

# Childhood Morgagni hernia: report of two cases

Kamal Nain Rattan<sup>1</sup>, Jasbir Singh<sup>2</sup>, Poonam Dalal<sup>2</sup>

<sup>1</sup>Department of Pediatric Surgery, PGIMS, Rohtak, India

<sup>2</sup>Department of Pediatrics, PGIMS, Rohtak, India

## Abstract

Morgagni diaphragmatic hernia is a rare congenital anomaly to be seen in the pediatric age group. We are reporting two cases of Morgagni hernia, which presented with non-specific symptoms and posed a diagnostic dilemma. One of the patients was 10 years old and associated with asplenia and Down's syndrome; the second case presented as isolated Morgagni hernia in an 8 month baby. The diagnosis was confirmed with radiography and computed chest tomography. Both cases were managed successfully with surgical repair of the diaphragmatic defect through trans-abdominal approach.

## Keywords

Congenital, diaphragmatic hernia, Morgagni hernia.

## Corresponding author

Jasbir Singh, Senior Resident, Department of Pediatrics, PGIMS, Rohtak, India 124001; email: jasbir2001@gmail.com.

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## Introduction

Described first in 1761, Morgagni hernia (MH) consists of herniation of abdominal contents into the thorax through a retro-sternal diaphragmatic defect. It is the rarest type of congenital diaphragmatic hernia (CDH) with reported incidence of 3-5% in all cases [1, 2]. The diaphragmatic defect formation occurs due to defective closure of fibero-tendinous portions of sternal and coastal parts of the diaphragm during embryogenesis [3]. MHs occur most commonly on the right (90%), may be bilateral (8%), with only 2% hernias observed on the left [4]. Pericardial attachment with diaphragm is

thought to be protective of the left side. Most cases of MH usually remain asymptomatic, sometimes even up to adulthood. But when it presents in pediatric patients, the clinical presentation may be atypical and consist of nonspecific respiratory and gastrointestinal symptoms, thereby causing diagnostic dilemma. A high incidence of associated anomalies such as congenital heart diseases, malrotation of gut and Down's syndrome have been observed with MH [5]. Surgical interventions are the treatment of choice even in asymptomatic cases because intestinal obstruction and incarceration, strangulation, or both, may ensue.

## Case presentation

### Case 1

A 10 year old male child presented as a pediatric emergency with cough and fever for 3 days. The patient was hemodynamically stable, febrile and well hydrated. He was found to have Down's syndrome phenotype. Previously he had been treated for similar episodes of recurrent cough at periphery. On auscultation of chest, bilateral air sounds were audible equally and crepitations were present. A chest radiograph (postero-anterior and lateral view) showed presence of dilated bowel loops in thorax in the retrosternal area. A computed tomography (CT) scan chest revealed a right side diaphragm defect immediately behind the sternum with herniation of gut loops, hence the diagnosis of MH was confirmed (**Fig. 1A-D**). Lung parenchyma was normal and there was slight shifting of the mediastinum towards the left side. In a significant finding, spleen was not visualized on computed tomography (**Fig. 1E**) and laterality of various abdominal organs was also not maintained. In view of Down's syndrome features, echocardiography and electrocardiogram were performed to rule out associated congenital heart disease.

After preliminary optimization, the patient was planned for corrective surgery. Vaccination with encapsulated bacterial vaccines was also done in view of asplenia. A supra-umbilical transverse incision was made and the diaphragm defect was identified. It was found to contain transverse colon, omentum and a small part of the liver left lobe. Hernia contents were reduced and the diaphragmatic defect closed with interrupted sutures. The abdominal incision was also closed in layers. Post-surgically, the patient remained stable

and started feeding orally on the 3<sup>rd</sup> postoperative day. Karyotyping was also performed, which confirmed the trisomy 21 chromosome. He was discharged after the 10<sup>th</sup> post-operative day and did well in follow-up.

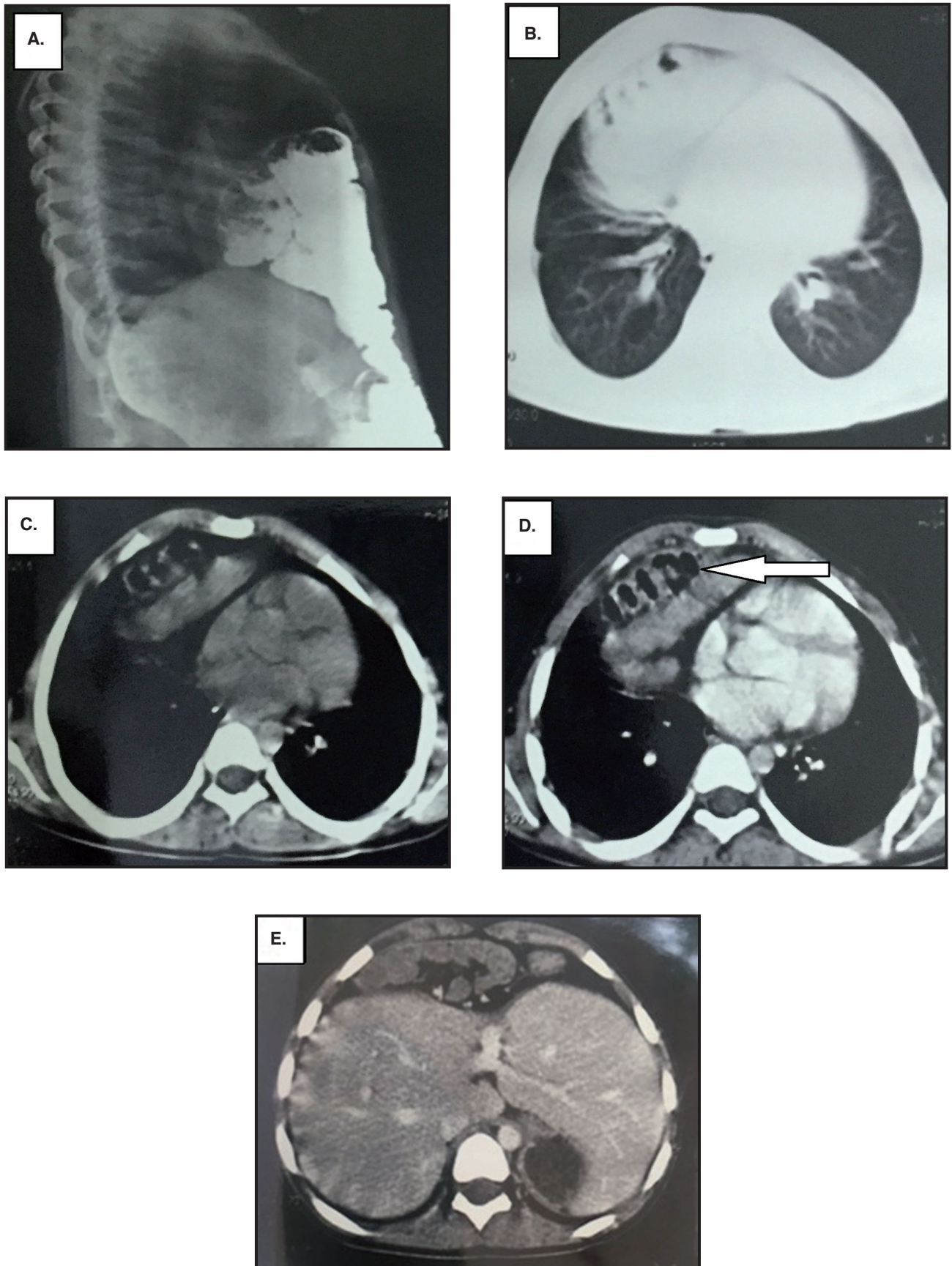
### Case 2

An 8 month old baby presented as a pediatric emergency with recurrent episodes of vomiting and coughs for 2 days. He had previously been treated for cough with salbutamol nebulisation. The baby had stable vital signs but was tachypnic, so investigated further. A chest radiograph showed dilated bowel loops herniating into the thorax via the retrosternal area (**Fig. 2A** and **Fig. 2B**). A CT scan of the chest depicted a well-defined defect in the retrosternal diaphragmatic area through which dilated gut loops were herniating (**Fig. 3**), thus leading to confirmation of the diagnosis of MH. The patient was started on intravenous fluids and broad spectrum antibiotics. After stabilization, he was managed with surgery through the trans-abdominal approach. The hernia sac was found to contain transverse colon with omentum. Hernia contents were reduced and the diaphragmatic defect was repaired with interrupted with non-absorbable sutures. All other abdominal viscera were found at their normal anatomical position. The baby was shifted to ICU for hemodynamic monitoring. Oral feeds were started on the 3<sup>rd</sup> post-operative day. He was discharged on the 8<sup>th</sup> day and did well in follow-up.

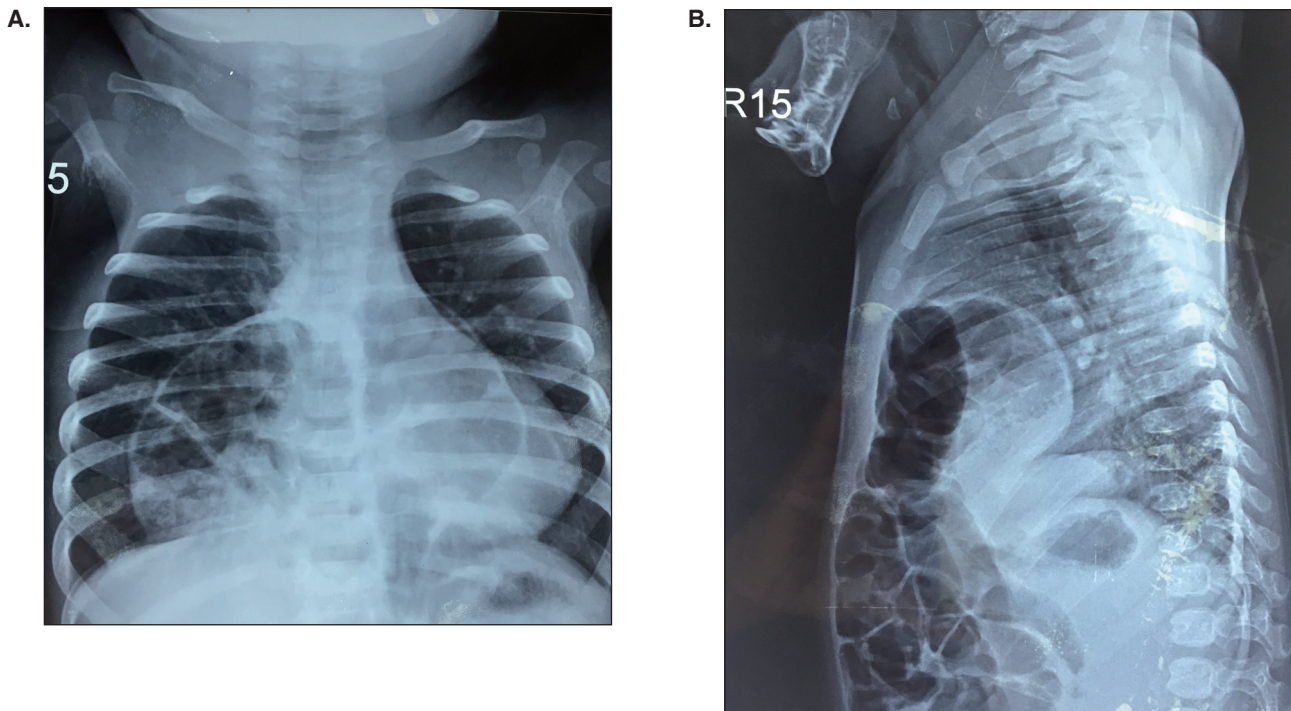
## Discussion

CDHs consist of herniation of abdominal contents in the thoracic cavity. The reported incidence of CDH is from 1 in 2,000 to 1 in 5,000 live births and it accounts for 8% of all major congenital defects [4]. The most common site for CDH is posterolateral Bochdalek hernia (78-90%) followed by esophageal hiatus hernia (14-24%) and retrosternally through the foramen of Morgagni (1.5-6%) [6].

Clinical presentation in MH is quite varied from asymptomatic cases discovered incidentally to life threatening surgical emergencies such as colonic perforation and acute intestinal obstruction. Isolated cases have been reported in neonates, where MH presented as mimicking congenital heart disease [7]. In a review of 36 patients with MH, Aghajanzadeh et al. reported that 50% of



**Figure 1.** Case 1. **A.** Chest radio-imaging (radiograph, barium meal) showing herniated gut loops in thorax retrosternally. **B-E.** Computed tomography (CT) (chest) showing situs ambiguous with asplenia and herniated gut loops in thorax (arrow) through retrosternal defect.



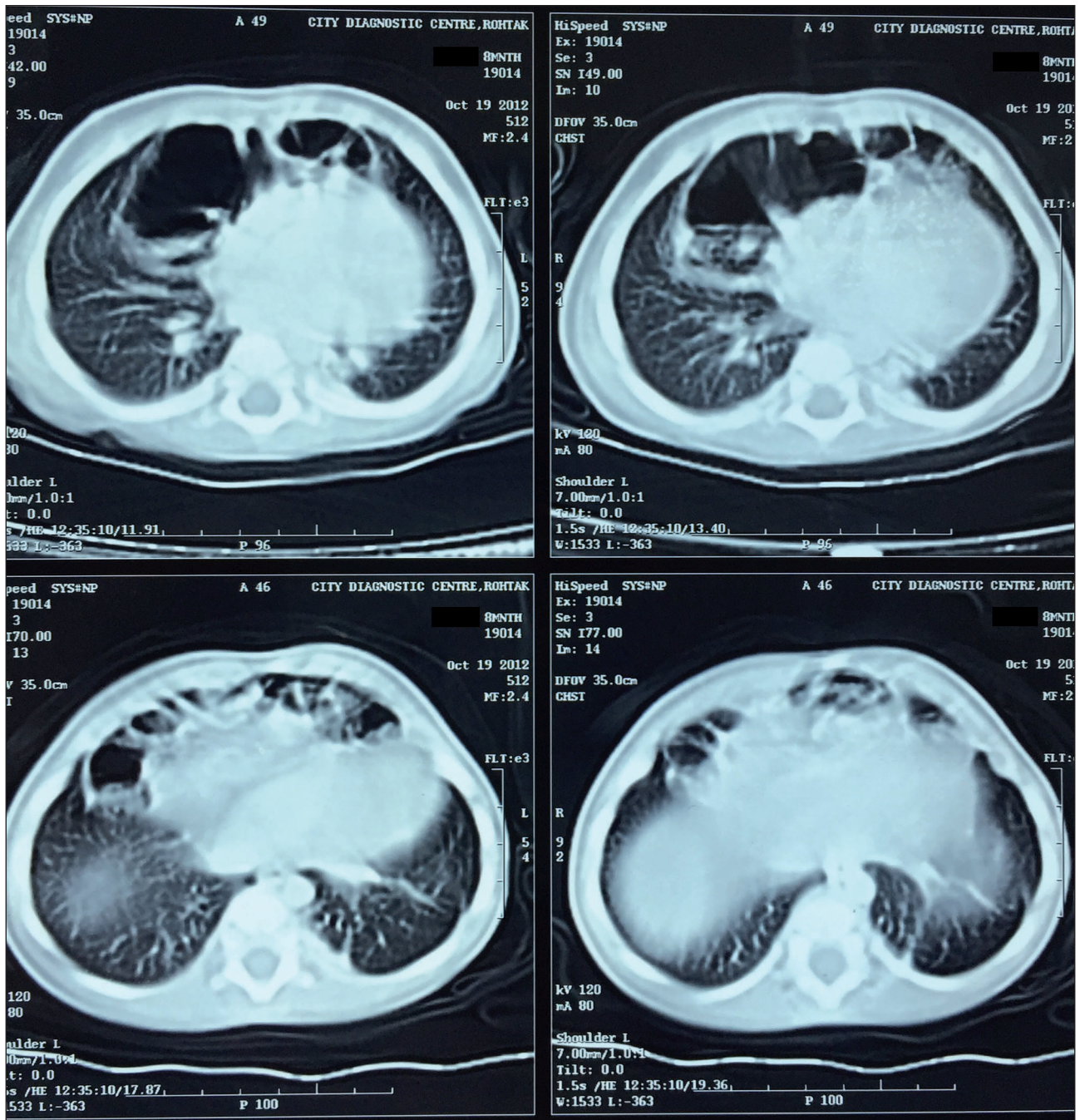
**Figure 2.** Case 2. **A.** A chest radiograph (anteroposterior view) showing herniated gut loops in thorax retrosternally in 8 month old patient. **B.** A chest radiograph (lateral view) showing herniated gut loops in thorax retrosternally in 8 month old patient.

patients remained asymptomatic, 17% presented with respiratory symptoms and 14% with gastrointestinal symptoms [8]. MH presenting as an intra-pericardial tumor has also been reported in literature. Most patients with MH present with repeated attacks of chest infection and nonspecific gastrointestinal symptoms. The diagnosis can thus be delayed and sometimes unnecessary therapeutic procedures may be contemplated in these patients. In some asymptomatic patients, trauma and rapid weight gain act as precipitating factors for various symptoms. The incidence of MH is higher in adulthood as compared to the pediatric age group because of extension of abdominal defects due to increased intra-abdominal pressure with advancing age. Transverse colon and omentum are the most common organs that herniate [9].

MH is also associated with other congenital anomalies with reported incidence as high as 80% [10]. Congenital heart diseases, especially ventricular septum defects, are the most significant anomalies in terms of prognosis. Other associated anomalies include central nervous system malformations, Down's syndrome and gut malrotation. An increased incidence of these anomalies has been observed with bilateral cases. Al-Salem found an incidence of associated CHD in 34.8%, Down's syndrome in 26% and

malrotation of gut in 21.7% of patients with MH [2]. Pokorney et al. reported associations of congenital anomalies in 34.8% in a review of 46 patients diagnosed with MH [11]. These anomalies are of importance in planning the surgical approach and also affect postoperative outcome. Other less common congenital malformations are undescended testis, pyeloureteral stenosis and diaphragm relaxation. Thus a multidisciplinary team approach is necessary to achieve a good outcome in presence of the above anomalies. In the first index case, MH was associated with Down's syndrome, asplenia and situs ambiguity. After a careful search of literature, we failed to find such a type of associated anomaly reported with MH. Asplenia is usually described as part of heterotaxy syndromes and commonly associated with complex cardiac diseases. In the index case with asplenia, situs ambiguous can be considered present as the thoracic and abdominal organs were positioned in such a way that their laterality was not maintained. Our case was also unique because even in presence of asplenia no serious congenital heart disease was detected in the patient.

Diagnosis of MH in the pediatric age may be difficult owing to atypical presenting features. A standard chest radiograph is very useful in arriving at diagnosis by depicting right, left, or bilateral



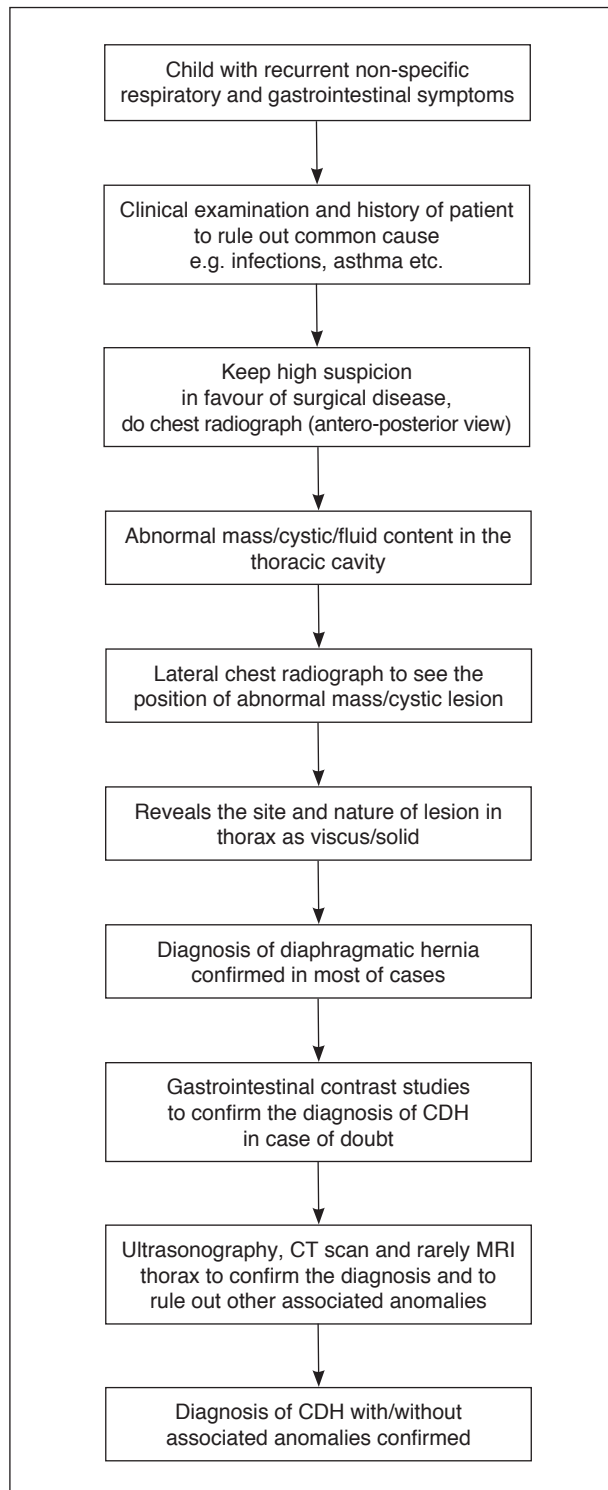
**Figure 3.** Case 2. Computed tomography (chest) showing herniated gut loops in thorax retrosternally in 8 month old patient.

pericardiophrenic angle density. As in index cases, a chest lateral view X-ray confirmed diagnosis of MH by revealing retrosternal herniation of gut loops. However, a differential diagnosis of MH would include cardiophrenic fat pad, diaphragmatic eventration, atelectasis, pneumonia, mediastinal lipoma, abscess, and pleuropericardial cyst. Importantly, a previously normal radiograph does not rule out diagnosis of MH as sometimes the hernia sac may remain empty initially. In the case of doubt, contrast barium studies can

be confirmative by revealing the presence of bowel loops in the thorax. Occasionally, the hernial sac contains liver or omentum, which can be confused with chest mass in radiography. In these situations, ultrasonography (USG), CT and magnetic resonance imaging (MRI) scan can provide useful information [2]. Since in our case asplenia was detected, these investigations have the advantage of diagnosing other associated congenital anomalies which may have treatment and prognostic implications.

Flowchart for suspecting and diagnosis of congenital diaphragmatic hernia is presented (Fig. 4).

Surgical interventions which may be either trans-abdominal or trans-thoracic are the treatment



**Figure 4.** Flowchart for suspecting and diagnosis of congenital diaphragmatic hernia.

CDH: congenital diaphragmatic hernia; CT: computed tomography; MRI: magnetic resonance imaging.

of choice. Most surgeons have preferred the trans-abdominal approach [2, 5]. This approach is easy and has the advantage of detecting associated anomalies such as malrotation of gut, which may be seen in 26% of patients, thereby correcting them simultaneously. Surgery is also recommended in incidentally detected asymptomatic and bilateral hernias, but the timing of surgical interventions is an issue of debate. Minimal invasive techniques are used at most centers nowadays.

Recently, with the emergence of better antenatal diagnostic and therapeutic interventions, management of MH is going to change in future. Zamprakou et al. identified 13 cases of MH prenatally by performing sonographic investigations and successfully managed two patients with antenatal interventions [12]. Antenatally detected lung hypoplasia is the most important factor determining the outcome. By using two dimensional antenatal USG or fetal MRI, we can detect the degree of lung hypoplasia and plan for antenatal intervention. Antenatal interventions are based on the observation that during the intrauterine period, lungs secrete fluid which causes adequate splintage of lung parenchyma. Chronic drainage of this fluid causes hypoplasia of lung and vice versa. Prenatal interventions by thoraco-amniotic shunting and fetal endoscopic tracheal occlusion (FETO) can lead to lung hyperplasia by retention of fluid. But these interventions require infrastructures which are available only in very few centers in developed countries.

In conclusion, congenital MHs are the rarest type of all diagnosed CDHs. They may remain asymptomatic for long periods and usually present in adults. The presentation may be atypical in the pediatric age group, hence MH should always be taken into consideration in the differential diagnosis whenever a child presents with recurrent respiratory and gastrointestinal symptoms.

#### Declaration of interest

The Authors declare that there is no conflict of interest.

#### References

1. Zani A, Cozzi DA. Giovanni Battista Morgagni and his contribution to pediatric surgery. *J Pediatr Surg.* 2008;43(4):729-33.
2. Al-Salem AH. Bilateral congenital Morgagni-Larrey's hernia. *World J Pediatr.* 2010;6(1):76-80.
3. Nasr A, Fecteau A. Foramen of Morgagni hernia: Presentation and treatment. *Thorac Surg Clin.* 2009;19(4):463-8.

4. Keijzer R, Puri P. Congenital diaphragmatic hernia. *Semin Pediatr Surg.* 2010;19(3):180-5.
5. Oleksii S, Kurinnyi S, Ponomarenko O, Migur M. Congenital hernias of Morgagni: Manifestations and treatment in children. *African J Pediatr Surg.* 2016;13(2):57-62.
6. Tibboel D, Gaag AVD. Etiologic and genetic factors in congenital diaphragmatic hernia. *Clin Perinat.* 1996;23(4):689-99.
7. Bassareo PP, Neroni P, Montis S, Tumbarello R. Morgagni hernia mimicking a severe congenital heart disease in a newborn: a case report. *J Med Case Rep.* 2010;4:395.
8. Aghajanzadeh M, Khadem S, Khajeh JS, Gorabi HE, Ebrahimi H, Maafi AA. Clinical presentation and operative repair of Morgagni hernia. *Interact Cardiovasc Thorac Surg.* 2012;15(4):608-11.
9. Roy M, Bala AK, Biswas S, Peters T. Morgagni hernia presenting as Obstructive jaundice. *Ind Pediatr.* 2011;48:59-61.
10. Cidgem MK, Onen A, Okur H, Otcu S. Associated malformations in Morgagni hernia. *Pediatr Surg Int.* 2007;23:1101-03.
11. Pokorny WJ, McGill CW, Harberg FJ. Morgagni hernia during infancy: presentation and associated anomalies. *J Pediatr Surg.* 1984;19(4):394-97.
12. Zamprakou A, Berg C, Strizek B, Müller A, Heydweiller A, Gembruch U, Geipel A. Morgagni hernia presenting with massive pericardial effusion and ascites: prenatal management by thoraco-amniotic shunting and fetal endoscopic tracheal occlusion (FETO) and review of the literature. *Arch Gynecol Obstet.* 2016 Apr 28. [Epub ahead of print].