CONGENITAL ANTERIOR DIAPRAGMATIC HERNIA (MORGAGNI) IN NEWBORN

(Case Report)

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INTRODUCTION

A Diaphragmatic hernia,acquired or congenital,is a protrusion of abdominal viscera through a defect in the diaphragm into the low pressure of the mediastinal space or pleural cavity from the high pressure of the abdominal cavity(1).

Morgagni hernias occur behind the sternum through defects in the diaphragm that are perhaps secondary to a developmental failure of the retrosternal segment of the transverse septum (1,2,3,4).

The fusion of the diaphragmatic membrane occurs during the tenth week of gestation and coincides with the return of the intestine into the abdominal cavity from the umbilical pouch(4).

Morgagni's hernia are the rarest type of congenital diaphragmatic hernia,accounting for one per 300 hernia (3).

The clinical manifestation (3,5) :

- significant respiratory distress and cyanosis
- decreased breath sound over involved part of thorax,often bowel sounds will be audible instead.
- shift of cardiac impulse
- scaphoid abdomen
- x-ray film of chest revealing loops of bowel in thorax with mediastinal shift away from involved side;at birth with no air yet in the bowel,an x-ray film may give appearance of dense opacification of involved side of chest.

Morgagni's hernia may have a more subtle onset,manifested by feeding problems and mild respiratory distress(6).

The principles of management of diaphragmatic hernia are to maintain ventilation as adequately as possible and to do surgical repair through either the abdomen or the chest;with reduction of intestine into the abdominal cavity(6).

Despite modern advances,mortality from diaphragmatic hernia remains very high,approaching 50 %.Although repair of the defect itself is relatively straight forward,the underlying pulmonary hypoplasia and pulmonary hypertension are largely responsible for overall mortality(6).

The purpose of this paper is to report a case of congenital anterior diaphragmatic hernia (Morgagni) in newborn.

CASE REPORT

S,a five day old male infant was admitted to the Neonatal Intensive Care Unit Department of Child Health Dr Sutomo Hospital on September 29,1992 with the main complaint of dyspnea.According to his mother,the baby had suffered from dyspnea since birth.The dyspnea was recurrent and intermittent,became more evident while the baby was crying or breast fed.When the dyspnea was severe,there was cyanotic circumoral ,too.

The baby was breast fed well and had no complaint of vomiting,restless sleep,fever,cough,coryza nor convulsion.The stool and urine passage were normal.

He was delivered at term,spontaneously,helped by a midwife with the birth weight of 3000 grams and cried immediately after

birth.He was the youngest of five siblings,the others are healthy.The occurance of any disease and the history of any drugs or traditional herbs taken in pregnancy were denied by his mother.

Physical examination on admission in NICU revealed a restless and dyspneic baby with a body weight of 3500 grams.The respiratory rate was 100/minute,the pulse rate was 144/minute and the temperature was 36,7 C.There were no jaundice nor anemia.There were flaring of the nose and circumoral cyanosis.The thorax was slight barreling with minimal subcostal retraction.There was no pericardial hyperactive,no murmur and the punctum maximum of the heart was still on the left.The breath's sound was decreased on the right hemithorax.No rales or rhonchi were found but sound of intestinal peristaltic was heard on the right chest.There were scaphoid abdomen ,normal intestinal peristaltic and the liver / spleen were not palpable.The extremity were normal,without edema or cyanosis.

Based on these findings,the working diagnosis was suspected congenital diaphragmatic hernia with differential diagnosis of eventration of the diaphragm.

The baby was fasted,a nasogastric tube was inserted and head up position was given.The treatment consist of oxygen administration 2 l/minute,ivfd of dextrose 10 % in 0,18 Saline 475ml/day and ampicillin 150 mgs twice a day .A chest and abdominal rontgenography, ECG , Blood Gas analysis and serum electrolyte were performed to confirm the diagnosis.

The chest and abdominal rontgenogram showed the shape and size of the heart were normal but there was shifting of the heart to the

left.In the right hemithorax,there was luscen appearance of air filled loops like intestine.

The ECG was normal , the blood gas analysis revealed pH 7.282 ,pCO2 46.9,pO2 37.1,HCO3 22.1, BE - 4.7,serum electrolyte Na 133 meq/l , k 4.58 meq/l.

As the diagnosis was confirmed,the baby was consulted to the surgery department on September 13,1992.The surgeon agreed to do urgent operation the day after and the baby was transfered to the anesthesiology department immediately.

On day 2 (September 13,1992) at Anesthesiology Department :

The dyspnea seemed to be diminished,the respiratory rate was 36 /minute,the pulse rate 120/minute.There was no flare of the nose.On the right hemithorax the breath's sound was still decreased and the peristaltic's sound was heard.The abdomen was still scaphoid.

The baby was fasted and given :

- oxygen 1 l/minute (nasal catheter)
- ivfd dextrose 10 % in 0.18 saline 300 ml/day
- ampisillin 150 mgs twice / day
- suction of the nasogastric tube every 2 hours
- serial blood gas analysis and serum electrolyte
- head up position

Laboratory examination revealed : hemoglobine 17.9 g/dl,leucocyte 3100/cmm,thrombocyte 260,000 , Hematocrit 63 %,blood glucose level 160 mg/dl,SGOT 3 , BUN 7.4 ,K 3.96 and Na 145.

On day 3,

The operation was performed using the abdominal approach.The finding on operation was herniation of right lobe of the liver,ileum and part of yeyunum through foramen Morgagni.The surgeon did reposition of the abdominal viscera;inserted the boulleau drainage and closed the defect in the diaphragm.The pressure of -5 cm H2O was given to the boulleau drainage.After being extubated,the baby was transfered to the ICU of Anesthesiology Department.

Early after operation,the baby breath spontaneously and adequately.The respiratory rate was 60/minute,without flaring of the nose,no cyanosis,the breath's sound was vesicular and symmetric, no intestinal peristaltic was heard.The heart rate was 132 /minute,regular without any murmur.

The Laboratory examination revealed: Hb 16 gr%,pH 7.3 , pCO2 46.7, pO2 67 , BE -2.6, Na 127 and K 3.97.Chest rontgenogram showed good inflation of both lungs and the heart position was normal.

The treatment was given :

- ivfd dextrose 10 % in 0,18 Saline 300 ml/day
- ampisillin 3 x 100 mg / day
- metamizol 0.1 ml (if needed)
- oxygen 3 l/minute (nasal catheter)
- boulleau drainage - 5 cm H2O
- periodic suction of the nasogastric tube
- head up position
- chest physiotherapy.

On day 4 (first day after operation),the baby was allert,cried loudly . The heart rate was 124/minute,the respiratory rate was 40/minute.There were no flare of the nose , no cyanosis,the breath's sound was vesicular on both side of hemithorax but decreased on the lower part of right hemithorax.The abdomen was normal,with normal intestinal peristaltic.There was a leakage (minimal)from the bolleau drainage.

The laboratory examination revealed :Hb 17.8 gr/dl,pH 7.38,pO2 142.1,pCO2 43.9, BE 1.5 ,Na 127.8 and K 4.9.

The treatment was given :oxygen 4 l/minute (ambient),ivfd dextrose 10 % in 0.18 Saline 360 ml/day,Ampicillin 3 x 100 mg,Becombion 1 ml drip/day,vitamin C 200 mg drip/day,periodic suction of the nasogastric tube ,Boulleau drainage - 5 cm H2O, chest physiotherapy, head up position and repair the leakage of the drainage.The chest rontgenogram showed a minimal pneumothorax on the lower lobe of right lung.

On day 5 (second day after operation),the baby's condition was good.The respiratory rate 32/minute and the pulse rate 120/minute.The normal intestinal peristaltic was heard on the abdomen.The baby was given oral nutrition,starting with dextrose 5 % continued with 10 % infant formula 12 x 10 ml through naso-gastric tube.There was approximately 5 ml liquid from leakage of the Boulleau drainage.Microlax was given to the baby to stimulate defecation .The other treatment was continued.

On day 6 ,the baby was allert,respiratory rate 60 /minute,pulse rate 120/minute.There were no flaring nor cyanosis with ambient oxygen 3 l/minute.The breath's sound on the lower right hemitho-

rax slightly decreased.The abdomen was normal with normal intestinal peristaltic without meteorism.

The laboratory examination revealed : Hb 19.6 gr/dl,Leucocyt 3700/cmm,differential count -/-/-/66/34/- , pH 7.414 , pO2 148.7,pCO2 24.9,BE - 7.3, Na 129.6 and K 3.63.

Chest rontgenogram showed the heart size and shape were normal,the pneumothorax on the lower lobe of the right lung decreased.

The baby was still treated with ivfd ,10 % infant formula 12x15ml/day with nipple ,Ampicillin,Becombion and Vitamin C

On day 7,theheart rate 120/minute,the respiratory rate 48/minute,temperature 37 C,the body weight was 3500 gr.The general condition was good.There was no flaring nor cyanosis,the heart's sound was normal,the breath's sound was vesicular and symmetric.The abdomen tender and intestinal peristaltic was normal,surgical wound was good.The liquid from the boulleau drainage was minimal.

The treatment was given : oxygen (ambient) 3 l/minute,ivfd dextrose 10 % in 0.18 Saline 240 ml/day,12.5 % infant formula 12 x 30 ml/day,Ampicillin 3 x 100 mg/day,Becombion and Vitamin C.

The chest rontgenogram still showed minimal pneumothorax on the lower lobe of the right lung.

The surgeon released the boulleau drainage and the baby was transfered to the NICU.

On day 8,the general condition was good,the ivfd and oxygen administration were stopped and the baby was given 15 % infant

formula 12 x 55 ml/day,Ampicillin 2 x 150 mg/day.

On day 10,the baby was breast fed well.There was no dyspnea,the stoll and urine passage were normal.Ampicillin was still given.

On day 11,The baby's condition was good,the surgical wound was good ,chest rontgenogram showed the normal shape and size of the heart and good inflation of both lung,without any pneumothorax. The baby was consulted to the surgery department and the surgeon agreed to discharge him and treated him as an outpatient.

On day 12,the baby was discharge in good condition,with body weight 3500 gr.

DISCUSSION :

The herniation of the abdominal organ through a defect on the diaphragm is usually congenital.

The most common type of congenital diaphragmatic hernia is the Bochdalek's hernia which represents failure of pleuroperitoneal canal to close completely during embryonic development.The defects occurs in the posterolateral aspect of the diaphragm,the left side is affected in 80 to 90 percent of cases(1).

Morgagni's hernia occurs in either side of the inferior end of the sternum,representing failure of midline fusion of the embryonic septum transversum,the lateral component of the diaphragm and the anterior thoracic wall.The defect on the left is usually obliterated by pericardium,therefore,most of the hernia are on the right(3).

Our patient had a Morgani's hernia and the defect was on the

right side,in accordance with the previous finding.

The incidence of congenital diaphragmatic hernia 1:2200 (5) to 1:4000(6), and constitutes 8 % of major congenital anomalies(3).Morgagni's hernia is the rarest type,occurs in 1 per 100 to 1 per 300 congenital diaphragmatic hernias(3).

The incidence of associated anomalies with congenital diaphragmatic hernia have varied from rare to 56 %(5), especially neural tube defects,cardiac defects,intestinal malrotation and chromosomal abnormality (trisomy,tetraploidy)(5,7).

There was no associated anomalies in our patient and this caused better prognosis.

The diaphragm develops from four embryonic components(1) :

Transverse septum

The growing head fold of the embryo brings a wall of mesoderm to a position cranial to the open midgut and caydal to the heart during the third embryonic week.This mesoderm forms the ventral component of the future diaphragm.

The cranial surface of the transverse septum also cotributes to the connective tissue of the pericardium,and the caudal surface contributes to the capsule and stroma of the liver.

Mediastinum

The mediastinum is the thick dorsal mesentery of the foregut,containing the future esophagus and the inferior vena cava.It is cotinuous anteriorly with the transverse septum and posteriorly with the axial mesoderm.Byposterior and caudal extension it splits to form the diaphragmatic crura.

Pleuroperitoneal Membranes

The pleuroperitoneal membranes close the right and left communication between the pleural and peritoneal cavities at about the eight embryonic week.Originally they form a large part of the developing diaphragm,but relative growth of other elements reduces their contribution to a small area.

Muscles of the Body Wall

Myotomes of the seventh to twelfth segments contribute the lateral component of the diaphragm by caudal excavation of the thoracic wall to form the costodiaphragmatic recesses.This process produces the final domed shape of the diaphragm.

In the third week,the transverse septum lies at the level of the third cervical vertebra,and the developing diaphragm descends to its final position at the level of the first lumbar vertebra by the eight week.The phrenc nerve,which originates from the third to fifth cervical levels,is carried caudad with the descending diaphragm.

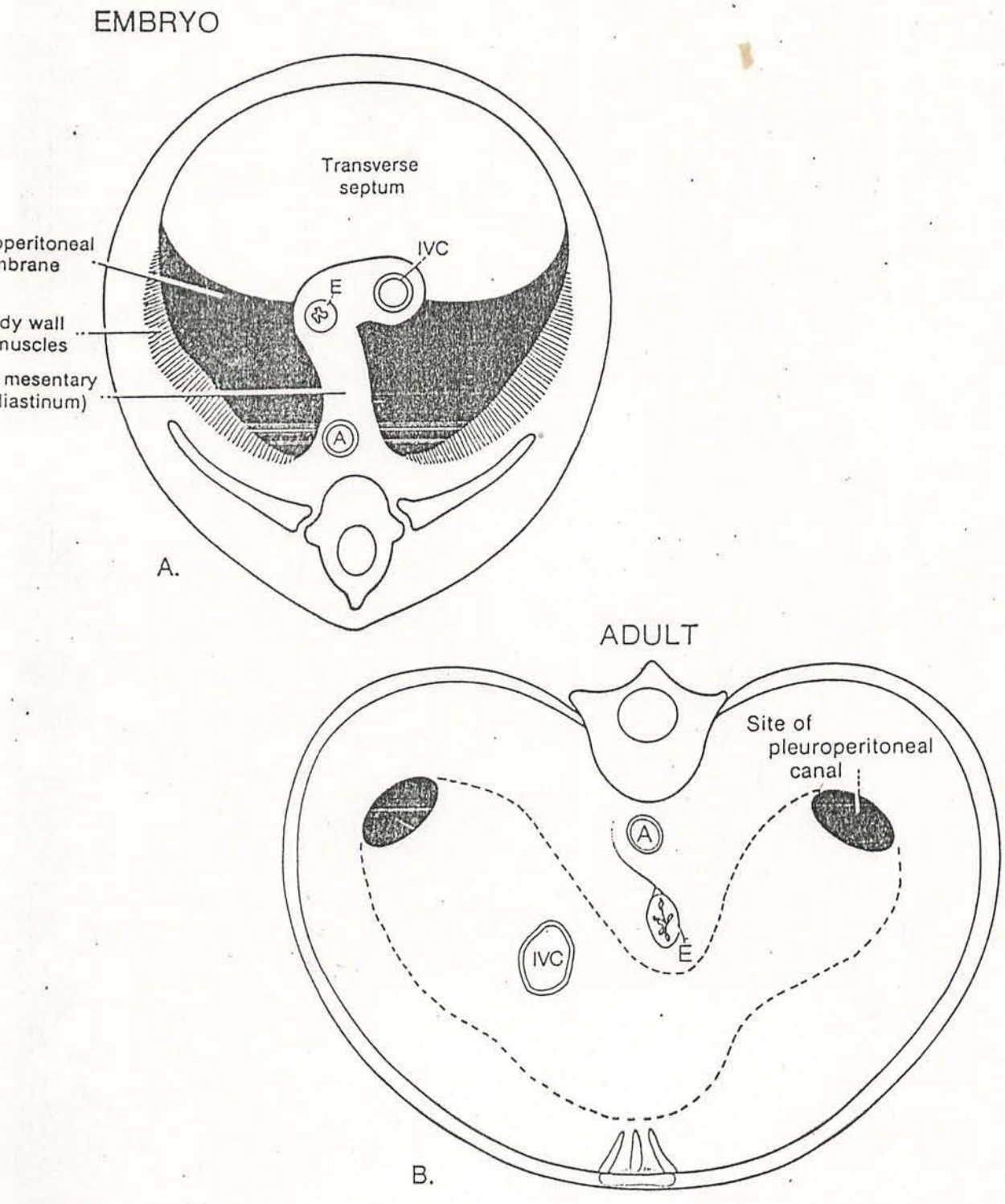
During the first 2 months of fetal life,there is no pressure on the developing diaphragm from above or below.Above,the lungs are not infated;below,the growth of the gut is talking place extraabdominally into the umbilical cord.The first mechanical pressure on the diaphragm comes during the tenth week when the intestine return from the umbilical cord to the abdomen.By that time all of the diaphragmatic components are normally in place and have sufficient strength to contain the abdominal viscera.This may not be the case if the normal development timetable is disturbed.

Embryology of the Diaphragm

A. The four embryonic components of the diaphragm.

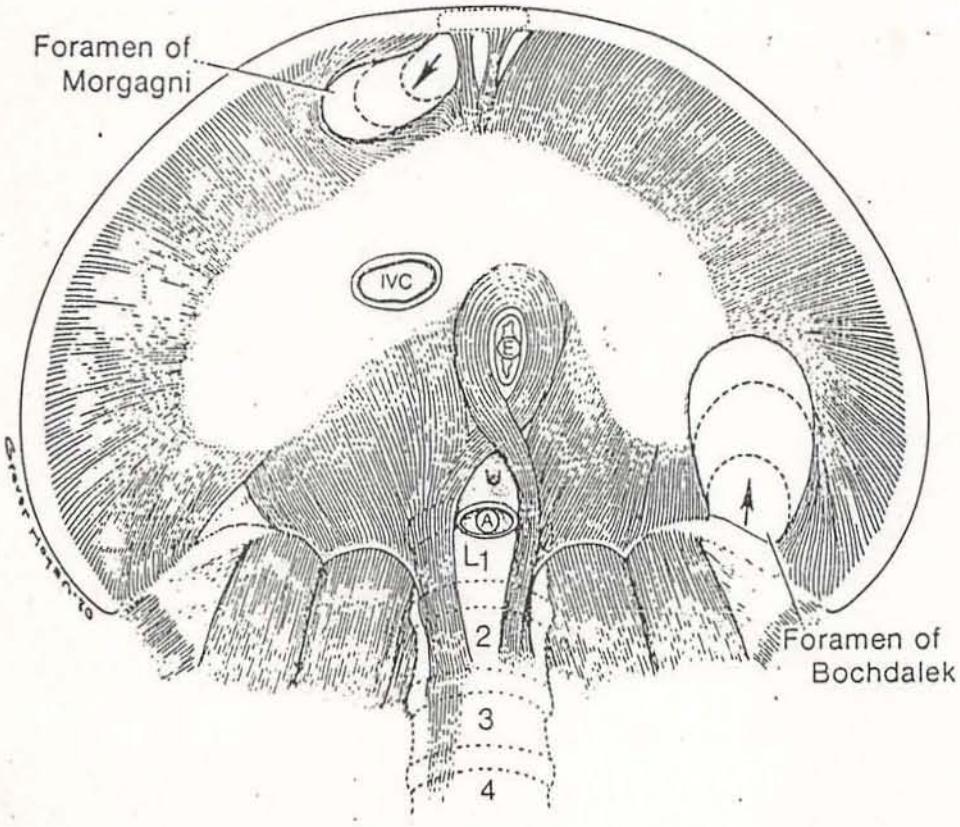
B. The adult diaphragm. The sites of the closed pleuroperitoneal canals occupy a relatively small area in the adult diaphragm.

Source: Plate 96A,B,C from Skandalakis JE, Gray SW, Rowe JS Jr.: Surgical anatomy of the diaphragm, vol. 1, in *Mastery of Surgery*, Nyhus LM, Baker RJ (eds). Little, Brown, Boston, 1984, p. 303, Figs 38-2A,B, Fig. 38-3.



The diaphragm from below, showing the foramen of Bochdalek and the foramen of Morgagni. Both are weak areas of potential herniation. Arrows indicate the direction of enlargement after herniation has begun.

Source: From Skandalakis JE, Gray SW, Rowe JS Jr.: Surgical anatomy of the diaphragm, in *Mastery of Surgery*, Nyhus LM, Baker RJ (eds). Little, Brown, Boston, 1984, p. 306, Fig. 38-5.



Since the formation of the diaphragm is complete by ninth week of intra uterine life,arrests in the development take place before this time. If a defect exists,the returning intestines at the tenth week may pass immediately into the thoracic cavity.Lung development normally continues until about the fourteenth to sixteenth week of life.If the intestines lie within the thoracic cavity and compress the ipsilateral lung by direct pressure and the contralateral lung indirectly by shift of the mediastinum,it is possible that this would account for the retarded pulmonary development(8).This hipoplasia is a major factor in the excessive mortality in the early neonate with or without operation(9).

The combined lung weights in non survivors are distinctly below the average similar weights for other stillborns of the same range of body weight( ).Actually,lung expansion and growth occur in most survivors either by an increase in the number of alveoly or increase in alveolar size or both but may take days or weeks,depending on the degree of pulmonary differentiation (10). The pathophysiologic features of the associated respiratory failure involve pulmonary hypoplasia as well as progressive pulmonary vascular hypertension (9).

The hypoplastic lungs cannot adequately ventilate or oxygenate,which leads to arterial oxygen desaturation,a mixed respiratory and metabolic acidosis and finally pulmonary hyper-tension.

This situation is compounded by the abnormal pulmonary arteial tree,which has more medial muscle and is thus more vasoreactive

(11).Hypoxia,hypercarbia and acidosis may all further stimulate vasoconstriction in the pulmonary arterial bed,raising pulmonary artery pressure and leading to right to left shunting through the foramen ovale or the ductus arteriosus(9,10,12).

Although many organ was migrated to the thoracal cavity,our patients did not suffered from pulmonary hypoplasia nor pulmonary hypertension.This condition may associate with the size of the defect,the affected side and the time of herniation takes place.
There are variation in symptoms,depends on the amount of the abdominal viscera which was migrated to the thoracal cavity.Large congenital diafragmatic hernia may present at birth wih cyanosis,respiratory distress,ascaphoid abdomen,decreased or absent breath sound on the side of hernia and heart sounds displaced to the side opposite the hernia.Small hernias,right sided hernias and substernal hernias of Morgagni may be asymptomatic or may have a more subtle onset,manifested by feeding problems and mild respiratory distress such as constipation,symptoms simulating those of gall bladder or peptic ulcer disease ,retroxiphoid pain,dyspnes,cough.
Our patient only suffered from intermittent and recurrent dyspnea,without any symptoms of gastrointestinal disturbances.Suctioning of the nasogastric tube was minimal. This condition may associated with temporary herniation of the abdominal viscera .The gaster was not involved and the intestine was temporary herniated,thus the gastrointestinal passage remain free.

The diagnosis is based on clinical findings, confirmed by x-ray showing a moderately dense tumor (usually at the rigt cardio

hrenic angle in the postero anterior film and in the anterior ediastinum on the lateral view) and loops of bowel in thorax ith mediastinal shift away from involved side of the hest(5,13,14).Hernia that do not contain bowel are the more ifficult diagnostic problem(3,5).

:linical findings in our patient was not typical or classical)ecause Morgagni's hernia was rare.With careful examination,signs ınd symptoms supporting the diagnosis was found.The rontgenogram :onfirmed the diagnosis.

Prenatally,the diagnosis is often made by ultrasonic studies,which may be precipitated by the occurence of polyhydram-ıions.However,a prenatal diagnosis should lead to delivery in a :enter equipped to handle the problem to optimize chances for survival as much as possible.Despite sophisticated perinatal management,the overall outcome was dismal.In the future,prenatal diagnosis may permitsurgical intervention before birth,permiting lung development to take place during the remainder of pregnancy with survival at birth.(15)

On rontgenogram,the lesions simulate congenital cystic adenoma-toid malformation,diffused congenital pulmonary cysts and pneuma-toceles.(3)

It may be impossible but is unnecessary to distinguish diaphrag-matic hernia,eventration and phrenic nerve paralysis(3,8).

When the diagnosis is made,additional oxygen ,ventilation by endotracheal tube(if necessary),continous intragastric suction and provision of glucose infusion & external warmth should be provided to all patients during transportation and until opera-

tion can be performed.Bag and mask ventilation is contraindicated and care must be taken with assisted ventilation to keep inspiratory pressure low to avoid damage or rupture of the contralateral lung.The supportive care was performed to our patient since admission ,without requiring any ventilation.

For many years it has been generally agreed that operation should be carried out without delay(8). Surgical repair is through either the abdomen or the chest,with reduction of abdominal viscera into the abdominal cavity.Small baby may require abdominal mesh if the abdominal cavity is too small for the intestines.

Bohn,et al(16) and Sakai,et al (17) suggest to delay the time of surgery for newborn infants with congenital diaphragmatic hernia.The reason is that pulmonary hypoplasia (not atelectasis) exists and will not be changed significantly by surgery;thus vigorous medical treatment of factors affecting the pulmonary vascular bed and increased pulmonary vascular resistance may improve the chance of the patient tolerating surgery and the adverse mechanical effects of surgery on the chest wall and diaphragm.

In our case, the surgical treatment was performed on the third day of admission .The consideration was the condition of the baby,and the second day was Sunday.The surgical correction was successful,with evidence of no respiratory problem thereafter except minimal pneumothorax,which was dissapeared on day 8 after operation.

Some authors reported the use of Tolazoline for persistent

pulmonary hypertension after congenital diaphragmatic hernia(18,19,20) and seemed to be successful only in patients with adequate pulmonary vasculature who suffered from pulmonary vasospasm.

The complication of congenital diaphragmatic hernia includes hypoxia,hypercarbia,acidosis,persistent pulmonary hypertension, hypoplasia of lungs,pneumothorax.

Despite modern advances,mortality from diaphragmatic hernia remains very high,approaching 50 % (6).Mortality is high because of the delay in recognition,failure to maintain respiratory support and pulmonary insufficiency from hypoplasia of lungs or too agressive replacement of viscera in a limited abdominal space.Newer therapies,including Extracorporeal membrane oxygenation (ECMO),offer the promise of improved survival.

The pronosis of our patient is good,since no complication was found after the successful operation.

SUMMARY

A case of congenital anterior diaphragmatic hernia (Morgagni) in a five day old baby has been reported.The etiology,patophysiology ,clinical picture ,diagnostic support,management and treatment were discussed.The patient was discharge in a good condition.

EFERENCES :

. Skandalakis, J.E; Gray, S.W;Mansberger, A.R;Colborn, G.L ; Skandalakis, L.J : Hernia . Surgical anatomy and technique. International ed.New York,M Graw-Hill Informations Services Co,1989;305-71.

:. Behrman,R.E and Speck,W.T :Peritoneum and allied structures, in Behrman,R.E and Vaughan,V.C. eds: Nelson textbook of pediatrics.12th ed.Philadelphia,W.B.Saunders Co,1983:986-90.

3. Salzberg,A.M and Krummel,T.M :Congenital malformations of the lower respiratory tract, in Chernick,V and Kendig,E.L. eds:Disorders of the respiratory tract in children.5th ed.Philadelphia,W.B.Saunders Co,1990:227-67.

4. Ziai,M :Pediatrics.4thed.Boston,Little,Brown and Co, 1990: 158-9.

5. Babson,S.G ;Pernol,M.L andBenda,G.I :Diagnosis and management of the fetus and neonate at risk.4th ed. St Louis,C.V.Mosby Co,1980:278-88

6. Ringer,S.A : Surgical emergencies in the newborn,in Cloherty, J.P and Stark, A.R. eds: Manual of neonatal care.3rd ed.Boston,Little,Brown and Co,1991:496-510.

7. Kashani,I.A ;Kimmons,H ; Valdescruz,L,M ; et al:Congenital right-sided diaphragmatic hernia and hypoplastic left heart syndrome.Am heart J 1985;109 :177-8.

8. Synder,W.H and Greaney,E.M : Congenital diaphragmatic hernia;77 consecutive cases.Surgery 1965; 57 :576-89

9. Naeye,R.L ; Shochat, S.J ;Whitman V ; et al :Unsuspected pulmonary vascular abnormalities associated with diaphragmatic hernia. Pediatrics 1976;58:902-6.

10. Wohl,M.E.B ;Griscom,N.T ;Strieder,D.J ;Schuster,S.R ;Treves,S ;Zwerdling,R.G : The lung following repair of congenital diaphragmatic hernia.J Pediatr 1977;90:405-14.

11. Levin,D.L :Morphologic analysis of the pulmonary vascular bed in congenital left - sided diaphragmatic -hernia.J Pediatr 1978;92:805-9.

12. Geggel,R.L;Murphy,J.D;Langleben,D;Crone,R.K;Vacanti,J.P;Reid,L.M :Congenital diaphragmatic hernia : Arterial structural changes and persistent pulmonary hypertension after surgical repair.J Pediatr 1985;107:457-64.

13. Marwood,R.P and Davidson,O.W : Antenatal diagnosis of diaphragmatic hernia.Case report.Br.J.Obstet Gynaecol 1981;88:71-2.

14. Benacerraf,B.R and Frigoletto,F.D:In utero treatment of a fetus with diaphragmatic hernia complicated by hydrops,Am J Obstet Gynecol 1986;155:817-8.

15. Benacerraf,B.R and Adzick,N.S : Fetal diaphragmatic hernia:ultrasound diagnosis and clinical outcome in 19 cases.Am J Obstet Gynecol 1987:156:573-6.

16. Bohn,D.B;Tamura,M;Perrin,D;Barker ,G;and Rabinovitch,M :Ventilatory predictors of pulmonary hypoplasiain congenital diaphragmatic hernia,confirmed by morphologic assessment.J Pediatr 1987;111:423-31.

17. Sakai,H;Tamura.M;Hosokawa,Y;et al:Effects of surgical repair on respiratory mechanics in congenital diaphragmatic hernia.J Pediatr 1987;111:432-8.

18. Shochat,S.J ;Naeye,R.L ;Ford,W.D.A ;Whitman V ;Maisels,M.J :Congenital diaphragmatic hernia.New concept in management.Ann Surgery 1979;190:332-40.

19. Levy,R.J;Rosenthal,A;Freed,M.D;Smith,C.D;Eraklis,A; Nadas,A.S:Persistent pulmonary hypertension in a newborn with congenital diaphragmatic hernia :Successful management with Tolazoline.Pediatrics 1977;60:740-2.

20. Bloss,R.S ;Turmen,T ;Beardmare,H.E and Aranda,J.V :Tolazo-linetherapy for persistent pulmonary hypertension after congenital diaphragmatic hernia repair.J Pediatr 1980;97:984-88.