Foramen of Bochdalek Hernia: *

A Review of the Experience at Children's Hospital of Denver, Colorado

G. James Cerilli,** M.D.

From the Surgical Service, Children's Hospital of Denver and the Department of Surgery, University of Colorado Medical School

Foramen of Bochdalek hernia is not a rare anomaly, and yet few surgeons have had extensive experience with this condition. Despite recent advances in pediatric surgery, the operative mortality rate is still 15 to 20 per cent.[2] This review of the experience in a large children's hospital is presented with the hope of demonstrating any possible recurring errors in management and diagnosis, and perhaps contribute to a better understanding of the current clinical problems.

Material

Thirty-five children (21 boys, 14 girls) with foramen of Bochdalek hernias were admitted to the private service of Children's Hospital from 1946 through 1962. Twenty-nine of the hernias were on the left side. The presenting age ranged from newborn infants to two years. Sixteen patients were seen within 24 hours of birth; 11 patients between the first and the thirty-first day; 6 patients from one month to one year; and 2 patients were not diagnosed until older than one year.

All foramen of Bochdalek hernias in this series represent defects of various sizes in the posterolateral area of the diaphragm, and only two of these had hernial sacs. This is in contrast to eventrations, which represent weakness of the membranous part of the diaphragm and always have a limiting membrane between the peritoneal and thoracic cavities.

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** Resident in Surgery, University of Colorado Medical Center, 4200 East Ninth Avenue, Denver 20, Colorado.

Diagnosis. The symptoms and physical signs of foramen of Bochdalek hernias are amply documented.[3] The classical picture of cyanosis, respiratory distress, mediastinal shift, and decreased breath sounds on the side of the hernia in an infant is well known, and few errors are now made when this clinical picture is present. However, only about half of the children in this series presented cyanosis that was definitely noticeable to the admitting physician (Table 1), although more than three-fourths of them had histories of episodes of cyanosis (Table 2). The often described physical finding of bowel sounds in the chest was present in only 8 patients, and is considered to be an unreliable sign.

Roentgenography of the chest continues to be the most dependable method of establishing the diagnosis of congenital hernia of the diaphragm. Without its liberal use, the diagnosis will be missed. Of the 34 cases of foramen of Bochdalek hernia, 29 were diagnosed by roentgenogram unequivocally. This was ultimately proved at operation, or autopsy. In two other cases, foramen of Bochdalek hernia was the primary diagnosis, with a lung cyst and probable pneumothorax listed as other possibilities. The three remaining patients had a preoperative radiological diagnosis of eventration, but at surgery proved to have Bochdalek hernias.

Analysis of associated congenital defects in these patients revealed a high incidence of cardiac and intestinal malformations (Table 3). This was more pronounced in

TABLE 1. *Physical Examination in Congenital Foramen of Bochdalek Hernias*

	%	No. Cases
Decreased breath sounds on side of hernia	68	24
Mediastinal shift	51	18
Cyanosis	51	18
Dullness on side of hernia	28	10
Bowel sounds in chest	23	8
Tachypnea	17	6
Scaphoid abdomen	11	4
Hyperresonance on side of hernia	8	3
Dead on arrival	3	1

patients who did not survive and probably reflects the higher mortality resulting from multiple major congenital anomalies, rather than being a reflection of the more complete examination that is possible at autopsy. It should be emphasized that malrotation and malfixation of the intestine to the posterior peritoneum occur often enough to deserve serious consideration in diagnosis and decisions as to treatment.

Treatment. Thirty-four of the 35 patients survived long enough so that repair of the defect was attempted. In all except the one case of recurrent hernia, repair was accomplished by bringing the edge of the defect together without using foreign material or flaps of tissue. The approach varied with the surgeon; thoracic, thoraco-abdominal, and abdominal approach were all used. In three children, intentional incisional hernias were created to facilitate bowel replacement.

TABLE 2. *Symptoms of Foramen of Bochdalek Hernia*

	%	No. Cases
Cyanosis	77	27
Respiratory distress	68	24
Vomiting	6	2
Fever	6	2
Diarrhea	3	1
Stridor	3	1
Abdominal pain	3	1
Incidental finding	3	1

The high incidence of preoperative and postoperative complications (Table 4) contributed significantly to the morbidity, but few of these complications directly contributed to the mortality. Only nine of the 34 patients had uncomplicated courses. The over-all mortality rate in the series was 28 per cent (Table 5). Six deaths occurred either during the operation, or on the first postoperative day. No specific cause of death was apparent to the physician except *cardiorespiratory failure;* as mentioned, there was a high incidence of associated cardiac anomalies in this group. Of the remaining four deaths, one child was dead on arrival, and another died of an iatrogenic tension pneumothorax. The third patient died from acute cardiac arrest secondary to anoxia when an endotracheal tube became dislodged during operation. The fourth child died on the sixth postoperative day; he had a persistent postoperative pneumothorax and had been vomiting for two days. No specific cause of death was found at autopsy.

Discussion

Bochdalek hernia in this series was more common in males than in females. As in other series,[6] there was a preponderance of left-sided defects. The increased incidence of left-sided lesions has been ascribed to later closure of the left pleuro-peritoneal canal, allowing the intestine, which had recently become intra-abdominal, to herniate through the defect.

About half of these patients presented as newborn infants, but almost one fourth were not diagnosed until older than one month. A foramen of Bochdalek hernia diagnosed in one patient at age eight months was not repaired until she was 16 years old. In the interim, she had remained completely symptom-free. At operation, the entire small bowel was found in the left chest; no hernial sac was present. Another patient, who had been asymptomatic since birth, presented at age two with nausea

and vomiting. Thus, it should be emphasized that the diagnosis of congenital diaphragmatic hernia cannot be excluded because a child has been asymptomatic for several months, or even for several years.

This series illustrates that the diagnosis of diaphragmatic hernia can be made with a high degree of reliability by roentgenogram. The other radiological diagnoses in patients in this series, who were ultimately shown to have foramen of Bochdalek hernias, were lung cyst, eventration, and probable pneumothorax. It cannot be emphasized enough that the mode of diagnosis of these hernias is by roentgen examination, as the clinical signs are nonspecific. It was rarely necessary to introduce radio-opaque material into the gastrointestinal tract to establish the diagnosis, but barium swallow may be helpful when there is a question whether the radiolucency in the chest is an air-containing bowel, a lung cyst, or pneumothorax.

Associated congenital anomalies are frequent in patients with foramen of Bochdalek hernia. Approximately half of the patients who died had both patent ductus arteriosus, and patent foramen ovale; 22 per cent had preductal coarctation of the aorta. In a patient with compromised respiratory capacity due to displacement of bowel into the chest, the superimposition of these cardiac anomalies probably is significant in the fatal outcome. Since the incidence of cardiac anomalies in the survivors was very low, but in the fatalities rather high, one might speculate that a more aggressive approach to the cardiac anomaly in patients with Bochdalek hernias might increase the survival rate. Whether this should be attempted at the time of the repair of the diaphragmatic defect is unknown; these infants are usually acutely ill, and a rapid procedure is desirable.

The high incidence of gastro-intestinal anomalies is also an important factor in proper treatment. Two-thirds of the chil-

TABLE 3. *Associated Anomalities in Foramen of Bochdalek Hernias*

	%	No. Cases
Survivors		
Malrotation of intestine	16	4
Malfixation of intestine	8	2
Ureteral reflux	4	1
Meckel's diverticulum	4	1
Deaths with Autopsy		
Malfixation of intestine	66	6
Patent ductus arteriosus	55	5
Patent foramen ovale	44	4
Undescended testicle	44	4
Hypoplasia of one lung	33	3
Pre-ductal coarctation	22	2
Malrotation	22	2
Cleft lip	11	1
Hypoplasia of lobe of liver	11	1

TABLE 4. *Complications of Operation or Preoperative Evaluation in Bochdalek Hernia*

	No. Cases
Operations	34
No complications	9
Deaths	10
Tension pneumothorax	4
Wound infection	3
Incisional hernia	3
Persistent postoperative vomiting	2
Pneumonia	2
Intestinal obstruction secondary to adhesions (late)	2
Volvulus	1
Pyloric obstruction	1
Cardiac arrest with resuscitation (endotracheal tube error)	1
Breakdown of diaphragm repair	1 (×2)
Mediastinal emphysema	1
Perforation of stomach secondary to trochar thoracotomy	1
Other	6

TABLE 5. *Age Distribution of Deaths in Bochdalek Hernia*

Age	Cases	Deaths	Mortality (%)
Less than 24 hours	16	7	44
24 hours to 1 month	11	2	18
Over 1 month	8	1	12
Total	35	10	28

dren who died had malfixation of the intestines, and about one-fifth of all patients in the series had malrotation of the intestine. Failure to correct malfixation or malrotation may necessitate a second operation, as in one patient who developed postoperative intestinal obstruction secondary to a volvulus and a malfixation.

The frequent presence of associated gastro-intestinal anomalies has a strong influence on the choice of surgical approach to the hernial defect. It is difficult to evaluate the presence of these gastro-intestinal anomalies when the hernia is approached through the chest. Another disadvantage of the thoracic approach is the greater difficulty in reducing the hernia. Of the six patients in this series who had thoracic incisions, three required additional abdominal incisions before the hernia could be reduced. The abdominal approach may require an intentional incisional hernia because of the difficulty in closing the abdomen with the return of the intestines to the peritoneal cavity. However, the creation of an incisional hernia may improve the postoperative respiratory function and venous return by decreasing intra-abdominal pressure. An increased survival rate in newborn infants has been reported when incisional hernias were intentionally created.[4] It would therefore appear that the abdominal approach has definite advantages if no cardiac anomaly is suspected, or if repair of a cardiovascular anomaly is not contemplated. The thoraco-abdominal approach permits evaluation and possibly correction of associated cardiovascular and gastro-intestinal anomalies, but has the disadvantage of being a more extensive procedure.

Evaluation and treatment of diaphragmatic hernia carried a high complication rate in this series. Four patients developed clinically significant pneumothorax. Three of these could have been prevented if a chest tube had been connected to an underwater seal drainage instead of just aspirating the air from the chest. In view of the sometimes delayed expansion of the lung after reduction of the abdominal contents, and in view of the precarious respiratory status of these children, a chest tube should be used. It may be difficult to appreciate the severe degree of respiratory insufficiency that these infants may be experiencing in the immediate postoperative period. Because of this, the use of a positive-pressure respirator with an endotracheal tube should be considered for any infant who shows any indication of postoperative respiratory distress.

The fourth infant who developed pneumothorax had a left foramen of Bochdalek hernia with a severe degree of mediastinal shift. An emergency wet roentgenogram of the chest was interpreted as showing a pneumothorax on the right side. This confusion arose because the cardiac shadow was displaced to the right and the cardiac shadow was used as an indicator of the left side of the chest. A needle was placed in the right chest, resulting in a puncture of the right normal lung and a secondary pneumothorax on the right. The patient became more acutely cyanotic and died. Care must be taken in identifying the side of the lesion on the roentgenogram, as the cardiac shadow is not a reliable guide. The confusion of a left Bochdalek hernia with congenital dextrocardia has been reported.[1]

The possibility of confusing an air-filled stomach or dilated intestinal loop with pulmonary lesions is illustrated by a second case. A two-year-old boy presented with nausea, vomiting, respiratory distress, and mild cyanosis. Physical examination revealed hyperresonance on the left and decreased breath sounds on the left, as well as a tracheal shift to the right. The initial interpretation of the chest film was tension cyst of the left lung, and a chest tube was inserted, using a trochar. The tension cyst of the lung proved to be an air-filled stomach, and gastric contents spilled into

the pleural cavity and out the chest tube. The possible similarity of a roentgenogram of a pneumothorax and that of a chest containing air-filled abdominal content must be recognized.

Disruption of the hernia repair occurred in only one patient in this series. A six-week-old boy presented with cyanosis and vomiting, and had a thoraco-abdominal repair of a left-sided hernia. When he was eight months old, a large eventration of the lateral left diaphragm was repaired. He was asymptomatic until he was two years old, when he began to have intermittent episodes of brief unconsciousness. At operation, it was necessary to insert a nylon prosthesis into a very large recurrent defect of the left diaphragm. Even large defects can usually be closed primarily without the aid of grafts of foreign material, and this case represents the only example in which a prosthetic material was required to close the defect.

Whether or not the lung expands at operation or in the immediate postoperative period has a definite prognostic value. Fifteen of the 21 surviving patients who underwent operation had complete expansion of the lung by the first postoperative day. No child whose lung expanded easily at operation died. Eight children died after operation; none of these had attained full expansion of the lung after operation, and most of the lungs did not expand at all during surgery. Autopsies of these eight patients revealed only three cases in which the lung was hypoplastic.

The mortality rate was particularly high in newborn infants whose symptoms led to the diagnosis of diaphragmatic hernia during the first 24 hours of life (Table 5). Whether this mortality reflects the physiological alterations produced by the foramen of Bochdalek hernia or reflects the high mortality rate of the associated congenital defects is unknown. If a diaphragmatic hernia is diagnosed at age two days to 30 days, the mortality rate is one-third of that among newborn infants, but is still approximately 20 per cent. It is in this group of patients that the greatest improvement in survival rate is to be expected. These children have sufficient respiratory capacity to live at least 24 hours, and have no rapidly fatal associated anomaly. Earlier diagnosis and more thorough cardiac and gastro-intestinal evaluation are mandatory for an improved survival rate. The over-all mortality rate in this series (28 per cent, which includes one patient dead on arrival) is similar to that reported recently.[5]

Summary

1. Diagnosis of foramen of Bochdalek hernia is predominantly by roentgenographic examination.

2. Incidence of cardiovascular anomalies was high in those children who failed to survive.

3. Ease of reduction and incidence of associated gastro-intestinal anomalies indicate the abdominal approach for repair.

4. Failure of the lung to expand early is a sign of a poor prognosis.

5. The mortality rate of children with foramen of Bochdalek hernia is 28 per cent in this series.

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