Bilateral congenital Morgagni-Larrey's hernia

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Background: **Congenital Morgagni-Larrey's hernia (CMLH) is rare and known to be associated with a high incidence of bilaterality and associated anomalies. This study aimed to review our patients with bilateral CMLH and evaluate their presentation, associated anomalies, diagnostic difficulties and therapy.**

Methods: **From January 1989 to December 2007, we treated 8 children with bilateral CMLH at our hospital. Their medical records were retrospectively reviewed for age, sex, symptoms, associated anomalies, diagnosis, operative findings, treatment and outcome.**

Results: **Among the 8 children, 4 were male and 4 female, with a mean age of 22.74 months (range: 0.93-108 months). Six of them sustained repeated attacks of pneumonia. In 5 children, bilaterality was diagnosed at surgery. Associated anomalies were observed in all children, including congenital heart disease in 5, Down's syndrome in 4, malrotation in 3, inguinal hernia in 2, umbilical hernia in 1, and pyloric stenosis in 1. All of these children were operated on transabdominally. All of them did well postoperatively and on follow-up ranging from 1.5 years to 5 years (mean: 3 years), one had recurrence of the hernia as well as an incisional hernia.**

Conclusions: **The diagnosis of bilateral CMLH may be difficult preoperatively, especially if one of the hernial sacs is empty. CT scan is valuable to diagnose bilateral hernias. To repair these hernias, we advocate a transabdominal approach, which allows easy reduction and inspection of contents, access and repair of bilateral hernias, and correction of associated malrotation if present.**

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Key words: **bilateral Morgagni-Larrey's hernia; congenital diaphragmatic hernia; diagnosis**

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Introduction

Congenital Morgagni-Larrey's hernia (CMLH) is a congenital herniation of abdominal contents into the thoracic cavity through a retrosternal diaphragmatic defect, the foramen of Morgagni. CMLH accounts for less than 5% of all types of congenital diaphragmatic hernias.[1-3] It has unique features in terms of clinical presentation, high incidence of bilaterality and associated anomalies.[3-5] The rarity of CMLH as well as the vagueness, variability and non-specificity of symptoms leads to the delayed diagnosis. In the majority of those with bilateral CMLH the diagnosis of bilaterality is made intraoperatively. This report describes our experience with 8 children with bilateral CMLH outlining aspects of presentation, radiological diagnosis and treatment.

Methods

From January 1989 to December 2007, we treated 23 children with CMLH. Of these patients, 8 (34.8%) had bilateral hernias. The medical records of patients with bilateral CMLH were retrospectively reviewed for age at diagnosis, sex, mode of presentation, associated anomalies, diagnostic X-rays, operative findings, treatment and outcome. The radiological investigations were also reviewed. Post-operatively, the patients were followed up regularly at the clinic for recurrence of symptoms as well as recurrence of the hernia. The follow-up ranged from 1.5 to 5 years (mean 3 years). During the follow-up, chest X-rays were done once every 6 months to rule out recurrence of the hernia.

Results

Of the 23 children with CMLH, 8 (34.8%) had bilateral hernias, 4 were male and 4 female. In all patients with bowel herniation into the chest, the diagnosis was suspected on chest X-ray, especially a lateral chest X-ray and the diagnosis was confirmed by barium enema or meal and follow-through. In those with herniation of solid organs namely part of the liver or omentum, the diagnosis was made on CT scan of the chest.

The clinical and operative findings of the 8 patients are shown in Table 1. The mean age at diagnosis was

22.7 months (range: 28 days to 9 years). All except two sustained repeated attacks of pneumonia, in whom hernia was discovered at surgery. In 5 (62.5%) children, the diagnosis of bilaterality was made at surgery. In the remaining 3, the diagnosis of bilaterality was made preoperatively either on plain chest X-ray (Fig. 1) or contrast studies (Fig. 2) and/or CT scan (Fig. 3). Patient 3 with Down's syndrome was 9 years old at diagnosis. He was asymptomatic and his hernia was discovered incidentally. He sustained a road traffic accident and suffered from a fracture of the left femur, fracture of the left pubic ramus, and a splenic injury. His chest X-ray showed bowel herniation into the left side of the chest, which was confirmed by CT scan. Traumatic diaphragmatic hernia was suspected but at the time of surgery he was found to have bilateral CMLH with hernial sacs (Fig. 4). Retrospectively, there was evidence of bilateral CMLH on CT scan of the chest (Fig. 5) as part of the left lobe of the liver herniated through the right defect. Patient 2, the youngest in our series, presented early at the age of 1 month with respiratory distress, vomiting, poor feeding and tachycardia. His chest X-ray showed bowel herniation into the left side of the chest and on the lateral chest X-ray, this bowel herniation was anterior, confirming the diagnosis of left CMLH. This was documented by barium enema, showing the transverse colon herniating into the left side of the chest. At surgery, he was found to have bilateral CMLH. Retrospectively, there was no evidence of bilaterality on plain chest X-ray or barium enema. Patient 4 was asymptomatic till the age of 6 months when she had pneumonia. She was readmitted 1 month later with a repeated attack of pneumonia. Her chest X-ray was normal at the age of 3 months, but during her last admission there was a right paracardiac opacity not investigated. At the age of 13 months, she was readmitted with another attack of pneumonia. The diagnosis of right CMLH was made on chest X-ray and confirmed by barium enema. Intraoperatively, she was found to have bilateral CMLH. Retrospectively, there was no radiological evidence of bilateral CMLH on chest X-ray or barium enema. Patient 7 was a 13- month-old male and admitted to our hospital with repeated attacks of pneumonia. She was found to have left CMLH which was confirmed by barium enema. By surgery she was found to have bilateral CMLH. Retrospectively,

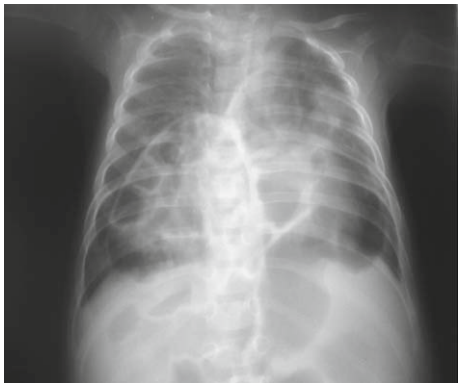

Fig. 1. Chest X-ray showing bowel loops herniating into both sides of the chest in an 8-month-old female with Down's syndrome.

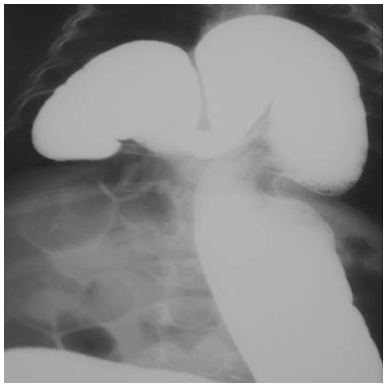

Fig. 2. Barium enema for an 8-month-old female with Down's syndrome showing colonic herniation into both sides of the chest.

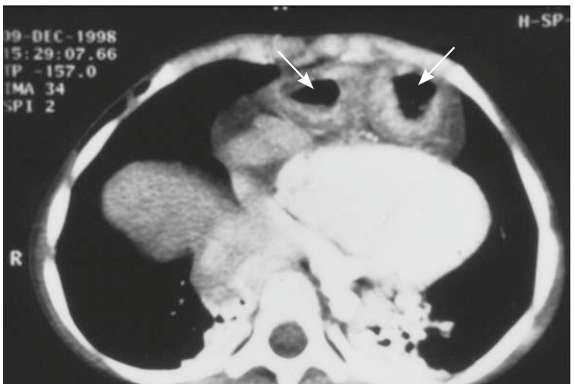

Fig. 3. CT scan of a 9-month-old female child who sustained repeated attacks of chest infection showing bilateral Morgagni's hernia.

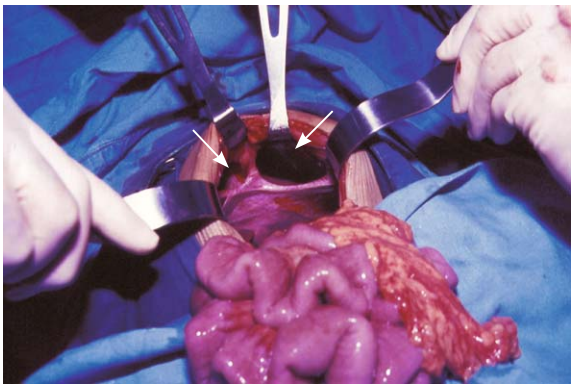

Fig. 4. Intraoperative photograph showing bilateral Morgagni's hernia in a 9-year-old male child who sustained road traffic accident.

Table. Clinical and operative findings in the patients with bilateral congenital Morgagni-Larrey's hernia (CMLH)

No.	Age	Sex	Associated anomalies	Content of hernia	Symptoms	Diagnosis of bilateral hernia	Follow-up and outcome
1	9 mon	Female	Atrial septal defect, pulmonary stenosis	Colon	Recurrent chest infection	Preoperative	No complications
2	28 d	Male	Atrial septal defect, ventricular septal defect	Colon + part of left lobe of liver	Respiratory distress, vomiting, poor feeding and tachycardia	Intraoperative	No complications
3	9 y	Male	Down's syndrome	Colon + small intestines + part of left liver lobe	Road traffic accident, suspected traumatic diaphragmatic hernia	Intraoperative	No complications
4	13 mon	Female	Down's syndrome, atrial septal defect, patent ductous arteriousus, umbilical hernia	Colon + small intestines + part of left liver lobe	Recurrent chest infection	Intraoperative	No complications
5	6 mon	Male	Down's syndrome, atrial septal defect, non-rotation of intestines	Colon	Recurrent chest infection	Preoperative	No complications
6	2 y	Female	Malrotation, left inguinal hernia	Colon + part of left liver lobe	Recurrent chest infection	Preoperative	No complications
7	13 mon	Male	Pyloric stenosis, malrotation, left inguinal hernia	Colon + small intestines	Recurrent chest infection	Intraoperative	No complications
8	8 mon	Female	Down's syndrome, atrial septal defect	Colon + part of left liver lobe	Recurrent chest infection	Intraoperative	Recurrent CMLH and incisional hernia

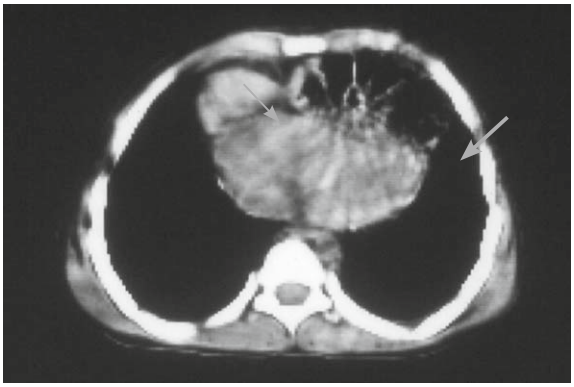

Fig. 5. CT scan of the chest of a 9-year-old male child who sustained road traffic accident showing bilateral Morgagni's hernias. The bowel loops herniated into the left side of the chest (thick arrow) and part of the left lobe of the liver herniated into the right side of the chest (thin arrow).

there was no radiological evidence of bilateral CMLH. Patient 8, an 8-month-old female with Down's syndrome and congenital heart disease, was admitted to our hospital with left CMLH discovered after two attacks of pneumonia, which was confirmed by barium enema. Intraoperatively she was found to have bilateral CMLH. Retrospectively, there was no radiological evidence of bilateral CMLH on chest X-ray or barium enema.

All the patients with bilateral CMLH had associated anomalies compared to 10 (66.7%) of those with unilateral CMLH. One of our patients had hypoplasia of the left lung on a lung perfusion scan and another had deformity of the anterior chest wall. In both, this was secondary to a large CMLH. The associated anomalies in other patients are listed in Table.

All 8 patients were operated on transabdominally (5 upper midline, 3 upper transverse). In all, there was a hernial sac which was excised and the defect was repaired using non-absorbable sutures. The contents of the hernial sac included the colon, part of the liver, and small intestines, with the colon being the most common (Table). There was no mortality and all patients did well post-operatively. The patient with associated lung hypoplasia required postoperative ventilation for 5 days. Follow-up for 1.5 to 5 years (mean 3 years) showed that one (12.5%) of the 8 patients had recurrence of the hernia on the right side 6 months after the initial repair. This patient with Down's syndrome also developed an incisional hernia, which was subsequently repaired. She is currently 2 years old after surgery with no recurrence.

Discussion

Anterior congenital diaphragmatic hernia through the foramen of Morgagni is very rare, comprising 3%-5% of all surgically treated congenital diaphragmatic hernias.[1-7] Embryologically, it results from failure of the fibrotendinous portions of the sternal and costal parts of the diaphragm to fuse. This can be on one side or both sides, more commonly on the right side than the left. It was first described by Giovani Morgagni in 1761.[8] The majority (90%) of CMLHs occur on the right side, 2% on the left side, and 8% are bilateral.[6] The rarity of CMLH on the left side is due to the pericardial attachment to the

diaphragm giving support and protection to that side. In our series, the majority of our patients had right CMLH (39%), but of interest was the finding of bilateral CMLH in 8 (34.8%) of our patients. The reason for the high incidence of bilaterality in our series is unknown.

In the pediatric age group, the presentation of CMLH can be variable. During infancy Morgagni's hernia can lead to acute respiratory distress indistinguishable from that of Bochdaleck hernia.[3] This was the case in one of our patients who presented at the age of 28 days with acute respiratory distress and was found to have bilateral CMLH. At times CMLH remains asymptomatic or discovered accidently during evaluation of other unrelated conditions. Rapid weight gain as well as history of trauma have been reported as contributing factors for the appearance of CMLH which was the case in one of our patients. Most commonly, CMLH patients present with repeated attacks of pneumonia or vague, unspecific gastrointestinal symptoms. Although Berman et al concluded that late presenting CMLH is relatively benign, it causes significant morbidity,[6,8-10] especially if the patient is not adequatly investigated, the diagnosis can be missed or delayed.

The majority of our patients suffering from repeated attacks of pneumonia received several courses of antibiotics and some of them were hospitalized several times. Physicians caring for these patients should be aware of this and infants and children presenting with Recurrent chest infection should be investigated.

CMLH is well known to be associated with other congenital anomalies particularly congenital heart disease, which is reported in up to 80% of patients.[6,7] Eighteen (78.3%) of our 23 patients had associated anomalies. Congenital heart disease was the commonest associated anomaly. This was the case in our series as 34.8% of our 23 patients had congenital heart disease and 5 (62.5%) of those with bilateral CMLH had congenital heart disease. It is well known that CMLH can be associated with Down's syndrome, but the reported incidence is variable. Pokorney et al[3] in a review of 22 infants with CMLH found 3 (14%) of them had Down's syndrome, but in a collective review of 46 children with CMLH, 16 (34.8%) of them had Down's syndrome.[11] Six (26%) of our 23 patients had Down's syndrome and 4 (50%) of those with bilateral CMLH had Down's syndrome. Other associated anomalies included malrotation in up to 26% of the patients.[6] Five (21.7%) of our 23 patients had associated malrotation and 3 (37.5%) of those with bilateral CMLH had malrotation. This is of great importance and must be kept in mind when considering the operative approach as well as intraoperatively to obviate the risk of postoperative volvulus.[10] An interesting finding was the remarkably high incidence of associated anomalies in those with bilateral CMLH compared to those with unilateral hernia. In our series, only 10 (66.7%) of those with unilateral CMLH had associated anomalies compared to a 100% incidence in those with bilateral CMLH. The reason for this variation is not known.

A variety of radiological investigations were used for the diagnosis of CMLH including plain chest X-ray, liver scan, ultrasound, barium enema, barium meal and follow-through, CT scan and MRI.[12-17] In the past, it was our routine in suspected cases of CMLH to do an antero-posterior chest X-ray and a lateral film to demonstrate anterior herniation of bowel loops into the chest. This is usuallly confirmed by a barium enema or meal and follow-through. We found barium enema a useful investigation since the colon is the most common organ to herniate in CMLH. This however is not always the case and at times the diagnosis can be difficult or delayed if the hernial sac is empty or contains omentum or part of the liver. At times, the hernial sac may be empty and so it is common for some of these patients to have a previously normal chest X-ray like in one of our patients which should not preclude a diagnosis of CMLH.[13,15] This is specially so in those with bilateral CMLH where one of the sacs may be empty. In such situations and where there is confusion regarding the diagnosis, ultrasound and/or CT scan proved useful in establishing the diagnosis. CT scan is useful in demonstrating bilateral CMLH even when the sacs are empty. This is specially true on the right side where CT scan can demonstrate continuity between the herniating organ in the paracardiac area whether omentum or part of the liver and the abdominal omentum and liver. We found CT scan a valuable investigation in demonstrating bilateral CMLH. In three of our patients with bilateral CMLH who had CT scan, there was evidence of bilateral hernia. This is of great importance if a transthorasic approach is used to repair the hernia.[18-20] Through a transabdominal approach, both hernial defects can be recognized and repaired. This however is not the case if a transthorasic approach is used. In all cases of CMLH, we advocate a transabdominal approach whether open or laparoscopic. Through the transabdominal approach it is not only easy to reduce the hernial contents and inspect them but also to discover and correct the associated malrotation which is present in up to 26% of patients. Adding to this the fact that bilateral hernias which at times are discovered only intraoperatively can be easily detected and repaired through the transabdominal approach.

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Contributors: Al-Salem AH proposed the study and wrote the first draft. Al-Salem AH is the single author of the paper.

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