ABSTRACT

Congenital diaphragmatic hernia (CDH) is an anomaly characterized by a discontinuity of the diaphragm, which allows the abdominal viscera to herniate into the chest during embryonic and fetal development. Almost certainly fatal if untreated. Hernia repair is now widely accepted and has become established as the primary treatment for CDH. There were three cases identified during six months that were diagnosed with CDH. These patients with respiratory distress, and radiology and laboratory evidence of CDH, need urgent surgical exploration of the hernia. These patients underwent hernia repair. All of the operations were succeeded although all experienced early post-operative complications, pneumothorax and pleural effusion. These patients were received different mode of ventilation. One patient died, while the others survived. The prognosis of these patients were poor without hernia repair.

Keywords: congenital diaphragmatic hernia, hernia repair, children


INTRODUCTION

Congenital diaphragmatic hernia (CDH) is an anomaly characterized by a discontinuity of the diaphragm, which allows the abdominal viscera to herniate into the chest during embryonic and fetal development. Congenital diaphragmatic hernia is a significant cause of neonatal morbidity and mortality that is often seen as a purely pediatric and surgical problem. Congenital diaphragmatic hernia is almost certainly fatal. Pulmonary hypoplasia, a hypoplastic left heart ventricle, and persistent pulmonary hypertension of the newborn (PPHN), with retention of the fetal circulation, lead to marked respiratory distress. Despite treatment in intensive care, survival is only 40-60% worldwide, or up to 80% in specialist centers. Early intrauterine diagnosis is important. Unfortunately, in Indonesia, CDH is often forgotten and accidentally diagnosed after formed by passing through these canals. The most influential surgical approach for CDH came from William Ladd and Robert Gross which advocated rapid diagnosis by chest X-ray and/or barium meal with early operative intervention.

Morgagni described the hiatal hernia through the foramina of Morgagni in 1761. In 1848, Bochdalek referenced the development of the pleuropertitoneal canals while describing the hernia that formed by passing through these canals. The most influential surgical approach for CDH came from William Ladd and Robert Gross which advocated rapid diagnosis by chest X-ray and/or barium meal with early operative intervention.
The purpose of this article is to remind and compare the evaluation, management, and determinant of outcome three patients with CDH who underwent hernia repair.

CASE ILLUSTRATION

Over a period of six months from April to September 2016, a total of three patients with CDH were considered retrospectively. Of these, two patients were survive and one patient was died. All of them presented as respiratory distress soon after birth.

First case

EKB, a three-hour-old baby, was referred to Sanglah Hospital because of respiratory distress after birth. He had already intubated when admitted to emergency room. He was born through cesarean section on the 38th week of gestation. Apgar scores were 6 and 7 at 1 and 5 minutes respectively. He began breathless, cyanotic and the chest wall became retracted predominantly on the right side.

There were tachycardia, tachypnea, nasal flare, decreased of respiratory sound on the left side accompanied by displacement of the heart sounds to the right side of the chest. The chest wall was asymmetrical on movement. Downes score was 8. The abdomen was excavated with sternal protrusion, no bowel sound, the liver and spleen were not palpable. Complete blood count, C-reactive protein (CRP) and immature to total (IT) ratio were normal. The chest X-ray showed heart pushed toward right side of chest, multiple cystic lesion in left lung (Figure 1). Echocardiography was normal.

He was diagnosed with respiratory distress and CDH. He was fasting and decompressive enteric tube was inserted by mouth. He admitted to Neonatal Intensive Care Unit (NICU) and started on pressure-controlled ventilation. Blood gas analysis 2 hours after ventilation was normal. He underwent urgent hernia repair at 4 days old. Intraoperative findings revealed a left posterolateral diaphragmatic defect with ileum, colon, stomach and spleen found in the hernial sac (Figure 2) and definitely diagnosed with left Bochdalek type diaphragmatic hernia. Chest X-ray a day after hernia repair assumed minimal pleural effusion and pneumonia.

Two days after surgery, he got fever and jaundice and the antibiotics were changed. Ventilator support was stopped 3 days after surgery. The blood culture was positive and antibiotic continued until 14 days before he discharged from hospital.

Second case

KSB, a newborn baby, suffered respiratory distress soon after birth in Sanglah Hospital. There was no spontaneous breath and he was cyanotic. He was born through vaginal delivery on the 37th week of gestation. Apgar scores 2, 4, 6, and 8 at 1, 5, 10 and 20 minutes respectively. Soon after birth, he was intubated along with chest compression. After 10 minutes, he began to breathe spontaneously. The Downes score was 8.

There were tachycardia with decreased respiratory sounds on the left side. The chest wall was asymmetrical on movement. The abdomen was excavated and no bowel sound was heard. The liver and spleen were not palpable.

Complete blood count, CRP, IT ratio, and blood glucose were normal. Prothrombin time, activated partial thromboplastin time, and international normalized ratio (INR) were slight prolonged. Chest X-ray revealed heart pushed toward the right side of chest and bowel gas appeared in the whole field of left thorax. The chest X-ray assumed left diaphragmatic hernia (Figure 3).

He was diagnosed with severe asphyxia, respiratory distress, and CDH. He got antibiotics, fasting.
and decompressive enteric tube was inserted by mouth. Reddish gastric juices was came out from the enteric tube. He admitted to NICU and started on pressure-controlled ventilation. Blood gas analysis 2 hour after intubated showed severe acidosis. He underwent urgent hernia repair at 2 days old that revealed a left posterolateral diaphragmatic
defect with small intestines, stomach and spleen found in the hernial sac (Figure 4). He definitely diagnosed with left Bochdalek type diaphragmatic hernia.

Postoperative chest X-ray showed left pneumothorax, minimal left pleural effusion differential diagnosis with hematothorax. A day after surgery, he got jaundice and planned for phototherapy with worsening of septic marker results. Four days after surgery, he experienced temperature instability and shock with oxygen desaturation many times a day. Fluid resuscitation was given and dobutamine was added as an inotropic support. The ventilation mode was changed to high-frequency oscillation ventilation (HFOV). He died 5 days after surgery because of severe sepsis.

Third case
MWB, a seven-hour-old baby, was referred to Sanglah Hospital because of respiratory distress after birth. He was born through cesarean section on the 41st week of gestation because of breech position. He was grunting, cyanotic around his mouth, and tachypnea. Apgar scores 4, 5, and 7 at 1, 5, and 10 minutes respectively.

Tachycardia accompanied with decreased of respiratory sounds on the left side of thorax. The chest wall was asymmetrical on movement. Downes score was 4. The fontanels were tense. The abdomen was excavated. The bowel sound was not heard. The liver and spleen were not palpable.

Complete blood count and CRP were normal with slight increase of IT ratio. Chest X-ray showed multiple radiolucent bubbles in the left hemithorax with suspicion of left CDH differential diagnosis with cysitic lung disease (Figure 5). He also underwent head computed tomography (CT) scan that revealed: minimal subdural hemorrhage in right occipital and minimal subarachnoid hemorrhage in posterior subfalcine.

He was fasting and decompressive enteric tube was inserted by mouth. He admitted to NICU and started on HFOV. Blood gas analysis showed respiratory acidosis. According to neurologic surgeon, no specific treatment was needed for the intracranial hemorrhage. He got hernia repair at 9 days old that revealed a left posterolateral diaphragmatic defect with organs included within the hernial sac were duodenum, jejunum and ileum and definitely diagnosed with left bochdalek type diaphragmatic hernia. After surgery, he did chest X-ray that assumed left pneumothorax (Figure 6).

Two days after surgery he got jaundice and the septic marker worsen. The antibiotic was changed. Five days after surgery, ventilation support weaned into continuous positive airway pressure setting for two days and continued to wean. He discharged from hospital after the antibiotic was stopped.

DISCUSSION
The diaphragm develops during weeks 4-12 of embryogenesis. It is composed of four components: the transverse septum, pleuroperitoneal folds, esophageal mesentery, and muscular body wall. A defect in fusion of the transverse septum to the lateral body wall leads to an anterior (Morgagni) hernia. Morgagni hernia constitute fewer than 10% of CDH. A posterior (Bochdalek) hernia represents a developmental defect of the pleuroperitoneal folds or failure of fusion of the folds and transverse septum with the intercostal muscles. Bochdalek hernia constitute 90% of CDH and more common on the left side (Figure 7 and Figure 8).

Neonates with symptomatic CDH, usually present with respiratory distress and cyanosis in the first few minutes to hours after birth with scaphoid abdomen, barrel-shaped abdomen, increased work of breathing, decreased aeration over the ipsilateral chest, with heart tones shifted to the contralateral side. Bowel sounds may be heard in the chest. Following delivery, swallowed air leads to intestinal distention that worsens lung compression. In severe cases, APGAR score at 1 and 5 minutes are low.

Using ultrasonography, most cases are identified prenatally. The characteristic finding of left-sided CDH is detection of the fluid-filled stomach within the lower thorax. Right-sided CDH is more difficult to diagnose antenatally because the herniated viscera consists predominantly of the right lobe of the liver, which has a similar echogenicity to the fetal lung. The lung-to-head ratio (LHR) is the most used prenatal predictor of survival in fetuses who have CDH. Furthermore, LHR may also dependent on the gestational age at measurement and may be less reliable in mid-gestation. In these cases, all were diagnosed postnatally because of respiratory distress with later suspicion to CDH.

A chest radiograph should be performed for the investigation of CDH. Chest X-ray had 100% sensitivity and 100% specificity to diagnose CDH. This will demonstrate an opacified hemithorax with a contralateral shift of the mediastinum. Bowel gas may be seen in the chest.

Echocardiography should be undertaken to exclude congenital cardiac lesions in infants with CDH and in any infant in whom pulmonary hypertension is suspected. It is important to determine right ventricular function, as the function of the right ventricle under increased afterload is an important determinant of illness severity. Early resolution of persistence of
pulmonary hypertension in 2 weeks, is an indicator of mild disease, with almost universally good outcomes. In these cases, one patient underwent echocardiography.

Immediate postnatal endotracheal intubation and mechanical ventilation can avoid the risk of intestinal distention. Bag-valve-mask ventilation should be avoided. An adequate-sized nasogastric tube should be placed and connected to continuous suction to allow intestinal compression. In the NICU, continuous pre- and post-ductal pulse oximetry is used to assess the degree of PPHN via right-to-left shunting at the level of the ductus arteriosus. In these cases, all of the patients had already intubated soon after delivery and also had decompressive nasogastric tube placement.

Surgical repair of the diaphragmatic defect should be performed after clinical stabilization, defined as mean arterial blood pressure normal for gestation, preductal saturation levels of 85-95% on FiO2 below 50%, lactate below 3 mmol/l, urine output more than 1 ml/kg/hour. Surgical repair can be performed while the patient is on ECMO. Repair via laparotomy or thoracotomy is the traditional treatment for patients with CDH. Since the first report by Silen et al (cited from 14), minimally invasive surgery (MIS) techniques (laparoscopic and thoracoscopic) have been considered as alternative approaches for CDH repair. Although the evident was insufficient, endoscopic surgery was clearly associated with more recurrence than was open surgery. Therefore, endoscopic surgery should not be the routine treatment for every neonate. In these cases, all patients underwent open surgery. None of them got ECMO because of limitation in our facility.

High-frequency oscillatory ventilation has also been utilized in the perinatal management of CDH both as a “rescue therapy” prior to ECMO and as a primary ventilatory modality in an attempt to reduce pulmonary barotrauma. Snoek et al found that there was no statistically significant difference in the combined outcome of mortality or bronchopulmonary dysplasia between HFOV and conventional ventilation groups in prenatally diagnosed CDH infants. In these cases, the second and the third cases treated with HFOV, and the first case was treated with conventional mechanical ventilation.

The leading cause of CDH related morbidity and mortality is respiratory failure resulting from pulmonary hypoplasia and pulmonary hypertension. Pneumothorax or chylothorax is the most common early post-operative complication. Diagnosis of a pneumothorax on a chest X-ray should be made with caution. The lung, on the side of a repaired CDH, usually become hypoplasia and will not fill the hemithorax thereby giving the false impression of a pneumothorax. Post-operative effusions are common, e.g. chylothorax (28% in some series), but no routine chest tube placement postoperatively. The majority of effusions are small. Overall recurrence rates are approximately 15% in the first two years of life. Risk factors for recurrence include large defect size and the need for a patch. In these cases, the first and second cases had pleural effusion and pneumothorax, and the third case had pneumothorax only. None of the cases had postoperative chest tube placement. The second case was died five days and the other two cases were alive. A careful monitoring of recurrence was commenced for the survivors.

SUMMARY

Three cases of CDH were reported. The diagnosis was established based on clinical manifestation, laboratory investigation, and intraoperative. One case died. All of the cases suffered from complications such as pleural effusion and pneumothorax but no one need additional chest tube placement. The survivor prognosis were good because of better preoperative conditions.

REFERENCES


