Sonography with color and pulsed Doppler in fetal pulmonary malformations

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Résumé

Apport de l’échographie avec Doppler couleur et pulsé dans les malformations pulmonaires du fœtus

J Radiol 2007;88:269-76

Objectif. Evaluer l’intérêt du Doppler couleur et du Doppler pulsé pour le diagnostic et le pronostic de ces malformations.

Matériels et méthode. Il s’agit d’une étude rétrospective de 24 fœtus explorés en échographie pour malformation pulmonaire. Il a été réalisé une classification en quatre groupes basée sur la présence de kystes et l’échogénicité de la lésion, puis une analyse des enregistrements vidéo comportant du Doppler avec une grille de lecture : vaisseau afférent, cartographie couleur, forme du spectre (vaissseau afférent et au sein de la malformation) afin de déterminer l’apport du Doppler au sein de chaque groupe.

Résultats. Quatre lésions étaient avasculaires : kystes bronchogéniques (3 cas), séquestrations sous diaphragmatiques (1 cas). Les autres lésions étaient vascularisées et le Doppler couleur a déterminé l’origine de la vascularisation alimentant la malformation : branches de l’artère pulmonaire dans 13 cas (8 maladies adénomatoïdes kystiques, 2 artérioses et 3 obstacles de l’axe aérien) ou de l’aorte dans 7 cas (séquestrations pulmonaires) avec retour veineux pulmonaire : 6 cas (séquestrations intra lobaires) ou retour veineux systémique : 1 cas (séquestration extra lorale). L’enregistrement au Doppler pulsé dans la malformation (6 cas) a complété le Doppler couleur en montrant des spectres de forme différente selon l’origine aortique ou pulmonaire du flux. La cartographie couleur était harmonieuse dans 3 cas (séquestration pulmonaire normale), et hétérogène dans 8 cas (dysplasies parenchymateuses). De plus, l’intensité de la perfusion était corrélée à l’évolution des lésions.

Conclusion. Le Doppler couleur et le Doppler pulsé permettent un diagnostic plus précis de ces malformations et semblent apporter des éléments pronostiques.


Abstract

Objective. Evaluate the advantages of color Doppler and pulsed Doppler in the diagnosis and prognosis of fetal pulmonary malformations.

Materials and method. This retrospective study of 24 fetuses explored pulmonary malformations using sonography. A classification into four groups was made based on the presence of cysts and the echogenicity of the lesion, then analysis of video recordings including Doppler with an interpretation chart: afferent vessel, color chart, spectrum shape (afferent vessel and within the malformation) to determine the contribution of Doppler within each group.

Results. Four lesions were avascular: bronchogenic cysts (three cases), subphrenic sequestration (one case). The other lesions were vascularized and color Doppler determined the source of vascularization feeding the malformation: branches of the pulmonary artery in 13 cases (eight cases of cystic adenomatoid disease, two cases of atresia, and three airway obstacles) or the aorta in seven cases (pulmonary sequestrations) with pulmonary venous return, six cases (intralobar sequestrations), or systemic venous return, one case (extralobar sequestration). The pulsed Doppler recording in the malformation (six cases) completed the color Doppler examination by showing the different spectrum shapes according to the aortic or pulmonary source of flow. The color chart matched in three cases (normal pulmonary segmentation) and was heterogeneous in eight cases (parenchymatous dysplasia). Furthermore, perfusion intensity was correlated with lesion progression.

Conclusion. Color Doppler and pulsed Doppler provided a more precise diagnosis of these malformations and seems to contribute prognostic information.

Key words: Color Doppler. pulsed Doppler. pulmonary malformations. prenatal diagnosis.


Bronchopulmonary malformations are rare (estimated at 0.3 per 1,000 pregnancies) but can easily be screened with sonography during the second trimester of pregnancy. Second-intention sonography, aiming not only to specify the diagnosis but also to determine prognostic factors, can be useful. Although most malformations spontaneously regress during the third trimester, some of them will progress and present complications (pulmonary hypoplasia, anasarca, and hydramnios), making it important to screen for these forms since they may need in utero treatment (drains). The second reason for screening is to specify the type of malformation to ascertain whether systemic vascularization is present, sometimes difficult to demonstrate postnatally, so as to guide later surgical management if necessary.

Abbreviations :

DC : color Doppler
DP : pulsed Doppler

Our study investigated the records of these malformations with a double objective:
- establishing a classification based on sonographic data;
- studying these malformations using color Doppler and pulsed Doppler to evaluate the contribution these two sonographic tools in the diagnosis and prognosis of these malformations.

Material and methods

Patients
This was a retrospective study conducted between 1999 and 2002 on 24 fetuses explored using sonography for the discovery of a bronchopulmonary malformation. All the examinations were done in the pediatric radiology department (University Hospital of Rouen, France, in the prenatal consultation unit). The inclusion criteria were the following:
- sonographic signs: pulmonary cystic malformations and parenchymal hyperechogenicity;
- presence of video recording including CD±PD study;
- known final diagnosis, based either on anatomical data for the children who were operated on or who died, or the comparison of prenatal data and postnatal imaging follow-up for the others.

Clinical data
The clinical data were studied retrospectively from the mothers’ records (obstetric records) and the newborns’ records (pediatric records):
- prenatally: the presence of a progressing or regressing lesion, the existence of hydramnios or anasarca, specification of whether in utero treatment was needed (puncture of amniotic fluid);
- postnatally: the existence of transitory neonatal respiratory distress (and whether respiratory assistance was required) or severe neonatal respiratory distress responsible for neonatal death as well as the possible presence of pneumothorax.

The final diagnosis was retained based either on the anatomical data for infants who were operated on (12 cases) or those who died (five cases) and on the prenatal and postnatal imaging data for the others (seven cases).

Technique
All sonographic images were taken with a new-generation device (Acuson Sequoia 512) by two operators.

The healthy pulmonary parenchyma as well as the malformation were studied for each fetus in mode B completed by a study of the mediastinum and the malformation using color Doppler.

Pulsed Doppler was used only to study the spectrum shape (given that angle correction was not systematic, flow speed measurement was not retained). All in all, eight recordings were analyzed, four of which corresponded to the discovery of systemic vascularization.

Analysis
Our studies proceeded in four steps. The first consisted in studying these malformations using standard sonography, which allowed us to classify them into four groups according to the signs and patterns identified:
- simple cysts;
- hyperechogenicity associated with anechogenic images at the hilum;
- pulmonary cystic masses (echogenic and cystic components);
- isolated pulmonary hyperechogenicity. In the second step, we studied the anomalies using color and pulsed Doppler based on video recordings (using an interpretation chart):
  - Vascularization:
    - Avascular lesions: was there a relation with the vascular structures?
    - Vascular lesions: what was the source of the vascularization feeding the malformation the aorta (systemic) or the pulmonary artery. When the source was systemic, was the venous return theazygos vein (systemic) or the pulmonary vein?
  - Analysis of color flow mapping:
    - Comparison of the density in the malformation in relation to the healthy lung.
    - Distribution: homogenous or heterogeneous.

In eight cases, pulsed Doppler spectrum shape was studied at the afferent vessel and within the malformation (wide window). The third step evaluated the diagnostic and prognostic contribution of the Doppler examination in each group.

Results
Clinical results
Of the 24 cases, 21 pregnancies went to term. One patient delivered prematurely at 28 weeks and two others terminated pregnancy because of the seriousness of the prognosis (one case of tracheal atresia and one case of cystic disease associated with a diaphragmatic hernia). In nine cases, we observed hydramnios: seven moderate regressive forms and two severe forms requiring repeated punctures. Anasarca was found in two cases: tracheal atresia with major mediastinal compression and one case of voluminous cystic adenomatoid disease responsible for vena caval obstruction.

Of the 22 infants born, three died early of respiratory distress: one at 28 weeks amenorrhea in a context of major pleural effusion associated with extralobar sequestration, the two others of pulmonary hypoplasia. The 19 other infants presented no respiratory distress at birth.

The final diagnoses were eight cases of isolated cystic adenomatoid disease, five cases of isolated bronchopulmonary sequestrations, three forms associating adenomatoid disease with sequestration, three cases of bronchogenic cyst, one case of bronchial atresia, one case of transitory airway obstacle, one case of tracheal atresia, one case of transitory airway obstacle (a probable mucous plug), one case of airway stenosis, and one case of supernumerary lung with compression.

Standard sonography results
The diagnoses within each group were the following:
- simple cysts: three cases (three bronchogenic cysts);
- hyperechogenicity associated with dilated bronchi: two cases (one case of bronchial atresia and one case of tracheal atresia);
- pulmonary cystic masses with both an echogenic and a cystic component: 12 cases (six cases of cystic adenomatoid disease, three bronchopulmonary sequestrations, and three combined forms);
- isolated pulmonary hyperechogenicity: seven cases (two cases of cystic adenomatoid disease, two sequestrations, one case of bronchial stenosis, one airway obstructed by a supernumerary lung, one transitory airway obstacle).
FIG. 1: Nonvascularized lesions on color Doppler.
Figure 1 shows nonvascularized lesions on color Doppler.

a Subdiaphragmatic echogenic and cystic image (cystic adenomatoid disease associated with bronchopulmonary sequestration).

b Subcarinal bronchogenic cyst clearly visualized in the concavity of the arch of aorta.

c Hilum bronchogenic cyst supported by the branches of the pulmonary artery.

FIG. 2: Visualization of the afferent vessel.
Figure 2 illustrates the visualization of the afferent vessel.

a The lesion (superficial) is fed by a branch of the pulmonary artery (cystic adenomatoid disease).

b Demonstration of a systemic vessel going toward a lesion from the inferior thoracic aorta, with diagnosis of bronchopulmonary sequestration.

FIG. 3: Visualization of venous return.
Figure 3 shows the visualization of venous return.

a Demonstration of pulmonary venous return from the lesion (intralobular sequestration).

b Demonstration of venous return toward the azygos vein, not modulated by the fetus hear beats (extralobular sequestration).

Doppler results

Color Doppler

Four lesions were not vascularized on the CD: these were bronchogenic cysts (three cases) and an extralobular sequestration associated with subdiaphragmatic adenomatoid disease (fig. 1).

The relation with the vascular structures was studied for the three bronchogenic cysts:

- The cyst in the left lower thorax was clearly visualized in contact with the aorta, with no sign of compression.
- The subcarinal cyst was found under the arch of aorta in contact with the pulmonary vessels.
- For the hilar cyst, the Doppler showed the mass supported by the proximal branches of the pulmonary artery.

The other lesions were vascularized and the color Doppler made it possible to determine the source of the vascularization in all cases, based either on the pulmonary artery (13 cases), i.e., eight cases of cystic adenomatoid disease, two cases of atresia, three cases of airway obstacle stenoses, or on a systemic vessel (seven cases), all corresponding to bronchopulmonary sequestrations (fig. 2).

For the seven bronchopulmonary sequestrations, the CD identified the venous return as pulmonary, six cases corresponding to intralobular or systemic sequestrations, and one case of extralobular sequestration (fig. 3).

Color flow mapping was heterogeneous in eight cases (five cases of cystic adenomatoid disease and three cases of bronchopulmonary sequestrations) and homogenous in three cases: one airway obstacle, one airway atresia, and one airway stenosis (fig. 4, 5).
The malformation was less perfused than in the healthy lung (nine cases), which corresponded to spontaneously regressing forms (fig. 6a). However, it was more intense in the healthy lung in one case. This was a progressive form, responsible for early neonatal death (fig. 6b).

**Pulsed Doppler**

At the proximal vessels, the shape of the spectrum was different depending on the aorta or pulmonary artery source of the flow (fig. 7).

When the malformation was fed by the aorta (seven cases), we found a wide systolic peak, followed by a positive diastolic flow. This spectrum was different from the pulmonary spectrum in its characteristic narrow peak followed by a dome (2). In all cases, the spectrum shape recorded within the malformation matched the spectrum shape found at the afferent vessel (fig. 8). Furthermore, in four cases, both types of spectrum were identified within the malformation:

- in three cases where a systemic vascularization was demonstrated, the recording showed a pulmonary spectrum at the top of the malformation;
- in one case where the systemic vessel was not identified, a systemic spectrum was found at the base of the lung. The vessel was demonstrated secondarily stemming from the celiac trunk.

In the four cases, these were shapes were associated with cystic adenomatoid disease and bronchopulmonary sequestrations (fig. 9).
Discussion

Prenatal diagnosis has revolutionized the management of bronchopulmonary malformations, until now revealed by complications in the neonatal period or in childhood (infections, hemoptysis). The only malformation that has given rise to classifications is cystic adenomatoid disease. The most frequently used of these classifications is Stocker’s, which is based on anatomical data. The main anomalies are grouped according to cyst size and the type of cells found in histology (3). This has given three groups, the first two with good prognosis and the third with a poor prognosis. Adzick simplified this classification, describing two forms according to the macro- or microcystic sonographic aspect (4). As for bronchopulmonary sequestrations, two forms are described: intra- and extralobular. The first shares envelopes and venous return with the lung in which it is lodged, whereas the extralobular form presents a pleural envelope of its own and a venous return toward the azygos vein.

However, the pathogenesis of these malformations remains poorly known, and many authors suggest a separate explanation for each type of malformation. Yet these anomalies are frequently found together, as evidenced by the classical association of cystic adenomatoid disease and bronchopulmonary sequestration (5-11) and the association of three types of malformation (bronchogenic cyst, cystic adenomatoid disease,
and bronchopulmonary sequestration) has also been described (12). The hypothesis of a single bronchovascular anomaly as the source of these malformations, as Clements and Warner suggest, is appealing (13). They believe the cause may be a lesion at the extremity of the bronchial tree, with a variable etiology, stemming from a localized injury, ischemia, or an infection. Furthermore, it is not only the type of damage, but first and foremost the date of onset and the severity that determine the morphology of the lesion.

This is clearly a spectrum of malformations as described by Achiron et al. (14), going from the normal lung vascularized by normal or abnormal vessels to the abnormal lung, i.e., the dysplastic lung vascularized by normal or abnormal vessels. We therefore decided to classify these defects based on their signs and patterns so as to compare the sonographic and Doppler data for diagnosis within each group.

Simple cysts corresponded to bronchogenic cysts and color Doppler was valuable in specifying the relation with vascular structures and the cysts’ topography. As for the hyperechogenicity associated with anechogenic images of an oblong area of the pulmonary hilum, standard sonography also diagnosed atresia in the two cases. The patterns were typical, associating distended and hyperechogenic lungs with anechogenic images that were lengthened at the hilum corresponding to the bronchi. These images showed airway dilatation caused by obstruction of pulmonary drainage upstream of the obstacle. Moreover, color Doppler was valuable in the case of tracheal atresia, because it confirmed that the mediastinal fluid structures were not vascularized but followed the branches of the pulmonary artery, which was later confirmed by the lung spectrum recording. A similar observation was found in the literature, describing increased arterial and pulmonary flow on color Doppler. This allowed the author to make the differential diagnosis with type III cystic adenomatoid disease (15).

The color flow topography of the airway atresia was studied, revealing regular arborization of the malformation in a normal pulmonary segmentation. In addition, the color mapping of the lung was less intense than on the side of the healthy lung, probably because of the hyperpressure.

However, the diagnosis was not always based on standard sonography data for the cases of cystic pulmonary disease and the cases of parenchymal hyperechogenicity; we therefore evaluated the utility of Doppler in these two groups in particular, which will be discussed together because they share the same diagnoses.

In the majority of cases, color Doppler made it possible to determine the source of the vascularization feeding the malformation, whether pulmonary branches or branches stemming from the aorta were involved. A slow transversal sweeping of the fetus, centered on the aorta, was used to search for a vascular structure going directly toward the malformation (16-18). Once this had been identified, it was easier to lengthen the search in the coronal or oblique sagittal plane. This technique was very sensitive: the only case not diagnosed with the Doppler was an extra-lobular subdiaphragmatic sequestration for which no abnormal vessel was detected using Doppler postnatally or during surgery. The diagnosis was made only when the anatomical abnormality was localized and visualized.

Doppler also contributed to the search for a systemic venous return. This was demonstrated in one case as a slow continuous venous flow moving toward the azygos vein. Contrary to the pulmonary venous flow, this flow was not modulated by the fetus’s heart contractions (19).

Pulsed Doppler was also valuable to distinguish the source of the flow within the malformation, notably in cystic adenomatoid disease associated with bronchopulmonary sequestration. This was observed in three cases of bronchopulmonary sequestration whose systemic vessel had been identified with color Doppler from the aorta. Pulsed Doppler of this systemic vessel showed a low-resistance arterial flow each time (flow speed was not taken into account because there was no angle correction). The flow recording within the malformation showed a different spectrum with characteristics of pulmonary flow and a high resistance index. In these three cases, this was indeed cystic adenomatoid disease associated with bronchopulmonary sequestration.

Recognizing these two types of spectrum can therefore identify the source of vascularization within the malformation, even if the systemic vessel was not previously identified on color Doppler. This was confirmed in the case where the lesion was highly vascularized on color Doppler: in the lower part of the malformation, the spectrum showed an arterial source with low resistance, whereas in the upper part, it was resistant and pulmonary. The systemic vessel was then identified with color Doppler. The spectrum shape was identical to that recorded within the malformation.

Another interesting aspect concerned color mapping of the malformation. All cases of pulmonary cystic disease presented heterogeneous flow maps, clearly contrasting with the homogenous mapping of the healthy ipsilateral or contralateral lung. This allowed us to suggest the hypothesis of parenchymal dysplasia showing up on the color Doppler as chaotic vascularization. On the other hand, homogenous vascularization may indicate normal bronchial tree arborization and therefore the absence of parenchymal dysplasia.

Based on this hypothesis, analysis of color flow mapping contributed additional information to the two cases of parenchymal hyperechogenicity:

- One presented a pulmonary source of vascularization, with homogenous distribution, as in the case of airway atresia, but with no dilated hilum fluid image. This raised the suspicion of airway stenosis, which was confirmed during surgery.

- The other presented low perfusion, but since it seemed regular, it suggested an airway obstacle. This was indeed an airway obstacle caused by a supernumerary bronchus compressing the origin of the upper lobe, diagnosed postnatally during fibroscopy.

In terms of prognosis, congenital lung malformations were considered serious lesions in the neonatal period, varying from 60% to 80% in older series. The prognosis seems to be much better in recent series (20), with spontaneous regression of the lesions in more than 50% of cases in the third trimester (21-24). In this study, we report 79% of survivors when combining all causes, the vast majority presenting no respiratory distress at birth.
Several authors have taken an interest in the physiopathological aspect of this phenomenon:

• In cystic adenomatoid disease, it may be a relative reduction in the secondary lesion when its growth stops, whereas the healthy lung pursues normal growth (25). The initial, very rapid growth of the mass may outstretch the vascular capacity and may then be followed by spontaneous involution.

• In bronchopulmonary sequestration, the possibility of thrombosis or fibrosis of the systemic vessel responsible for an infarction of the sequestrated segment, and therefore a reduction in the lesion’s size has been suggested (26). The possibility of torsion of the vascular pedicle has also been suggested in cases of extralobar sequestration (27). Smulian’s description of spontaneous resolution of a bronchopulmonary sequestration with progressive reduction in the size followed by vascularization as seen on color Doppler does not confirm this hypothesis (21).

All in all, the hypotheses are varied but are all based on a vascular source. In our study, the analysis of the color flow mapping of the malformations was very useful. Indeed, we observed that the lesions that were only slightly vascularized regressed spontaneously. Only one lesion presented intense vascularization, greater than in the healthy lung. This was a lesion progressing with the pregnancy with an unfavorable outcome because of contralateral lung hypoplasia. This suggests that the lesions’ potential for progression may parallel their vascularization. This hypothesis is particularly interesting, because it may mean that hypervascularized lesions need to be followed to search for signs of decompensation (anasarca and hydramnios), which may require in utero treatment. However, this is the first observation of this nature and a study on a larger scale should be conducted to verify this hypothesis.

Conclusion

Color Doppler and pulsed Doppler contribute additional diagnostic arguments compared to standard sonography. They have identified the source of aorta or pulmonary artery flow feeding the malformation as well as the venous return. Color flow mapping is also valuable in predicting the existence of parenchymal dysplasia. Overall, the prognosis of these malformations is good in the majority of cases, with favorable progression in the third trimester. Perfusion intensity seems related to the natural progression of these lesions. This remains to be confirmed by a prospective study.

References

