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Morgagni Hernia

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Morgagni Hernia

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Introduction

A Morgagni hernia is one of four types of diaphragmatic hernias; the other types include a Bochdalek hernia, where the defect is posterolateral, a hiatal hernia, where the defect is at the esophageal hiatus, and a paraesophageal hernia, where the defect is located adjacent to the esophageal hiatus.[1] The Morgagni hernia, in which the defect is found in an anterior and retrosternal location, was first described by Morgagni in 1769.[2] It is rarer than the other type of congenital diaphragmatic hernia (Bochdalek hernia) and comprises only 2% to 5% of all congenital diaphragmatic hernias.[3][4][5][6] Morgagni hernias tend to be less symptomatic as pulmonary hypoplasia is uncommon, leading to a delayed diagnosis of these defects.[4][2]

Etiology

Morgagni hernias, especially when discovered in infancy or early childhood, are known to be associated with other congenital anomalies, with the incidence ranging from 34% to 50%.[7][8][4][2][9][10][11] The most common anomalies include cardiac defects (25% to 60%)[7][8][9] and trisomy 21 (15% to 71%)[7][8][9][12], while malrotation[11][8][9][12], anorectal malformations[8], omphalocele[11], skeletal anomalies[11], and pentalogy of Cantrell have also been associated with a Morgagni hernia. The association with trisomy 21 is thought to be due to a defective dorsoventral migration of rhabdomyoblasts that is caused by increased cellular adhesiveness that is seen in patients with trisomy 21 and may also be why those with Down syndrome are more likely to develop a recurrence after repair.[2][10] In contrast to patients with a Bochdalek hernia, where these patients tend to become symptomatic soon after birth due to pulmonary hypoplasia, up to 50% of patients are asymptomatic upon diagnosis of a Morgagni hernia.[12] Patients who are under two years of age are more likely to be symptomatic at the time of diagnosis. However, often the diagnosis may not be made until adulthood when chest imaging was performed for unrelated reasons.[2] Due to the pericardial attachments that provide support to the left side of the diaphragm, up to 91% of Morgagni hernias are found on the right side, with only 5% found on the left side and 4% found to be bilateral.[2][13]

Epidemiology

The estimated incidence is between 1 in 2000 and 1 in 5000 live births, although the true incidence is unknown.[3] [12][5] It comprises 2% to 5% of all congenital diaphragmatic hernias.[3][5]

Pathophysiology

A Morgagni hernia is located posterolaterally to the sternum and is caused by a failure of the pars tendinalis part of the costochondral arches to fuse with the pars sternalis.[7][14][2] Failure of fusion on the right side is a Morgagni hernia, while a failure of fusion on the left is often called a Larrey hernia. Although left-sided and bilateral hernias occur, 90% of Morgagni hernias occur on the right side due to the pericardial attachments to the diaphragm that provide protection and support to the left side.[6][8] The defects originally are small, with over 90% of defects having a hernia sac, but can grow over time due to increases in intra-abdominal pressure causing weakness of the diaphragm.

[7][14][2] The hernia most often contains large intestine (54% to 72%) or omentum (65%), but can also contain small intestine, stomach, and liver.[12][5][15][4][16][4]

History and Physical

The presence of a Morgagni hernia usually presents later in life than a Bochdalek hernia, with patients noting respiratory and upper gastrointestinal symptoms as their main complaint.[13][10][5][2][8] However, up to 50% of patients may be asymptomatic on presentation with diagnosis occurring during the workup for an unrelated problem. [14][17][1] In children, respiratory complaints, such as respiratory distress or tachypnea (20% to 73%)[5][10][18] and recurrent pulmonary infections (29% to 55%) are most common, although poor feeding, failure to thrive, and coughing and choking with feeds can also be seen.[10][18][15][5][14][19][20][6] Likewise, in adults, both gastrointestinal and respiratory symptoms can be experienced, including retrosternal or chest pain that is often relieved by standing, dyspnea, flatulence, indigestion, or cramping.[21][2][1][13][14][17] The presence of trauma, obesity, pregnancy, chronic constipation, or chronic cough may be precipitating factors in the onset of symptoms due to the increase in intra-abdominal pressure causing herniation of omentum and intestine into the defect.[2][13] [10] The physical exam may find bowel sounds on auscultation of the chest if the intestine is present in the hernia. [21] One must also be aware of acute strangulation or volvulus, as evidenced by rebound tenderness, tachycardia, persistent vomiting, and blood on the rectal exam.[18][22][15] These patients require prompt resuscitation, diagnosis, evaluation of the bowel with possible bowel resection, and repair of the hernia defect.

Evaluation

The definitive diagnosis is made radiologically and first includes an anterior-posterior and lateral chest radiograph.[2] [6][14] With bowel herniation, a radiolucent paracardiac shadow is seen that is found to be retrosternal on lateral radiographs.[10] Chest radiograph alone can make the diagnosis approximately 71% of the time, usually when the bowel is seen in the chest.[8] In patients where omentum or solid organs have herniated through the defect, a rounded opacity is seen at the right cardiophrenic angle.[2] In these instances, further imaging, such as a chest computed tomography (CT) scan, swallow study, or barium enema might be used to confirm the diagnosis.[5][8][7][10][14] [2] A CT scan is often the next step in the diagnosis, as it is easy to obtain and has up to a 100% rate of correctly identifying the defect.[2][17] The CT scan will show a retrosternal mass of fat density when omentum is included, or an air-containing viscus will be seen if the bowel has herniated through the defect.[2] A swallow study can be performed if a hiatal hernia is part of the differential diagnosis, and a barium enema has also been employed, where an upward angulation of the mid transverse colon can identify herniated colon into the defect.[2]

Treatment / Management

Due to the risk of incarceration, it is recommended that all Morgagni hernias should be surgically repaired; however, the approach to repair and type of repair is still under debate. The hernia defect can be approached either through an abdominal approach or through a thoracic incision. A posterolateral right thoracotomy is made through the 6 intercostal space when utilizing the thoracic approach.[21] The advantages of this approach are a more accurate vision of the right-sided defect, easier dissection of the hernia sac off the pericardium, mediastinal, and pleural structures, and potentially a safer reduction of the abdominal viscera such as the liver.[6][21][13][2] The hernia sac may or may not be excised, and then 0-silk or 0-polypropylene sutures are used to close the defect.[21] Disadvantages of this approach include risk of missing a bilateral defect due to the inability to see the left side, a higher likelihood of requiring postoperative ventilation depending on the preoperative respiratory status of the patient and the risk of a chest wall deformity in children who undergo the procedure at a young age before most of their growth has taken place.[13][9]

When approaching repair from an abdominal approach, the repair can be performed either through an open laparotomy incision or with minimally invasive laparoscopic techniques. The advantages of an abdominal approach include the ability to identify and repair bilateral defects and evaluate and repair other intra-abdominal pathology, such as malrotation, during the same operation. [6][14][2][9] An open laparotomy is most often reserved for emergent

cases, in patients who would not tolerate laparoscopy, in those with severe scoliosis or extensive adhesions if extensive bowel resection is required or the bowel cannot be retrieved from the incarcerated hernia.[13][9] An upper abdominal incision is performed, and the hernia contents are reduced.[22] There is debate about whether the hernia sac should be excised or incorporated into the sutures; however, the repair should be under minimal tension if performing a primary repair, with Nasr recommending that a mesh be placed for all hernias greater than 20 to 30 cm. [2][10][15] The defect is repaired with nonabsorbable sutures in a mattress fashion, incorporating the costal margins. [17][22][2] The open laparotomy approach has been shown to have shorter operating times compared to laparoscopy, but the minimally invasive approach confers a shorter recovery time and faster return to normal activities and eating with no difference in complication rates while providing more space for dissection and better visualization.[13][8][23] [12]

In 1997, Georgacopulo was the first to report a successful laparoscopic repair of a Morgagni hernia in a child. [24] With the laparoscopic approach, the patient is placed in reverse Trendelenburg with the surgeon located at the foot of the bed. [9][25][20][5] The camera port is placed through the umbilicus, and two working ports are positioned in the right and left upper abdomen in the mid-clavicular line.[4][25][9][26][27][20][19][5] The falciform ligament may need to be divided for adequate exposure, depending on the size of the defect. [25][4][9][27] The hernia contents are reduced; if the sac is to be excised, it is done at this time. As the defect often has a greater transverse than the anterior-posterior diameter, closure with sutures being brought through the abdominal wall can be used. [27] Placement of a patch in large defects occurs first by suturing the patch to the posterior rim of the hernia defect. [9] [4] The patch is then incorporated with the nonabsorbable sutures that travel full-thickness through the anterior abdominal wall and are tied in the subcutaneous tissue. [9][4] For defects that do not require a patch, closure occurs in a similar fashion with the placement of "U"-type stitches that transverse the abdominal wall, includes the hernia sac if it remains, incorporates the posterior rim of the diaphragmatic defect and comes out the abdominal wall with securement in the subcutaneous tissue.[11][25][8] This type of repair is beneficial in patients who do not have an anterior diaphragmatic rim and is easier to perform than intracorporeal sutures.[15][4] Recovery is usually uneventful, with most patients being discharged within three days of surgery. [27][20][19][4] The robotic technique has also been described, with improved ergonomics, articulation, and tremor filtration sited as benefits to this minimally invasive version.[28][19][26]

Differential Diagnosis

If a loop of bowel is seen herniating through the defect on chest radiograph, the differential could also include a pericardial cyst, loculated pneumothorax, or a hiatal hernia.[1][2] If part of the omentum or liver has herniated into the defect, as seen as a solid structure on the chest radiograph, the differential should also include atelectasis, pneumonia, pericardial fat pad, intrathoracic lipoma, bronchial carcinoma, pleural mesothelioma, or an atypical mediastinal tumor.[10][2][1] Further imaging with a lateral chest radiograph or CT scan should be able to confirm the Morgagni defect.

Prognosis

For defects found in infancy, the risk factors for mortality are those associated with prematurity, such as low birth weight, early gestational age, and low APGAR scores, or due to the other congenital anomalies seen with a Morgagni hernia. Although pulmonary hypoplasia is not commonly seen, identification prenatally and the size of the defect, including the presence of a sac, may make a difference in outcomes; however, the lifelong implications for children are unknown.[6][2][13][15] After repair, most patients recover well with a resolution of preoperative symptoms and a low recurrence rate.[15][4][13]

Complications

Due to concern for bowel obstruction, strangulation, volvulus, and/or necrosis, which can occur in up to 10% of cases, it is recommended that even asymptomatic patients should be referred for surgical correction.[1][2][14][5] [6] Complications seen after surgical correction are those common after any surgical procedure and include a low

incidence of wound infections, incisional or port site hernias, stitch abscesses, and bowel obstruction.[12][9][27][17] [9][16][6][15][18] Recurrence rates have been reported from 2% to 42%[11][5][16][10], although many studies have reported no recurrences with follow-up time as long as 10 years after repair.[21][4][17] Risk factors for recurrence include the closure of the defect under tension without the use of a patch, leaving the sac in place without resection, use of absorbable suture for repair, and a patient history of Down syndrome.[5][23][10][11] As the approach to repair varies, one must be aware of certain pitfalls that are associated with each technique. Those who choose to repair the defect through the transthoracic route should be aware of the potential for bilateral defects that may be missed.
[13] This approach is suboptimal for sac removal and is associated with requiring ventilator support postoperatively and the potential for chest wall deformity in infants and children.[13][9] An open laparotomy incision, while optimal for complicated or emergent cases, has been associated with increased recovery time, a higher incidence of wound complications and worse cosmesis compared to the minimally invasive approach.[13] All approaches lead to good outcomes as long as one remains aware of both its benefits and risks.

Deterrence and Patient Education

A Morgagni hernia is a rare type of congenital diaphragmatic defect. [2] Patients who experience recurrent pulmonary infections, shortness of breath, or worsening abdominal pain or vomiting should seek healthcare attention. [15] The defect is closed by performing surgery either through the chest or the abdomen. [13] A patch made of synthetic material may need to be placed if the defect is large. [2] There is a low recurrence rate after surgery, and most patients experience full relief of their symptoms. [15][4]

Pearls and Other Issues

Patients who are symptomatic from a Morgagni hernia can range in presentation from repeated chest infections or intermittent gastrointestinal complaints to persistent vomiting, tachycardia, and rebound tenderness consistent with acute strangulation.[18] Even in patients who are asymptomatic upon diagnosis, surgical repair is indicated to prevent emergent repair of strangulated or volvulized bowel.[14] Patients with Down syndrome have a higher risk of recurrence, and this should remain in the differential if a patient returns with similar symptoms or signs of obstruction. [10]

Enhancing Healthcare Team Outcomes

Patients with Morgagni hernias may present with respiratory symptoms, gastrointestinal complaints, or may be asymptomatic.[15] [Level 4] Workup includes a chest radiograph that may show air-containing viscus; if the diagnosis is inconclusive, a CT scan can be obtained to confirm the diagnosis.[2] [Level 3] Timely referral to the general surgery service allows for repair on a semi-elective basis prior to the incidence of complications such as obstruction or intestinal perforation.[5] [Level 4] Prognosis is good, with a low recurrence rate.[2] [Level 3] Good communication and collaboration between interprofessional can limit complications, leading to improved outcomes.

Questions

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