SAS Journal of Medicine
Abbreviated Key Title: SAS J Med
ISSN 2454-5112
Journal homepage: https://saspublishers.com

OPEN ACCESS

Pediatric Radiology

Imaging of Congenital Diaphragmatic Hernias

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DOI: https://doi.org/10.36347/sasjm.2025.v11i04.009 | **Received:** 08.03.2025 | **Accepted:** 12.04.2025 | **Published:** 15.04.2025

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Abstract **Review Article**

***Background*:** Congenital diaphragmatic hernias are complex and life-threatening lesions that are not just anatomic defects of the diaphragm, but represent a complex set of physiologic derangements of the lung, the pulmonary vasculature, and related structures. Imaging plays an increasingly important role in the care of these infants. ***Materials and Methods*:** Prenatal ultrasound and MRI have enabled early and precise identification of the anomaly and associated abnormalities. These tools have also made it possible to define the degree of pulmonary hypoplasia, and to predict newborn survival and the need for aggressive respiratory rescue strategies. In the postnatal period, conventional radiography supplemented by cross-sectional imaging in selected cases can be very useful in establishing the differential diagnosis of intrathoracic masses, detecting associated abnormalities and managing complications associated with pulmonary hypoplasia. ***Conclusion*:** Fetal intervention such as, fetal tracheal occlusion, is being trialled and may change the outcomes in the future.

Keywords : Congenital Diaphragmatic Hernia, Imaging, Ultrasound, Scan.

INTRODUCTION

Congenital diaphragmatic hernia (CDH) is the consequence of a defect in the development of the diaphragm. The defect allows the abdominal contents to herniate into the thoracic cavity. They are serious and life-threatening anatomic lesions that are frequently associated with additional anatomic malformations, syndromes, and chromosomal anomalies. The incidence is approximately per 10,000 live births [1], making it one of the more common birth anomalies. CDH is associated with high mortality but with improvement in detection and perinatal management the overall survival rates are now up to 70% [2-4]. Thus, familiarity with the pathogenesis and prenatal and postnatal imaging appearance of the various types of diaphragmatic hernia is important for the multidisciplinary medical and surgical team taking care of these patients. In this essay, we review and update the imaging features of diaphragmatic defects with emphasis on early and accurate identification of high-risk and complex lesions.

Pathogenesis

The development of the diaphragm occurs before 10 weeks gestation and separates the pleuro-peritoneal canal with a pleuro-peritoneal membrane to form the diaphragm. Two lateral segments (left and right) and a central segment coalesce together to form the diaphragm. When the fusion is not complete a diaphragmatic hernia results. **(figure 1a)** Pulmonary development is also very important in the understanding of CDH. The lungs begin their development as a ventral outpouching of the primitive foregut just after the third week of gestation, and they undergo successive branching until the sixteenth week. Thereafter, terminal bronchioles, acini and alveolar air sacs continue to develop throughout the remaining gestation and into childhood until approximately 8 years of age. Development of the pulmonary vasculature closely follows that of the airways and can also be affected by maldevelopment of the diaphragm. Although knowledge of the embryologic causes of abnormalities of the diaphragm is useful in the understanding of the pathogenesis of diaphragmatic hernias, it does not form a clinically useful classification system. Hernias are more practically classified by their morphologiccharacteristics and location [3].

The abdominal contents herniate into the thoracic cavity and compress the lungs and often cause a shift of the mediastinum.

The diaphragmatic defect occurs on the left side in up to 90%, on the right side in 10% 5 and rarely, bilaterally 6 Approximately 70% of defects involve the posterolateral (Bochdalek) region of the diaphragm, with

anterior (Morgagni) involvement in 25–30% [7, 8]. Central regions are rarely affected **(figure 1b).**

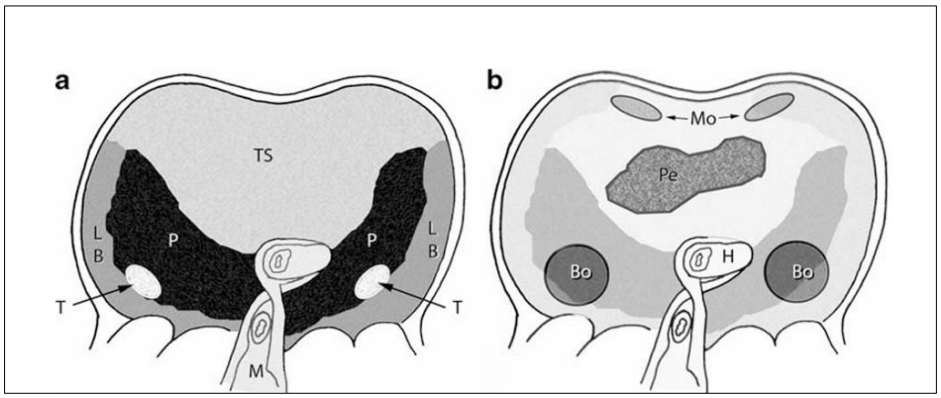

Figure 1:

a: Diagrams of developmental components of the fetal dia phragm. a Developmental components of the diaphragm include the transverse septum (TS), mediastinum (M), pleuroperitoneal membrane (P), lateral body wall musculature (LB), and the lumbocostal trigone

b: Relationship of diaphragmatic hernias to developmental components of the diaphragm. Foramen of Bochdalek (Bo), foramen of Morgagni (Mo), pericardial defect (Pe), hiatal foramen (H) [9]

Prenatal Imaging Evaluation

Sonography has been the mainstay of prenatal diagnosis of CDH for at least 20 years. Although CDH can be detected on routine sonography as early as 18 weeks, the mean gestational age at the time of detection is approximately 24 weeks [10]. **(Figure. 2a and 2b).** The sensitivity of sonography for prenatal detection of CDH varies from 18% to 87% in published series and appears to improve in the presence of associated, more readily observed anomalies, with advancing gestational age, and with increasing experience of the individual performing the examination [11]. In one multiinstitutional study in Europe, the detection rate for CDH increased from 51% in uncomplicated cases to 72% when CDH was associated with other malformations [10]. The typical sonographic features of a left CDH include the presence of a stomach bubble or left lobe of the liver at the level of the fetal heart, bowel loopsin the fetal chest, and a right mediastinal shift **(Fig 3).** In addition, the abdomen is scaphoid, and the gallbladder and hepatic or umbilical veins might be in an abnorma lposition within the abdomen [11]. The echogenicity of the fetal lung and liver can be similar and difficult to distinguish on gray-scale sonography. Color Doppler sonography might be helpful in detailing the abnormal position and course of intrahepatic vessels. Bowing of the umbilical segment of the portal vein to the left of the midline and portal branches to the lateral segment of the left hepatic lobe coursing toward or above the diaphragmatic ridge are good predictors for liver herniation into the fetal thorax (positive predictive values of 85% and 100%, respectively) [12, 13]. However, the fetal stomach is not always displaced into the chest, especially in fetuses with right-side CDH. These fetuses might have herniation of the gallbladder into the right chest, abnormal position of the intrahepatic portion of the umbilical vein, and mediastinal shift to the left [11].

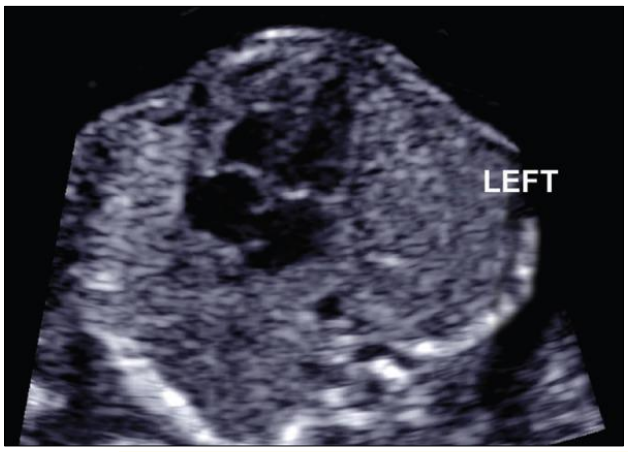

Figure 2 : Normal four chamber axial view of the heart

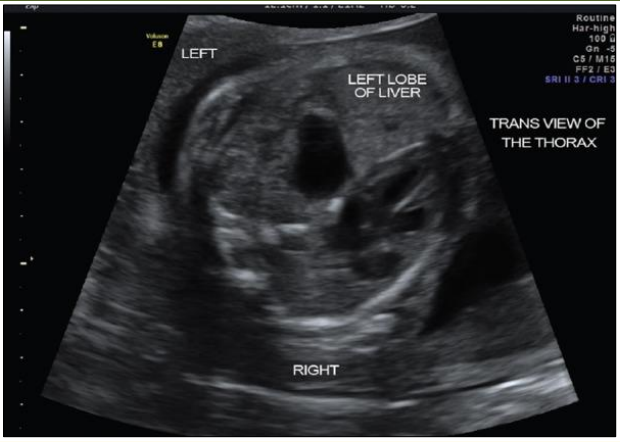

Figure 3 : Left sided CDH showing stomach and liver in the four chamber view and mediastinal shift of the heart to the right side

MRI In fetuses suspected of having a CDH, MRI is often recommended to assess herniation of the liver and associated anomalies **(Fig. 4).** MRI has been shown to provide additional information to that obtained sonographically in 38% to 50% of fetuses with chest lesions [14, 15], and MRI affects clinical care in selected cases [14–17]. One of the important contributions of prenatal MRI in this condition is the assessment of fetal lung volumes (FLV). Fast spin-echo T2-W sequences have been reported to have a high degree of reproducibility and interobserver agreement in estimating FLV in normal fetuses and in fetuses with CDH [18, 19].

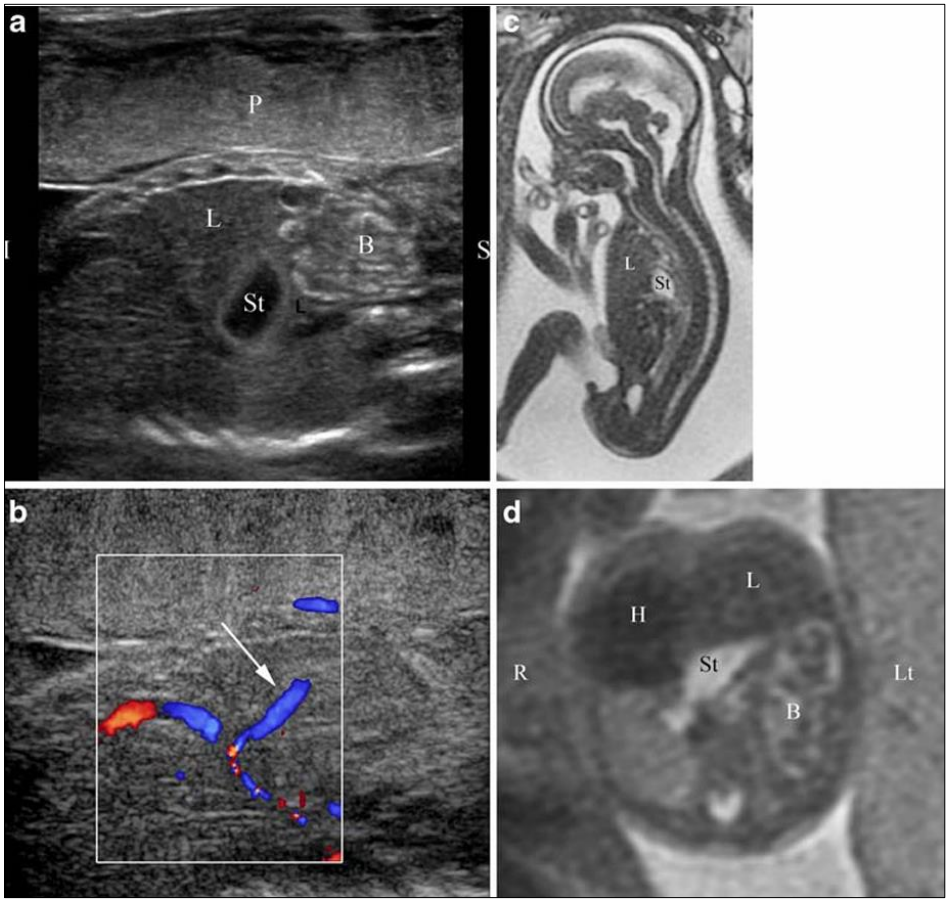

Figure 4 : Left Bochdalek hernia in a 20-week fetus. a Sagittal sonogram of the left fetal chest shows herniation of echogenic bowel (B), stomach (St) and liver (L) into the fetal chest (P placenta). b Color Doppler sonogram shows superior dis placement of hepatic vessels (arrow) into the fetal chest. c, d Sagittal (c) and axial (d) FIESTA and T2-W MRI images of the fetus obtained on the same day show intrathoracic herniation of the liver (L), bowel (B) and stomach (St) with displacement of the heart (H) to the right side (R right, Lt left)

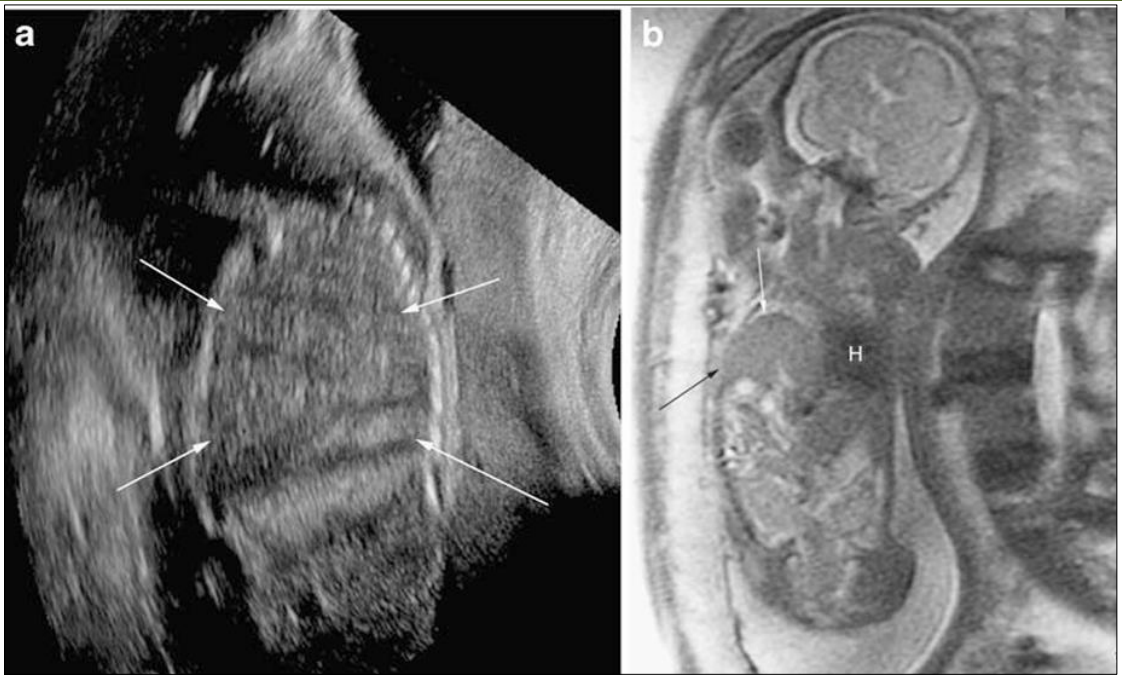

Figure 5 : Right Bochdalek hernia in a 20-week fetus. a Sagittal sonogram of the right fetal chest shows the liver filling the right hemithorax (arrows). b Coronal T2-W MR image obtained the same day confirms the presence of a right-side Bochdalek hernia with herniation of the liver and bowel (arrows) and displace ment of the fetal heart (H) to the left

Differential Diagnosis

At times the diagnosis of CDH is not clear, and other diagnoses should be considered, such as bronchopulmonary foregut malformations, congenital cystic adenomatoid mal formations, sequestrations, and primary pulmonary agenesis or hypoplasia. These lesions might coexist with CDH and influence surgical planning (Fig. 6).

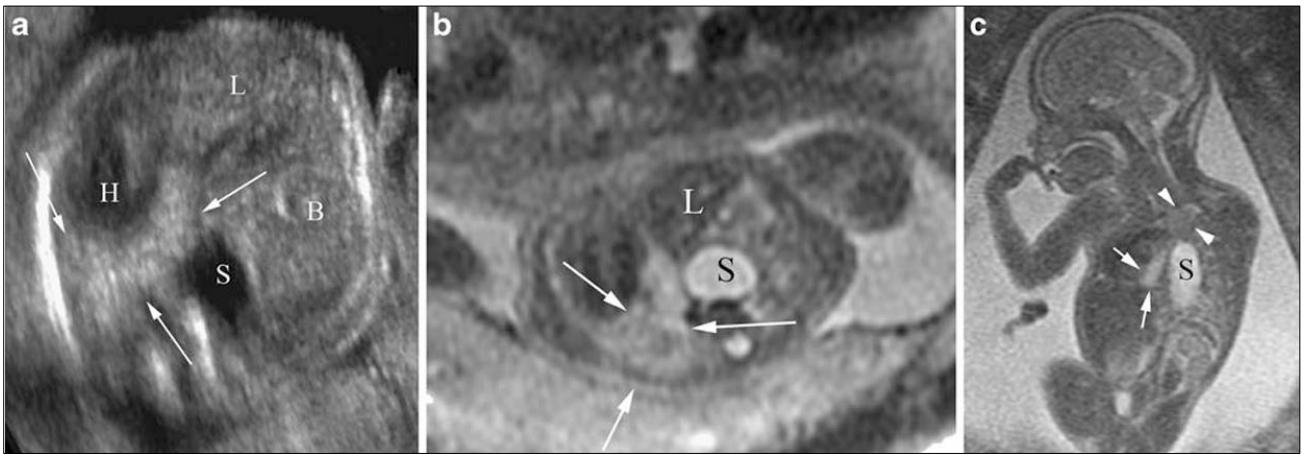

Figure 6 : Coexisting left Bochdalek hernia and sequestration in a 22-week fetus. a Axial sonogram of the fetal chest shows herniation of bowel (B), stomach (S), and liver (L) into the fetal chest, with displacement of the heart (H) to the right chest. An echogenic mass (arrows) is interposed between the heart and stomach. a, b Axial (b) and coronal (c) T2-W MR images of the fetus obtained on the same day confirm the presence of a thoracic mass (arrows) with signal intensity higher than that of compressed lung (arrowheads). A left lung sequestration was removed at postnatal repair of a CDH

Postnatal Imaging

Initial chest radiographs often show an opacified hemi thorax with mass effect and contralateral shift of the mediastinum. Once air is introduced into the gastrointestinal tract, bowel gas is lacking in the abdomen and can be identified in the chest. The location and course of vascular catheters and the nasogastric tube is altered in a high proportion of patients with CDH. The esophageal portion of the nasogastric tube deviates to the right in left-side CDH and to the left in right side hernias. An intrathoracic nasogastric tube strongly suggests displacement of the stomach into the hernia. The umbilical venous catheter is also affected by the degree and direction of liver displacement into the chest. Conversely, the location of umbilical artery catheters is rarely affected by CDH because of the retroperitoneal location of the aorta and its limited mobility (Figs. 7)

[20].

Findings on initial chest radiograph, including the amount of aeration of the ipsilateral or contralateral lung, degree of mediastinal shift and hernia content, are not predictive of clinical outcome [21]. Other intrathoracic masses can mimic CDH (Fig 8). When the diagnosis of CDH is in question, a small amount of contrast material or air can be injected via the nasogastric tube to outline the location of the bowel.

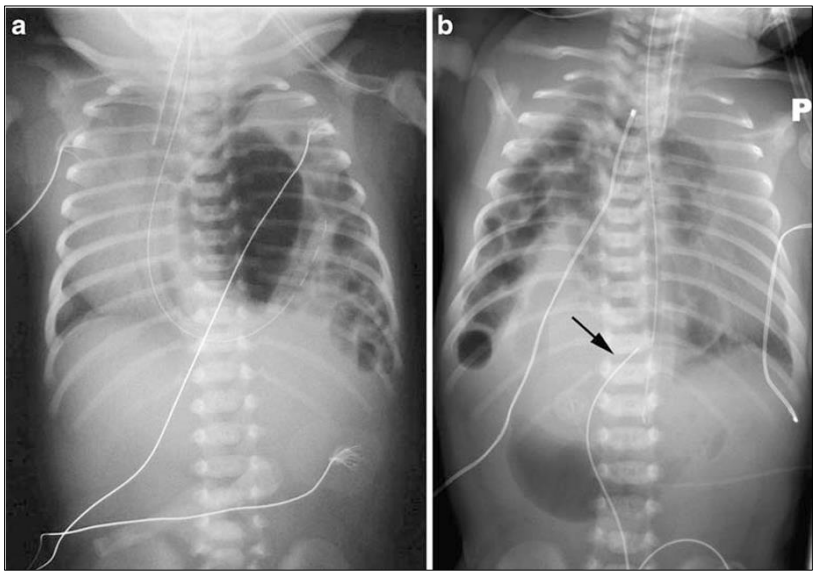

Figure 7 : Neonatal Bochdalek hernia. a A 1-day-old boy with left Bochdalek hernia. Chest radiograph shows deviation of the endotracheal tube and esophageal portion of the naso gastric tube to the right. Intra thoracic displacement of the nasogastric tube tip, bowel and stomach confirm the diagnosis of diaphragmatic hernia. b A2 day-old girl with right Bochdalek hernia. Chest radiograph shows deviation of the nasogastric tube and umbilical venous catheter (arrow) to the left

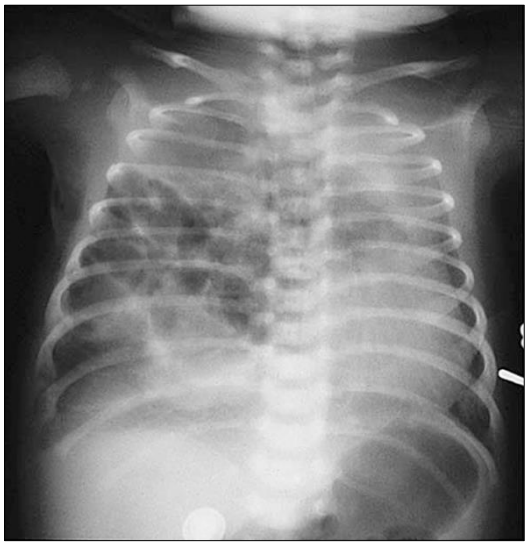

Figure 8 : A 3-day-old boy with congenital cystic adenomatoid malformation of right lung. Chest radiograph shows air-filled cysts that mimic intrathoracic bowel herniation

CT with multiplanar reconstruction is useful to elucidate associated lung masses and broncho pulmonary foregut malformations. The use of intravenous contrast agent and CT arteriography should be considered for depiction of the vascular supply of lung lesions. However, the administration of oral contrast material should be avoided because it can increase the degree of mediastinal shift and subsequent respiratory compromise.

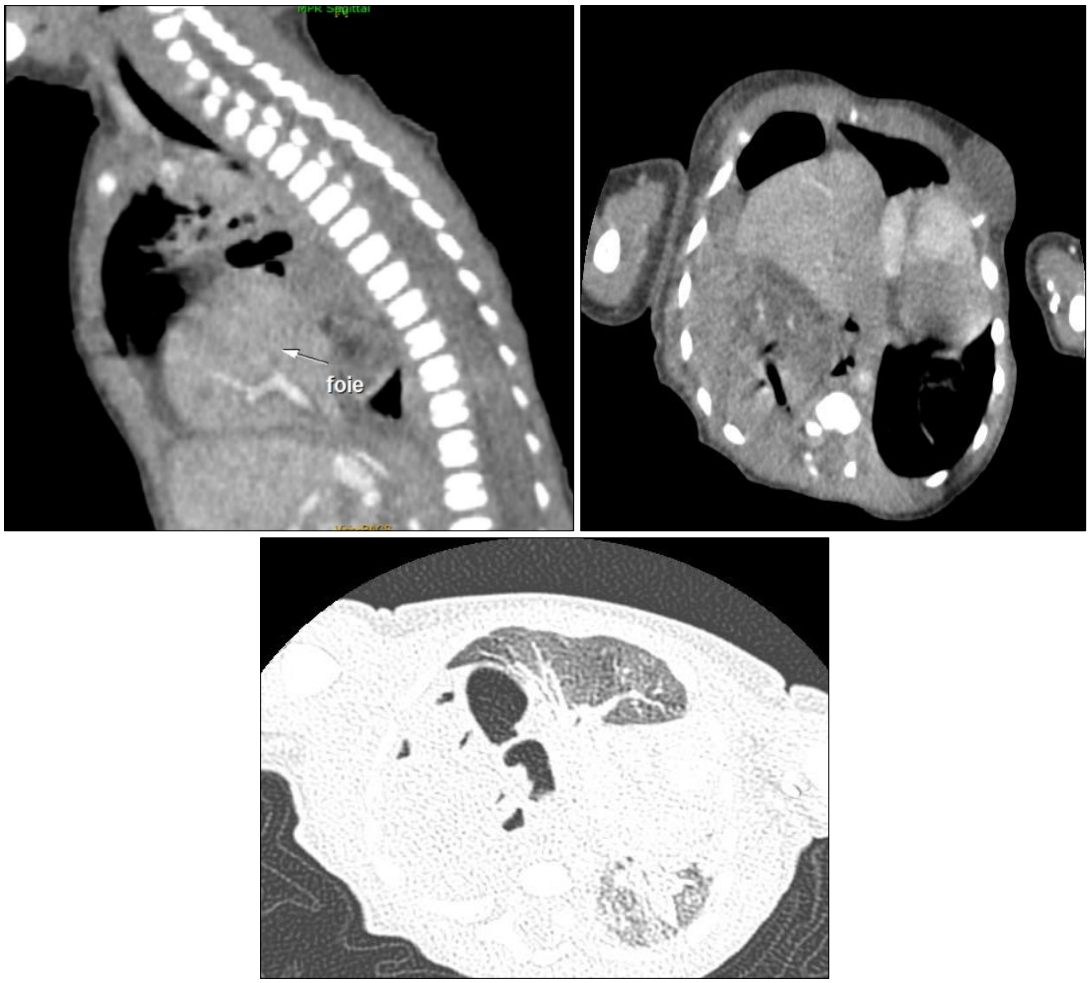

Figure 9 : **CT scan with contrast injection showing intra-thoracic liver herniation. Presence of digestive structures in the thoracic cavity**

Hiatal Hernia

A hiatal hernia is defined as the protrusion of a portion of the stomach into the mediastinum through the esophageal hiatus of the diaphragm [3]. (Fig 10). Three types of hiatal hernia are currently recognized, sliding hiatus hernia, in which the esophagus moves freely through an enlarged hiatus into the chest; paraesophageal hernia, in which the gastroesophageal junction remains in its normal location and a portion of the stomach bulges through a hiatus anterior to the stomach; and congenital short esophagus, in which the stomach is irreducibly fixed in the chest. One potential explanation for the presence of congenital hiatal hernias relates to a delay in the descent of the stomach from the chest. The resulting esophageal hiatus is larger than normal, leaving a space between the esophagus and diaphragm [3]. Although hiatal hernias account for almost half of all hernias, the great majority of these are acquired lesions. Hiatal hernias account for only 9% of diaphragmatic hernias in infants younger than 1 year. Prenatal diagnosis of a hiatal hernia might be possible by identification of a hypoechoic mass in the posterior mediastinum in continuity with the intraabdominal portion of the stomach [22]. Some of these cases are associated with a congenitally short esophagus that might cause a distended stomach to be located in the thoracic cavity.

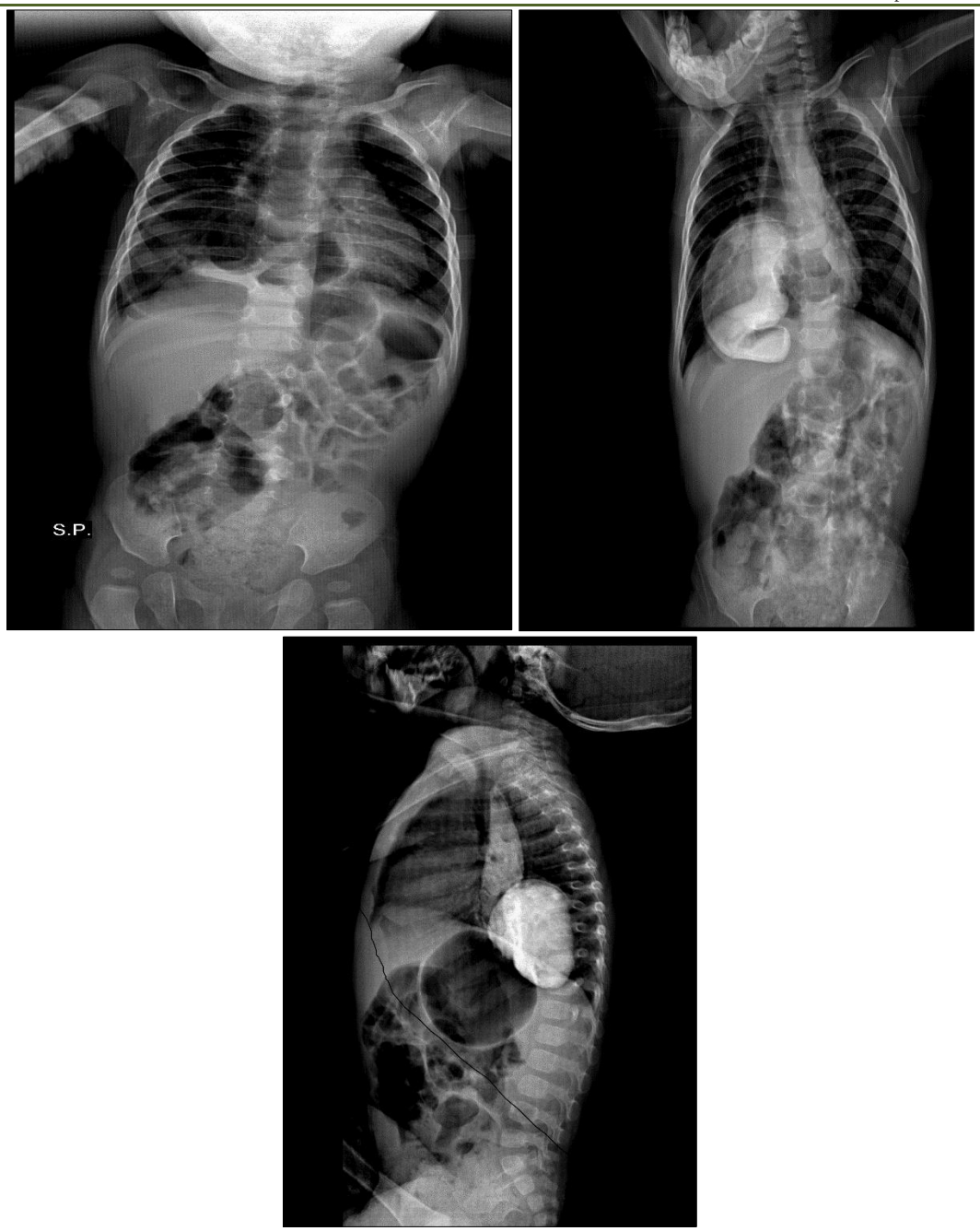

Figure 10 : Esophagogastric Transit:
A: Digestive-like lucencies in the thoracic cavity, located in the midline and the right hemithorax. Leftward displacement of the cardiac opacity
B: After contrast ingestion: The esophagus is dilated and remains open. Intrathoracic esophagus. Retrocardiac intrathoracic portion shifted to the right hemithorax

SUMMARY

CDH is a fetal abnormality that is seen frequently in tertiary centres. The treatment for CDH is an uncomplicated postnatal surgery. However, the pulmonary hypoplasia associated with the CDH in moderate and severe cases makes the prognosis poor. Antenatal assessment at a tertiary centre with fetal intervention when indicated could change the outcomes in the near future.

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