Diaphragmatic Hernia

A Five Year Period Presentation of Cases at St. Luke's Hospital

MR. ARTHUR FELICE MD FRCS
CONSULTANT SURGEON
DIRECTOR DEPT. OF SURGERY

DR. CECIL VELLA MD
HOUSE PHYSICIAN-SURGEON
DEPT. OF SURGERY

Definition

A diaphragmatic hernia is the term used to define the herniation of abdominal organs through a congenital defect in the diaphragm. This term is thus not used to indicate a hiatus hernia or traumatic hernias.

Statistics

In the United Kingdom the incidence of diaphragmatic hernias is one in every 2000 live births. In Malta the incidence calculated over a five year period is one in every 1500 to 2000 live births. It has been estimated to be the cause of 8% of perinatal deaths and this figure demonstrates the high risk of the infant with such a hernia.

Embryology and Pathology

The separation of the pleural and peritoneal sacs of the embryo is completed between the eighth and the ninth week. The middle part of the forming diaphragm is composed of a mesodermal plate known as the Septum Transversum, while the lateral spaces known as the Pericardio-peritoneal canals are closed by the pleuro-peritoneal membranes.

If the diaphragm fuses late and the mid-gut returns early into the abdomen, then it would be possible for the elongated mid-gut to slip through the persistent gap in the diaphragm into the pleural cavity. This event will prevent the closure of the diaphragm and the gut will develop in length and calibre within the pleural space. The development of the lungs is thus effected.

The normal lung continues to develop new bronchial branches up to the 14th to the 16th week of intrauterine life. Pressure on the lung by the herniated mid-gut from the 8th to the 14th week may then have a considerable effect on the lung development. This happens both on the side of the hernia as well as on the contralateral side because of the mediastinal shift and the pressure on the contralateral lung bud. This provides an explanation for the frequent occurrence of bilateral pulmonary hypoplasia, which carries such a bad prognosis when it occurs to a severe degree.

Anatomy

The commonest hernia in the newborn occurs in the postero-lateral part of the diaphragm, on the left side more often than on the right. (Bochdalek 1848). There are no sacs in the Bochdalek hernia, the pleural and peritoneal cavities being in direct communication. This type of hernia must be distinguished from congenital eventeration of the diaphragm in which a sac of mesothelial tissue actually separates the abdominal from the thoracic viscera.

Finally, the anterior retrosternal hernia of Morgagni presents at birth very infrequently.

Cases

There were seven cases of diaphragmatic hernias over the five year period 1980-1985 at St. Luke's Hospital. Six of the hernias were of the Bochdalek type, all affecting the left diaphragmatic dome, while the seventh hernia was of the Morgagni type and this presented at the age of two years. Five of the Bochdalek hernias were operated at birth while the sixth died before surgical intervention was possible.

The calculated incidence over the five years was of 1 in 1500 - 2000 live births. The average mortality of the six cases including the case which was not operated was around 65%. The following table compares the sex, birth weight, gestation, one minute Apgar score, presentation and associated anomalies in the seven cases.

Sex	Gestation (weeks)	Weight (kg)	Apgar (1 min)	Presentation	Anomalies
(F)	39	3.06	6	Respiratory distress Floppy infant	Transposition of great arteries
(M)	39	3.36	5	Respiratory distress	Hypoplastic (L) pulmonary artery
(F)	39	3.00	8	Respiratory distress	Hypoplasia of lungs
(M)	40	2.95	9	Respiratory distress	Dandy-Walker Cyst
(M)	39	2.80	9	Asymptomatic	Marfan's syndrome
(M)	40	3.00	8	Respiratory distress	Nil
(F)	40	?	?	Respiratory distress	No postmortem

Discussion

The calculated incidence was of 1 per 1500-2000 live births. The male to female ratio was approximately 1:1 and all of the cases involved the left side of the diaphragm. In all the Bochdalek hernias there were no relevant findings in the family history nor in the antenatal examination. Two mothers had rheumatic fever in childhood but this was incidental. On the other hand in the Morgagni hernia there was a family history of Marfan's syndrome, which is known to be associated with defects of connective tissue including diaphragmatic hernia (McKusick V.A., Pyeritz R.E. 1979). Up to now there has been no established cause of this congenital anomaly but evidently diaphragmatic hernias of both types are very commonly associated with other congenital anomalies.

Presentation

Diaphragmatic hernias usually present immediately after birth. The presenting features result from the mechanical effects which the herniated abdominal organs have on the already hypoplastic lungs. Increase in the amount of intra-thoracic contents also causes shifting of the mediastinum to the opposite side. The commonest presenting problem is therefore cardio-respiratory embarrassment. The infants present with a low Apgar score, tachypnoea, cyanosis and shifting of the heart to the opposite side which may give the impression of a dextrocardia. The diagnosis is confirmed by a plain X-Ray of the chest which shows the presence of air-containing abdominal viscera in the chest, together with mediastinal shift. In cases where the amount of herniated viscera is large, the abdomen has a scaphoid appearance. In two of the described cases, which had a relatively elevated Apgar score, the diagnosis was suspected by slight tachypnoea and the presence of bowel sound within the thorax. The diagnosis was confirmed by chest X-Ray.

Management of Diaphragmatic Hernia

Diaphragmatic hernias must be considered as emergencies of the first order. Although the treatment is ultimately surgical there are a number of resuscitation procedures which should be performed in order that the infant reaches the operating theatre in the best possible condition.

Oxygen must *never* be given by face mask or bag as the swallowing of air into the intestine will cause further distension and deterioration in respiratory function. The infant should be intubated as soon as possible and oxygen then given by IPPV. An infant nasogastric tube should be inserted and frequent aspiration performed to prevent fluid accumulation and to decrease the risk of aspiration pneumonia. In cases where a pneumothorax develops before surgery the chest should be drained via a tube drain in the second intercostal space.

Indications for Operation

The exact degree of lung hypoplasia cannot be assessed pre-operatively and one is practically duty bound to repair any such hernia, well knowing that a proportion of these children will not have the respiratory potential to survive. This can only be found by clinical testing during convalescence and subsequent post-mortem in those that die.

Operative Treatment

The choice of incision depends on the surgeon but a sub-costal incision (usually on the left) gives good exposure, provides an opportunity to inspect the abdominal viscera and perform a gastrostomy for gastric decompression.

The abdominal viscera are gently pulled out of the chest, a Malecot catheter passed through the ninth intercostal space and connected to an underwater seal. The defect in the diaphragm is then closed

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using a double - breasting technique. If there is a complete absence of the left diaphragm, a flap of the lower rib-cage, connected by the intercostals, is fashioned and is sutured to the thin rim of diaphragm on the posterior wall.

Post-Operative Care

Intermittent positive pressure ventilation is usually required for the first few hours after operation and moreover in those cases where hypoplasia of the lung is marked. In these high pressure ventilation is usually required and ventilation-induced pneumothorax is a common complication. Regular post-operative chest X-Rays are thus required to assess reinflation of the lung and also the possibility of contralateral pneumothorax. The timing for removal of the infant from the ventilator is vital and in general the return to spontaneous breathing with well maintained blood gases is the principal indication for extubation. Meanwhile continuous gastric aspiration and intravenous therapy are essential till the infant is well enough to resume oral feeding.

Sequelae in Surviving, Post-Operative Cases

The most common complications in the early post-operative period are related to pulmonary function and have already been outlined. Other complications may arise from the variety of associated anomalies affecting the heart and large vessels. In general infants who require prolonged ventilation are those who have marked hypoplasia of the lungs, having therefore a smaller chance for survival. The overall mortality is in the region of 65%. The long term morbidity results from other congenital anomalies frequently associated.

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