a]Department of Pediatric Surgery, Hopitaux pediatriques de Nice CHU Lenval, Nice, Departments of [b]Pediatric Surgery and [c]Pediatric Radiology, Hopital Timone enfant, Marseille, France

Correspondence to Jean-François Lecompte, MD, Department of Pediatric Surgery, Hopitaux pediatriques de Nice CHU Lenval, 57 Avenue de la Califonie, 06300 Nice, France
Tel: +33 492 030 316; fax: +33 492 030 486;
e-mail: lecompte.j@pediatrie-chulenval-nice.fr

Received 9 October 2015 **accepted** 26 January 2016

Introduction

Congenital lung malformations are traditionally discovered by means of prenatal ultrasonography (US) [1]. The natural history of these lesions is variable based on their histological nature, and they can grow more or less rapidly (even leading to a hydrops), remain stable or regress during the prenatal or postnatal period. Prenatal diagnoses have greatly modified the evolution of these lesions, permitting treatment before the onset of complications.

Other prenatal and postnatal imaging techniques can be proposed to characterize these lesions to predict their evolution and select the most appropriate treatment for the suspected diagnosis. Unlike radiographic computed tomography (CT), which can only be used after birth, the MRI can be performed during antenatal development to characterize the lesion [2]. Discrepancies are often found between the suspected diagnosis and the definitive diagnosis, which appear only after an anatomopathological examination is conducted. The objective of this study was to compare data from prenatal imaging (US and MRI), postnatal radiographic CTs, and anatomopathological analyses to specify the sensitivity and specificity of these exams in case of antenatal diagnoses of thoracic lesions.

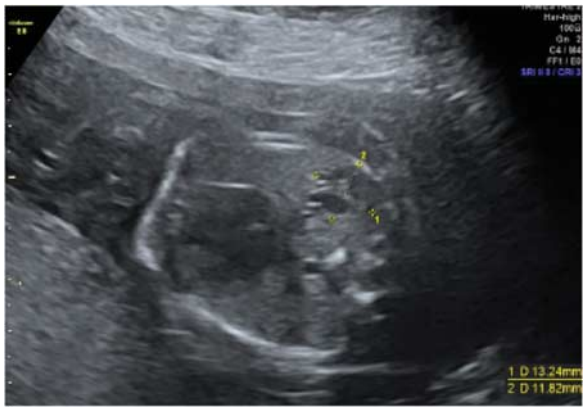
Materials and methods

Patients receiving a prenatal diagnosis of a pulmonary malformation by means of prenatal US were discussed in a multidisciplinary meeting (with pediatric surgeons, obstetrician, geneticist, prenatal radiologists and pediatricians) to determine the most appropriate treatment. A prenatal MRI was proposed between the 28th and 30th week of amenorrhoea. In case of a strong suspicion of a congenital cystic adenomatoid malformation (CCAM), a second MRI was performed 4 weeks later to evaluate the dynamics of the evolution. Patients were examined in a consultation by a paediatric thoracic surgeon who led the treatment of the patient. Thus, patients were subjected to at least one US by an ultrasonographer referred by the prenatal diagnostic centre, one prenatal MRI, and one CT with contrast agent at 1 month of life.

The study was conducted retrospectively, and the records of patients who had undergone operations for congenital pulmonary malformations with a prenatal diagnosis were

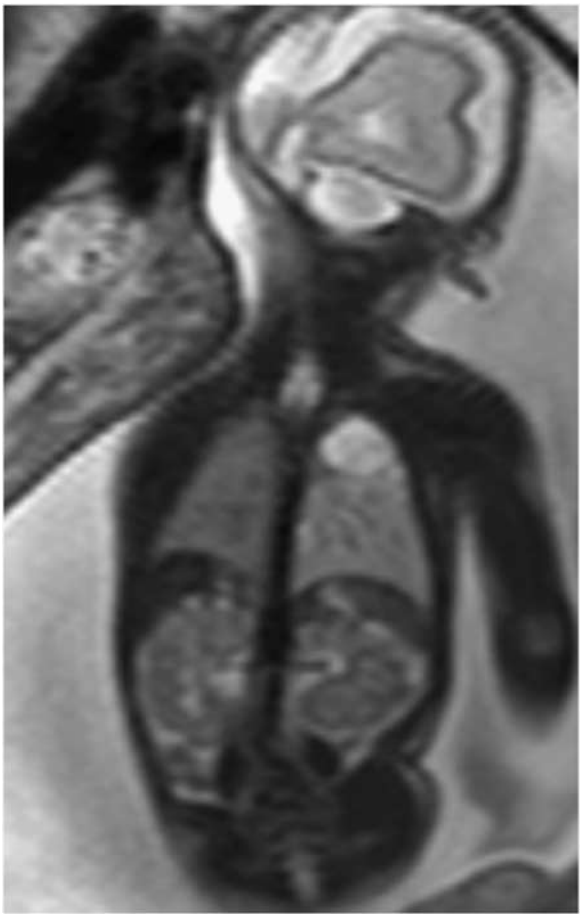
1687-4137 © 2016 Annals of Pediatric Surgery DOI: 10.1097/01.XPS.0000480963.75083.91

Fig. 1



Prenatal ultrasonography: congenital cystic adenomatoid malformation.

Fig. 2



Prenatal MRI: congenital cystic adenomatoid malformation in the right upper lobe.

examined. The definitive diagnosis was made on the basis of the results of anatomopathological analyses.

The following information was collected: the ultrasound characteristics of the lesions (cystic indications, echogenicity and vascularization) and the suspected diagnosis from this imaging; the MRI characteristics of the lesion and the suspected diagnosis; and the data from the postnatal CT. These data were compared with the operative report and the histological analysis of the lesion.

The study was approved by Ethics committee.

Results

From February 2007 to August 2013, 39 patients were diagnosed prenatally with congenital pulmonary malformation and underwent thoracoscopy in our unit. Twenty-two (56%) of these patients were male and 17 (44%) were female. US leading to a diagnosis was conducted on average at 23.5 weeks of gestational age (18–32, median: 22.6). The last US in the reference centre was conducted at 31.6 weeks (24–39, median: 31.7). Prenatal MRI was performed on average at 30.6 weeks (26–36, median: 32). Seventeen patients suspected of CCAM during the prenatal period underwent two MRI examinations at the end of 26.5 and at 33.2 weeks, on average. The postnatal CT was conducted at a median of 30 days of life (12–358, average 74 days). All 39 patients were subjected to at least one prenatal US by an ultrasonographer specialized in congenital lung malformation (Fig. 1). Thirty-six patients underwent a prenatal MRI with HASTE, TRUFISP and T1 sequences in three planes, without sedation or a contrasting agent injection (Fig. 2). Three patients did not undergo an examination due to refusal from the family. Thirty-seven patients underwent a CT with contrast agent at 1 month of life. One patient received a postnatal MRI due to respiratory distress related to the malformation. The others underwent thoracoscopy (four conversions) at a mean of 5.8 months (2 days–22 months; median, 5 months). After a histological analysis of the lesions, we found 18 CCAMs, nine pulmonary sequestrations, six hybrid lesions (associated with CCAM and pulmonary sequestration), five bronchogenic cysts, and one bronchial atresia. The diagnostic hypotheses from prenatal US and MRI and postnatal CT were compared with histological data (Table 1). We then calculated the sensitivity and specificity of the different imaging exams for each pathology using histological analyses as the gold standard (Table 2).

Discussion

Our study demonstrates that the different prenatal and postnatal imaging examinations evaluated possess an imperfect sensitivity and specificity when evaluated against anatomopathological data. These limitations strongly impact the treatment of these pulmonary pathologies diagnosed during the antenatal period. The advancement of prenatal imaging has modified the treatment of congenital pulmonary malformations. These lesions are only treated in case of complications or development of symptoms related to the pathologies. As it is possible to make a diagnosis before the onset of clinical manifestations, monitoring and/or resection are often proposed as preventative strategies. In these cases, the treatment strategy depends on the suspected diagnosis.

Prenatal US is typically the first examination conducted and permits a screening of congenital pulmonary malformations.

Table 1 Correlations between histological analyses and prenatal and postnatal imaging data

Diagnosis an pathological exam and number		Correlation with pathological finding		
		Postnatal CT	Prenatal MRI	Prenatal US
CCAM	18	17/17	10/15	17/18
Bronchogenic cyst	5	3/5	3/5	3/5
Pulmonary sequestration	9	7/9	5/9	4/9
Hybrid lesion	6	4/5	3/6	2/6
Bronchial atresia	1	1/1	0/1	0/1
Total	39	32/37	21/36	26/39

CCAM, congenital cystic adenomatoid malformation; CT, computed tomography; US, ultrasonography.

Table 2 Sensitivity and specificity (%) of the imaging techniques for several pathologies

	Postnatal CT	Prenatal MRI	Prenatal US
CCAM			
Sensitivity	1	0.66	0.89
Specificity	0.85	0.66	0.5
Bronchogenic cyst			
Sensitivity	0.6	0.6	0.6
Specificity	1	0.96	1
Pulmonary sequestration			
Sensitivity	0.77	0.55	0.44
Specificity	0.96	1	1
Complex lesion			
Sensitivity	0.8	0.5	0.16
Specificity	0.96	0.83	0.9

CCAM, congenital cystic adenomatoid malformation; CT, computed tomography; US, ultrasonography.

However, the sensitivity of this technique is rather poor for bronchogenic cysts (0.6), pulmonary sequestrations (0.6) and especially complex lesions (0.16). Prenatal US allows for an effective detection of malformations without characterizing them [3]. Our results indicate a very high specificity of this technique for bronchogenic cysts (1), pulmonary sequestrations (1), and complex lesions (0.9). Therefore, prenatal US may be sufficient upon initial diagnosis of these malformations.

Prenatal MRI possesses roughly the same sensitivity and specificity as US. The sensitivity of prenatal MRI is quite poor for bronchogenic cysts (0.6) and pulmonary sequestrations (0.55). However, MRI permits a better screening for complex lesions, with improved sensitivity as compared with prenatal US (0.5 vs. 0.16). MRI also offers an improved diagnostic specificity for CCAM (0.66 vs. 0.5), but not for other lesions. MRI may be useful principally for screening of these complex lesions that represent 20 to 40% of congenital pulmonary malformations [4,5].

Postnatal CT had the highest reported sensitivity for CCAM. However, its specificity does not exceed that of US or prenatal MRI for bronchogenic cysts, pulmonary sequestrations, and complex lesions. In contrast, it offers a greater diagnostic sensitivity for these lesions. In all cases, it appears essential to evaluate the lesion during the postnatal period and determine the most appropriate treatment [6,7].

Given our findings, upon diagnosis of a bronchogenic cyst or pulmonary sequestration during an US examination, prenatal MRI and postnatal CT may not be useful to establish the diagnosis. However, prenatal MRI and postnatal CT may be useful for evaluating the impact of the lesion on the rest of the parenchyma. Similarly, when this diagnosis is not made by means of US, but only by means of prenatal MRI, CT is not always necessary for a more accurate diagnosis. A postnatal MRI may be performed, but its usefulness must be evaluated in future investigations. However, in case of a suspicion of CCAM, the most frequently suspected lesion during the prenatal period, the three exams together allow for a more precise diagnostic approach [3].

For pulmonary sequestrations, US and prenatal MRI possess an equally weak sensitivity (0.44 and 0.55, respectively); however, their specificity is excellent (1). In contrast, CT only possesses a sensitivity of 0.77. As a result, the discovery of an aberrant artery is frequently realized only during surgery. Therefore, this malformation must be systematically sought at the beginning of the intervention, regardless of the suspected preoperative diagnosis. This exploration is particularly important in inferior lobar malformations in which frequency of this malformation is the greatest.

The three imaging techniques do not differ in their sensitivity in diagnosing bronchogenic cysts, which is uniformly poor (0.6). In contrast, their specificity is high (0.96–1). When this diagnosis is suspected, an initial surgical exploration must be conducted to better characterize the bronchogenic cyst, and determine whether a cystectomy is possible. The diagnosis is refined perioperatively (all of our patients underwent a cystectomy in these cases, avoiding a parenchymal resection). All these treatments are based on the suspected prenatal diagnoses using different imaging strategies. Surgical indications of resection are stricter in case of cystic lesions due to the risk for infection and degeneration [3,8]. The ability to make a preoperative diagnosis is important for the preoperative and perioperative treatment strategy and for informing parents. This study identifies important deficiencies in each imaging technique examined. We found that the sensitivity of prenatal US and MRI is relatively poor. Postnatal CT with intravenous contrast agents remains the examination that best optimizes sensitivity and specificity for all investigated congenital pulmonary malformations. Nevertheless, a single imaging examination is not always sufficient to make a precise diagnosis. US allows for a screening of congenital pulmonary malformations. MRI offers an improved detection of complex lesions (with CCAM +/– sequestration +/– bronchogenic cystic components), but these examinations alone are not sufficient to make an accurate diagnosis.

Regardless of the suspected prenatal diagnosis and the monitoring or proposed resection by the medical team, a complementary postnatal CT with intravenous contrast agent must be conducted. This approach allows for the most precise diagnosis and a re-evaluation of the proposed therapeutic strategy. This strategy is essential for complex lesions undiagnosed by means of prenatal imaging (due to the poor sensitivity of MRI and US for these lesions). In such cases, the CCAM or bronchogenic

cystic components must be treated thoroughly compared with the sequestration components, because of the greater frequency of complications in the former. Nevertheless, despite the reliability of CT exams, given our results, five patients out of 37 (14%) had a definitive diagnosis that differed from that made with postnatal CT imaging.

Therefore, the treatment of these patients must be discussed on a case-by-case basis, using the results of different imaging examinations to make the most accurate lesional diagnosis. However, the diagnosis cannot be certain before the excision of the lesion. This limitation must be taken into account during the prenatal consultation so as to have the most prepared discussion about the predicted evolution of the lesion and the possible need for preventative excision. Using these various imaging techniques together and by recognizing the relative strengths and limitations of each in diagnosing a range of possible pathologies, physicians will be better equipped to make the most accurate diagnosis and propose the most appropriate treatment strategy for patients.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

References

1 Gajewska-Knapik K, Impey L. Congenital lung lesions: prenatal diagnosis and intervention. *Semin Pediatr Surg* 2015; **24**:156–159.
2 Parikh DH, Rasiah SV. Congenital lung lesions: postnatal management and outcome. *Semin Pediatr Surg* 2015; **24**:160–167.
3 Conforti A, Aloi I, Trucchi A, Morini F, Nahom A, Inserra A, Bagolan P. Asymptomatic congenital cystic adenomatoid malformation of the lung: is it time to operate? *J Thorac Cardiovasc Surg* 2009; **138**:826–830.
4 Alamo L, Reinberg O, Vial Y, Gudinchet F, Meuli R. Comparison of foetal US and MRI in the characterisation of congenital lung anomalies. *Eur J Radiol* 2013; **82**:e860–e866.
5 Zeidan S, Gorincour G, Potier A, Ughetto F, Dubus JC, Chrestian MA, *et al.* Congenital lung malformation: evaluation of prenatal and postnatal radiological findings. *Respirology* 2009; **14**:1005–1011.
6 Davenport M, Warne SA, Cacciaguerra S, Patel S, Greenough A, Nicolaides K. Current outcome of antenally diagnosed cystic lung disease. *J Pediatr Surg* 2004; **39**:549–556.
7 Kotecha S, Barbato A, Bush A, Claus F, Davenport M, Delacourt C, *et al.* Antenatal and postnatal management of congenital cystic adenomatoid malformation. *Paediatr Respir Rev* 2012; **13**:162–170; quiz 170–171.
8 Hancock BJ, di Lorenzo M, Youssef S, Yazbeck S, Marcotte JE, Collin PP. Childhood primary pulmonary neoplasms. *J Pediatr Surg* 1993; **28**:1133–1136.