

The bilateral Morgagni hernia with Down's syndrome: Rare association

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Abstract

Congenital diaphragmatic hernias are a rare condition among paediatric practitioners. Diaphragmatic Morgagni hernia is also a rare condition and its association with Down's syndrome is further rarer.

We present a 18 months old child who presented to emergency department of Kind Abdullah Hospital, Bisha, Kingdom of Saudi Arabia with respiratory tract infection and dyspnoea. There was absence of breath sounds both lung bases. Plain chest radiograph suggested bilateral diaphragmatic defect with suspected loops of gut on both sides of pleural cavity.

Conclusion: To diagnose a rare condition like bilateral Morgagni hernia in childhood, we need to have high index of suspicion. Early diagnosis and treatment may save the life of these children.

Keywords: Congenital diaphragmatic hernia, Bilateral Morgagni hernia, Down's syndrome, Atrial septal defect

Introduction:

Diaphragmatic Morgagni Hernia is an uncommon condition. Bilateral Morgagni hernia is still more rare and its association with Down syndrome is unique.

Case report:

We present this very rare case in an 18 months old male child who was presented to emergency department with history of chest infection and with respiratory distress. He had repeated chest infections in his past history. There were absent respiratory sounds at both lung bases. Plain radiograph of chest was suggestive of bilateral diaphragmatic defects with suspected gut shadow (Figure 1). This bilateral Morgagni hernia was confirmed by barium enema with the presence of bowel loops in the pleural cavities (Figures 2a & 2b). This patient also had atrial septal defect (ASD). The patient had a corrective operative surgery at the age of 02 years (Figures 3,4 & 5). The recovery was uneventful.

Discussion:

Congenital Morgagni hernia defect is less frequent in occurrence. It is one of the form of retrosternal hernia. It results from the failure of fusion of sternal and crural portions of diaphragm and a well defined hernia sac is usually present. The defect in the diaphragm is generally located on the right side 90% or Bilateral 7%; occasionally it may be on the left side, although the presence of both heart and pericardium are a barrier against herniation¹, and rarely cardiac tamponade has been described with this hernia. The lesion rarely presents during the neonatal period, in which case it leads to severe respiratory distress, sometime associated with anomalies in the other organs, including heart.² The rarity of CDH and the non-specific symptoms may lead to a delay in diagnosis, particularly in childhood.³ Half of the Morgagni hernia patient show non-specific symptoms other than mild respiratory discomfort and vague gastrointestinal manifestations.⁴ Although these cases, represents only small percentage with Morgagni

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Figure 1: Plain X-ray of chest



Figure 2: (a) Barium Enema in same child showing retrosternal large bowel loops; (b) Showing with bowel gas Shadows.

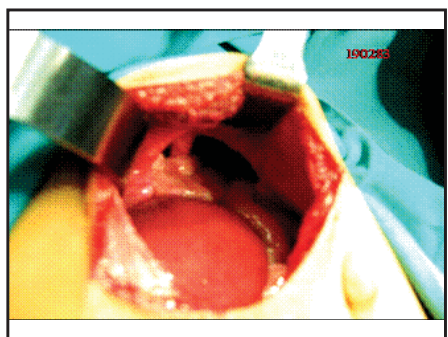


Figure 3&4: Photograph of same patient showing bilateral defect with bowel loops

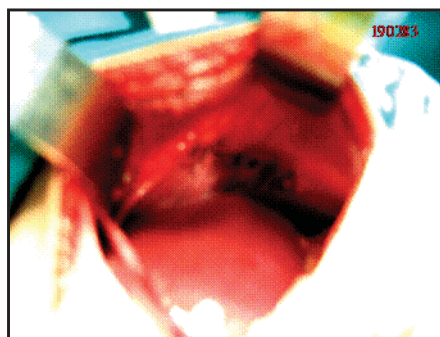
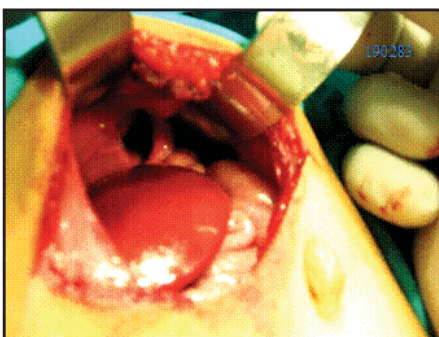


Figure 5: Defects closed

hernia, this suggest that there may be a genetic component involved. Morgagni hernia occurs in fraternal and identical twins, it suggested that genetic factors play a role in diaphragm development.⁵ Newborns may present with respiratory distress at birth similar to Bochdalek hernia, additionally recurrent chest infections and gastrointestinal symptoms have been reported in those with previously undisgnosed Morgagni hernia.^{6,7}

Conclusion:

Bilateral Morgagni Hernia with Down syndrome and associated atrial septal defect is very rare clinical entity. In order to diagnose such condition, we should have high index of suspicion. Physicians caring for these patients should be aware of this, and a high index of suspicion is recommended to obviate delay in diagnosis with its associated morbidity. We advocate surgical repair even in asymptomatic patients. This is to obviate the risk of strangulation and colonic

perforation. We also advocate a transabdominal approach via either an upper midline or an upper transverse incision. This allows easy reduction and inspection of contents, allows access and repair of bilateral hernias, and corrects an associated malrotation if present.

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