DIAPHRAGMATIC HERNIA

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Diaphragmatic herniae are being recognized with increasing frequency. Herniation of abdominal contents into the thoracic cavity may occur through sites of embryological fusion, through the oesophageal hiatus, or following traumatic rupture of the diaphragm.

Congenital Herniae

The development of the diaphragm is complicated (Fig. 1). The anterior portion is formed from the septum transversum. The septum transversum originally arises in the upper part of the cervical region, and in the course of its caudal migration it receives its innervation from branches of the 3rd, 4th and 5th cervical nerve roots (the phrenic nerve) which accompanies it in its descent. In the 17 mm. embryo, the septum transversum has reached its permanent position.

The posterior part of the diaphragm is formed by the fusion of the pleuro-peritoneal membrane and the dorsal mesentery. The pleuro-peritoneal membrane is the last structure to complete the formation of the diaphragm. This takes place earlier on the right side, at about the 17 to 18 mm.

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**Fig. 1.**—Embryological formation of the diaphragm (Modification from Harrison's 'A Textbook of Human Embryology'). 1 and 2—Left and right pleuro-peritoneal membranes. 1a and 2a—Contributions from body wall. 3—Septum transversum.
embryonic stage, and later on the left side at about the 19 to 20 mm. stage. Before the formation of the pleuro-peritoneal membrane, there is a free communication between the pleural and peritoneal cavities, the 'hiatus pleuro-peritonealis'. In about the 10th week of fetal life, the herniated intestines return from the umbilical cord to the abdominal cavity, and if the pleuro-peritoneal canal is still open, loops of bowel will herniate into the pleural cavity (Bochdalek hernia). Embryological sites of fusion of the diaphragm are sites of potential weakness and it is at these points that congenital herniae occur.

The following cases are examples of these types of herniae.

**Case No. 1. Congenital Diaphragmatic Hernia in the Newborn (Bochdalek Hernia)**

This baby was found to be dyspneic and cyanosed at birth. X-ray of the chest (Fig. 2) showed the right chest to be filled with intestines, which were displacing the mediastinum to the left. At thoracotomy his chest was found to be filled with loops of small and large intestine. These were reduced, and the hernial orifice, which was located in the postero-lateral part of the diaphragm, was sutured. He made an uneventful recovery, and an X-ray showed satisfactory expansion of his lungs.

Herniae of the postero-lateral part of the diaphragm (through the foramen of Bochdalek) are the most common type of congenital diaphragmatic hernia in the newborn. Surgical repair should be carried out preferably within the first 48 hours of life. The operative mortality, unfortunately, is high, this being partly due to the frequent presence of a hypoplastic lung, which does not inflate after reduction of the contents of the hernia. This type of hernia contains no sac, and is therefore a 'false' hernia.

**Case No. 2. Congenital Diaphragmatic Hernia (Bochdalek Hernia) in a Boy aged 2**

This boy was referred for further investigation because of failure to thrive, and attacks of abdominal colic. His chest X-ray (Fig. 3) showed gas-containing shadows suggestive of intestines at the left base. However, it was not possible to assess whether these were above or below the diaphragm. Barium enema (Fig. 4) demonstrated colon in the chest. A thoracotomy was carried out and herniated loops of small and large intestines were found in the chest; there was no hernial sac present. These were reduced, and the hernial orifice, which was found to be at the site of the foramen of Bochdalek, was repaired. His post-operative X-ray was satisfactory.

The great majority of infants born with these congenital defects, if untreated, die in the first few hours or days. A few, however, may survive to adult life, and the hernia be discovered incidentally at routine X-ray examination.

**Case No. 3. Congenital Retrosternal (Parasternal) Diaphragmatic Hernia (Morgagni Hernia)**

This 72-year-old man had had a productive cough with increasing dyspnea for 2 years. X-ray of his chest (Fig. 5) showed a rounded cystic opacity in the right cardiophrenic angle. A lateral film showed that the air-containing opacity was situated retrosternally. Barium meal showed the stomach and small intestine to be normal. Barium enema (Fig. 6) demonstrated part of the large bowel to be herniated into his chest. Further questioning elicited the information that he had suffered from flatulence for the past 10 years. A right thoracotomy was carried out, the hernia reduced, and the hernial orifice repaired. His post-operative convalescence was uneventful, and an X-ray of his chest was satisfactory.

This type of congenital hernia is due to the abnormal development of that portion of the diaphragm which is formed from the fusion of the embryonal septum transversum, and the body wall components. This hernia is probably not as rare as previously thought, since symptoms are present in only a small proportion of patients. If the contents of the hernial sac are extraperitoneal fat or omentum, not adherent to the sac, symptoms are unlikely. If these structures become adherent, symptoms, particularly pain, may occur due to traction. Transverse colon in the sac produces symptoms which are predominantly abdominal. The patients usually complain of intermittent attacks of colic, bouts of abdominal distension, and sometimes vomiting. If the contents of the hernia are extraperitoneal fat or omentum, then the shadow seen radiologically is opaque; if bowel is present, then the typical air-containing space is seen (Figs. 5 and 6).

The majority of cases do not require surgery, as they have no symptoms. When colon is present, however, operation should be carried out whether symptoms are present or not, because of the danger of strangulation.

The following two cases demonstrate congenital malformations of the diaphragm which may be mistaken for congenital diaphragmatic herniae.

**Case No. 4. Eventration of the Diaphragm**

This man of 49 years of age was admitted with a 10-year history of dyspepsia, which did not fit any typical pattern, and was not related to meals. His X-rays (Figs. 7 and 8) showed an elevated left diaphragm, which moved sluggishly on screening. Barium meal (Fig. 9) showed no evidence of hiatus hernia, but the stomach was inverted, giving the characteristic 'fish-hook' appearance, recognized as being typical of an eventration. At thoracotomy the diaphragm was found to be thin, lax, and lying in folds; it was incised, overlapped and sutured. He made a good recovery, and was relieved of his symptoms.

Eventration is included here because it is easily confused with diaphragmatic hernia, as the symptoms and radiological appearances are similar. In its literal meaning, eventration should signify actual herniation, but the accepted meaning of the term is used to describe a condition of an abnormal elevation of the intact diaphragm. True eventra-
HOFFMAN: Diaphragmatic Hernia

Fig. 10.

Fig. 11.

Fig. 12.

Fig. 13.
tion is of congenital origin. The term eventration should not be used in conditions where there is an elevated diaphragm due to phrenic nerve paralysis. Eventration results when there is failure of muscle development; whereas in phrenic nerve paralysis, the diaphragm shows a normally developed muscle which is atrophic. In eventration movements of the affected leaf of the diaphragm may be greatly diminished, but there is no paradoxical movement as when the phrenic nerve is paralysed.

In the majority of cases, treatment is unnecessary, but in people who have symptoms, surgery should be considered. Considerable relief may be obtained by plication of the affected diaphragm.

Case No. 5. Partial Eventration of the Right Diaphragm. ‘Herniation’ of the Liver into the Chest

This 43-year-old man was admitted for investigation of a large ovoid homogeneous opacity at the right base (Fig. 10). Radiologically it was not possible to decide if the lesion was in the chest, in the abdomen or arising from the diaphragm. He had a three years' history of productive cough and shortness of breath. Barium meal showed no evidence of a diaphragmatic hernia. A diagnostic pneumoperitoneum was therefore induced (Fig. 11). The lateral two-thirds of the lower surface of the right diaphragm was clearly outlined, but the air failed to show the upper margin of the mass. From this it was concluded that the opacity was formed by the liver. Pneumoperitoneum is not capable of differentiating between a diaphragmatic hernia and an eventration, due to the fact that adhesions between the diaphragm and visceral pleura seal off the communication.

At thoracotomy it was found that the liver was protruding into the chest through the intact diaphragm, which was stretched over its upper surface, and adherent to it. There was no evidence of hernia or tear in the diaphragm. Nothing further was done and the chest was closed.

The differential diagnosis which had to be considered in this case was a true hernia of the liver. A hernia of the liver usually follows severe trauma to the chest or abdomen. The injury has often taken place so many years ago that the patient fails to recall it, or feels that it has no connection with his present complaints, especially as the condition can be entirely asymptomatic, and may be discovered only at routine X-ray examination. Since these lesions eventually produce symptoms, and also because of the possibility of missing a tumour, it is necessary to explore all these conditions.

Diaphragmatic Hernia of Traumatic Origin

The diaphragm may be torn by compression effects of crush injuries, or by stab or gunshot wounds.

Case No. 6. Traumatic Rupture of the Diaphragm due to a Car Accident

This 48-year-old man was admitted to a general surgical ward following a road accident. In the crash he was thrown forward injuring the left side of his chest and abdomen on the steering wheel. On admission he was slightly shocked and complained of severe pain in the left chest and abdomen. His abdomen was tender with rigidly below the left costal margin. A diagnosis of ruptured spleen was made and a laparotomy carried out. The spleen was, however, found to be uninjured, but the stomach was found to be protruding into the chest. The condition at the time was thought to be a congenital diaphragmatic hernia, and no further action was taken. He was discharged home still complaining of dyspnoea on exertion. A month later he was referred to the Thoracic Unit and his X-ray (Fig. 12) showed the left chest occupied by a large air space, absent lung markings, and the heart and mediastinum displaced to the right. At thoracotomy a grossly distended stomach and the spleen were found to occupy most of his left chest. The herniated viscera were reduced, and a transverse tear in the diaphragm, about 6 inches long, was repaired. His convalescence was uneventful, and X-ray of his chest a fortnight later was satisfactory.

The diaphragm may be ruptured by trauma at any point. The left diaphragm is more frequently injured than the right, because the right has the protection of the liver. Traumatic herniae have no hernial sac, and properly speaking this constitutes an eversion of the abdominal contents into the thorax. The most common cause of diaphragmatic rupture is some severe form of impact such as occurs in automobile accidents. When the disruptive force is applied to the chest wall or upper abdomen, the diaphragm, being a structure of relative weakness, tears.

If following an accident, the victim is shocked, dyspnoeic and cyanosed, the possibility of ruptured diaphragm should be borne in mind, and X-ray examination carried out. A pneumothorax is the most likely condition to be considered as a differential diagnosis. This may be ruled out by passing a gastric tube, or getting the patient to drink some barium and observing it on an X-ray screen. There is also an obvious irregularity or absence of diaphragmatic continuity. The clinical picture of diaphragmatic rupture can be divided into an acute phase and a chronic phase. In the chronic phase the symptoms may be mild, or absent. Once the diagnosis has been established, surgical repair should be carried out, and the results are usually satisfactory.

Acquired Diaphragmatic Hernia

This group of cases deals with œsophageal hiatus hernia, and its complications, and includes a case of spontaneous rupture of the diaphragm.

Case No. 7. œSophageal Hiatus Hernia

This 59-year-old lady was admitted with a history
of dysphagia of three years' duration. She complained of heartburn and stated that food appeared to stick behind the sternum. Regurgitation, which was frequently accompanied by vomiting, occurred when she was stooping to do her gardening, and when she was lying in bed. The latter was well-controlled by sleeping upright.

Barium meal (Fig. 13) showed an oesophageal hiatus hernia involving the cardia of the stomach, and screening showed a good deal of gastro-oesophageal reflux. It was decided to treat the condition surgically. At thoracotomy the hernial sac was cut away, the herniated portion of the stomach reduced, the muscle bundles of the right crus approximated, and the acute gastro-oesophageal angle restored. Her convalescence was complicated by persistent diarrhoea from colonic diverticulitis which responded satisfactorily to hydrocortisone enemas and oral prednisolone. She eventually made a good recovery, and a barium meal showed a satisfactory repair. Screening showed no evidence of regurgitation.

Herniae through the inguinal canal have been well-understood for over a century, hernia through the oesophageal hiatus, on the other hand, have received attention only recently. Under normal conditions the stomach cannot be displaced into the mediastinum because of the obliquity of the gastro-oesophageal junction, and the effective sphincteric action of the crural muscles which surround the hiatus. The hiatal orifice is vulnerable, and can be disrupted by the prolonged action of increased intra-abdominal pressure such as occurs e.g. in pregnancy, obesity, flatulent dyspepsia or by the wearing of constricting corsets. Once a hiatus hernia develops, most of the symptoms and complications are due to the reflux of gastric juice into the oesophagus. This may lead to acute inflammation and ulceration of the oesophageal mucosa, and eventually to stricture formation.

The symptoms in the above case were fairly typical of the condition. Besides pain, regurgitation and vomiting, severe anemia and hematemesis may occur. Anginal attacks, cholecystitis and peptic ulcer are conditions most likely to be confused with hiatus hernia. In the absence of symptoms conservative treatment may be justified, but the complications, especially stricture formation, are so difficult to manage, that in the presence of symptoms, early surgical repair should be advised.

Case No. 8. Ulceration and Stricture of the Oesophagus due to Hiatus Hernia

This 56-year-old man was referred with a recent history of increasing dysphagia and retrosternal pain. Three years prior to this he had suffered from epigastric pain, nausea and vomiting for several months. On admission, barium meal showed a hiatus hernia, with an ulcer crater and marked stenosis. At oesophagoscopy a stricture was seen at the 29 cm. mark, with severe oesophagitis above. At thoracotomy the oesophagus was found to be shortened, its walls thickened, and the gastro-oesophageal junction embedded in a mass of fibrous tissue. It was possible to dissect the lower end of the oesophagus, reduce the hernia, and repair the defect. A post-operative barium meal showed a satisfactory repair, and no evidence of ulceration or stenosis. He has remained symptom-free since.

Apart from the intact mucous membrane of the stomach and first part of the duodenum, the human tissues are readily inflamed by gastric secretions. An example is gastro-jejunostomy, which in the presence of high gastric acidity, often results in stomal ulceration. If the sphincter mechanism at the cardia, especially during sleep in the recumbent position, is deficient, acid pepsin may easily flow back into the oesophagus and cause oesophagitis, ulceration and eventually fibrosis with stricture formation. Although hiatal herniation is the cause of the condition, all patients with hiatal deficiency do not develop oesophagitis and its sequelae, just as all patients who have undergone gastro-enterostomy do not necessarily develop stomal ulceration.

The above patient had a fairly typical history. He first complained of upper abdominal dyspepsia of a few months' duration, suggestive of cholecystitis or peptic ulceration. This was followed by a silent period which gradually gave way to increasing dysphagia.

Other symptoms of peptic ulceration of the oesophagus are heartburn, regurgitation, vomiting and retrosternal pain. Surgical management of these cases is very difficult. If after preliminary dilatations of the stricture it is possible to dissect the oesophagus, reduce and repair the hernia, the results are satisfactory. Such was the case in our patient, but frequently the hernia is irreducible. In these cases the methods of choice are excision of the stricture with either oesophago-jejunostomy, or the interposition of a loop of jejunum or colon. These operations are unfortunately not free from complicating side-effects.

Case No. 9. Oesophageal Stricture due to Hiatus Hernia following Gastro-enterostomy

This man of 70 years of age had an operation for a perforated peptic ulcer 15 years ago. Recently his symptoms became very troublesome and a gastro-enterostomy was carried out. A few days after his discharge from hospital, he noticed some difficulty in swallowing, which gradually got worse, until eventually he could swallow only fluids. Barium meal (Fig. 14) showed a hiatus hernia with a severe stricture at the gastro-oesophageal junction. As it was not possible to dilate the stricture, it was decided to explore him. At thoracotomy the lower end of the oesophagus was mobilized, its wall incised, and a pin-point stricture found at the gastro-oesophageal junction. A cardioplasty was carried out. This procedure involves incising the stricture longitudinally and re-suturing the cut edges horizontally. Bilateral vagotomy was also done to reduce the amount of gastric secretion. His post-operative course was satisfactory, and he remained symptom-free. A recent barium meal showed no evidence of stricture, but screening demonstrated free
gastro-oesophageal reflux. This operation does not restore the normal gastro-oesophageal anatomy, and hence would be unsuitable for a younger person.

The occasional development of an oesophageal stricture following partial gastrectomy or gastro-enterostomy has been recognized for many years, particularly so if an in-dwelling gastric tube has been left in situ for some time. Less often the condition has followed gastric intubation which has been carried out for other surgical or medical conditions. Oesophagoscopy in such cases usually showed appearances similar to those found in reflux oesophagitis, and barium meal often demonstrated the presence of a hiatus hernia.

Reflex oesophagitis in the presence of a hiatus hernia occurs more readily in the recumbent position, and is probably aggravated by the presence of an in-dwelling gastric tube. These complications might be prevented if patients, even if the presence of a hiatus hernia is not suspected, be nursed in an upright position following abdominal operations, especially if a gastric tube has to be left in situ.

**Case No. 10. Carcinoma complicating a case of Hiatus Hernia**

This 68-year-old female was admitted for investigation with a history of intermittent episodes of dysphagia going back for two years. Barium meal (Fig. 15) showed a hiatus hernia, and a moderately severe oesophageal stenosis. On screening there was marked gastro-oesophageal reflux. At oesophagoscopy an ulcer was seen and routine biopsy was taken. Histological examination showed this to be an adeno-carcinoma arising in gastric mucosa. She was explored through a left thoraco-abdominal incision and resection of the lower third of the oesophagus, and the upper portion of the stomach was carried out, and continuity restored by gastro-oesophageal anastomosis. Macroscopically the lesion in the resected specimen looked like a chronic ulcer. Histology showed the presence of a chronic gastric ulcer which showed malignant change of an adeno-carcinoma at the periphery. Her post-operative recovery was uneventful, and a barium meal was satisfactory.

There are two varieties of ulceration which may occur in the presence of a hiatus hernia: (1) peptic oesophagitis involving the squamous epithelium of the oesophagus which is secondary to regurgitation of gastric juices. Peptic ulceration and a fibrous stricture may subsequently develop. The ulcers tend to remain superficial, may ooze blood, but rarely cause severe haemorrhage, and do not perforate. (2) If ulceration occurs in gastric mucous membrane, the ulcers arising from this behave like gastric ulcers, in that they may heal, bleed, perforate or cause stenosis. They differ from peptic ulcers of the oesophagus in that they may respond to medical treatment. Carcinoma arising from a chronic gastric ulcer in a hiatus hernia is rare, but there is no reason why such chronic gastric ulcers should not become malignant as they do in the stomach. Our patient was fortunate in that the symptoms of her stenosis led to investigations which revealed the carcinoma at an early stage. The possibility of malignant change occurring in association with gastro-oesophageal reflux in hiatus hernia should be kept in mind, and biopsy carried out whenever possible.

**Case No. II. Spontaneous Rupture of the Diaphragm with Strangulation of the Stomach**

This female patient of 48 years of age gave a history of sudden pain in her chest whilst she was dancing. She walked home and stayed in bed for a week. During this time the intensity of the pain varied, and it was at times accompanied by vomiting. She was admitted to a general medical ward, and her chest X-rays (Fig. 16) were first interpreted as being due to a tension pneumothorax. On the day following admission, she collapsed with signs of profound shock. Offensive fluid was aspirated from the chest, which was found to have an acid reaction. This temporarily relieved her condition, and at this stage the diagnosis of intra-thoracic stomach was made. A further X-ray (Fig. 17) showed two fluid levels—one in the stomach, and the other in the pleural cavity. She was transferred to the Thoracic Unit, where a thoracotomy was carried out. The stomach, which was greatly dilated, was found to occupy most of the left pleural cavity, and was congested, oedematous and purple. The spleen, part of the transverse colon and omentum were also found in the chest. There was no hernial sac present. After reducing the herniated viscera into the abdomen, a tear about 4 to 5 inches long was seen in the posterolateral part of the diaphragm, which was repaired. She made a good recovery, and her chest X-ray showed her left lung to be expanded.

Whereas strangulation is a common complication of most herniae, it is rare in the diaphragmatic variety. Most of the recorded cases of strangulation have occurred either in already existing diaphragmatic hernia or following traumatic rupture of the diaphragm. A sudden increase of intra-abdominal pressure such as occurs in vomiting, physical exertion, over-eating or post-operative distension, are thought to be the causative factors of a diaphragmatic hernia proceeding to strangulation. In our case, however, there was no previous history of a hernia, and no hernial sac was found. In addition to this there was no history of injury. The effort which caused this rupture to occur was trivial, and it may therefore well be classified as a spontaneous rupture of the diaphragm.

Signs of strangulation of a diaphragmatic hernia are a sudden onset of severe pain, vomiting and respiratory embarrassment. Late symptoms include profound shock, cyanosis and dehydration. These late symptoms should be forestalled by early operation, as once they have occurred, mortality is high.
Summary

Diaphragmatic herniae are classified into (a) congenital, (b) traumatic and (c) acquired. 11 cases are discussed in order to demonstrate these various types of herniae and their treatment. In addition the current views regarding the etiology and management of each of the above groups are reviewed.

(1) Congenital Herniae

(a) The embryological development of the diaphragm is described.
(b) Examples of the two most usual forms, i.e. Bochdalek and Morgagni herniae, are given.
(c) Most of these herniae are met in infancy and death will occur in most cases if surgical treatment is not initiated rapidly. Only very occasionally are they discovered in adults.
(d) Eventration of the diaphragm as a differential diagnosis is considered.

(2) Traumatic Herniae

(a) Traumatic rupture of the diaphragm is occurring more frequently, principally due to the rising incidence of car accidents.
(b) Traumatic pneumothorax and hemothorax are the conditions most often confused with the above.

(c) The condition clinically presents initially as an acute phase associated with severe shock and this may end fatally particularly if strangulation of the herniated viscera has occurred. This stage may subside and gradually merge into the chronic phase when symptoms may be mild or even absent.

(3) Acquired Herniae

(a) Oesophageal hiatus herniae and its complications are the commonest type of diaphragmatic hernia which are likely to be encountered in clinical practice.
(b) If severe symptoms and gastro-oesophageal reflux with oesophagitis are present surgical repair should be carried out whenever possible because of the dangers of complications.
(c) Examples of cases suffering from such complications which are described include stricture formation, stenosis following gastric surgery and a case of malignant degeneration occurring in a stricture with ulceration.
(d) An unusual case of spontaneous rupture of the diaphragm with strangulation of the stomach is included.

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