Global Scientific JOURNALS

GSJ: Volume 8, Issue 2, February 2020, Online: ISSN 2320-9186
www.globalscientificjournal.com

ANTENATAL DIAGNOSIS OF CYTIC LUNG ADENOMATOID MALFORMATION (CAML) ABOUT TWO CASES

GHADDAB IMEN 1, MHABRICH HOUDA 2, HAJJAJI AWATEF 1 , FALEH RAJA 1.

1 Department of Gynecology and Obstetrics, University Hospital of Monastir, Tunisia

2 Department of Radiology B, University Hospital Fattouma Bourguiba of Monastir, Tunisia

Abstract

Cystic adenomatoid lung malformation (CALM) is one of the most common lesions of lung malformations. They typically represent 25% of congenital lung lesions and 71% of pulmonary malformations diagnosed in utero. The aim of this work is to specify the interest of antenatal imagery in the positive, differential diagnosis and the prognosis of CALM, by presenting two cases of the latter supported in the departments of Radiology and Obstetrics Gynecology of the Maternity Center of Monastir. The antenatal diagnosis of CALM was made by obstetric ultrasound in the two cases. In the first observation, the discovery of this malformation was made at 24 weeks' gestation and was confirmed by magnetic resonance imaging (MRI) at 28 weeks. The ante and post natal evolution was favorable with surgical treatment at 1 year and the histology had confirmed the diagnosis, it was a type III of the classification of Stocker. Whereas in the second case, the prenatal diagnosis was made at a later stage, motivated by the installation of a severe hydramnios. The fetus carried a complicated macrocystic CALM of fetal anasrca, severe hydramnios. The evolution was marked by the occurrence of an intrauterine death (IUD) at 36 weeks' gestation.

INTRODUCTION:

Many bronchopulmonary anomalies of a malformative nature can be encountered in the newborn. They are generally the consequence of a disorder of the development of the respiratory system at a given stage of intrauterine growth.

The respiratory unit will be greatly disturbed, leading to respiratory distress charts at birth.

Antenatal ultrasound screening for these malformations is possible. It allows a care whose immediate nature at birth sometimes conditions the survival of the newborn. This shows the importance of a precise prenatal diagnosis.

One of these pathologies is congenital adenomatoid cystic lung malformation (CALM) [1].

Very rare, its prenatal diagnosis can be made from the sixteenth week of egg development [2]. Post-natal surgical excision of the lobe concerned is life-saving for the newborn. This justifies the place of imagery in the diagnosis and antenatal monitoring of fetal malformations with a view to postnatal treatment of the pathology.

We present in this work two cases of CALM supported in the departments of Radiology B and Obstetric Gynecology of the Maternity and Neonatology Center of Monastir. In this study, we will define the elements of ultrasound diagnosis and the value of MRI in the management of CALM.

Case n°1:

This is a patient aged 36 years old, primigravida, having in his family history a grandfather carrying a dextrocardia. The course of the current pregnancy was initially normal, without dysgravidie.

Early ultrasound was consistent with the term with normal nuchal translucency. A morphological ultrasound performed at 24 weeks revealed an evolutionary mono-fetal pregnancy, eutrophic for the term. She noted the presence of left pulmonary hypertrophy (Figure 1). It presented a heterogeneous echostructure of peripheral microcystic aspect with presence of a homogeneous and more hyperintense internal range. She drove her heart to the right side. The right lung appears hypointense, also pressed against the chest wall. A fetal MRI was then performed at 28 weeks gestation (Figure 2). It confirmed the presence of an intrathoracic mass syndrome that drove the heart to the right side and lowered the ipsilateral diaphragmatic cupola and kidney. The left lung was very hypertrophied and hyper-intense but heterogeneous. The coronal sequences performed in T2 SE showed two parenchymal zones of different signal and morphology: a heterogeneous peripheral zone and a more homogeneous internal zone towards which a small hypointense linear structure reminiscent of a vessel whose origin was ably aortic was directed. The residual right lung volume was 159 mm3.

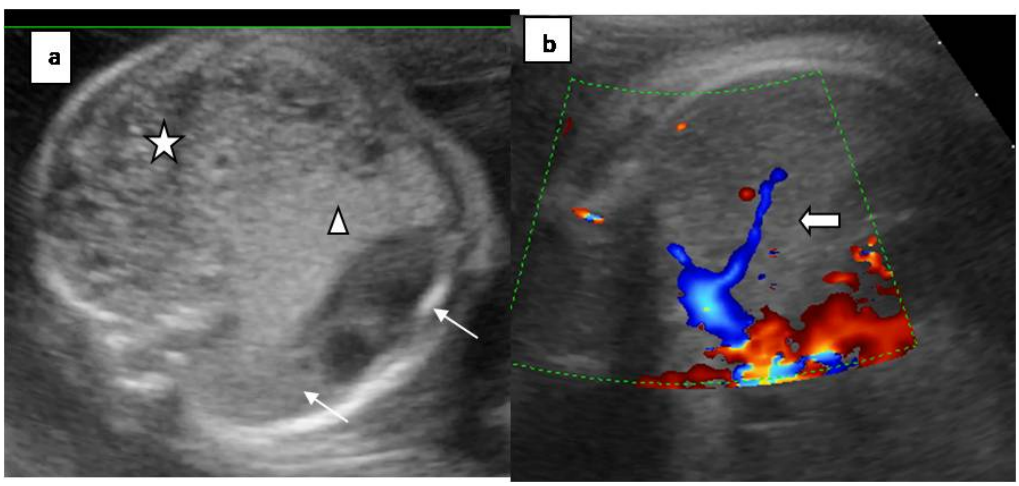

Figure 1: Obstetrical ultrasound performed at 24 W (a) and color Doppler mode (b). Left pulmonary hypertrophy heterogeneous. It is the site of a peripheral mass of micro-cystic echostructure (star ☆) and a relatively homogeneous, hyperechoic inner mass (triangle △) with systemic vascularity in the color Doppler, originating from the abdominal aorta (thick arrow). The heart and right lung are pushed back and compressed against the contralateral chest wall (arrows).

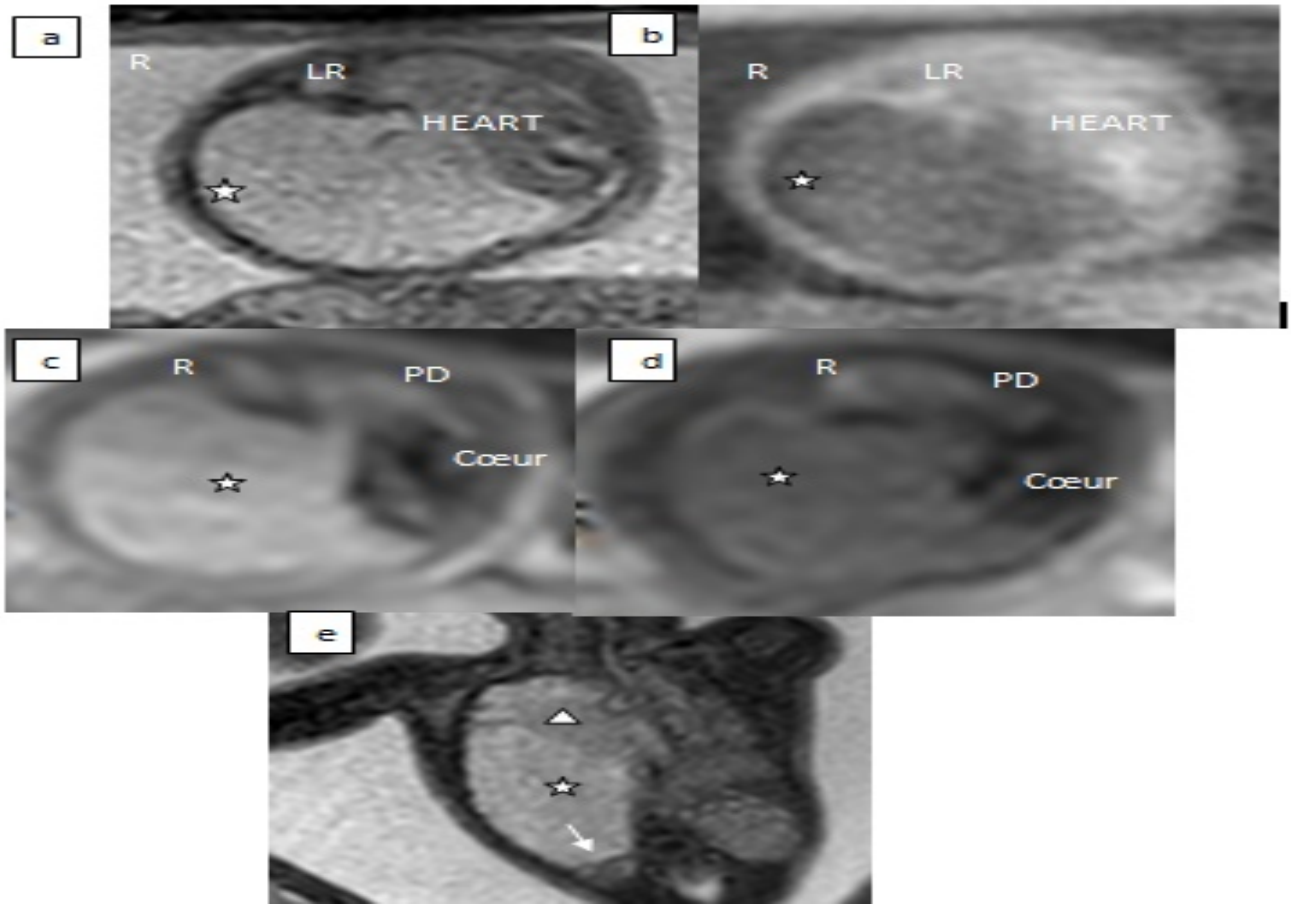

Figure 2 : Fetal MRI performed at 28 weeks' gestation in T2 (a), T1 (b), T2 (a), diffusion (c) with ADC (d) and T2 coronal views: hypertrophy left (star) hyperintense in T2, hypo intense in T1 translating the water content of this pulmonary mass. The latter pushes the heart and the right lung (RL) towards the contralateral side. The coronal section objectifies the downward movement of the diaphragmatic cupola (arrow) and the right kidney. Presence of a triangular shaped beach, in discrete hyper signal pressed against the mediastinum (triangle) In front of the presence of two thoracic malformations, a pluridisciplinary consultation meeting and after talks with the parents had decided to continue the pregnancy. Close ultrasound monitoring was performed. It aimed to monitor the vitality and fetal growth as well as the appearance of complications including cardiac and pleuropulmonary.

The evolution was marked by vaginal delivery at 38 weeks of amenorrhea and 5 days of a newborn female weighing 3250 gr, Apgar 9/10. The newborn was hospitalized in the first hour of life for mild respiratory distress with good airflow to ambient while keeping oxygen saturation at 99%.

He had undergone a thoracoabdominal CT scan that had confirmed the presence of a micro-cystic CALM associated with pulmonary squestration. The infant was operated at the age of 1 year with good subsequent evolution.

Case n°2:

It was a 34-year-old patient, G1P0, with no particular pathological history. Her current pregnancy was poorly followed. She consulted the emergency department at 33 weeks' gestation for rapid increase in the volume of her abdomen occurred in 2 days, causing her breathing. Obstetric ultrasound thus performed (Figure 3), objectified a severe hydramnios associated with a fetus with anasarca. Indeed, all these integuments are infiltrated. Bilateral pleural effusion was more marked on the right. The left lung was hypertrophied. His echostructure was heterogeneous. It was home to several cystic formations, the majority of which were voluminous. The heart was pushed back to the right side. He associated with it a lowering of the left diaphragmatic cupola. The presence of moderate ascites was also noted.

A fetal MRI was then performed at 34 weeks' gestation (Figure 5). It confirmed the fetal anasarca, the severe hydramnios. She found the bilateral pleural effusion, the left lung mass which was the seat of multiple macrocytes, the largest of which measured 5 cm long axis. The contorateral lung was 159 mm3. The evolution was marked by the occurrence of an MFIU at 36 weeks' gestation. The patient had a caesarean for presentation of the transverse with the birth of a macerated dead weighing 3200g.

©GSJ

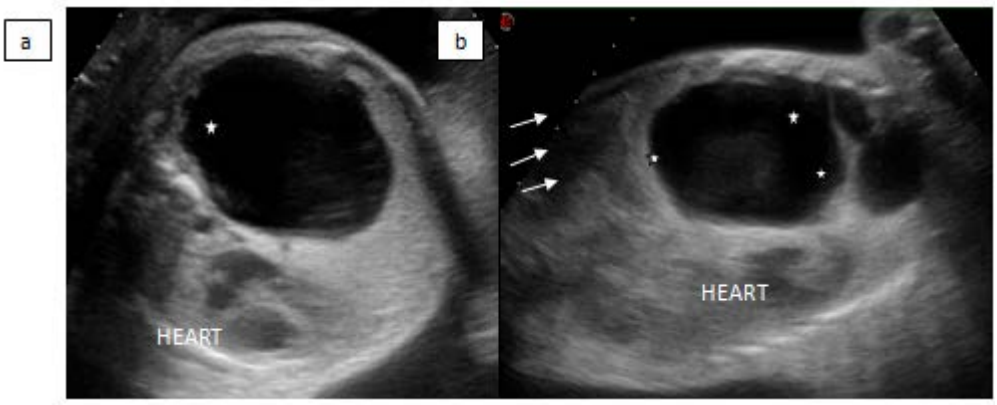

Figure 3: obstetric ultrasound performed at 33 weeks in axial (a) and coronal (b) section: Right pulmonary cystic masses (star) driving the heart towards the contralateral side and lowering the homolateral diaphragmatic dome (arrows).

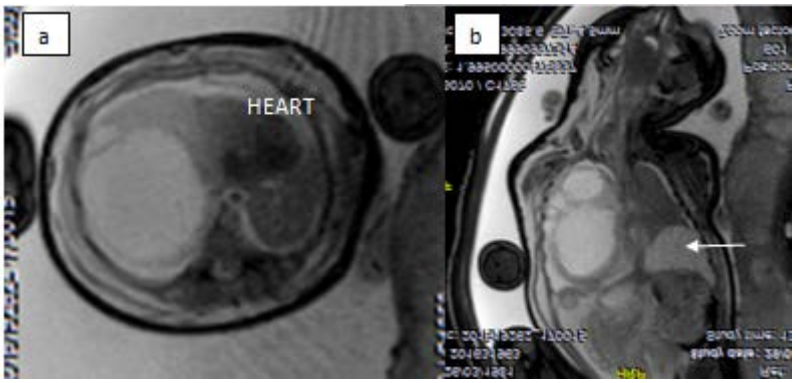

Figure 4: Fetal MRI performed at 36 weeks' gestation in axial (a) and coronal (b) section in SE T2: Right macrocystic lung mass (star) associated with a more marked pleural effusion on the left (arrow) and diffuse cutaneous infiltration (curved arrow).

DISCUSSION

Adenoid cystic malformation of the lung or adenomatoid cystic malformation (CALM) of the lung is a dysplastic pathology of the lung, consecutive to a stop of normal bronchiolar development by bronchial atresia, probably of vascular origin.

As a result, cystic dilations of the immature downstream pulmonary parenchyma occur which remains in communication with the tracheobronchial tree explaining the possible occurrence of a hydramnios.

It is a pathology whose frequency is difficult to quantify, all the cases being neither recognized nor published. The frequency would be of the order of 1 / 25,000 to 35,000 pregnancies [3]. They typically represent 25% of congenital lung lesions [4] and 71% of pulmonary malformations diagnosed in utero.

The discovery of this sporadic anomaly is generally fortuitous either, in antenatal, in 90% of the case, during the systematic ultrasound examination of the second trimester, in front of an intra-parenchymal thoracic mass, or, in postnatal, in front of distress neonatal respiratory system.

This malformation probably occurs at a later stage of embryogenesis than sequestration or bronchogenic cysts. It concerns the terminal bronchioles and the alveolar canals. Bronchiolar maturation is stopped by bronchial atresia leading to lung dysplasia. Cystic dilations of the pulmonary parenchyma then occur downstream of the cessation of maturation [5, 6]. This dysplasia is characterized by the absence of the mucus and cartilage glands and by the abundance of elastic tissue.

Lung lesions characterized, in case of CALM, by increase in volume, weight and size of the lobe concerned with presence at the cut of multiple intra parenchymal cavities of variable size which can reach several centimeters in diameter and communicating with each other. The mass is irrigated by pulmonary vessels. There is no systemic irrigation.

CALM is a hamartomatous malformation, of which Stocker proposed an anatomo-pathological classification based on macroscopic criteria [7] and which is currently the most used.

It distinguishes 3 types:

• Type I: macrocystic: Multiple cysts with a diameter greater than 2 cm or a single large cyst with small cysts surrounding it containing air and / or liquid. This type represents 50% of cases and has the best prognosis.

• Type II: Presence of numerous cysts less than 1 cm in diameter, It represents 40% of cases.

• Type III: microcystic. Small cysts less than 0.5 cm in diameter presenting as a solid-looking mass. It represents 10% of cases and has a worse prognosis.

CALM is a segmental malformation that can affect one or more lung segments, but most often it is unilobar [8]. It is conventionally more often located on the left, in the lower lobe. It is rarely bilateral. It communicates with the bronchial tree.

In our 2 observations, we found that CALM is localized in the left lung.

Concerning obstetric ultrasound: It is not always easy to classify by ultrasound the lesions of a MAKP according to the classification of Stocker.

Adzick therefore proposed an ultrasound classification into two categories [9]:

• Macrocystic: with one or more cysts whose diameter is greater than or equal to 5 mm, (therefore visible on ultrasound), of anechoic appearance. These lesions represent 59% of CALM [10]. Groups types I and II of Store. This is the case with our second patient.

• Micro-cystic: with cysts of diameter less than 5 mm, giving a hyperechoic appearance to the lesion. These lesions represent 41% of CALM [10]. Includes type III. This aspect was present in the fetus of our first patient.

It is often after a morphological ultrasound at the 22 weeks' gestation that the diagnostic hypothesis is frequently raised.

The signs of call are variable can be direct or the radiologist will then endeavor to highlight cystic masses intrathoracic [3]:

• Homogeneous, hypoechogenic masses, of variable size,

• Heterogeneous masses, associating a hyperechoic component in which there are anechoic, rounded zones, of variable size and number.

The color doppler ultrasound does not find any flux inside these cysts. He also did not find aberrant vascularization coming directly from the aorta. These direct signs can be associated with: repression of the mediastinum with deviation, or even compression of the heart and this was the case in the first observation, an eversion of the diaphragm and hydrops . Indeed, the fetus of our second patient had presented an anasarca associated with a severe hydramnios.

• Healthy lung hypoplasia.

Fetal growth is usually normal [3]. Once the positive diagnosis of CALM has been made, a complete ultrasound assessment must be carried out in search of associated malformations. These can be: renal (bilateral renal agenesis, multicystic dysplasia), cardiac (common arterial trunk, tetralogy of Fallot, interventricular communication), digestive (duodenal atresia, jejunal, anal imperforation, omphalocele) and thoracic (diaphragmatic hernia, pulmonary sequestration).

MRI provides little additional information compared to ultrasound with limited exploration of the aerated pulmonary parenchyma.

Macrocystic forms appear as a hyperintense heterogeneous lobulated mass without nourishing vessels, microcystic forms as a homogeneous lobulated mass. Mediastinal and diaphragmatic displacement are also visible [2].

In mixed forms, MRI also makes it possible to make and confirm the diagnosis of pulmonary sequestration. It appears, in its typical form, as a homogeneous triangular mass of the left posterobasal, and hyperintense. The diagnosis of certainty is made by the identification of the systemic feeding artery in MRI [2]. This aspect was observed in our first case.

About prognosis in utero, a synthesis of the review of the literature allows us to propose these lesions as elements of poor prognosis in the evolution of CALM:

-Ultrasound size of lung lesions: small lesions, microcystic according to Adzick or type III according to Stocker.

-Ultrasound localization of pulmonary lesions: bilateral pulmonary localization and multilobar localization in a single lung.

-The ultrasound volume of the lesion occupying the entire hemithorax with contralateral mediastinal deviation.

-The type of pathological image: mixed forms (cysts and areas of hyperechogenicity or pseudo-tumor forms) are of poor prognosis.

-The associated ultrasound signs which are pleural effusion, fetal ascites and hydramnios have a very poor prognosis. These various complications appeared in the fetus of the second case. An IUD then occurred at 36 weeks.

For perinatal management, when a diagnosis of CALM is made, it is necessary to carry out a careful morphological assessment in order to confirm the diagnosis, to eliminate differential diagnoses, to look for associated malformations, and to evaluate the repercussions of the malformation. on the fetus. The morphological assessment will specify the location of the lesion, its type, its size, possibly the size of the cysts. He is looking for a mediastinal deviation or compression, an eversion of the diaphragm and discreet signs of fetal decompensation.

A karyotype is carried out in principle: a few cases of chromosomal abnormalities associated by chance have been described (T21, T18) [8].

After the diagnosis, in the absence of an hydrops, it is in all cases necessary before making a decision to allow an observation period, certain lesions regressing dramatically [14], 56% according to Laberge [3].

Prenatal therapeutic indications are not frequent [15]. They concern fetuses with a macrocystic lesion with a predominant cyst, those in anasarca and major hydramnios.

Simple thoracocentesis can be a time delay maneuver. The liquid quickly re-accumulates [10].

Thor The thoracoamniotic shunt can be offered in forms with a predominant cyst, when the vital prognosis of the fetus is at stake [16, 17].

Surgical resection in utero could have been considered in early and complicated types III in some centers.

There is no indication to remove the fetus prematurely, except in the event of signs of decompensation appearing in the absence of the possibility of drainage.

About delivery, vaginal delivery has been advocated by most authors as a method of delivery. Immediate management in the postnatal period is the attitude of several authors. Therapeutic attitudes range from rehabilitation to surgical lobectomy, including intubation ...

Rapid surgical intervention is essential when the newborn presents respiratory distress (segmentectomy, lobectomy) [18]. In the other cases, a post-natal morphological assessment is carried out: chest x-ray, chest scanner. The lesion may not be seen on a standard X-ray [19, 14].

The lesion is unlikely to regress after birth [18].

If the chest scanner does not find any lesion, the child will be monitored clinically.

When the scanner finds a lesion, the attitude is more discussed [10, 20, 21]. Some teams recommend removing the lesion, even in the absence of symptoms, before complications such as infection or even cancer occur. This attitude makes it possible to intervene in healthy children, in good conditions, but it leads to the operation of children whose CALM may never have appeared. The other advantage is to be able to operate on children before the age of 2, in which the healthy lung can still develop to compensate for the loss of substance. Other teams only operate on children who have already presented complications [10].

Conclusion

Congenital cystic adenomatoid malformations of the lung are rare and generally have a good prognosis. Careful follow-up and delivery must be carried out in a tertiary center and a postnatal investigation must include clinical and radiological evaluation. Minimally invasive intervention in utero considerably improves the prognosis of fetuses with severe lesions.

© GSJ

REFERANCES

1.Berteloot L , Bobbio A, Millischer-Bellaïchea A , Lambot K, Bretona S, Brunellea F. Congenital malformations of the lung, the radiologist's point of view. Respiratory Disease Review 2012 ; 29: 820—835.

2. Hourrier S, Salomon L.J, Bault J.P, Dumez.Y . Prenatal diagnosis and management of foetal lung lesions. Respiratory Disease Review 2011;28:1017—1024.

3. Laberge JM and al. Outcome of the prenatally diagnosed congenital cystic adenomatoid lung malformation: a Canadian experience. Fetal diagn ther 2001;16 : 178-86.

4. Dommergues M and al. Congenital adenomatoid malformation of the lung : when is active fetal therapy indicated. Am J Obstete Gynecol 1997; 177: 953-8.

5. Berrocal T, Madrid C, Novo S and al. Congenital anomalies of the tracheobronchial tree, lung, and mediastinum: embryology, radiology, and pathology. Radiographics 2004;24:17

6. Wilson RD and al. Cystic adenomatoid malformation of the lung: review of genetics, prenatal diagnosis, and in utero treatment. Am J Med Genet A 2006;140: 151—5.

7. Stocker JT, Madewell JE, Drake RM. Congenital cystic adenomatoid malformation of the lung: Classification and morphologic spectrum. Humain Pathologie 1977; 8:155-71.

8. Duncombe GJ, Dickinson JE, Kikiros CS. Prenatal diagnosis and management of congenital cystic adenomatoid malformation of the lung. Am J Obstet Gynecol 2002;187: 950-4.

9. Adzick NS, Harrison MR, Crombleholme TM, Flake AW, Howell LJ. Fetal lung lesions; management and outcome. American Journal of Obstetric and Gynecology 1998;179;884-9.

10.Katarzyna GK, MRCOG, Lawrence I. Congenital lung lesions: Prenatal diagnosis and intervention Obstetrics and Fetal Medicine. Seminars in Pediatric Surgery2015; 24:156–159.

© GSJ

11. Vergnes P. Antenatal diagnosis of lung malformations: A propos of 9 cases reports. Chir Pediatric 1989; 30:185-92.

12. Haddon MJ, Bowen A. Bronchopulmonary and neurenteric forms of foregut anomalies. Imaging for diagnosis and management. Radiol Clin North Am 1991;29:241—54.

13. Stovin PG. Early lung development. Thorax 1985;40:401—4.

14. Dommergues M., Mandelbrot L, Aubry M.C, Aubry JP, Dumez Y. Adenomatoid formations of the lung. Médecine fœtale 1992;11:15-6.

15. Higby K, Melendez BA, Heiman HS. Spontaneous resolution of nonimmune hydrops in a fetus with a cystic adenomatoid malformation. J perinatol 1998;18: 308-10.

16. Bruner JP, Jarnagin BK, Reinisch L. Percutaneous laser ablation of fetal congenital cystic adenomatoid malformation: too little, too late?. Fetal Diagn Ther 2000;15: 359-63.

17. D'Ercole c , Lévy-Mozziconaccli A, Shojai R , Piéchon L,Bretelle R, Boubli L. Recent progress in prenatal diagnosis: what remains of invasive fetal samples?. 2000.

18.Mackenzie TC, Guttenberg ME, Nisenbaum HL, Johnson MP, Adzick NS. A fetal lung lesion consisting of bronchogenic cyst, bronchopulmonary sequestration, and congenital cystic adenomatoid amlformation : the missing link ?. Fetal diagn ther 2001;16: 193-5.

19. Cha I, Adzick NS, Harrison MR, Finkbeiner WE. Fetal congenital cystic adenomatoid malformations of the lung : a clinicopathologic study of eleven cases. Am J Surg Pathol 1997; 21: 537-44.

20. Huang YH. Prenatal diagnosis of congenital cystic adenomatoid of the lung: four cases report. Changgeng Yi Xue Za Zhi 1997;20:207-13.

21 .Keidar S, Ben-Sira L, Weinberg M, Jaffa AJ, Silbiger A, Vinograd I. The postnatal management of congenital adenomatoid malformation. ISR Med Assoc Journal 2001;3:258-61

22. Entezami M, Halis G, Waldschmidt J, Opri F, Runkel S. Congenital cystic adenomatoid malformation of the lung and fetal hydrops, a case with favourable outcome. Europen Journal of Obstetric Reproduction and Biologie 1998;79: 99-101.

23. Mahle WT . Echographic evaluation of the fetus with congenital cystic adenomatoid malformation. Ultrasoun obstet gynecol 2000;16: 620-4.

© GSJ

24. Maria D.C.DF. Antenatal ultrasound diagnosis of adenomatoid cystic malformation of the lung. Thesis for obtaining the ultrasound certificate 1986-1987:5-8.

© GSJ