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Original Articles

Congenital Morgagni's hernia: A national multicenter study

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ABSTRACT

Background: Congenital Morgagni's hernia (CMH) is rare and represents less than 5% of all congenital diaphragmatic hernias. This is a national review of our experience with CMH outlining clinical presentation, methods of diagnosis, associated anomalies, treatment, and outcome.
Patients and methods: The medical records of all patients with the diagnosis of CMH treated at four pediatric surgery units in Saudi Arabia were retrospectively reviewed for age at diagnosis, sex, presenting symptoms, associated anomalies, diagnosis, operative findings, treatment, and outcome.
Results: During a 20-year period (January 1990–December 2010), 53 infants and children with CMH were treated. There were 38 males and 15 females. Their age at diagnosis ranged from 1 month to 9 years (mean 22.2 months). Forty-three (81%) presented with recurrent chest infection. Twenty-two (44.5%) had right CMH, 15 (28.3%) had left-sided hernia and 16 (30.2%) had bilateral hernia. In 7, the diagnosis of bilaterality was made at the time of surgery. Associated anomalies were seen in 38 (71.7%). Twenty-one (39.6%) had congenital heart disease, 8 (15%) had malrotation, and 15 (28.3%) had Down syndrome. All were operated on. Twenty-nine (54.7%) underwent repair via an open approach. The remaining 24 (45.3%) underwent repair using minimal invasive surgery, laparoscopic-assisted hernia repair (19 patients) or totally laparoscopic approach (5 patients). At the time of surgery, the hernia sac content included the colon in 33 (62.3%), part of the left lobe of the liver in 13 (24.5%), the small intestines in 11 (20.75%), the omentum in 5 (9.4%), and the stomach in 4 (7.5%). In 12 (22.6%), the hernia sac was empty. When compared to the open repair, the laparoscopic-assisted approach was associated with a shorter operative time, an earlier commencement of feeds, less requirement for postoperative analgesia, a shorter hospital stay, and better cosmetic appearance. There was no mortality. On follow-up, 2 (7%) of the open surgical group developed recurrence.
Conclusions: CMH is rare and in the pediatric age group commonly presents with recurrent chest infection and has a high incidence of associated anomalies, commonly congenital heart disease and Down syndrome. We advocate a laparoscopic-assisted approach to repair CMH. This is a simple technique that produces a sound repair, and when compared with the open approach it takes less operative time, requires less analgesia, allows earlier commencement of feeds, is associated with a shorter hospital stay, and has a better cosmetic outcome.

Congenital Morgagni's hernia was first described by Giovani Morgagni in 1761 [1,2]. It is a congenital herniation of abdominal contents into the thoracic cavity through a retrosternal diaphragmatic defect. CMH is relatively rare and make up about 1%–5% of all types of congenital diaphragmatic hernia, and although commonly diagnosed in the pediatric age group, it can remain asymptomatic till adulthood [3,4].

In the pediatric age group, CMH has unique features in terms of clinical presentation, high incidence of bilaterality and associated anomalies. CMH commonly presents with repeated attacks of chest infection and rarely vague, nonspecific gastrointestinal symptoms but at times it remains asymptomatic or discovered accidently during evaluation of other nonrelated conditions. The rarity of CMH and the vague nonspecific symptoms are two important factors leading to delayed diagnosis. CMH nevertheless can cause significant morbidity [2]. This is specially so if the child is not adequately investigated. Awareness of this is of paramount importance. During infancy CMH however can lead to acute respiratory distress indistinguishable from that of Bochdalek hernia [5].

The treatment of CMH is surgical repair either conventionally by open abdominal or thoracic approaches, or more recently, by using

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minimal invasive surgery [2,6–11]. This study is a national review of our experience with CMH highlighting aspects of presentation, diagnosis, associated anomalies and their influence on outcome. The methods of treatment are discussed and a comparison between the conventional open technique and the laparoscopic-assisted repair of CMH is presented.

1. Patients and methods

Four pediatric surgery units in the kingdom of Saudi Arabia participated in this study. The medical records of all patients with the diagnosis of CMH during the study period from January 1990 to December 2010 were retrospectively reviewed and the following information was collected: age at diagnosis, sex, clinical presentation, associated anomalies, site of hernia, and methods of repair, postoperative complications and outcome.

2. Results

During the study period, 53 infants and children with CMH were treated. There were 38 males and 15 females (M/F of 2.5:1). Their mean age at diagnosis was 22.2 months (1 month–120 months). The majority (81%) presented with recurrent chest infection (Table 1). Five had nonspecific upper respiratory tract symptoms and two were treated as gastroesophageal reflux disease prior to diagnosis. In one, the hernia was discovered incidentally. One of our patients had a road traffic accident with blunt abdominal trauma and an abdominal CT scan noted bilateral CMH. This was confirmed intraoperatively where bilateral CMH with hernia sacs were found containing intestines. The last patient had beta-thalassemia major and underwent splenectomy because of frequent blood transfusion. Following this and as a result of progressive liver enlargement, the left lobe of the liver herniated into an already existing left Morgagni's hernia. Twenty-two (41.5%) patients had right-sided CMH, 15 (28.3%) had left-sided hernia and 16 (30.2%) had bilateral hernia. In those with bilateral hernias, the diagnosis of bilaterality was made at the time of surgery in 7.

Associated anomalies were seen in 38 (71.7%) as shown in Table 2. A single associated anomaly was seen in 17 while 21 had more than one anomaly. Congenital heart disease was the commonest associated anomaly seen in 21 patients (39.6%). Down syndrome was noted in 15 patients (28.3%) and all of them had congenital heart disease. Eight (15%) had associated inguinal hernia, 4 (7.5%) had umbilical hernia and 2 had hydrocephalus.

In the majority (50 patients), plain chest radiographs (anteroposterior and lateral views) were diagnostic showing anterior herniation of bowel into the chest (Fig. 1). Additional diagnostic investigations included barium enema (12 patients), barium meal and follow-through (7 patients) and CT scan of chest (5 patients) (Figs. 2 and 3).

All patients underwent operative repair. The method of repair changed over the years. In the initial stages of the study, 29 (54.7%) underwent repair via an open approach. The remaining 24 patients underwent repair using a minimally invasive approach with laparoscopic-assisted hernia repair (19 patients) or totally laparoscopic approach (5 patients). A comparison between the open approach and the laparoscopic-assisted approach is shown in Table 3. At the time of surgery, the hernia sac content included the colon in 33 (62.3%), part of the left lobe of the liver in 13 (24.5%), the small intestines in 11 (20.75%), the omentum in 5 (9.4%) and the stomach in 4 (7.5%). In 12 (22.6%), the hernia sac was empty. In all patients who had open repair, the hernia sac was excised and the repair was done using nonabsorbable sutures. The hernia sac was not excised and plicated in those who had minimal invasive surgery. Postoperatively, all did well except two patients in the open-surgery group (7%) who developed recurrence and required reoperation. One of them also had incisional hernia which was repaired. Both of them had Down syndrome.

Table 1
Clinical features of patients with congenital Morgagni's hernia.

Clinical features	No. of patients	%
Recurrent chest infection	43	81
Nonspecific upper respiratory tract symptoms	5	9.4
Gastroesophageal reflux disease	2	3.8
Discovered incidentally	1	1.9
Following road traffic accident with blunt abdominal trauma	1	1.9
Following splenectomy	1	1.9

Table 2
Associated anomalies.

Associated anomalies	No. of patients	%
Congenital heart disease	21	39.6
Down's syndrome	15	28.3
Inguinal hernia	8	15
Malrotation	8	15
Umbilical hernia	4	7.5
Hypospadias	4	7.5
Anorectal malformation	2	3.8
Hydrocephalus	2	3.8
Undescended testes	1	1.9
Hemangioma	2	3.8
Scoliosis	1	1.9

3. Discussion

Congenital diaphragmatic hernia through the foramen of Morgagni is an interesting condition from several aspects including its rarity, high incidence of bilaterality, presentation, and high incidence of associated anomalies [12–15]. It is relatively rare comprising 3%–5% of all surgically treated congenital diaphragmatic hernias. Over a period of 40 years, Berman et al. [15] treated only 18 cases of CMH and Pokorney et al. [5] saw only 4 cases over a period of 25 years. Cigdem et al. [12] over a period of 23 years treated 16 cases of CMH. Al-Salem [13] over a period of 18 years treated 23 cases of CMH. The exact incidence of CMH in Saudi Arabia is not known. In a previous study, CMH made up about 11% of all types of congenital diaphragmatic hernia [16]. This is relatively high when compared to that reported internationally. One contributing factor for this is the high rate of consanguinity in this part of the world.

The majority (90%) of CMH occur on the right side and 2% on the left side and 8% are bilateral. The rarity of CMH on the left side is caused by the pericardial attachment to the diaphragm which is more on the left side giving support and protection to that side. Twenty-two (41.5%) of our patients had right-sided hernia, 15 (28.3%) had left-sided hernia and 16 (30.2%) had bilateral hernia. The reason for the high frequency of bilateral hernias in our patients is not known. An interesting point in our series is the high frequency of bilateral hernias diagnosed intraoperatively. One reason for this is the presence of an empty sac at the time of radiological evaluation. This must be kept in mind and looked for intraoperatively.

Another interesting point is the increased incidence of associated anomalies in patients with CMH but the incidence is variable ranging from 34% to 50% [5,12,13]. Thirty-eight (71.7%) of our patients had associated anomalies. This is higher than that reported internationally. Congenital heart disease continues to be the commonest associated anomaly. This was the case in our series as 21 (39.6 %) of our patients had congenital heart disease. The severity of associated congenital heart disease was variable but none of them had major life-threatening defects and atrial septal defect and ventricular septal defect were the commonest associated heart defects. This however