Chronic traumatic and congenital diaphragmatic hernias: Presentation and surgical management

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BACKGROUND: The diagnosis of chronic diaphragmatic hernias, whether due to congenital defects or trauma, may be difficult to make and may rely on clinical suspicion in the setting of persistent nondiagnostic radiographic findings. Repair is indicated to avoid catastrophic cardiopulmonary compromise and/or incarceration of abdominal organs.

STUDY OBJECTIVES: To review the varied presentations and treatment of chronic diaphragmatic hernia.

DESIGN: Retrospective review.

SETTING: University of Washington and Harborview Medical Center, Seattle, Washington.

PATIENTS: Between 1997 and 2001, nine patients presented with chronic diaphragmatic hernia (two congenital cases, seven post-traumatic cases). Four cases involved the right diaphragm. The following clinical features were noted: asymptomatic, chest radiograph showing bowel herniation (n=1); chest wall mass (n=1); asymptomatic with the chest radiograph showing marked elevation of hemidiaphragm (n=1); dyspnea with the chest radiograph showing marked elevation of hemidiaphragm (n=1); diarrhea and heartburn (n=1); generalized gastrointestinal upset (n=1); recurrent pneumonia (n=2); recurring effusions (n=4); and dyspnea on exertion (n=5).

INTERVENTIONS: Diagnosis was confirmed by chest radiograph in two patients, chest computed tomography scan in one patient, barium studies in three patients and thoracoscopy in three patients. All hernias were repaired via thoracotomy, and two hernias were repaired with artificial patch.

CONCLUSIONS: Patients with chronic diaphragmatic hernias present with a variety of symptoms and radiographic findings. When radiology or symptoms suggest bowel involvement, barium studies are appropriate. In other cases, chest computed tomography scans and/or thoracoscopy are useful. Repair is accomplished through the ipsilateral chest, with primary repair of the diaphragm preferred over patch repair.

Key Words: Chronic; Diaphragm; Hernia

Hernies diaphragmatiques chroniques, traumatiques ou congénitales : tableau clinique et traitement chirurgical

CONTEXTE : Le diagnostic des hernies diaphragmatiques chroniques, d’origine traumatique ou congénitale, peut être difficile à poser et parfois on se remet à l’intuition clinique dans les cas où les radiographies laissent

Résumé a la page suivante
The presentation of chronic diaphragmatic hernias may vary from an asymptomatic chest radiograph abnormality to septic shock due to intestinal infarction (1-4). They may be a consequence of congenital diaphragmatic herniation presenting in a delayed fashion or missed traumatic injuries. Because of the passage of time, patients often present not to surgeons, but to family practitioners, internists or respirologists with recurring pulmonary and/or gastrointestinal complaints. Making the diagnosis is important because of the potential for complications, and repair should be considered, even in asymptomatic patients. Confirming the diagnosis by noninvasive means may be difficult, however, because radiology has relatively low sensitivity and specificity. Nine cases that illustrate the varied presentations are presented, and diagnosis and treatment are discussed.

### Case Presentations

Nine cases of chronic diaphragmatic hernia were compiled between 1997 and 2001 at the University of Washington Medical Center and Harborview Medical Center, Seattle, Washington (Table 1).

Two patients with congenital defects (patients 2 and 5) presented with respiratory complaints; in both patients, incarcerated small and large bowels were present. Both were repaired via thoracotomy, one with a patch, and patient 5 also required a laparotomy to correct malrotation. After four years of follow-up, both patients remain symptom-free.

Seven patients presented after blunt trauma, with presentation ranging from five months to 40 years after the event. All but one patient had respiratory complaints (Table 1). Both patients were repaired via thoracotomy, one with a patch, and patient 5 also required a laparotomy to correct malrotation. After four years of follow-up, both patients remain symptom-free.

Seven patients presented after blunt trauma, with presentation ranging from five months to 40 years after the event. All but one patient had respiratory complaints (Table 1). All patients were repaired via thoracotomy. In
one patient (patient 9), an attempt had been made to reduce the defect through laparotomy, but the attempt failed due to extensive adhesions. In two patients, both with morbid obesity, the diagnosis could not be made by chest radiography, but was confirmed by computed tomography (CT) scan in one case and by thoracoscopy in the second case. These patients were the only two to experience complications. One patient (patient 8) developed an acute pulmonary saddle embolism on the second postoperative day, requiring urgent surgical clot removal. Patient 7 experienced progressive right heart failure complicated by pre-existing sleep apnea, and after refusing any further intervention, died two months after repair. All of the other patients are doing well, with resolution of respiratory complaints noted at follow-up visits ranging from six months to four years after repair.

DISCUSSION
Pathophysiology of diaphragmatic hernias
As opposed to hiatus hernias, congenital and traumatic diaphragmatic hernias result from defects in the musculotendinous membrane rather than from dilation of normally occurring diaphragm openings. The diaphragm is subjected to radial forces such that a small defect tends, over time, to enlarge, leading to two broad classes of complications. The first is related to the inability to generate normal, effective diaphragmatic contraction, leading to weak inspiratory and expiratory strength, and thus, to decreased exercise capacity in conjunction with increasing dyspnea. This is similar to the problems encountered with diaphragmatic eventration and phrenic nerve paralysis. The second complication relates to migration of abdominal contents into the chest (liver and colon on the right; stomach, spleen, small bowel and colon on the left). A pressure gradient of 7 to 20 cm H2O normally exists between the abdominal (more positive) and thoracic compartments. This gradient can increase to as much as 100 cm H2O in deep inspiration or with other conditions, including pregnancy, that increase intra-abdominal pressure (1-7). As abdominal contents fill the thoracic cavity, cardiac and respiratory functions become compromised. In addition, the colon and/or small bowel can become incarcerated, leading to a pattern of abdominal complaints, bowel obstruction, and eventually, perforation and septic shock.

Chronic traumatic diaphragmatic hernias: Clinical presentation
Delayed post-traumatic chronic diaphragmatic hernias are usually defined as those in which the diagnosis is made more than one month after the injury (8), and the onset of symptoms can be delayed by as much as 40 years. The true incidence of this entity is hard to define with certainty. Shah and associates (8) reviewed 980 patients with traumatic diaphragmatic injuries reported in the literature, and noted that the diagnosis was made preoperatively in 43.5% of patients, and at surgery or autopsy in 41.9%; in the remaining 14.6%, the diagnosis was delayed. Maekawa and associates (9) noted that over an 18-year period in Japan, 297 cases of blunt traumatic diaphragmatic hernias were reported, of which 47 cases (32 in left hemidiaphragm, 15 in right hemidiaphragm) were delayed (9). Chronic post-traumatic diaphragmatic hernias are a consequence of blunt injury in three-quarters of cases (10). A number of similar cases related to penetrating injuries have been described as well.

When due to trauma, the left hemidiaphragm is more often affected (1). This has been variously attributed to the ‘protective’ effect of the liver in blunt trauma and to congenitally weak attachments of the left diaphragm. Mansour noted that of 15 patients with chronic post-traumatic diaphragmatic hernias, 13 had hernias that involved the left side and four had hernias that followed penetrating trauma (11). When penetrating diaphragmatic injuries, left-sided lesions may be associated with a preponderance of right-handed assailants (11-14).

The broad range of patient complaints in the setting of chronic traumatic diaphragmatic hernias reflects the variety of herniated structures found in these patients. Traumatic chronic diaphragmatic hernias may be asymptomatic in up to 20% of cases, may occasionally masquerade as acute pneumonia and may present with gastrointestinal symptoms of varying severity in up to 70% of cases, including incarcerated abdominal viscera in 30% to 50% of cases (1,2,11,15). In the series reported by Hegarty et al (2), 72% of patients presented with herniated colon, 40% presented with herniated stomach, 16% presented with small bowel and 8% presented with spleen herniations. Most patients present with hernias of mixed content, as demonstrated in the present series (Table 1).

Chronic congenital diaphragmatic hernias: Clinical presentation
There is no widely accepted definition for a chronic congenital diaphragmatic hernias. Congenital diaphragmatic hernias recognized in a delayed fashion can present after the first few days of life to several months or years after birth. Congenital diaphragmatic hernias are diagnosed beyond the first hours of life in only 10% to 20% of recognized cases (4). The present series includes two patients with chronic congenital diaphragmatic hernias – an eighteen-year-old child and a forty-year-old woman (Table 1).

Congenital diaphragmatic hernias include the anterior foramen of Morgagni and the more common posterolateral foramen of Bochdalek defects. The left side is much more commonly affected than the right. In three series reported in the 1990s comprising 23 patients, 17 defects were left posterolateral, four were right-sided and two were anterolateral in the area of the foramen of Morgagni (16-18). Malone and associates (3) described a series of 22 patients with chronic congenital diaphragmatic hernias; 46% had a Bochdalek hernia, 23% had a central hernia related to agenesis of the diaphragm, 13% had a Morgagni hernia and 18% had an evagination of the diaphragm.

Virtually all patients with chronic congenital diaphragmatic hernias present with pulmonary complaints, in addition to suffering from a high incidence of gastrointestinal
Symptoms varying from failure to thrive to frank bowel obstruction. Intestinal obstruction is less likely with chronic congenital diaphragmatic hernias compared with chronic traumatic diaphragmatic hernias because of an associated hernia sac in congenital lesions. Children with gastrointestinal complaints tend to be older than the age of one year. Younger patients present with acute respiratory problems. Virtually all patients with posterolateral diaphragmatic defects have respiratory complaints, ranging from 'pneumonia' to 'tension pneumothorax' (16-19). In sharp distinction, an anterior Morgagni hernia is more likely to present as an asymptomatic mass or a cardiophrenic shadow (3).

Recognizing chronic congenital and traumatic diaphragmatic hernias
A number of factors contribute to delayed diagnosis of chronic diaphragmatic hernias, including lack of documentation of previously normal radiographic studies; prolonged period of time between the inciting event (in the case of trauma) and presentation; failure to recognize that the entity exists (as in congenital diaphragmatic hernias); and the misdiagnosis of radiographic findings. The key to diagnosing post-traumatic diaphragmatic hernias is obtaining a history of trauma. It is important to note that acute diaphragmatic injuries can be missed at laparotomy (8,11). The diagnosis of chronic congenital diaphragmatic hernia rests on the exclusion of other causes for the diaphragmatic hernia. If no history of abdominal trauma can be elicited, the case for a congenital etiology is supported. Surgical exploration and repair confirm the diagnosis if the position of the lesion appears to be consistent with diaphragmatic eventration, or a Morgagni or Bochdalek hernia.

The diagnosis of chronic diaphragmatic hernia relies heavily on radiography. Chest radiographs are diagnostic in only 25% to 50% of cases (11,19). Patients who present with primarily intestinal complaints and a radiological pattern suggesting 'gas' above the diaphragm (including a nasogastric tube in the chest) should be evaluated with a barium enema and/or upper gastrointestinal series to evaluate the stomach and small bowel (19,20). Upper gastrointestinal series should be included in all cases of congenital chronic diaphragmatic hernia to exclude associated malrotation (3). Patients who present with less specific complaints and who only have suggestive radiographic changes (pleural mass, recurrent effusions, elevated diaphragm, cystic lower lobe mass) or who present with 'complex pleural space' problems with no gastrointestinal signs or symptoms should be studied by CT scans of the chest with oral contrast (Figure 1). Studies of acute traumatic diaphragmatic injury suggest that CT scans have a sensitivity of approximately 80% for detecting left-sided lesions, but only 50% for detecting right-sided lesions. This is because the major diagnostic sign is constriction of abdominal viscera (the 'collar sign') or presence of hollow viscera in the chest. Diaphragmatic defects can be detected but are difficult to clearly delineate by CT scan (21).

In the setting of chronic diaphragmatic hernias, there is an increased incidence of abdominal visceral herniation recognizable on a CT scan. If the CT scan is suggestive of such a hernia, magnetic resonance imaging has proven to be effective in diagnosing small defects. Fluoroscopy is useful in determining abnormal diaphragmatic excursion, which may suggest an injury or defect, but it has a similar sensitivity and specificity to chest radiography (8,11). For patients who may have a post-traumatic chronic diaphragmatic hernia, thoracoscopy (and occasionally laparoscopy) has proven to be an effective, minimally invasive method of confirming the diagnosis. Thoracoscopy is used in patients who have soft findings on a chest radiograph, often after an equivocal CT scan. Thoracoscopy can be used to repair defects in some cases in which adhesions are not severe.

Operative approach to chronic diaphragmatic hernias
When a chronic traumatic diaphragmatic hernia is diagnosed, repair is usually performed via the ipsilateral chest, because there are usually dense adhesions between the viscera and lung that preclude safe transabdominal reduction. Reducing a herniated viscus through an abdominal approach risks excessive pulling and tearing of the viscus, which could potentially spill intestinal contents into the pleural space. Rarely, a thoracoabdominal incision is needed, although it is best to avoid this because of the risk of phrenic nerve injury and of creating another diaphragmatic defect (14).

The operative approach in the congenital setting depends on whether malrotation is present (present in up to one-third of patients), in which case, an abdominal approach is also needed to divide Ladd's bands (3). Primary repair can be performed in the majority of cases (with or without pledgets), but in some cases, artificial material is required (8,9,16-18). There is some experimental evidence suggesting that repairing the defect with prosthetic material may result in better respiratory muscle function, but this has not been studied clinically, and in general, we prefer to...
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CONCLUSIONS
Chronic diaphragmatic hernias, whether of traumatic or congenital origin, have a variety of presentations, ranging from asymptomatic radiographic changes to shock. The broad range of patient complaints in the setting of chronic diaphragmatic hernias reflects the variety of herniated structures found in these patients. Patients with congenital defects tend to present with respiratory symptoms and chest radiographic findings that are confused with pneumonia or other processes. Traumatic injuries are often asymptomatic, and chest radiographs reveal only nonspecific findings. Patients with either type of chronic diaphragmatic hernias are at risk of developing either severe cardiopulmonary compromise or complications related to incarceration of abdominal organs. CT scans of the chest are used to evaluate patients who present with respiratory or gastrointestinal symptoms and a chest radiograph suggestive of an intestinal gas pattern in the thorax. For patients with persistent respiratory or abdominal complaints and unclear CT scan findings, diagnostic thoracoscopy should be considered. Repair of a chronic diaphragmatic hernia is safest through the ipsilateral thorax, and primary repair of the diaphragmatic defect is preferred, although patch repairs can be used for larger defects.

Of note, many patients do not present to surgeons, but rather, to internists or pediatricians. In the present series, all but two patients were managed initially by nonsurgeons, underlying the need for all physicians to be aware of the possibility of this condition.

REFERENCES