

# Fetal lung lesions diagnosis: the crucial role of ultrasonography

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Fetal lung lesions may cause significant effects of mass may evolve into a non-immune hydrops and lead to the death of the fetus or the child (1). Treatment options for these severely affected infants are constantly evolving. The widespread use of ultrasound in prenatal diagnosis, in tertiary center like ours, allows us to identify the fetus, including lung lesions more 'small. Prenatal diagnosis and possible therapeutic intervention in the immediate prenatal or postnatal period has significantly changed the quality of life (14) (15) (20) and the survival of fetuses and infants, especially those who were completely asymptomatic at birth (2) (13).

Object of our interest is the pulmonary sequestration and congenital pulmonary malformation is the second in order of frequency, with an incidence between 0.15% and 6.4% of cases (3).

## Definition

The pulmonary sequestration is a rare congenital malformation characterized by the formation of an island of anomalous non-ventilated lung tissue, and therefore non-functional, that is not connected with the tracheo-bronchial tree and an abnormal arterial blood supply, supplied by the systemic circulation ( thoracic or abdominal aorta) rather than the lung (4) (5) (17). The vase abnor-

mal traction during the development of the lung, capture or "seizure" of this body suddenly separated from the lobe. It thus forms an area in which the abnormal alveoli do not come to differentiate themselves and observe the appearance of a single or multilocular cysts (4, 5).

## Classification (6)

We distinguish two main types of seizure:

- a) intralobar sequestration
- b) extralobar sequestration

## Embryogenesis

In fact in order to understand this breakdown, one must know the origin embryological tissue polmonare. Tra the 5th and 6th week of embryonic development known as "lung bud" originates from the ventral side of the primitive upper intestine (foregut) and from that lung bud, with a series of divisions, originates the tracheo-bronchial tree (4, 6).

The hypothesis that, in fact, explain the origin of pulmonary sequestration are two:

- From primary laryngo-tracheal tissue was originally a supernumerary lung bud, so that the seizure would occur simultaneously with the smooth development of the lung parenchyma (4, 6).
- Alteration of the normal process of development of lung parenchyma in the absence of a supernumerary bud or ancillary. In this case, an island of lung tissue would develop independently of tracheo-bronchial tree, but arise from it (4, 6).

It 'obvious, then, as the period in which embryological origin of the defect is important for the genesis and the type of pulmonary sequestration. If it happens before the formation of the pleura, it remains close to the lung tissue and is covered by the pleura, causing seizure "intralobar." If you place after the formation of the pleura causing the seizure "extralobar." Moreover, the cells that make up the extralobar pulmonary sequestration can migrate at points distant from the tissue of origin, reaching close to the diaphragm or below that. This type of dissemination of the islands explain how pulmonary sequestration can enter into relationships with lung and gastrointestinal organs have their own independent pleural lining (4, 6, 7).

Potter has proposed an alternative classification, distinguishing three varieties (6):

- Intralobar sequestration: mass of tissue in the context of a lung lobe (75% of cases) diagnosed mainly in children and adults (6) (8)
- Extralobar sequestration: mass of tissue outside the tracheo-bronchial tissue, but that shrinks relationship of

contiguity with the lung, (4) (6). Form more 'is frequently found in fetuses and at birth (8).

– Complete seizure: mass of tissue that will not create relationships with the lung parenchyma (6).

In all cases the blood supply is ensured by the primitive splanchnic vessels of the intestine above.

The INCIDENCE in both sexes is the same for intralobar sequestration, whereas the extralobar form occurs with a frequency four times higher in men than women (4).

Has not been documented familiarity (8).

LOCALIZATION of both forms is more common to load dell'emitorace of sx (60% intralobar variety; for 90% of the extralobar variety) (4).

The explanation of the most 'frequent localization all'emitorace left is that the channel is closed most primitive pleuro peritoneal late, left to right (right) (4).

Moreover, the lack of liver tissue left to provide additional space for the development of abnormal tissue (4).

The largest segment of 'hit, obviously in intralobar sequestration is the rear baseline of both lower lobes; exception is the involvement of an entire lung (4).

Seizures extralobari, however, have more 'frequently located between the left lower lobe and diaphragm, but sometimes can be localized even below it, the same diaphragm, the mediastinum and even into the pericardium . This' explains how easily extralobar sequestration is associated with ipsilateral diaphragmatic defect (about 60% of cases) (4).

As for spraying, both seizures receive their arterial supply descending aorta or one of its branches, both above and below the diaphragm (4).

In the variant is more common than extralobar sottodiframmatica spraying, where the vessel passes through the aortic or esophageal hiatus or through a diaphragmatic defect separate (4).

The variety has intralobar arteries more 'or less large size compared to the volume of tissue sprayed. (branch of the descending thoracic) (4) (7).

Price (7), in relation spraying, describes three types of intralobar sequestration based on the anatomy of the aberrant vessel:

Type 1: abnormal artery vascularized only the normal lung tissue;

Type 2: the anomalous artery vascular tissue and normal lung lobe seized;

Type 3: abnormal artery vascularized lobe only sequestered (7).

The intralobar sequestration usually drains into the blood through the pulmonary vein into the left pulmonary veins, whereas extralobar drains into the azygos vein or the portal system, establishing a left-right shunts of varying size to lead to severe heart failure (3) (7).

In fact, intralobar sequestration, while constituting 45% of cases, it will make a diagnosis in adulthood, it is made very difficult to diagnose in utero. On the other hand, the extralobar variety, although they constitute only 25% of cases, remains the variant 'commonly diagnosed during pregnancy (3).

From the perspective of ANATOMIC PATHOLOGY intralobar sequestration appears macroscopically as a well circumscribed reddish gray color, which is well distinguished from the healthy lung, but in close contact with this being located usually within the parenchyma of the posterior basal segment of either lower lobe. The extrapulmonary sequestration, however, is clearly sepa-

rate from normal lung and its surface appears shiny and homogeneous by having their own pleural covering. We have just now 'described that both seizures receive blood from the systemic arterial circulation and although this comes from' aorta, the vessels tend to have the elastic structure of a pulmonary artery rather than the muscular structure of the branches of the normal aorta. Histologically, the bronchial contents in both varieties, have ciliated lining and appear dilated with focal development of cellular tissue. The expansion stems from the retention of bronchial secretions and edema, giving quite voluminous cysts (4).

ULTRASOUND, sequestration appears as a homogeneous mass, usually hyperechoic compared to the residual pulmonary parenchyma free from well-defined contours, while the sizes vary from a few mm to 6-7 cm. The location is almost always chest. The shape is roughly spherical in intralobar sequestration, extralobar triangular one. Often, the intralobar form, there are small cystic areas in the context of tissue mass (16). Given the particular topography of the lesion, it is usually recognized at the thoracic transverse scan at the level of the heart chambers, but rather 'is displayed in the axial scan with a plan sottocardiaco, ie in the longitudinal paramedian thoracic scan (8). At the Color Doppler highlights the systemic arterial vasculature (6), which is the pathognomonic finding (3), while the mapping of the district venous flow lesions can 'demonstrate the specific features of vascular, allowing a correct prenatal diagnosis. (9). Ultrasonography, therefore, allows a diagnosis very early (even before 16 weeks) (3).

It is important to formulate the differential diagnosis of lobar sequestration CAM type III and congenital lobar emphysema and one extralobar from kidney or adrenal malformations and teratomas. (4) (6). Sometimes, it becomes divisive for the enforcement of the pregnant patient MRI (Magnetic Resonance Imaging) (3) (4).

If the seizure escaped the prenatal diagnosis, it is formulated in the early diagnosis months or years of life (7). Not been reported until now no significant association with chromosomal abnormalities (8).

### Associated malformations

Other sonographic signs that can accompany both types of seizure are (4)

- polyhydramnios (for reduced fetal swallowing) due to mechanical obstruction sull'esofago from esophageal atresia with or without tracheoesophageal fistula, esophageal duplication and / or megacolon (4);
- shift mediastinal and pleural effusion in 70% of cases (4) (17) (19);
- bronchogenic cysts (4);
- pulmonary hypoplasia especially in the seat thoracic extralobar sequestration (4);
- tricuspid atresia and severe heart disease (4);
- Hydrocephalus (4);
- diaphragmatic hernia, which alone accounts for 50% of the anomalies associated with extralobar variety (4) (8);
- polydactyly, fingers drum sticks and funnel chest (4);
- congestive heart failure (4) (6)
- non-immune fetal hydrops to etiology (6)

Heart failure is easy to explain both the initiation of significant shunts that encourage the abnormal vascular-

ization of the seizure that mechanical compression directly on the heart and great vessels.

The same heart failure may cause fetal hydrops which is the complication most 'fearful of the seizure.

According to some authors, the pathogenic mechanism dell'idrope be sought in structural changes to the media and adventitia of the veins (venous arterializzazione), which blocks the drainage, causing a transudate, responsible dell'idrope primary fetal (10) associated with the seizure.

Made by ultrasonography, the diagnosis, management provides obstetricians (6) (20):

– Accurate ultrasound examination to exclude any associated anomalies;

– Fetal echocardiography to exclude congenital heart disease and to monitor the hemodynamic changes caused by the seizure;

– Ultrasound every 2-3 weeks to assess the amount of amniotic fluid and the possible occurrence of pericardial effusion, pleural chest, lung hypoplasia or the appearance of non-immune fetal hydrops (6).

In the presence of polyhydramnios, the risk of premature delivery, is indicated given tocolitici.

And 'interesting to note that about 30% of seizures in isolation, ie without associated malformations (favorable prognostic factor) (8) shows signs of regression after the 24th week of gestation and complete remission in the third quarter (3).

Therefore, it is clear that it is extremely risky to the simultaneous occurrence of fetal hydrops and how it is already a very unfavorable prognostic factor (19). If, unfortunately, were to establish the hydrops in utero or ex utero mortality reaches 100%.

The PROGNOSIS depends on (4):

– Presence and severity of pulmonary dell'ipoplasia

– Amount of lung tissue residue

– Presence and severity of associated anomalies

– Development of heart failure until fetal all'idrope (mortality close to 100%)

– Operability of the mass.

In case of large pleural effusion and/or fetal hydrops, Weiner and other authors (8) have proposed that intrauterine intervention (18) thoraco-amniotic catheter in order to decompress the lung. Unfortunately the results were not encouraging (6).

Excellent results were obtained, however treatment with surgical resection rather than lobectomy of the segment in all cases where there is total regression of the lesion to the controls performed in the neonatal period. In both varieties, although asymptomatic (13), is decided by the second year of life, for the surgical approach for subsequent histological verification of the mass, to reduce the risk of bleeding, infection and malignancy. This 'explains the choice of lobectomy (12) instead of segmentectomy because rarely is free adjacent lung tissue (7) (8). Experienced hands (11) enter the isolation of the afferent vessel, systemic, whose section would lead to exitus child. The artery should be 'treated with care because of seizure undergoes structural changes, such as atherosclerosis, become extremely brittle (7) (10). A few brave attempt was made using the anomalous systemic artery embolization (14), but raises the rate of morbidity (fever and pain), leaving the lesion susceptible to all the risks of complications (infection and neoplasia) (7).

In conclusion, mortality is 100% in cases of dropsy and

associated decreases in the absence of associated malformations, while survival is the rule in the absence of hydrops and for those fetuses in which the lesion regresses in utero (8) (15).

After the intervention the quality of life of the unborn child is normal! (8)

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