CASE REPORT

# Intrathoracic Renal Ectopia with Congenital Diaphragmatic Hernia

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**ABSTRACT**

Congenital thoracic ectopic kidney is a very rare developmental disorder. It is often asymptomatic and discovered incidentally on chest radiography or ultrasonography. Symptoms can be present if it is associated with diaphragmatic defect and respiratory compromise. Herein is a case of a newborn male with cyanosis and respiratory distress since birth. Imaging eventually revealed the presence of intrathoracic ectopic left kidney with congenital diaphragmatic hernia. Radiological knowledge and increased awareness of this rare condition can avoid undue investigations and reduce patient morbidity.

**Keywords:** congenital, diaphragmatic hernia, renal ectopia

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## INTRODUCTION

Thoracic kidney is the rarest form of renal ectopia, with only about 200 cases reported and published in medical literature, and majority of them are in adults [1]. Most ectopic kidneys are found in the lower lumbar or pelvic region secondary to failure of ascent during fetal life. Intrathoracic kidney has the lowest frequency rate among all renal ectopias with a prevalence rate of about less than 0.01% [2]. The reported incidence is less than 5 per 1 million births [3]. It accounts for 5% of all renal ectopias and its association with congenital diaphragmatic hernia has been reported to have an incidence of only 0.25% [4,5].

Due to its rarity, literature on intrathoracic kidney, its management, prognosis and complications is sparse and is confined to case reports [5]. This paper presents a case of intrathoracic left kidney with congenital diaphragmatic hernia with review of literature on this condition.

## THE PATIENT

Clinical Data

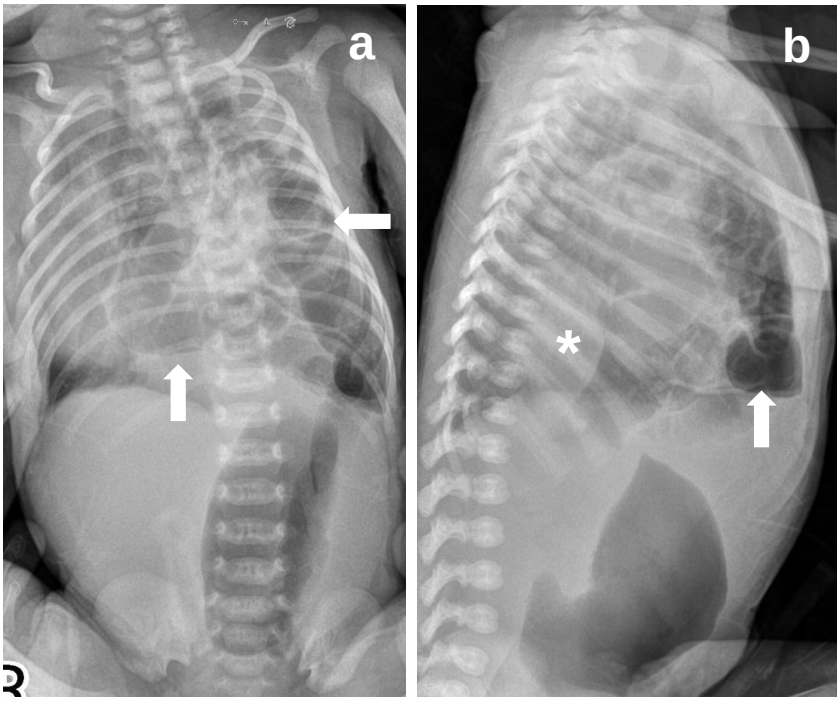
A full-term, male infant, from Leyte, Philippines, presented with respiratory distress. He was delivered via Cesarean section with appropriate weight for gestational age at a maternity clinic. Upon birth, the patient presented with cyanotic episodes, especially during crying, and had poor suck. Persistence of cyanosis prompted medical consultation and subsequent admission.

Physical examination showed decreased breath sounds in the left hemithorax and an apex beat heard at the right midclavicular line, level of $4^{th}$–$5^{th}$ intercostal space. Other physical examination findings were unremarkable. Patient was transferred to the neonatal intensive care unit for monitoring. Laboratory investigations were done and revealed *Staphylococcus haemolyticus* on blood culture and the patient was managed as a case of sepsis.

Diagnostic Imaging and Management

Chest and abdominal radiographs at 7 days old (Fig. 1a) showed multiloculated, large cystic lesions at the left hemithorax. Contralateral deviation of the mediastinal structures was noted. There is a focal, markedly dilated bowel loop in the abdomen. Lateral radiograph revealed a rounded opacity in the posterior part of the lower chest (Fig. 1b). Differential considerations at that time were congenital diaphragmatic hernia and congenital cystic adenomatoid malformation.

Ultrasound examination, at 12 days old, revealed absence of kidney in the left renal fossa and a normally placed right kidney. No ectopic kidney was visualized in the lower




**Figs. 1a–b** Babygram at 7 days old. Frontal (**a**) and lateral (**b**) projections revealed multi-loculated large cystic lesions (arrows) in the left hemithorax with marked right-sided deviation of the mediastinal structures. A rounded opacity (asterisk) was also noted at the posterior part of the lower hemithorax

abdomen. Ultrasound was carried out in the intercostal region and revealed the left kidney in the left hemithorax (Fig. 2a) showing abnormal rotation, with its hila facing inferiorly, and has a lobulated echostructure. Medial deviation of the lower pole of the left kidney is also noted (Fig. 2b).

Contrast-enhanced computed tomography (CT) of the thorax and abdomen was also performed. This revealed a large left-sided diaphragmatic defect through which bowel loops are seen herniating through and occupying most of the left hemithorax, slightly extending to the right. The displaced left kidney is seen within the thoracic region (Fig. 3). The spleen is seen in the lateral aspect of the left hemiabdomen. No normal-looking reniform structure is appreciated in the left renal fossa. The right kidney is normal in size, shape, and position. The visualized collecting systems are not dilated (Fig. 4a–b).

The patient was referred to the pediatric surgery department and surgical intervention was done, with the herniated bowels reduced into the abdominal cavity. The ectopic left kidney was carefully reduced to the retroperitoneal cavity and the diaphragm was repaired. Post-operative course was uneventful with normal laboratory findings. The patient was discharged well and was advised for long-term follow-up.

**Figs. 2a–b** Ultrasound of the left hemithorax at 12 days old. Gray-scale sonographic images showed a left kidney located superior to the spleen with bowel loops in between (**a**). The left renal hilum is rotated inferiorly with medial deviation of the lower pole of the kidney (**b**)

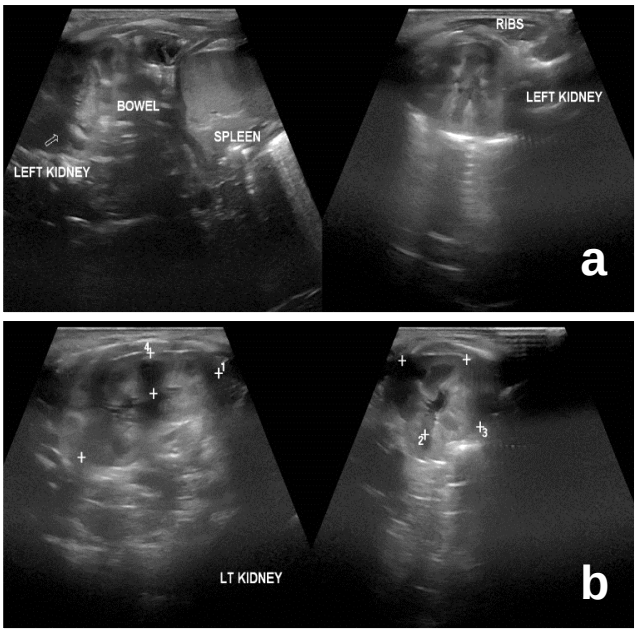
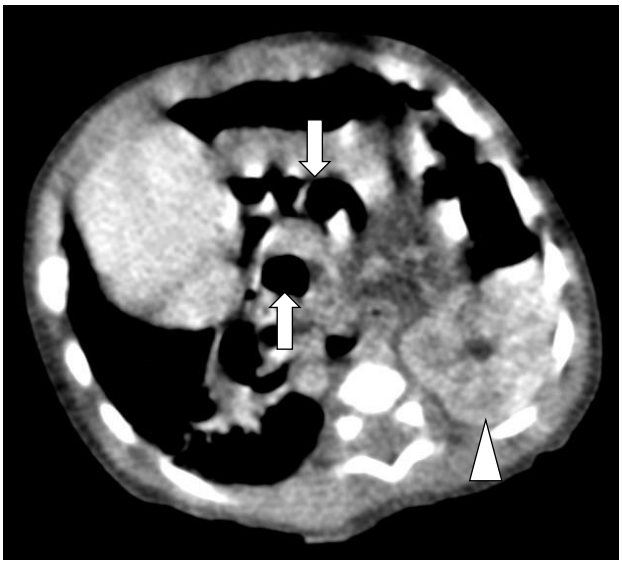


**Fig. 3** Chest CT scan at 14 days old. Axial image in soft tissue window setting revealed herniated bowel loops (arrows) occupying left hemithorax. The displaced left kidney (arrowhead) is seen posteriorly



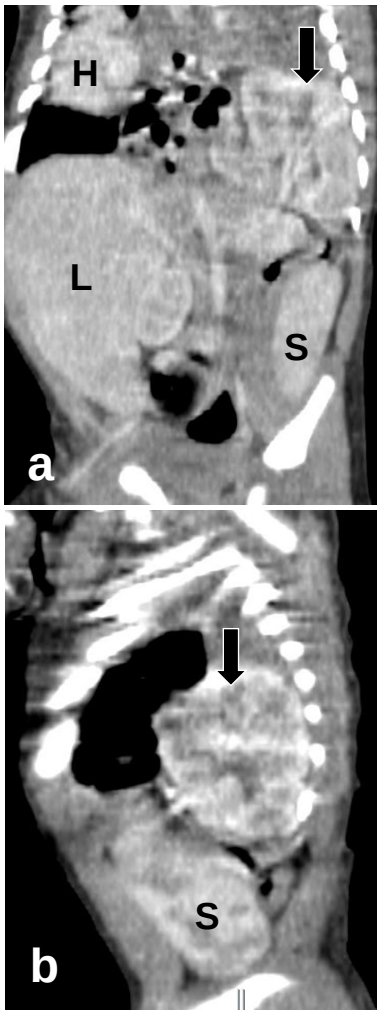
## DISCUSSION

Ectopic kidney refers to a kidney located at any site other than the renal fossa [2]. The abnormality is found more commonly on the left (62%) than the right side (36%) and is rarely bilateral (2%). Males (63%) are affected more frequently than females (37%) [2]. About 60% of ectopic kidneys are placed in the pelvis minor, 35% paravertebrally, and less than 5% in the thoracic cavity [4]. With a prevalence rate of less than 0.01%, intrathoracic kidneys are extremely rare [2].

Intrathoracic kidney is not always associated with diaphragmatic hernia [2]. Studies showed an ectopic thoracic kidney that has not herniated through the diaphragmatic defect, but has been responsible for preventing closure of the diaphragm, which resulted in a diaphragmatic hernia [6]. Herniation of the kidney through the diaphragm is only possible if the kidney has a long renal artery and ureter. The renal vasculature and ureter on the affected side are usually significantly longer than those in the normally positioned kidney [7]. Intrathoracic ectopic kidneys do not show dysplastic parenchymal architecture and renal function is usually normal [8].

Various techniques have been used for the diagnosis of intrathoracic kidney. The radiographic appearance of a thoracic kidney may be similar to that of posterior mediastinal masses. In this case, a smooth round mass is seen extending into the chest near the midline on frontal chest radiograph and at the posterior aspect of the diaphragm on the lateral view [9]. Ectopic kidney is generally discovered accidentally during routine abdominal ultrasound [10]. In every case, when the kidney cannot be found in its typical location on ultrasound, it is necessary to look for it outside the lumbar area. Radiologists should consider examining the thorax before labeling a patient with unilateral renal agenesis [11]. On prenatal sonography, the kidneys can be seen as early as 12 weeks of gestation. True intrathoracic ectopic kidney presents during fetal life and has four characteristics: (1) a rotation anomaly with the hilum facing inferiorly, (2) structural abnormalities, such as lobulated or deformed shape (3) a long ureter, (4) anomalous high derivation of the renal vessels from the thoracic aorta, and (5) medial deviation of the lower pole of the kidney [13].

**Fig. 4a–b.** Abdominal CT scan at 14 days old. Coronal (**a**) and sagittal (**b**) images in soft tissue window setting show the liver (L) occupying most of the right hemiabdomen. The spleen (S) is seen in the lateral aspect of the left hemiabdomen and inferior to the displaced intrathoracic left kidney (arrows). Note the heart (H) is displaced to the right



CT and MRI provide not only detection of a posterior mediastinal lesion but also visualization of its contour, extent, and size. CT urography is also helpful in confirming the diagnosis and in demonstrating the collecting system, vascular supply, and complications [9]. Renal scintigraphy with dimercaptosuccinic acid (DMSA) provides functional and anatomic information as well as perfusion of the kidneys [12].

Intrathoracic ectopic kidneys have been classified into four subtypes with different associations and approach to management: Group 1: Intrathoracic ectopic kidney associated with a closed diaphragm; Group 2: Intrathoracic ectopic kidney associated with diaphragmatic eventration; Group 3: Intrathoracic ectopic kidney associated with traumatic diaphragmatic rupture; and Group 4: Intrathoracic ectopic kidney associated with a congenital diaphragmatic hernia. The first two groups are usually asymptomatic and have a benign course, while the other two are often symptomatic and require therapeutic intervention [14].

Congenital diaphragmatic hernia associated with intrathoracic ectopic kidney have been reported to have ureters and vascular structures of adequate length, allowing reduction of the ectopic kidney into the abdominal cavity without hemodynamic impairment. Due to the ectopic nature of the kidney and the risk of complications, patients would benefit from long-term close monitoring and follow-up [15].

## CONCLUSION

The association between intrathoracic renal ectopia and congenital diaphragmatic hernia is a very rare occurrence that poses diagnostic and management dilemmas for clinicians. When managing cases of intrathoracic kidney, it is crucial to accurately identify the specific subtype based on radiologic studies. This information is vital for determining the most appropriate treatment approach and devising an effective management plan. A meticulous approach and careful handling of the intrathoracic kidney are necessary to ensure optimal outcomes and preserve function. Radiologic imaging remains the main diagnostic tool for its detection.

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