Cent. Eur. J. Med. • 6(1) • 2010 • 131-135
DOI: 10.2478/s11536-010-0061-2

VERSITA

Central European Journal of **Medicine**

Congenital herniation through the foramen Morgagni – clinical presentation, diagnosis and treatment in pediatric population

Case Report

Slavković Anđelka[1], Marjanović Zoran[1], Đorđević Ivona[1*], Jovanović Zorica[1], Budić Ivana[2]

[1] *Pediatric Surgery Clinic, 18000 Niš, Serbia*

[2] *Pediatric Surgery Clinic, Department of Anestesiology, 18000 Niš, Serbia*

Received 15 December 2009; Accepted 11 October 2010

Abstract: Congenital Morgagni hernia is a rare entity, accounting for less than 1,5% of all types of congenital diaphragmatic hernias. The majority of Morgagni hernias are diagnosed late because patients can be asymptomatic or present with non-specific respiratory and gastrointestinal symptoms and signs. The medical records of all patients diagnosed with CMH and treated in our hospital were retrospectively reviewed for age at diagnosis, sex, site of hernia, clinical symptoms, associated anomalies, operative findings and treatment. Over a 20-year period, 5 cases with CMH diagnoses were hospitalized and operated. The age of diagnosis ranged from nine months to 11 years. Male to female ratio was 3:2. Associated anomalies were seen in 2 patients (40%). Most patients had transabdominal operations. There were no complications in the postoperative period. Morgagni hernia is a rare condition. The rarity is due to the nonspecific presentation of symptoms, which contributes to a delay in diagnosis. We advocate surgical repair even in asymptomatic patients, because of the risk of strangulation. There is a low rate of complications.

Keywords: *Retrosternal hernia • Foramen Morgagni*

1. Introduction

Herniation of abdominal organs into the thoracic cavity through a congenital defect in the retrosternal space (foramen of Morgagni) between the 7th rib and processus xyphoideus is a rare phenomena in children, called congenital Morgagni hernia (CMH). Normal fusion of the pleuroperitoneal membrane with septum transversum in the 8th fetal week makes up the diaphragm. Morgagni hernia occurs as the result of an embryological defect in the septum transversum between the lateral aspect of the diaphragm and the anterior chest wall. In the pediatric age group the presentation of CMH is variable, but it can remain asymptomatic until it is discovered accidentally in adulthood. This rare entity accounts for less than 1,5% of all types of congenital diaphragmatic hernias (CDH) [1].

In severe cases CMH may present with symptoms of bowel obstruction or strangulation. Rarely, it may present in the neonatal period (with acute respiratory distress syndrome, cyanosis, cough, vomitus, and repeated episodes of chest infection), although the majority of Morgagni hernias are diagnosed later in life because patients can be asymptomatic or present with non-specific respiratory and gastrointestinal symptoms and signs. CMH can be diagnosed accidentally on standard plain radiograph of the lateral chest. High incidence of associated anomalies (32-50%) is described with hernia of Morgagni: congenital heart anomalies, Turner's syndrome, pentalogy of Cantrell and omphalocele. Patients with Down's syndrome have increased risk of CMH [2].

Depending on the hernia contents – omentum, stomach, small intestine, colon or liver – CMH can

* *E-mail: ivonadj74@gmail.com*

Table 1. All patients with congenital diaphragmatic defects admitted to our hospital from 1988 to 2008.

Type of hernia	No of patients	Percentage(%)
Posterolateral	8	44,5
Morgagni hernia	5	27,8
Eventeration of diaphragm	4	22,2
Paraoesophageal hernia	1	5,5

Table 2. Presentation of CMH by sex, age, site, signs, associated anomalies and surgical approach.

	Age/sex	Clinical symptoms	site	Associated anomalies	Contents of hernia sac	Surgical approach
1	M/11years	Recurrent chest infection	Right	/	Omentum,colon transversum	Thoracotomia anterolateralisdex.
2	F/4 years	Asimptomatic	Right		Omentum,colon transversum	Thoracotomia anterolateralisdex.
3	F/2 years	Recurrent chest infection, vomiting	Right	ASD	OmentumColon transversum	Laparotomiasupraumbilicali stransversalis
4	M/ 1 year	Recurrent chest infection	Right	/	Colon transversumIleum	Laparotomia Subcostalis dex.
5	M/3 years	ARDS, vomitus	Right	Sy Down,Malrotation	Omentum,Colon transversum,Colon ascendens,Part of liver	Laparotomia subcostalis dex.

appear differently on chest radiography. Diagnosis of CMH is much more obviously made if there are abdominal organs in the hernia sac. On standard plain AP radiography, the hernia sac would present with shadow resembling air, highly suspect for intestine. On lateral chest radiography, there would be anterior herniation of bowel loops (located retrosternally). Contrast examination (barium enema) confirms the diagnosis. In rare cases of intermittent herniation, radiography can be absolutely normal. In these cases chest echosonography examination is useful. Computered tomography can also be considered as a non-invasive method to diagnosis CMH. MRI and radionucleotide liver scan may help with diagnosis, especially when the liver is in the hernia sac [3].

On differential diagnosis pneumonia, atelectasis, intrathoracic tumor or echinococcal cyst should be considered.

The need for surgery depends on presentation. Although the majority of hernias are asymptomatic, repair is recommended to avoid future complications. In general, all symptomatic patients should receive surgical repair, but operating on asymptomatic patients is controversial. In cases when the colon is in the hernia sac, operation is indicated to prevent perforations and decrease risk of obstruction and strangulation [4]. Detection of intestinal herniation through an original or recurrent CMH defect is often delayed or missed because its radiologic appearance on routine chest X ray may be normal.

Treatment options include transabdominal, transthoracic or laparoscopic repair. Most surgeons prefer the transabdominal approach because of the possibility of missing other preexisting diseases in the abdominal cavity. The transthoracic approach provides wide exposure and easy repair of hernia sac [5]. The first laparoscopic repair was performed by Kuster in 1992 [6].

2. Material and Methods

This is a retrospective review of 18 infants and children admitted to the Pediatric Surgery Clinic, Nis, from 1988 to 2008, with different types of congenital diaphragmatic defects (Table 1). In five cases diagnosis of Morgagni hernia was confirmed. Diagnosis was made on clinical symptoms, plain radiography, barium contrast enema and CT. Medical charts were reviewed for age at diagnosis, sex, site of hernia, clinical symptoms, operative treatment and contents of hernia sac (Table 2).

3. Results

Over the period of 20 years, 5 patients with a diagnosis of CMH were treated in the Pediatric Surgery Clinic, Nis. The relevant demographic, clinical and operative findings are presented in Table 2. Male to female ratio was 3:2. Their ages at presentation ranged from 9

Figure 1. Standard chest radiography with bowel loops in the right hemithorax.

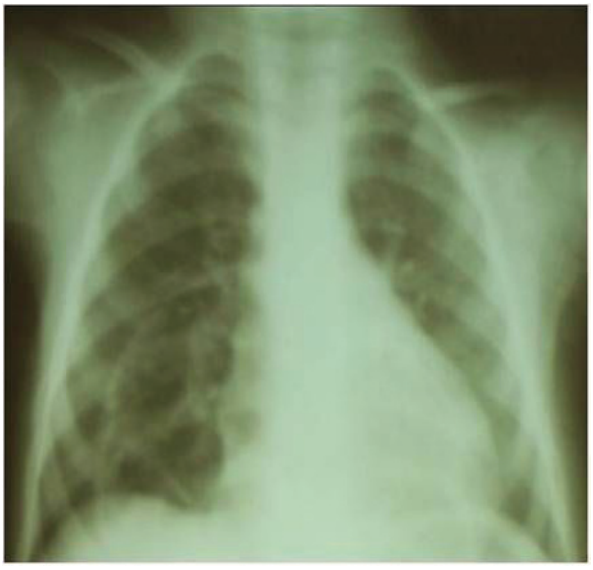
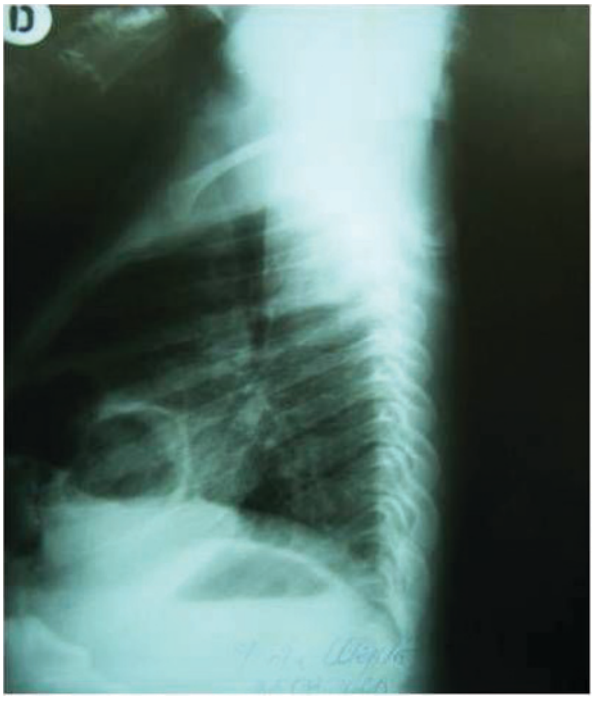


Figure 2. Lateral chest radiography confirms bowel loops retrosternally.



months to 11 years (mean 4,9 years). All of the patients had right-sided hernia. The majority (60%) presented with large number of recurrent chest infection, since early infancy.

Patients 1 and 3 had repeated chest infections since birth, one of which necessitated admission to the intensive care unit. Plain chest and lateral chest radiography suggested CMH (Figure 1 and 2). Definitive diagnoses were made by barium enema examinations and CT scans.

Patient 4 was asymptomatic until 8 months of age, when he developed pneumonia. Two months later, the patient was readmitted to the hospital with the same diagnosis. Plain chest radiography and barium enema confirmed the diagnosis.

Patient 5 had Down's syndrome and had ARDS, poor feeding and vomiting at 3 years of age when he was referred to our hospital. Routinely performed plain radiography showed highly suspicious bowel loops in the right thoracic cavity, from the level of the 2nd rib to the right phrenocardial space. Lateral chest radiography presented polycystic tumor-like structures filled with gas and located retrostrenally. Diagnosis of CMH was confirmed by barium enema examination and CT scan.

In all 5 cases hernia was right-sided. Diagnosis was suspected on plain chest X ray films when bowel loops were located anteriorly in the thoracic cavity, behind the sternum. Barium enema examinations were performed in all patients. In those with herniation of solid content, namely part of the liver or omentum, diagnosis was made based on CT scan of the chest.

Associated anomalies were seen in 2 patients (40%), as showed in Table 2. One patient had congenital heart disease, and one had malrotation of bowel and Down's syndrome.

Three patients (60%) were operated on transabdominally (2 subcostal and 1 transversal supraumbilical laparotomy). Two patients (40%) had right anterolateral thoracotomy. A hernia sac was present in all patients. In 3 cases the hernia sac was ligated and the diaphragmal defect repaired with non-resorptive sutures, although in the other 2 cases, after ligation of the hernia sac, diaphragm repair was reconstructed by posterior sternal fascia. There were no complications in the postoperative period.

4. Discussion

Diaphragmatic hernia is a congenital defect, well known since the 17th century, with incidence of about 1 in 2000-5000 live births. Most diaphragmatic hernias occur posterolaterally, but about 5% of them present retrosternally and are called Morgagni hernia, named after the Italian anatomist Govani Morgagni who first described the phenomena in 1761. The foramen of Morgagni is a small anterior subcostosternal defect that extends from the sternum medially to the 8th costal cartilages laterally. Hernias result from failure of the

fibrotendinous portion of the pars sternalis to fuse with the fibrotendineus part of the chondral arches. This space is usually filled with adipose tissue, covered with peritoneum from above and pleura as the upper layer. The space is small, but a rapid rise in intraperitoneal pressure can cause prolapse of abdominal viscera into the thoracic cavity, giving rise to a hernia.

Fusion of the pleuroperitoneal membrane with the septum transversum in the 8th fetal week makes up the diaphragm. Morgagni hernia occurs as a result of the embryological defect in the septum transversum between the lateral aspect of the diaphragm and the anterior chest wall. Embryology of the diaphragm can explain morphological changes that can lead to CMH: 1. pre-hernial lipoma, which is always present in early embryologic stages, is considered to penetrate the retrosternal space dragging the peritoneum along; and 2. CMH is a result of malformation in the non-muscular mesenchymal component prior to altered myogenesis [7].

CMH is an extremely rare condition in the pediatric age group, with few cases reviewed in literature. Berman et al. reported only 18 patients with CMH over a 40 year period [8]. Pokorney et al., in series of 74 patients with congenital diaphragmatic hernia over a period of 25 years, reported only 4 cases with CMH [9]. CMH can be associated with other anomalies, but the reported incidence is variable. Patients with Down's syndrome in particular have an increased risk of Morgagni hernia, caused by generalized hypotonia and muscle weakness that can be associated with ventral hernia and rectal diastasis. In our study we presented one patient with Down's syndrome and malrotation.

Morgagni hernia occurs more commonly in males than in females. The majority occurs on the right side (90%), about 2% are bilateral, and 8% are left-sided. Left-sided CMH is rare because of the reinforcing effect of the heart, pericardium and its diaphragmatic attachment [10]. Hernia sac contents include colon, small intestine, part of the liver, omentum, and stomach, as the most common abdominal viscera to herniate.

The majority of CMH are diagnosed late because patients can be asymptomatic or present with non-specific respiratory and gastrointestinal symptoms and signs. The symptoms and signs in obstructive recurrent CMH may be non-specific unless bowel complications occur. Misdiagnosis is not uncommon. In the pediatric age population, clinical presentation can vary from asymptomatic cases to those with ARDS in the early neonatal period. Fortunately in most cases, clinical signs are presented late in childhood with recurrent chest infections, vomitus, bloating and indigestion, so we presume it has a benign form with a low mortality rate.

The diagnosis of CMH is often established by routine plain chest radiography, and on lateral films to show anteriorly placed bowel loops. On standard plain AP radiography, hernia sac presents with shadow like air, highly suspect for intestine. On lateral chest radiography, there is anterior herniation of bowel loops (located retrosternally). More than half of CMH cases are detected when patients are being investigated for unrelated problems. In rare cases of intermittent herniation, radiography can be absolutely normal. In these cases chest echosonography examination is useful. Contrast examination (barium enema) confirms the diagnosis in cases when the colon is the content of the sac. Computer tomography can also be considered as a non-invasive method of diagnosing CMH. CT scan and ultrasound are advocated for difficult diagnoses with liver and omentum in the sac. MRI and radionucleotide liver scan may help with diagnosis, especially when the liver is in the hernia sac.

Once CMH is diagnosed in symptomatic patients, early surgical repair is needed to avoid the risk of bowel strangulation and perforation. Treatment of asymptomatic cases is still controversial. We think it is reasonable to operate on patients even when the hernia content is only omentum or liver.

Operative approach includes transabdominal, transthoracic or laparoscopic repair. The transabdominal approach is advocated in certain diagnoses, when abdominal viscera are in the hernia sac. By this approach, abdominal organs can be easily pulled down to their normal positions in the abdomen, especially in bilateral hernias that can be diagnosed and repaired during the operation. Other associated anomalies (malrotation) that follow can also be easily corrected [6]. The transthoracic route is favored due to greater exposure and easy repair of the hernia sac [11], freeing of the pleural adhesions from the hernia sac, and mediastinal mass characterization [12].

Repair through laparotomy is the method of choice, but laparoscopy is a useful diagnostic tool before an attempted repair if there is any uncertainty. Since the first laparoscopic treatment of CMH was described in 1992 [6], Kuster's technique has been rapidly popularized by many surgeons. Easy access to the retrosternal space, minimal trauma and a short hospital stay are factors that contribute to the popularization of this technique. Laparoscopic repair is favored in adults especially in non-acute cases. Richardson describes it as a new method of repairing CMH, with improvements in laparoscopic hernia treatment allowing for shorter recovery times than open surgery [13].

There is no general consensus whether to remove the sac or to use mesh intraoperatively. In all of our

five patients, the hernia sac was ligated, followed by diaphragmatic defect repair using non-resorbable sutures. Ligation and removing the sac is meant to avoid leaving a loculated space-occupying lesion in the chest that might result in recurrence or mesothelial-lined cyst formation [6,10]. The use of prosthetic mesh is becoming more popular, especially if the defect is large. Mesh overlapping the edges of the defect provides a good tension-free repair, even used laparoscopically [14,15].

Postoperative complications have rarely been described. Recurrence of CMH is extremely unlikely to occur after proper closure of a small-sized gap using interrupted nonabsorbable sutures.

Although complications are rare, thay have been well described in literature. High incidence of associated anomalies significantly increases the index of mortality in the pediatric group of patients [16].

References

[1] Al-Salem AH, Nawaz A, Matta H, Jacobsz A. Herniation through the foramen of Morgagni: Early diagnosis and treatment. Pediatr Surg Int 2002;18:93-97

[2] Ceylan E, Onen A, Sanli A, Yilmaz E, Ucan ES. Morgagni hernia: late diagnosis in a case with Down Syndrome. Respiration 2004;71: 641

[3] Colie DA, Turnbull CM, Shaw TR, Price WH. Case report: MRI appearances of left sided Morgagni hernia containing liver. Br J Radiol 1996;69:278-803

[4] Cacmaki O, Pektas O, Baskin D. Retrosternal (Morgagni) hernia with small bowel obstruction secondary to Richter's incarceration. Pediatr Surg Int 1990;5:274-275

[5] Kiliç D, Nadir A, Döner E, Özdemir N, Kavukçu S, Akal M, et al. Transthoracic approach in surgical management of Morgagni hernia. Eur J Cardiothorac Surg 2001;20:1060-1069

[6] Kuster GG, Kline LE, Garzo G. Diaphragmatic hernia through the foramen of Morgagni: laparoscopic repair case report. J Laparoendoscopic Surg 1992;2:93-100

[7] Arráez-Aybar LA, González-Gómez CC, Torres-García AJ. Morgagni-Larrey parasternal diaphragmatic hernia in the adult. Rev Esp Enferm Dig. 2009 May;101(5):357-66

[8] Berman L, Stringer DA, Ein SH, Shandling B. The late presenting pediatric Morgagni hernia: a benign condition. J Pediatr Surg 1989;24:970-2

[9] Pokorney WJ, McGill CW, Herberg FJ. Morgagni hernias during infancy: presentation and associated anomalies. J Pediatr Surg 1984;19:394-7

[10] Federico JA, Ponn RB. Foramen of Morgagni hernia. In: Shields TW, LoCicero III J, Ponn RB, (Eds). General thoracic surgery. 5th ed., Philadelphia: Lippincott Williams and Wilkins; 2000

[11] Loong TP, Kocher HM. Clinical presentation and operative repair of hernia of Morgagni. Surgery Today 2003;32:902-5

[12] Minneci PC, Deans KJ, Kim P, Mathisen DJ. Foramen of Morgagni hernia: changes in diagnosis and treatment. Ann Thorac Surg 2004;77:1956-9

[13] Richardson WS, Bolton JS. Laparoscopic repair of congenital diaphragmatic hernias. J Laparoendosc Adv Surg Tech A 2002;12(4):277-80

[14] Huntington TR. Laparoscopic transabdominal preperitoneal repair of a hernia of Morgagni. J Laparoendosc Surg 1996; 6:131-3

[15] Rau HG, Schardey HM, Lange V. Laparoscopic repair of Morgagni hernia. Sur Endosc 1994;8:1439-42

[16] Skari H, Bjornland K, Haugen G, Egeland T, Emblem R. Congenital diaphragmatic hernia: a meta-analysis of mortality factors. J Pediatr Surg 2000;35(8):1187-97