

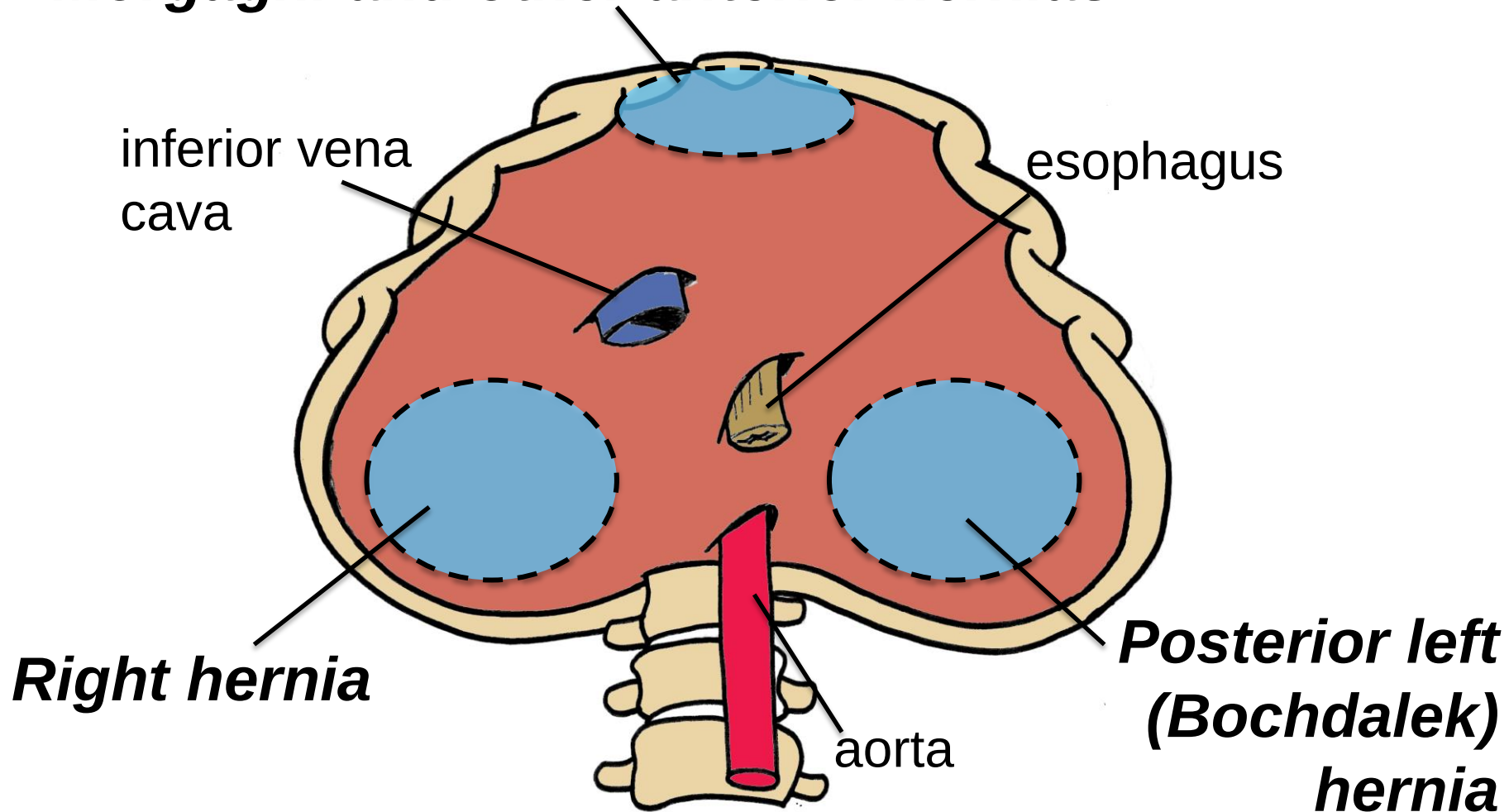
Congenital diaphragmatic hernia

The congenital diaphragmatic hernia (CDH) is a developmental defect resulting in a partial or complete absence of the diaphragm associated with lung hypoplasia.



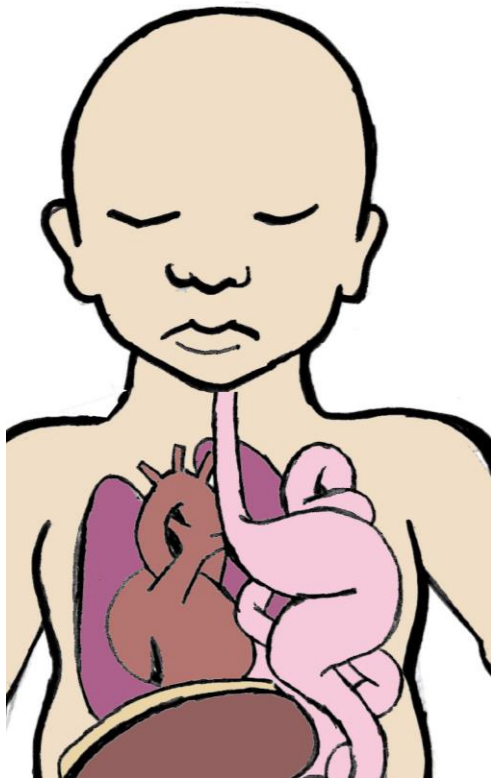
Schematic representation of the most common types of CDH

Morgagni and other anterior hernias

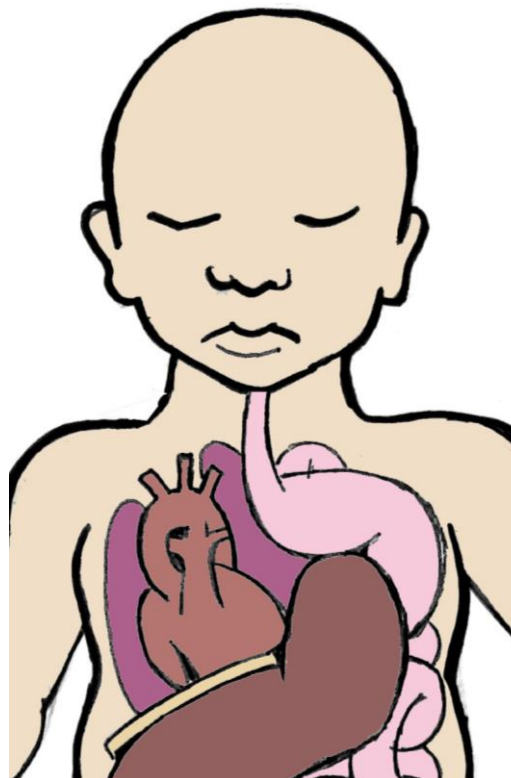


CDH and intrathoracic displacement of abdominal organs

LEFT CDH

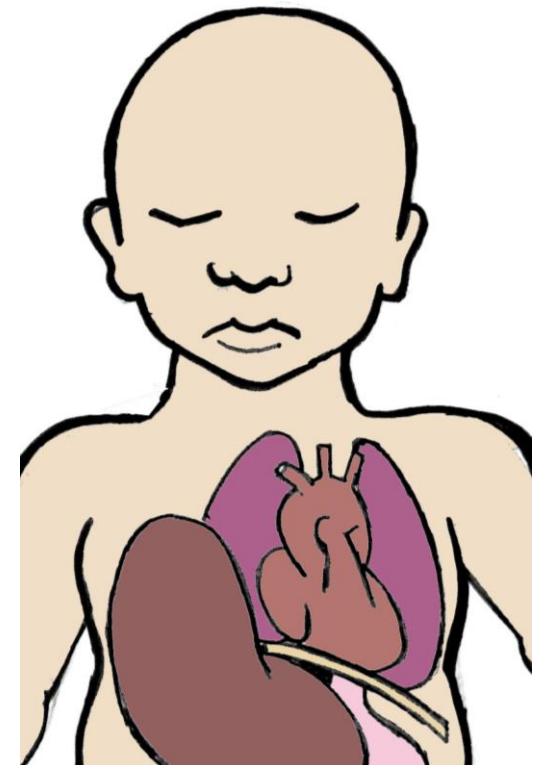


stomach, bowel



stomach, bowel,
left lobe of liver

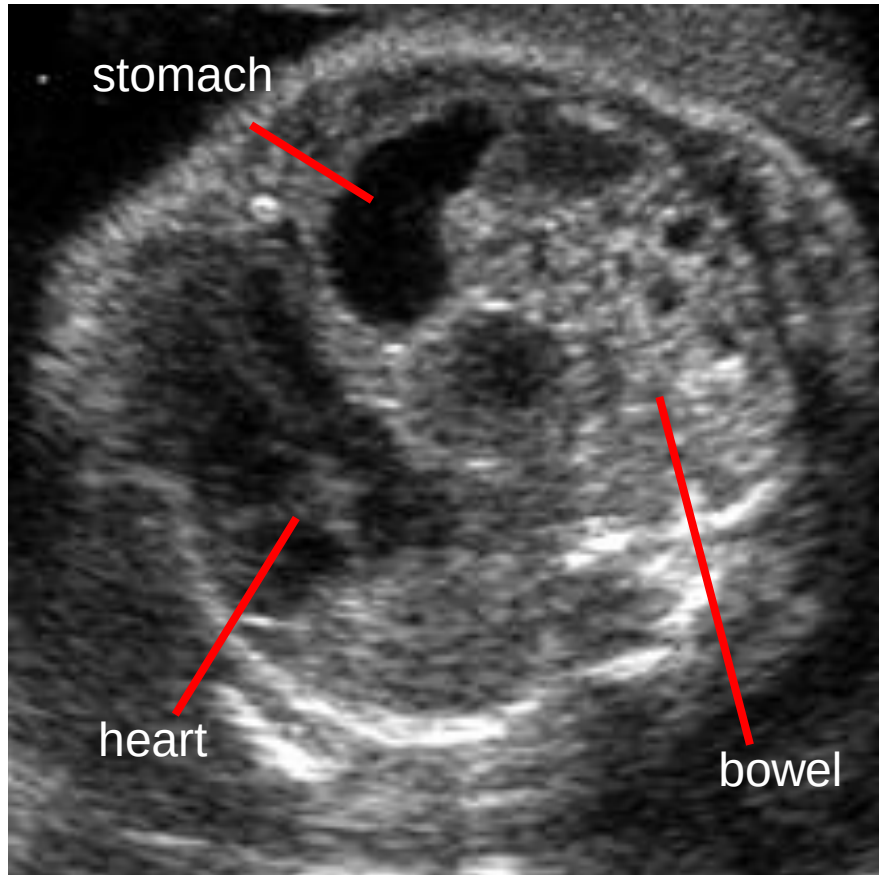
RIGHT CDH



liver

Sonographic diagnosis of left CDH

30 weeks



20 weeks



The diagnosis of left CDH is easy in the third trimester of gestation, when the bowel is distended by meconium, or in cases with intrathoracic displacement of the stomach

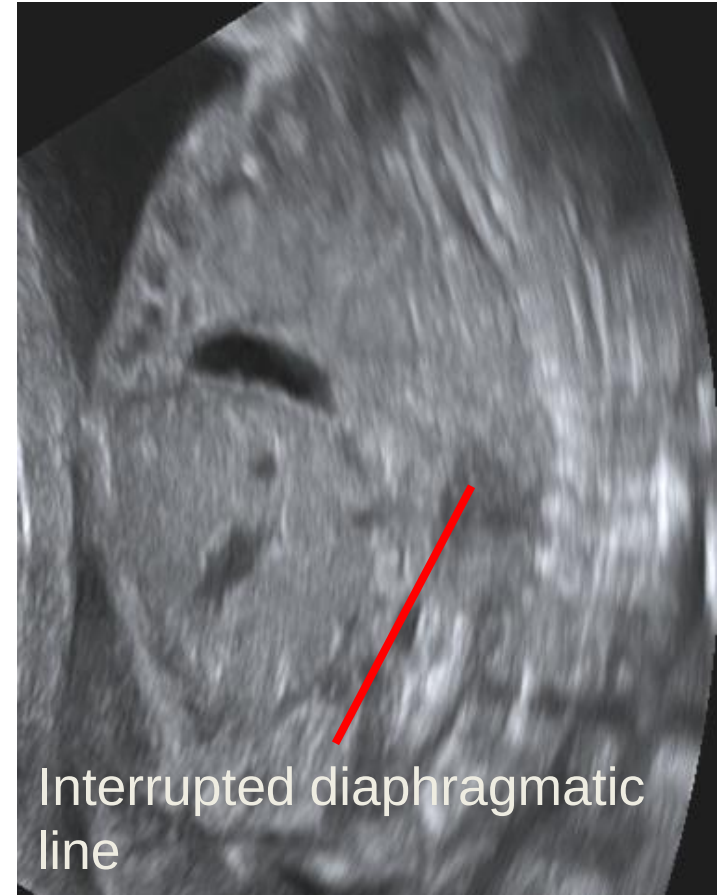
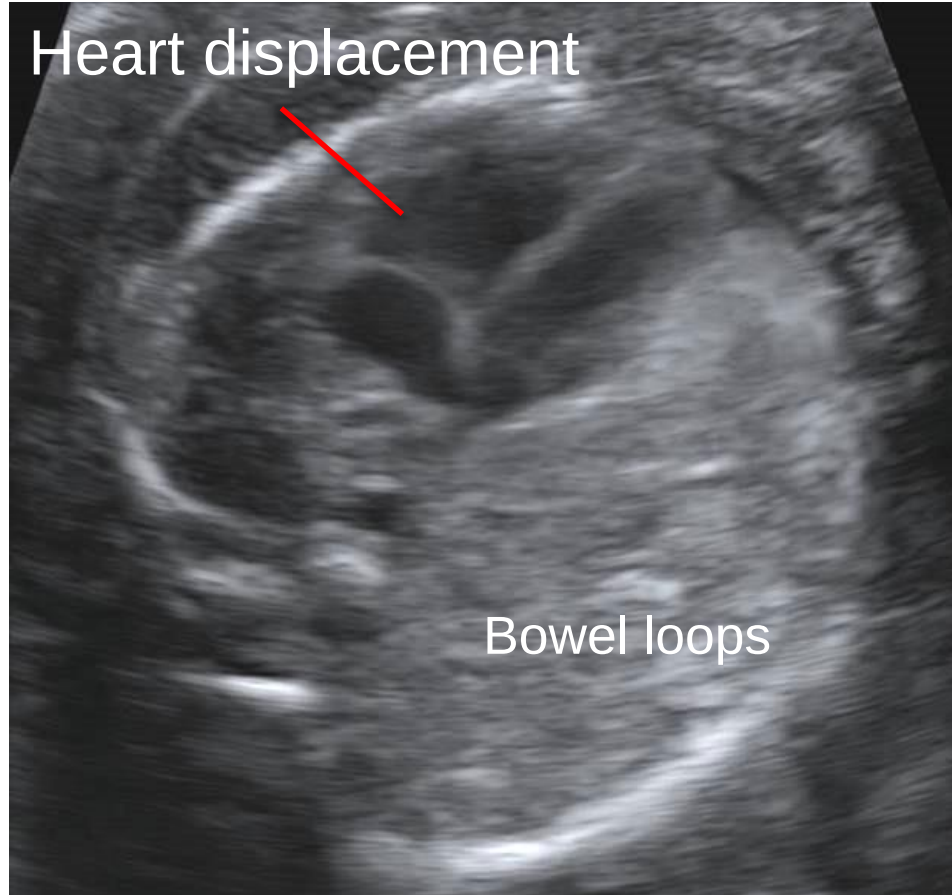
Sonography of Left-sided CDH with stomach displacement

A cross-sectional view of the fetal thorax in a left-sided CDH case characterized by a cardiac displacement to the right side and identification of the stomach into the fetal thorax.

CLIP 01



Sonography of left CDH in the second trimester without stomach displacement

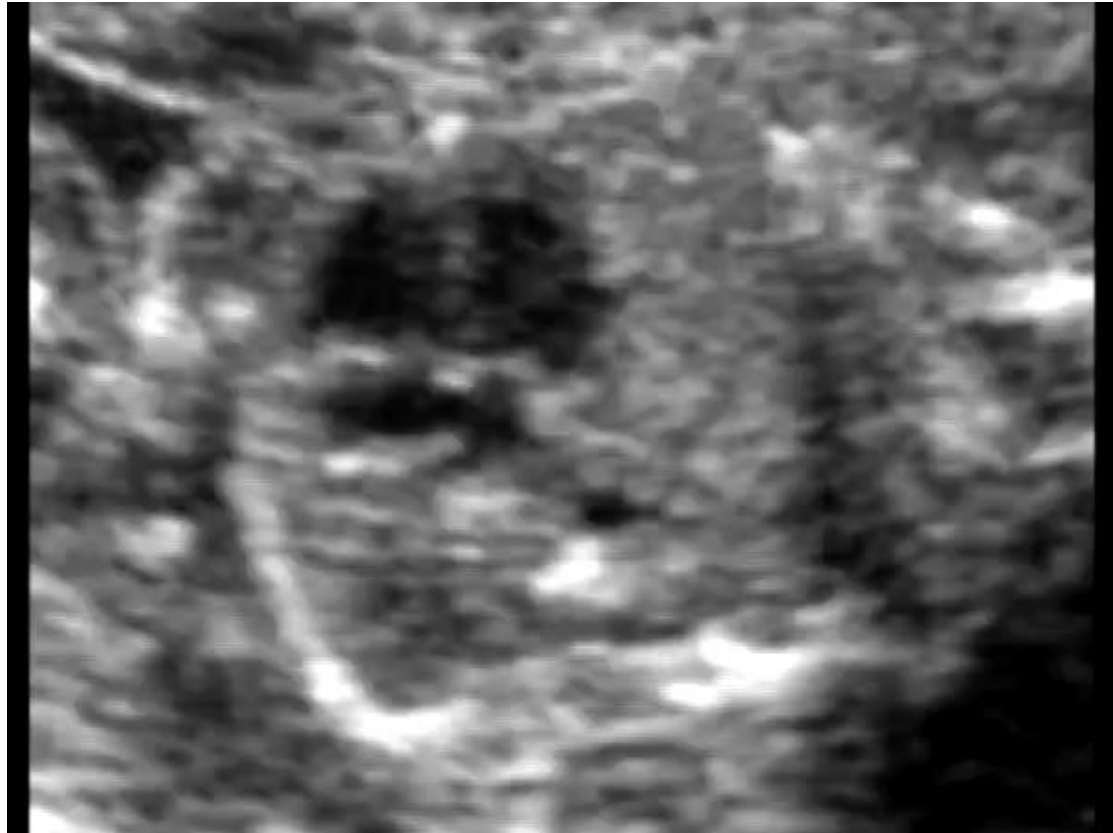


In these cases the appearance may be elusive and the main clue to the diagnosis is the dextroposition of the heart; in dubious cases a longitudinal view of the chest demonstrating the interruption of the diaphragmatic line is helpful

Left sided CDH with intrathoracic displacement of bowel

In this fetus at 20 week gestation the meconium free bowel loops have a echogenicity very similar to the lung and the main clue to the diagnosis is the displacement of the heart

Clip 22



Longitudinal views of the trunk in fetal diaphragmatic hernia

In a fetus with left CDH a longitudinal view of the trunk confirms the interruption of the diaphragm



Clip 24

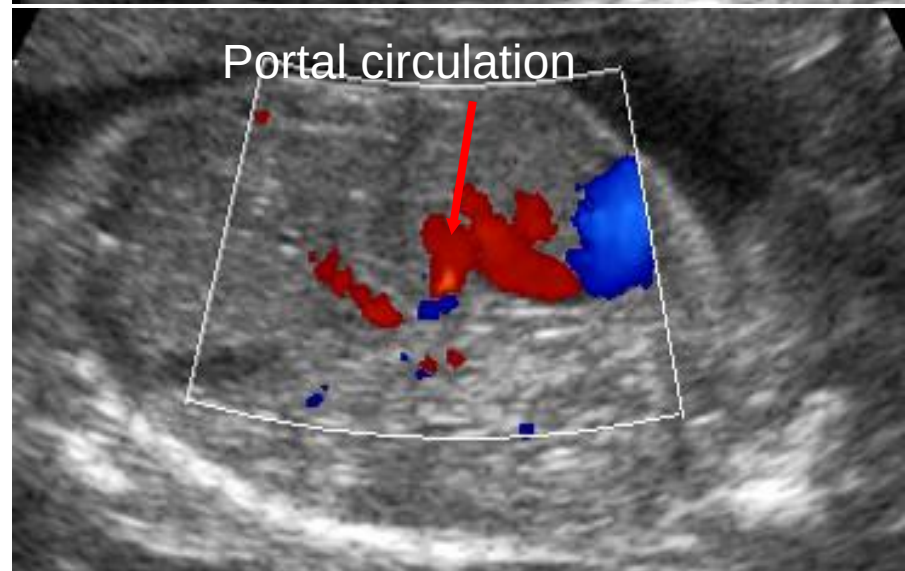
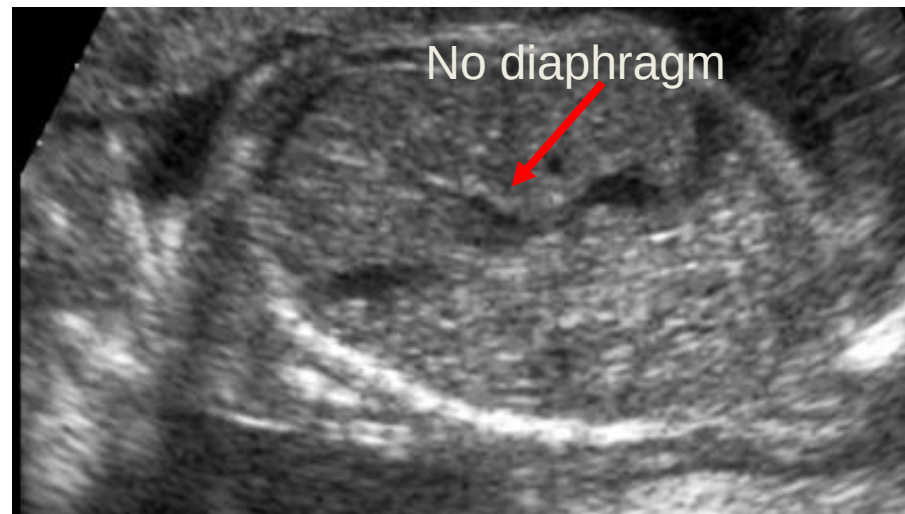
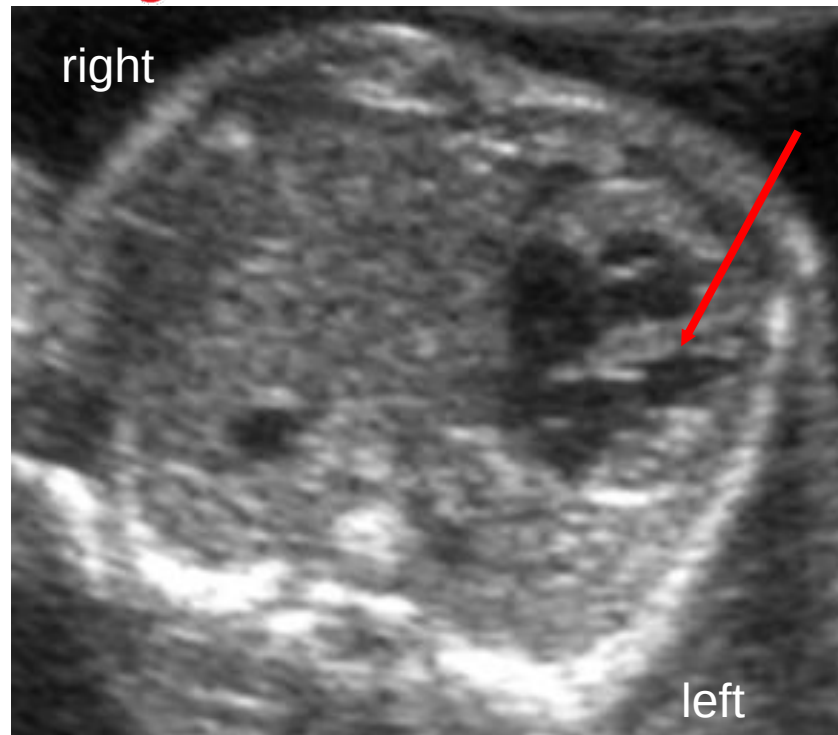
Left sided CDH with intrathoracic displacement of the left lobe of the liver

In this fetus at 32 week gestation meticulous scanning was required to demonstrate that the reason for the displacement of the heart was a left CDH with intrathoracic displacement of the left lobe of the liver

Clip 23



Sonography of right CDH



Right CDH has frequently an elusive sonographic appearance; the main clue is the cardiac displacement; longitudinal views demonstrating the abnormal disposition of the liver are also useful

Sonography of right - sided CDH

A right-sided CDH case with the heart displaced to the left-sided and the liver into the right side of the fetal thorax.

Clip 02



Sonography of right sided CDH with pleural effusion

The diagnosis of right CDH required a longitudinal view of the fetal trunk

Clip 26



Diagnosis during first trimester screening

With the improvement in new ultrasound devices, it is not as difficult to differentiate lung echogenicity for that of the herniated viscera and therefore, CDH cases can be diagnosed even during first trimester screening

Clip 03



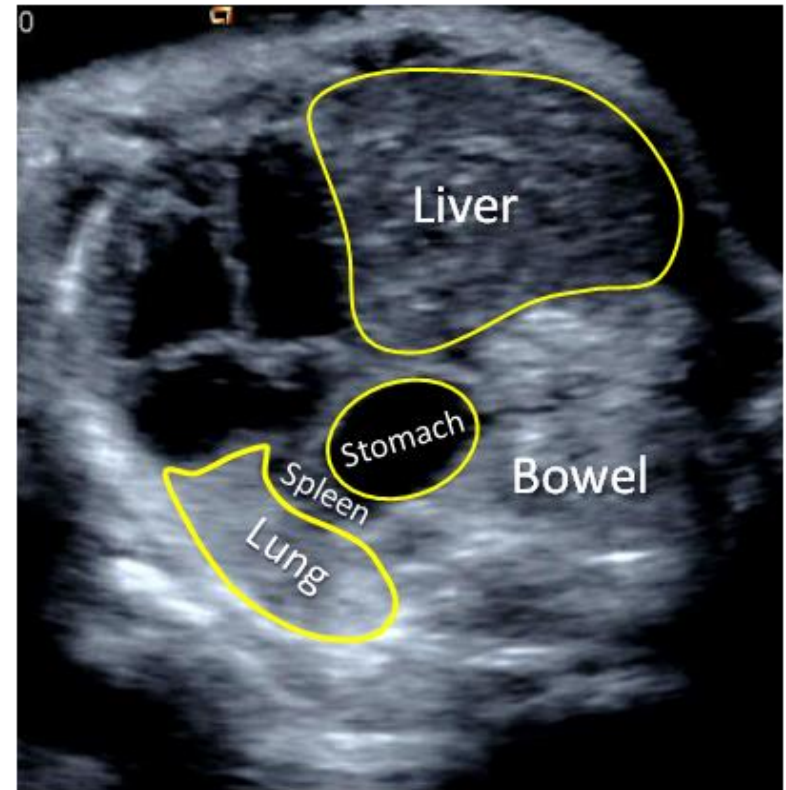
Vaginal sonography of right CDH at 13 weeks' gestation



Clip 21

Identification of the herniated organs

A cross sectional view of the fetal thorax in a case of left-sided CDH demonstrating the shift of the heart to the right side and the presence of the stomach, spleen, bowel and liver into the fetal thorax.



Identification of the liver herniation

In left-sided CDH, the liver herniation can be directly identified in a transversal view of the fetal thorax as the presence of a hypoecogenic area below the stomach

Clip 04



Identification of the liver herniation

In the absence of liver herniation the stomach and bowel occupy the left side of the thorax.



Clip 05

Identification of the liver herniation

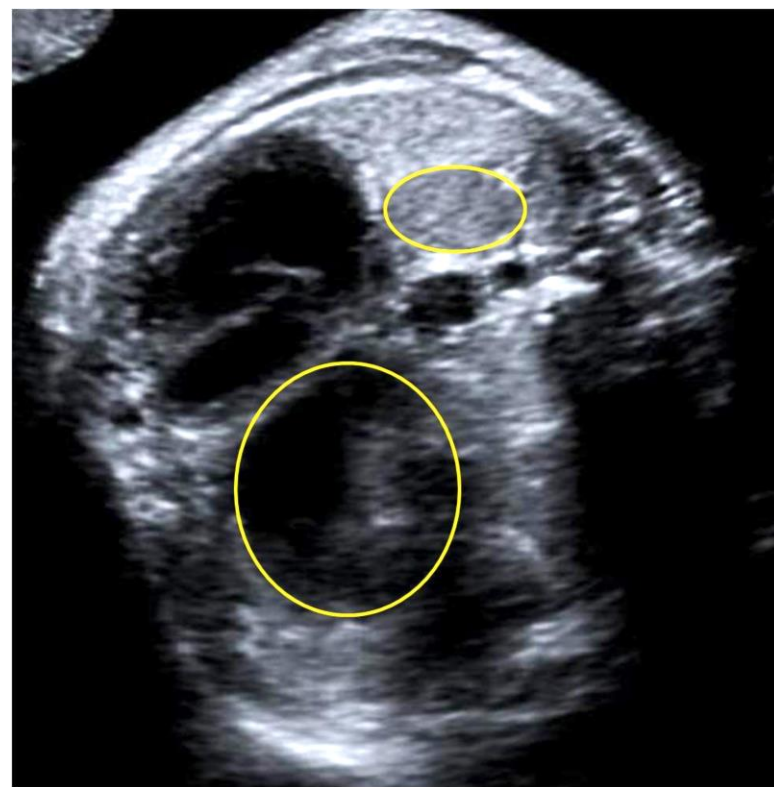
Other indirect sign suggesting the presence of liver herniation is the deviation of the umbilical vein demonstrated in a cross-sectional view of the abdomen. Color or power Doppler ultrasound could be useful in its identification.



Clip 06

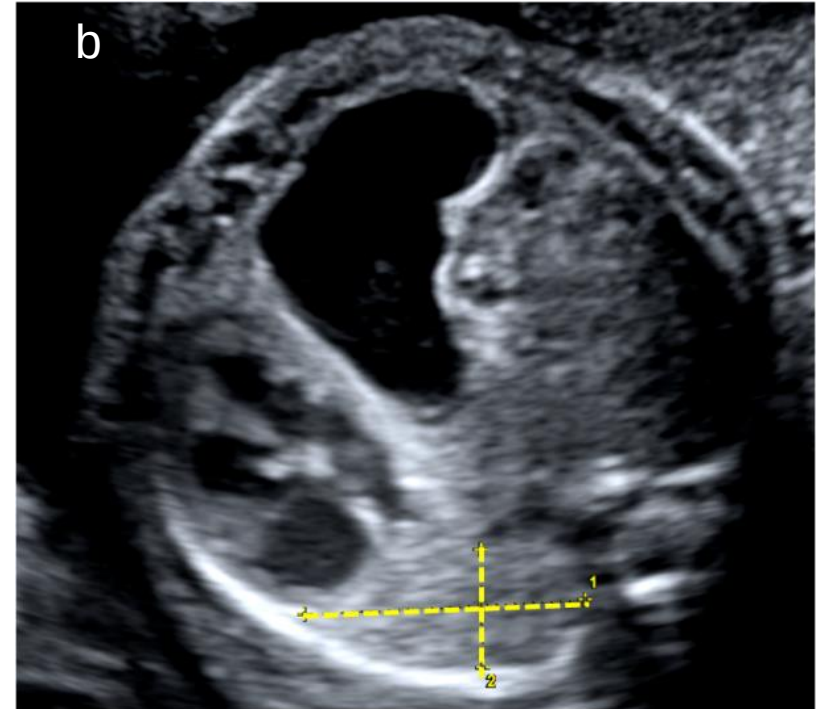
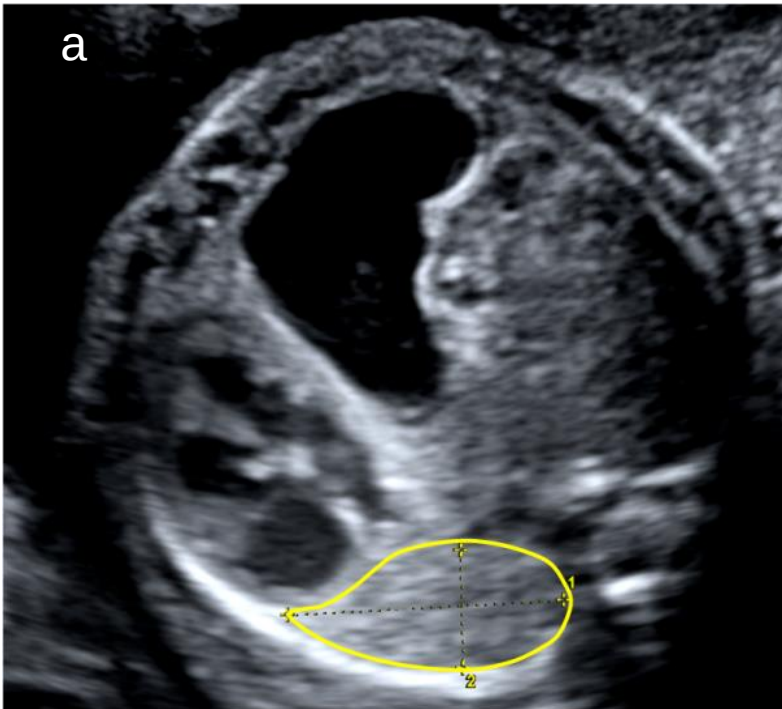
Identification of the spleen into the thorax

The spleen is usually located between the stomach and the contralateral lung to the side of the hernia and could be identified by its hypoecogenic density in comparison to the lung. Its identification is important in order to avoid including the spleen area into the lung area measurement.



Lung to head ratio measurement

A case of left-sided CDH without liver herniation. The evaluation of the lung area to head circumference ratio (LHR) requires a cross-sectional view of the thorax at the level of the four chamber view of the heart and can be measured by the lung borders (a) or by the longest diameter approach (b).



Calculation of the LHR

Demonstration of the lung area measurement by the longest diameter approach. The LHR is calculated by the multiplication of the longest diameter by its widest perpendicular diameter and then divided by the head circumference.

The observed LHR value is compared to the expected LHR in order to calculate the observer to expected (O/E) LHR as: $\text{LHR observed} / \text{LHR expected} \times 100$.

Clip 07



Ipsilateral lung in CDH

The ipsilateral lung to the side of the hernia can be identified in a cross-sectional view of the fetal thorax at the level of the three vessel trachea view



Clip 08

Severe left – sided CDH case

Illustrative picture of a left-sided CDH case with severe pulmonary hypoplasia

The stomach and spleen are located posterior to the heart with a extremely low lung size

Clip 09



Mild left – sided CDH case

Left-CDH case without liver herniation, and LHR of about 70%. Accordingly, it was classified as mild pulmonary hypoplasia and good prognosis.

Clip 10



Prenatal management

Fetoscopic placement of an endotracheal balloon in a CDH case with severe pulmonary hypoplasia.

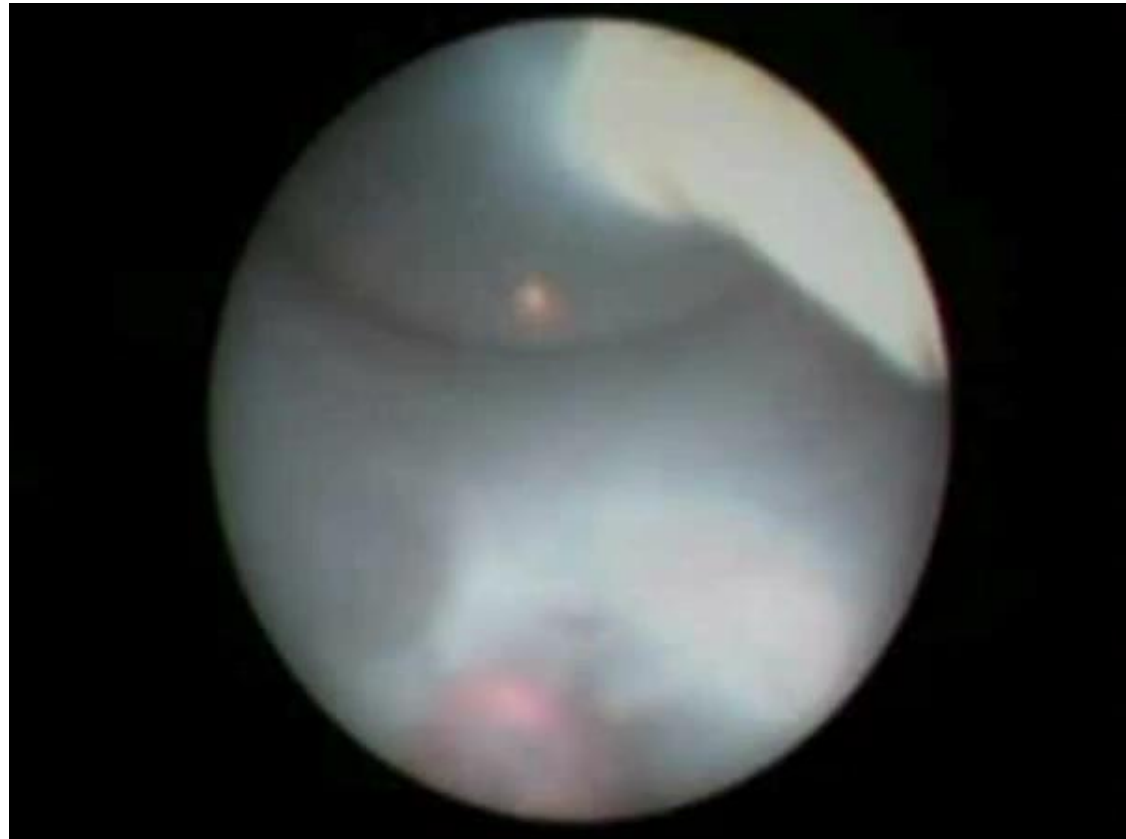
Clip 11



Prenatal management

In utero unplugging of the balloon by fetoscopic laser coagulation

Clip 12



Prenatal management

In utero unplugging of the
balloon by echographic
guide puncture

Clip 13



Evaluation of the endotracheal balloon

After the procedure, the balloon could be echographically visualized into the trachea in a sagittal view of the fetal neck.

Clip 14



Fetal response to tracheal occlusion

Illustrative picture of a case managed with tracheal occlusion.

The LHR was 26% before TO

Clip 15



Fetal response to tracheal occlusion

Illustrative picture of a case managed with tracheal occlusion.

The LHR increased to 55% 24 hours after TO

Clip 16



Fetal response to tracheal occlusion

Illustrative picture of a case managed with tracheal occlusion.

The LHR increased eventually to 98% 1 week, after TO

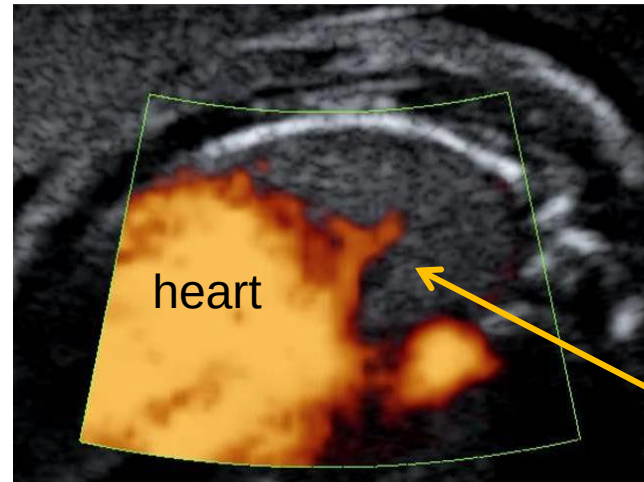
Clip 17



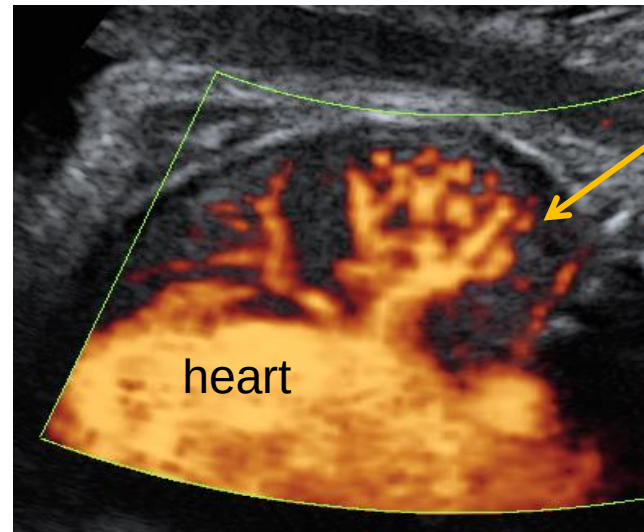
Lung perfusion changes

The same case with a lung tissue perfusion increment from 10% (a) before TO to 60% (b) 1 week after the procedure.

a)



b)

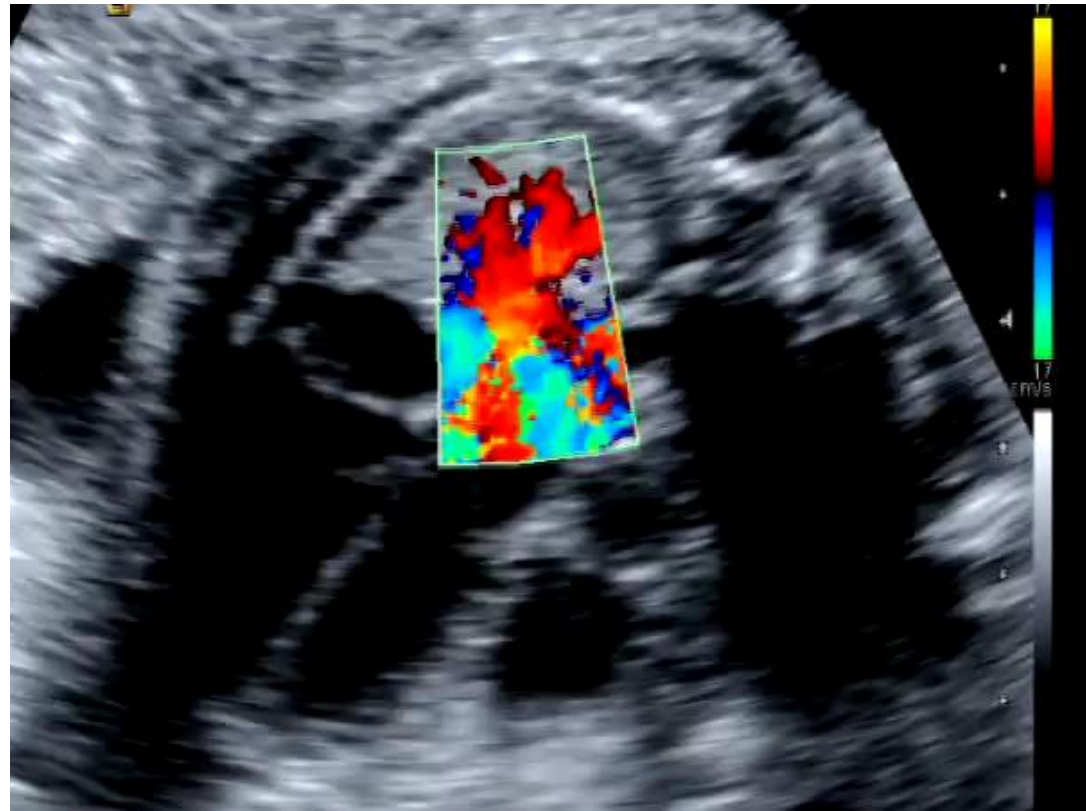


Intrapulmonary
vessels

Intrapulmonary artery Doppler evaluation

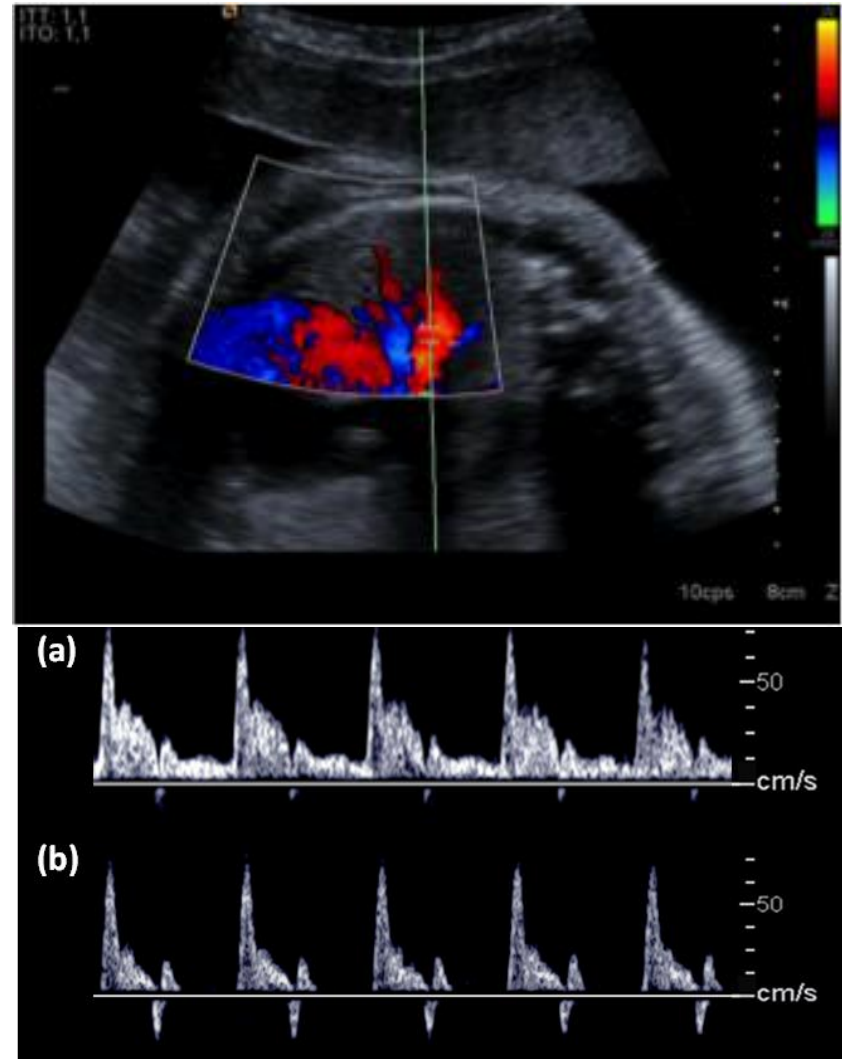
Lung Doppler evaluation in a cross-sectional view of the fetal thorax where the proximal branch of the pulmonary artery is visualized

Clip 18



Waveform of the intrapulmonary artery Doppler

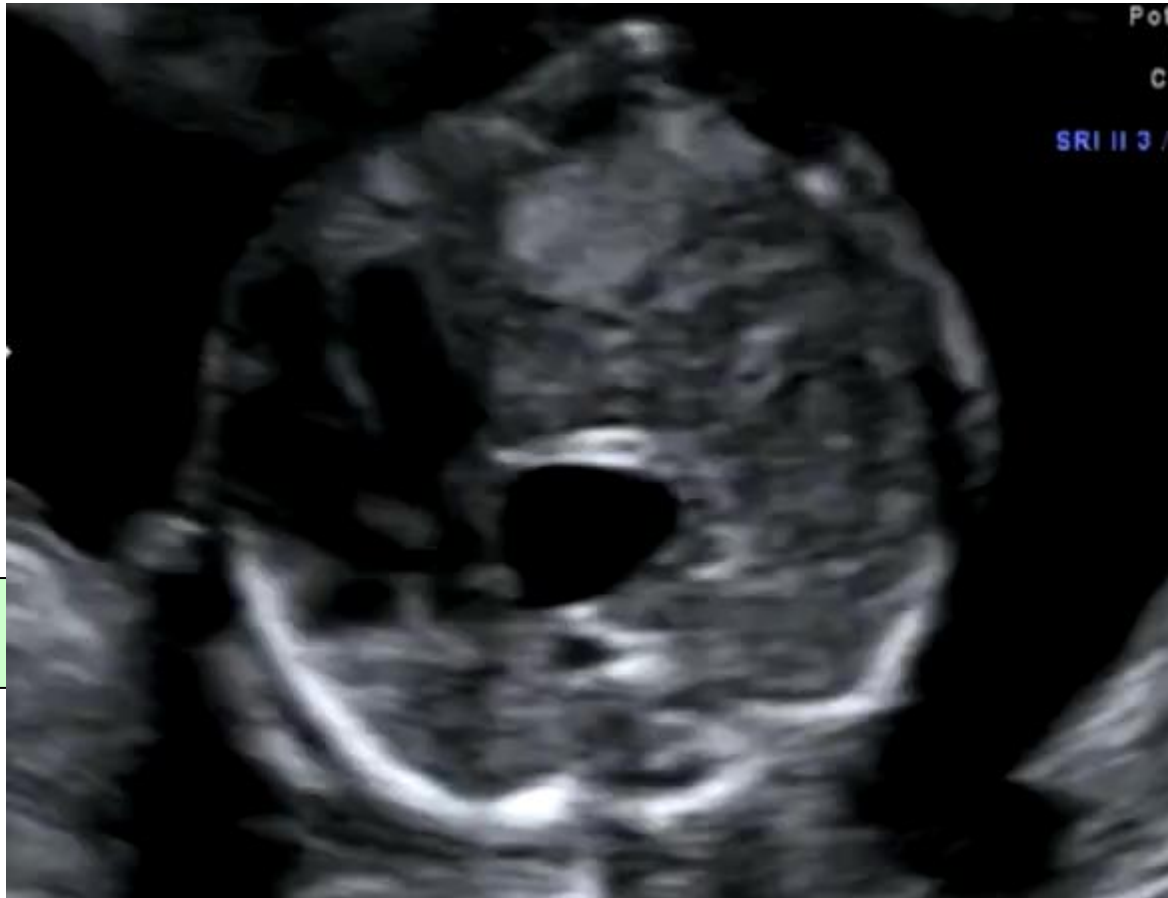
Insonation of the intrapulmonary artery and its typical normal needle waveform (a) and a case with increased lung impedance and absence of diastolic flow (b).



Additional malformations

A case of left-sided CDH with herniation of the stomach, spleen and bowel but without liver herniation coexisting with a bronchopulmonary sequestration in the ipsilateral lung to the side of the hernia.

Clip 20



Additional malformations

The major associated anomalies are congenital heart malformations that could be identified in up to 32% of the CDH cases.

In this particular case an atrioventricular canal is observed coexisting with a left-sided CDH with a huge liver herniation and a poor prognosis as a consequence.

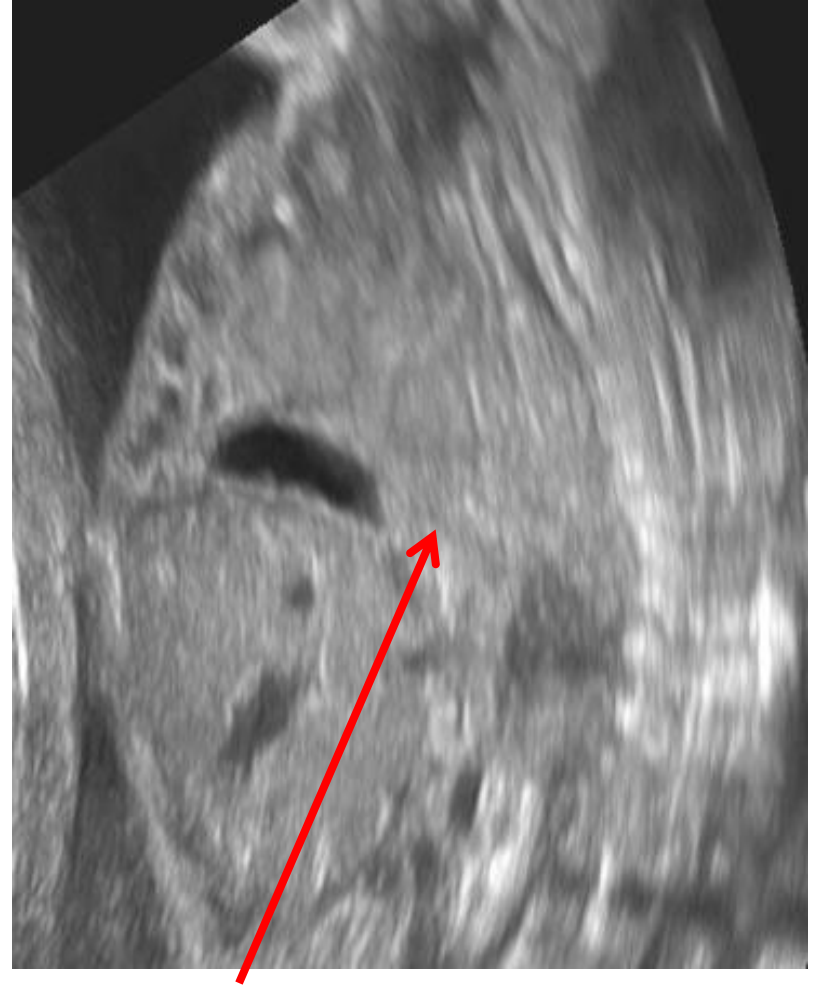
Clip 26



Differential diagnosis: Echogenic lungs vs CDH: ultrasound



Enhancement of diaphragmatic line



No diaphragmatic line

Differential diagnosis Echogenic lungs vs CDH: MR

