

LATE-PRESENTING CONGENITAL DIAPHRAGMATIC HERNIA WITH TENSION GASTROTHORAX: A CASE SERIES

Khairon Diyana Mohamed Nahrudin^{1,2}, Norhafiza Ab Rahman^{2,3}, Mohd Yusran Othman², Mohd Yusof Abdullah²

¹Department of Surgery, Hospital Serdang, Selangor

²Department of Paediatric Surgery, Hospital Tunku Azizah, Kuala Lumpur

³Department of Surgery, Kulliyah of Medicine, International Islamic University Malaysia, Kuantan Pahang

INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a rare birth defect with an overall prevalence of 2.3 in 10,000 births. Late-presenting CDH is defined as CDH diagnosed after neonatal age. We are sharing three cases of life-threatening tension gastrothorax as a manifestation of a late-onset CDH.

CASE SERIES

Case 1: A 10-month old boy presented with respiratory distress requiring intubation. Chest radiograph revealed a huge lucency in the left hemithorax with mediastinal shift which improved after nasogastric tube insertion. CT thorax confirmed the diagnosis of left CDH. Laparotomy revealed a 5x6cm posterolateral defect with stomach and spleen content which was completely reduced and primarily repaired (figures 1-4).

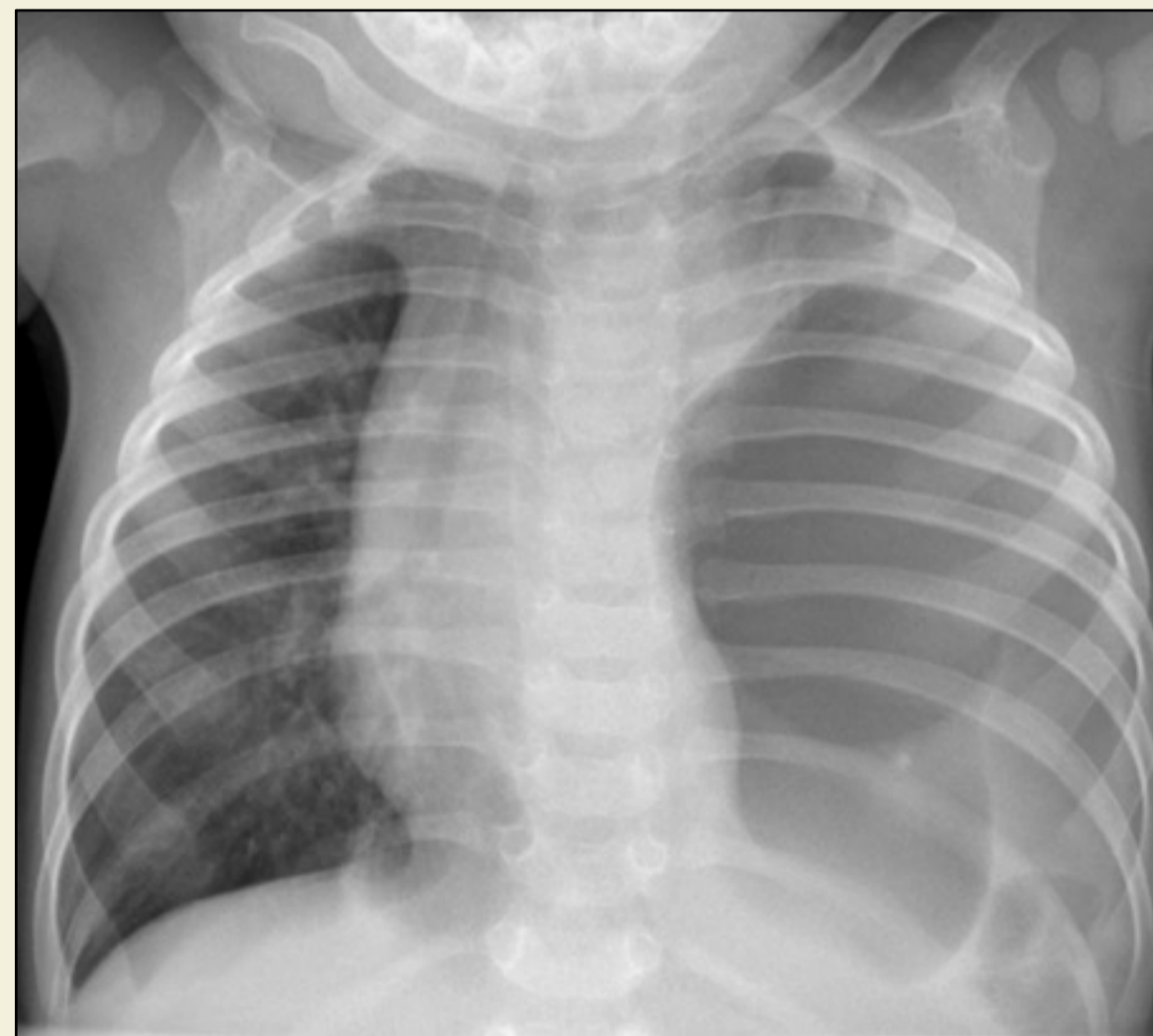


Figure 1: Chest x-ray on arrival showed a huge lucency in left hemithorax with a marked mediastinal shift

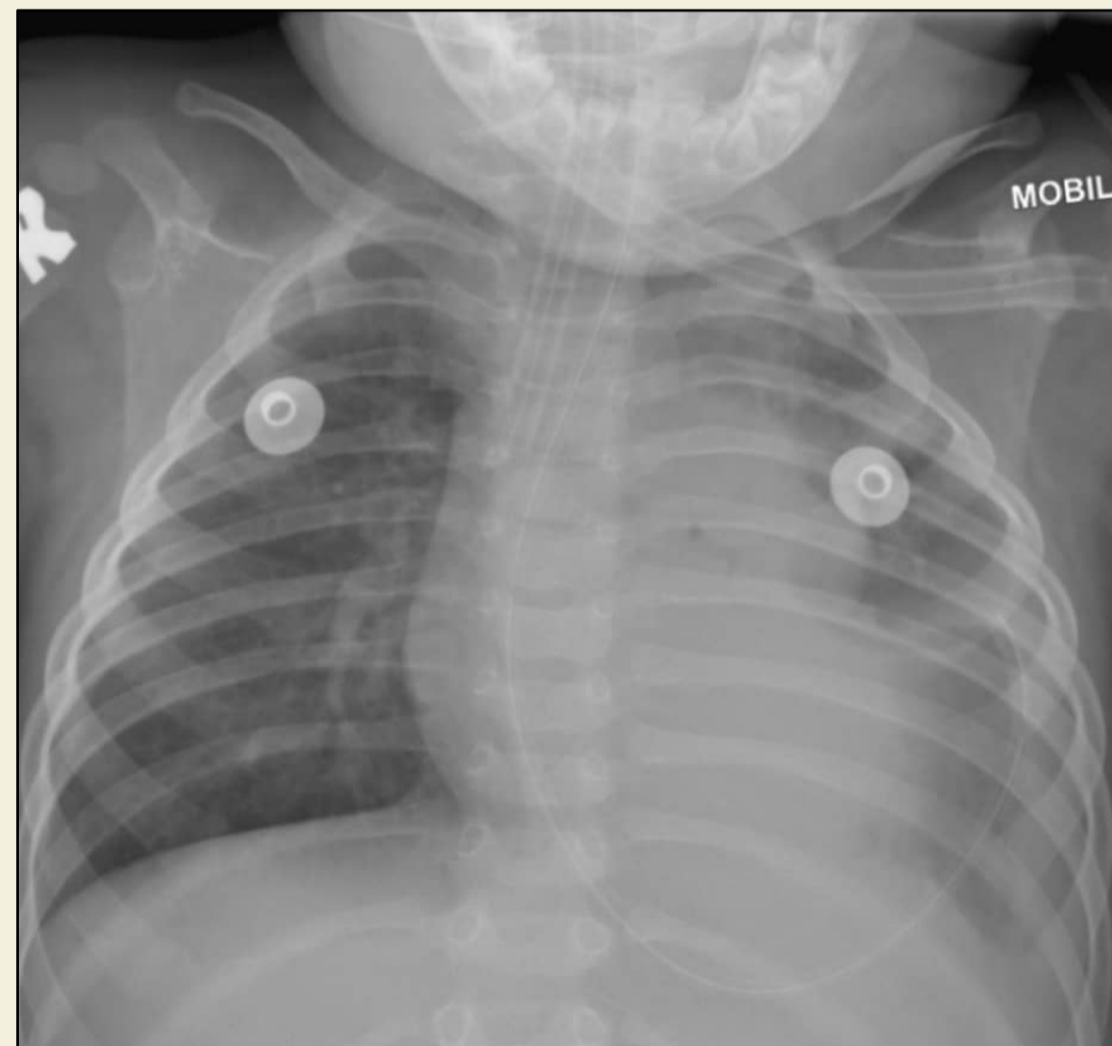


Figure 2: Chest x ray after insertion of nasogastric tube shows resolution of mediastinal shift

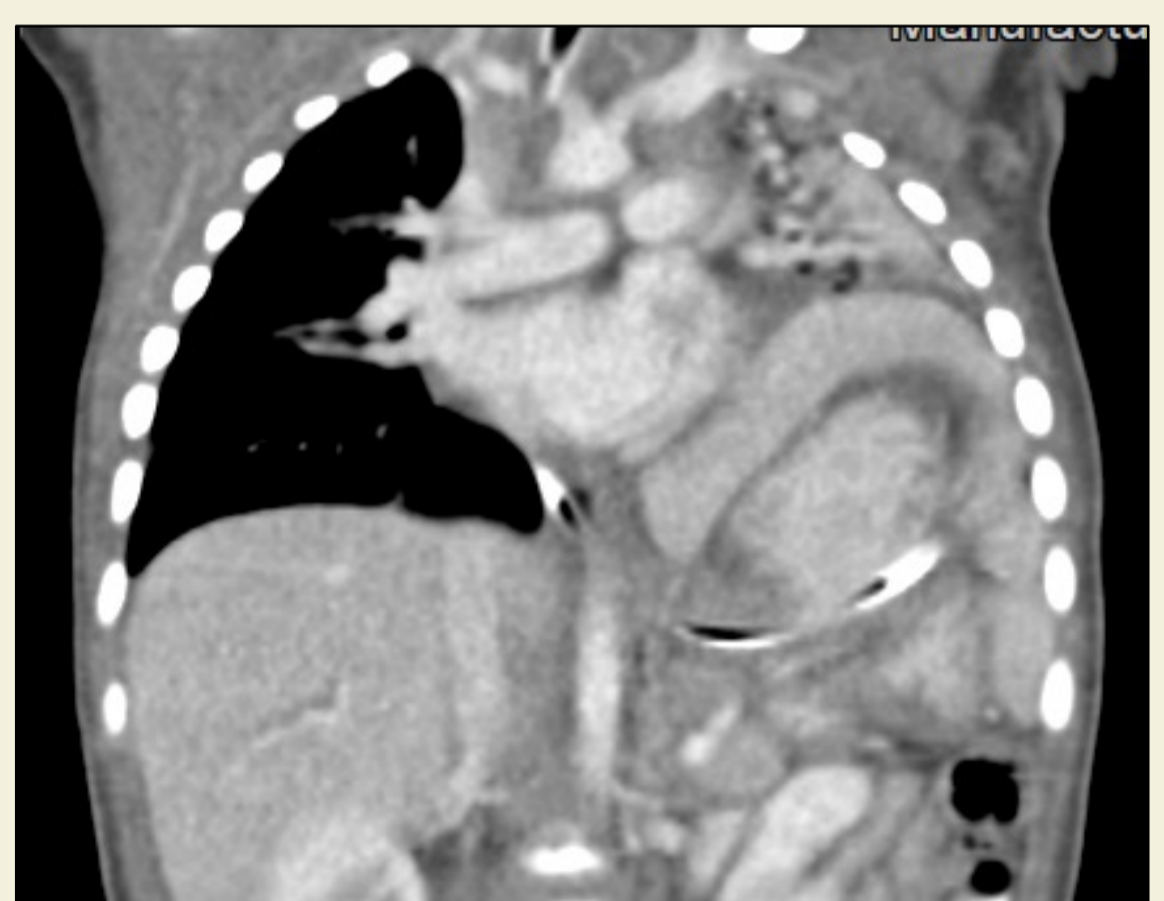


Figure 3: CT scan image showed stomach (with nasogastric tube within) and spleen in the left hemithorax

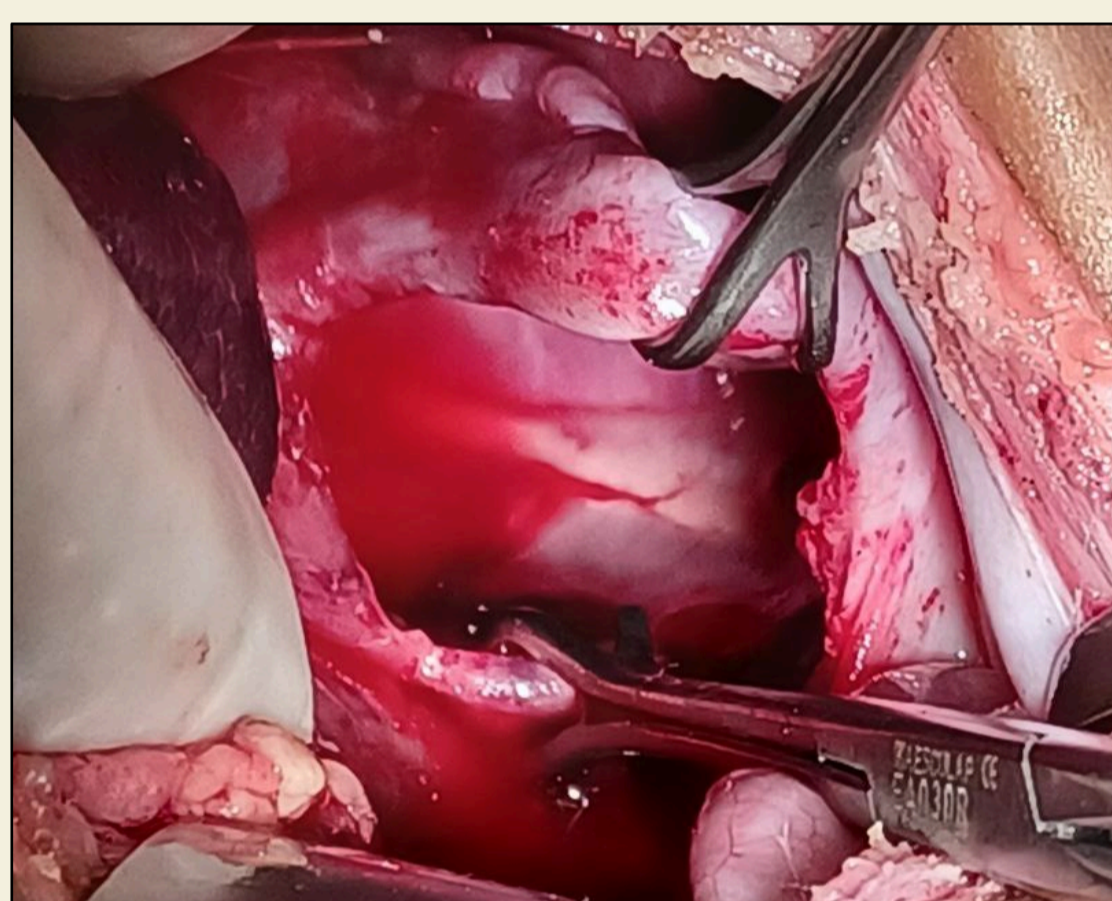


Figure 4: Operative photo shows a left diaphragmatic posterolateral defect seen upon laparotomy

Case 2: A 22-month old boy presented with rapid breathing with reduced air entry on the left side. Chest radiograph revealed a lucent left hemithorax causing mediastinal shift. Patient was misdiagnosed as tension pneumothorax and tube thoracostomy was performed in the emergency department. CT thorax confirmed presence of CDH and surgical referral was made for primary repair. The chest tube was removed safely with no stomach injury (figure 5).

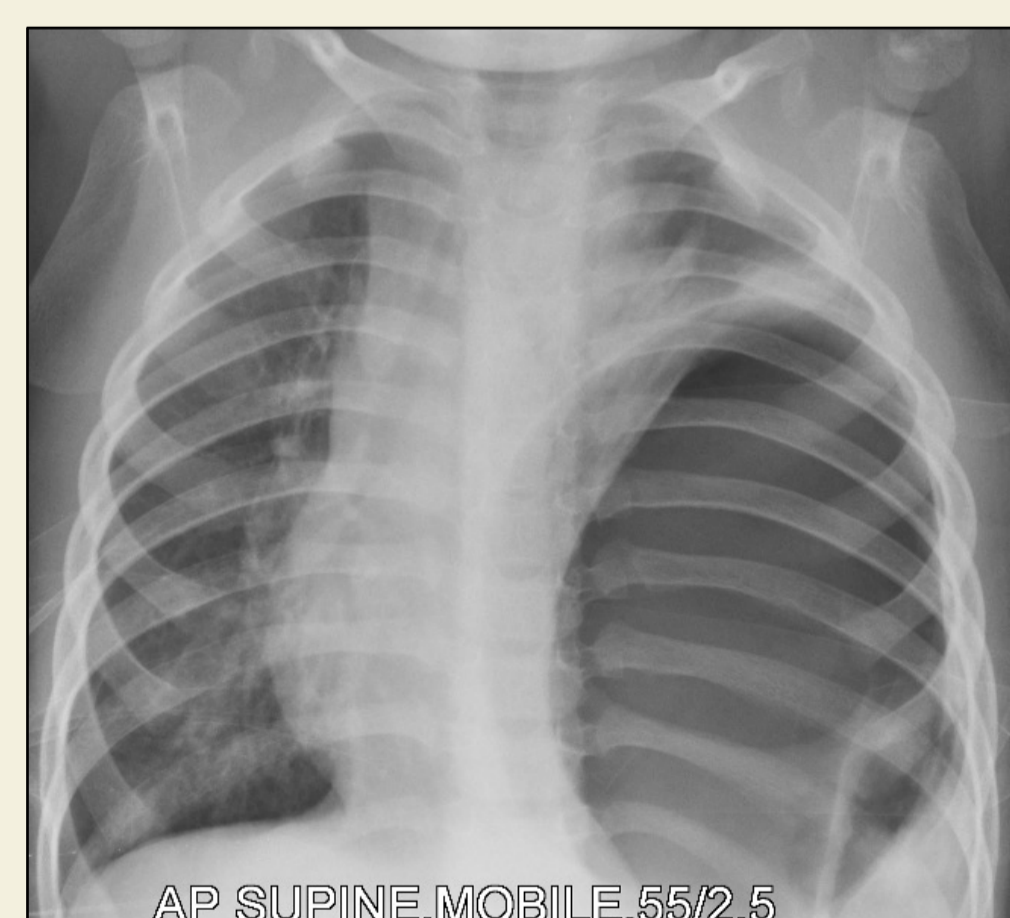


Figure 5: Chest x-ray on arrival which was misdiagnosed as tension pneumothorax

Case 3: A 12-month old boy presented with abdominal pain, rapid breathing and intractable crying. Chest radiograph was suggestive of left CDH. Ultrasound assessment showed a suspicion of left CDH with possible gastric volvulus. Emergency laparotomy and primary repair was performed. Content was reduced completely and the stomach was healthy (figure 6).

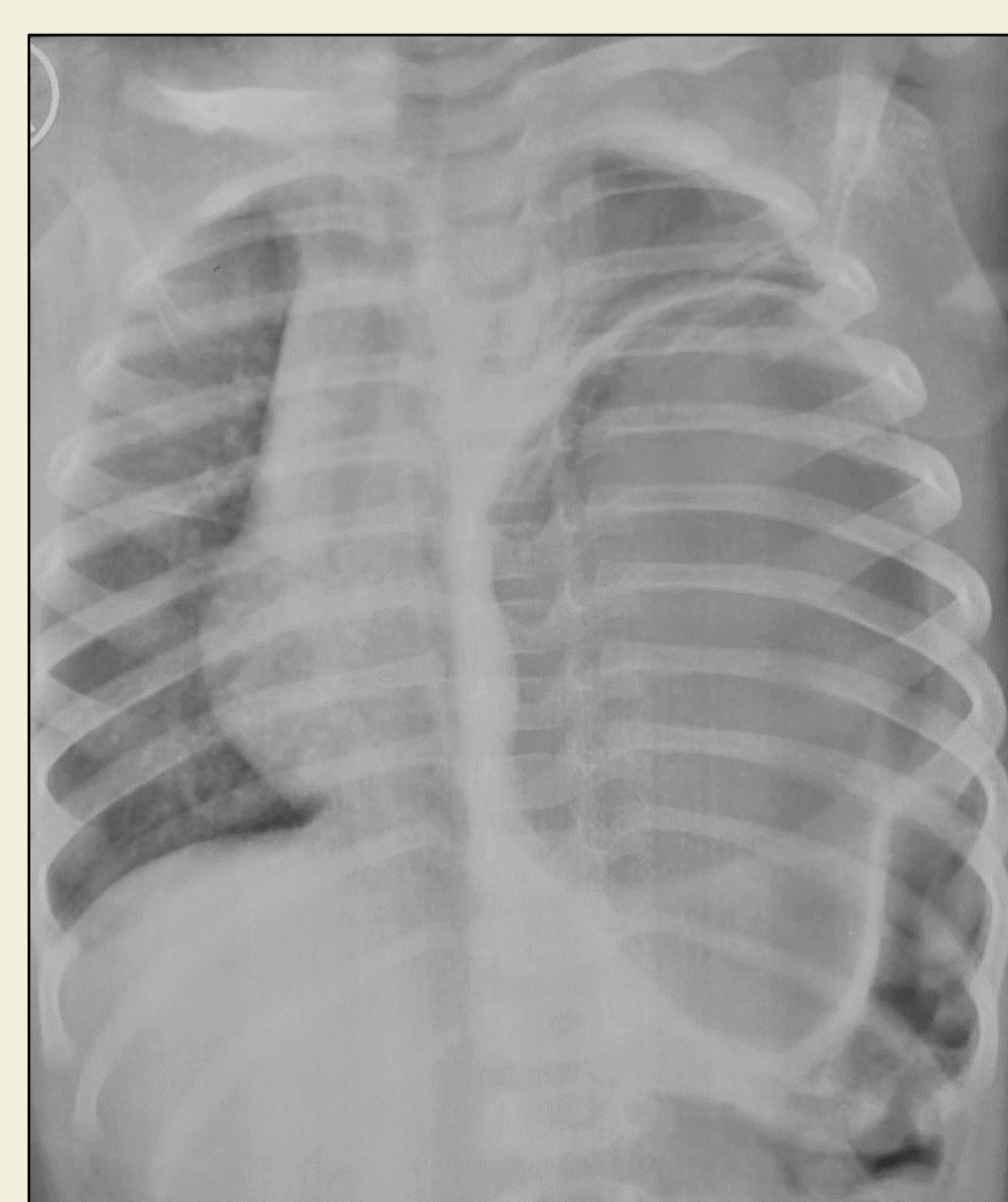


Figure 6: Chest x-ray at presentation suggestive of left diaphragmatic hernia

DISCUSSION

Congenital diaphragmatic hernia is one of the major congenital malformations with high mortality rate of around 30% (1) for the last 2 decades despite recent advancement of medical technology. Patients usually present during the neonatal period. Late-presenting CDH is defined as CDH which are diagnosed after the neonatal period with an incidence as high as 5-45.5% of all cases of CDH (2). It can be complicated with tension gastrothorax. Tension gastrothorax occurs when the stomach that herniated through a congenital diaphragmatic defect into the thorax becomes massively distended by trapped air (3). Majority of gastrothorax in children is contributed by a life-sided CDH and it may evolve into a tension gastrothorax once it is gradually filled up with either air, fluid or food by a one-way valve mechanism, leading to a mediastinal shift, respiratory distress and even cardiac arrest.

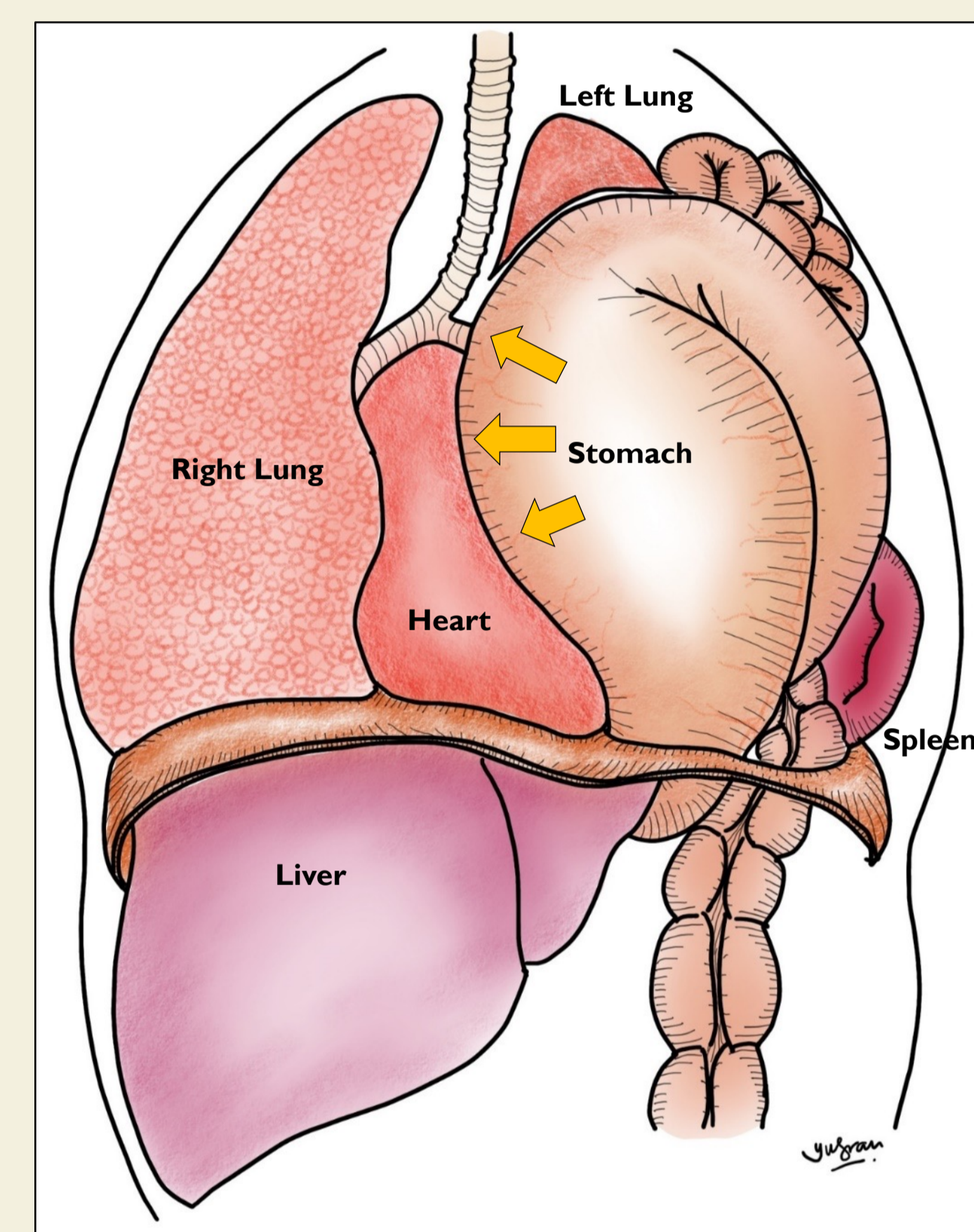


Figure 7: Illustration represents a dilated air-filled stomach within the left hemithorax causing compression to the mediastinal structure (yellow arrow)

Diagnosis of gastrothorax is commonly misinterpreted as pneumothorax and insertion of tube thoracostomy causing stomach perforation has been reported (4). The inadvertent placement of tube thoracostomy may cause organ perforation leading to disastrous consequences. Presence of obstructive symptoms with a clinical finding of audible bowel sounds in the chest may give clues to the attending clinician to carefully correlate with the chest radiograph findings. Correct diagnosis may allow an immediate relief of the mediastinal shift by inserting a large-bore nasogastric tube to decompress the stomach. Definitive surgery can then be performed in a well-planned setting.

CONCLUSION

A late-onset CDH with a tension gastrothorax must be recognised promptly to allow a rapid relief of the life threatening mediastinal shift and also prevent the morbidity of its misdiagnosis.

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