Mediscope The Journal of GMC

ORIGINAL ARTICLE

Study on management of congenital diaphragmatic hernia

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Abstract

Congenital diaphragmatic hernia (CDH) is a defect in the dome of diaphragm, more often in left and posterior-lateral that permits the herniation of abdominal contents into the thorax. Treatment requires stabilization prior to surgical correction. The best hospital series report 80-100% survival. The objective of the study was to present the experience regarding management of selected respiratory stable cases of CDH in non intensive care setup. Retrospective case series analysis was conducted on total 17 stable acyanotic patients with or without oxygen support and left sided defect were planned for surgical correction. Surgery was done per abdominally through left subcostal incision. In postoperative ward, patients received oxygen with nasal cannula and assisted ventilation with artificial manual breathing unit (AMBU) bag through ETT (endotracheal tube) if required. Patient's vital parameters; pulse, respiration, oxygenation (SpO2) and hydration were monitored throughout postoperative period. Oral feeding was started after bowel movement on 2nd or 3rd postoperative day. Plain X-ray of the thorax and abdomen was repeated on 4th or 5th postoperative day to asses lung expansion. Postoperative follow up was given at one week and one month after discharge. The age of the patients ranged from 2 days to 2 year 6 months and the mean (SD) age and body weight was 1.2 (0.6) and 5.0 (1.2), respectively. The male/female and vaginal/cesarean delivery ratios were 12:5 and 10:7, respectively. Associated congenital anomalies found were 3 (17.7%): 1 (5.9%) cleft lip and palate, 1 (5.9%) undescended testes and 1 (5.9%) hypospadias. Respiratory distress was found in 15 (88.2%) patients and 2 (11.8%) patients with recurrent abdominal distension and vomiting. One baby needed assisted ventilation with endotracheal tube and AMBU bag for 24 hours postoperatively. One case with pneumothorax required chest drain for 5 days. All other patients had good lung expansion, correction of mediastinal shifting and no evidence of any pleural effusion. All babies tolerated feeding well postoperatively after bowel movement. Survival rate was 100%. The higher survival rate among the more mature babies suggests natural selection of those with minimal respiratory impairment. In our short series survival was 100% where surgical correction was made on selective 17 cases of left sided CDH in a non intensive care setup.

Key words: Congenital diaphragmatic hernia, perioperative management.

Introduction

Congenital diaphragmatic hernia (CDH) consists of a posterior-lateral defect of the diaphragm, generally located on the left side that allows passage of the abdominal viscera into the thorax. CDH was described many years ago but survival after repair was not achieved until the 20th century.[1,2]

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Pioneers of pediatric surgery reported amazingly low mortalities until the actual severity of the condition surfaced when abortions, stillbirths and pre-hospital deaths were considered, adding a "hidden mortality" to operative and postoperative demises.[3,4] The pathophysiology of lung insufficiency and persistent pulmonary hypertension that threaten survival are currently better understood. CDH is a rare condition that occurs in <1-5:10000 births.[5] It seems to be slightly more frequent in males and less frequent in blacks and left-sided CDH is more common than right-sided, with a ratio of 6:1, bilateral lesions are reported, but they are invariably fatal.[6,7]

The pathogenesis of CDH is still unclear. Two theories exist both supported by animal experimental data. One describes CDH originating as a hole in the diaphragm which causes the pulmonary sequelae. The second theory suggests CDH results from an abnormal mesenchymal plate.[8,9]

The live born incidence of co-existent abnormalities are 40-50%.[10,11] Cardiac defects occur along with trisomies, neural tube defects, renal anomalies and duodenal atresia.[10-14] Midline defects, genitourinary problems and syndromes such as Fryn's and Pentology of Cantrell have also been reported.[15,16]

The severity of the condition varies widely, the degree of pulmonary hypoplasia and pulmonary hypertension largely determining outcome. Respiratory and cardiovascular functions are severely compromised at birth and this, together with the frequently associated malformations, cause considerable mortality and morbidity. The symptoms of insufficient gas exchange are associated with those of persistent pulmonary hypertension caused by arteriolar constriction and closure of the pulmonary arterial bed.[17,18] In some cases without neonatal symptoms, CDH may manifest itself at any age by mild respiratory distress or it can even be an unexpected finding during a medical check-up for other reasons.[19] In these cases, a hernial sac is more often present.[20]

The classic presenting picture of the more common left sided CDH is of a newborn with severe respiratory distress and a scaphoid abdomen with breath sounds reduced on the left and the heart sounds best heard on the right side of the chest. About 5% of CDH are present at less than 24 hours of age with mild tachypnoea or later still with failure to thrive, recurrent chest infections, pleural effusions or as an incidental finding on chest X-ray. Surgical repair of CDH used to be in the past a life-saving emergency. It is presently accepted that it should be undertaken only after cardio-respiratory functions are stable. Repair is usually achieved via an abdominal incision with gentle reduction of the abdominal viscera from the thorax. The diaphragmatic defect is either closed by primary repair or in the case of a large defect, using a prosthetic patch through standard open procedure or by minimal invasive surgery.[21]

In this study we present our experience regarding management of selected stable cases with left sided CDH in non intensive care setup.

Materials and Method

This retrospective case series analysis was done in Khulna Shishu Hospital and a private clinic over a period of 7 years from January 2008 to December 2014. A total of 17 patients with CDH were referred from pediatric practitioners to surgical unit. All patients got resuscitation after admission, nasogastric suction and prophylactic broad-spectrum antibiotics preoperatively, and blood was ready for transfusion prior to surgery. All clinically suspected cases were diagnosed preoperatively by abdominal radiology. Only stable patients who are acyanotic with or without traditional nasal tube oxygen inhalation and left-sided defect was planned for surgical correction. The hernia was explored per abdominally through left subcostal incision after adequate counseling. Diagnosis was confirmed, defect was identified and isolated. All the herniated abdominal contents were reduced from thoracic to abdominal cavity. Diaphragmatic defects were closed by interrupted prolene stitches.

No water-seal drainage or abdominal stretching was done in any case. In postoperative ward patients received oxygen with nasal cannula and assisted ventilation with artificial manual breathing unit (AMBU) bag if required. Patient's vital parameters; pulse, respiration, oxygenation (SpO_2) and hydration were monitored throughout postoperative period. Plain X-ray thorax was done just after operation to diagnose pneumothorax and managed accordingly. Oral feeding was started after bowel movement on 2nd or 3rd postoperative day. Plain X-ray of the thorax and abdomen was repeated on 4th or 5th postoperative day to assess lung expansion. Postoperative follow up was given at one week and one month after discharge. Data were collected from patient's hospital records and analyzed for age, sex, clinical features, diagnosis, surgical procedure performed, complications and their outcome. Results were reported using descriptive statistics and expressed as mean (SD) or percentage (%) where appropriate.

Results

Patients demographic data are shown in Table 1. The age of the patients ranged from 2 days to 2 year 6 months and the mean (SD) age and body weight was 1.2 (0.6) and 5.0 (1.2), respectively. The male/female and vaginal/cesarean delivery ratios were 12:5 and 10:7, respectively. All CDH were posterior-lateral (Bochdalek's type) on left side and complete except one where the defect was covered by a thin membrane. Associated congenital anomalies are shown in Table 2. Associated congenital anomalies found were 3 (17.7%): 1 (5.9%) cleft lip and palate, 1 (5.9%) undescended testes and 1 (5.9%) hypospadias. Presenting symptoms in patients are shown in Table 3. Respiratory distress was found in 15 (88.2%) patients and 2 (11.8%) patients with recurrent abdominal distension and vomiting. Postoperatively oxygen was given to all patients through nasal catheter for 24 hours to 72 hours. After a transient attack of cyanosis, one baby needed assisted ventilation with endotracheal tube and AMBU bag for 24 hours postoperatively. Postoperative X-ray showed pneumothorax in one case who required chest drain for 5 days. All other patients had good lung expansion, correction of mediastinal shifting and no evidence of any pleural effusion. All babies tolerated feeding well postoperatively after bowel movement. All the cases of CDH survived. So, survival rate was 100%. No wound complication was reported. During follow up weight gaining was smooth and uneventful.

Table 1. Demographic data, n = 17

Variables	Values
Age, years	1.2±0.6
Body weight, kg	5.0±1.2
Male/Female	12/5
Vaginal/cesarean delivery	10/7
Defect site, left/right	17/0

Table 2. Associated congenital anomalies

Congenital anomalies	Number	%
Cleft lip and palate	1	5.9
Undescended testes	1	5.9
Hypospadias	1	5.9
Total	3	17.7

Table 3. Presenting symptoms in patients

Presenting symptoms	Number	%
Respiratory distress	15	88.2
Abdominal distension and vomiting	2	11.8

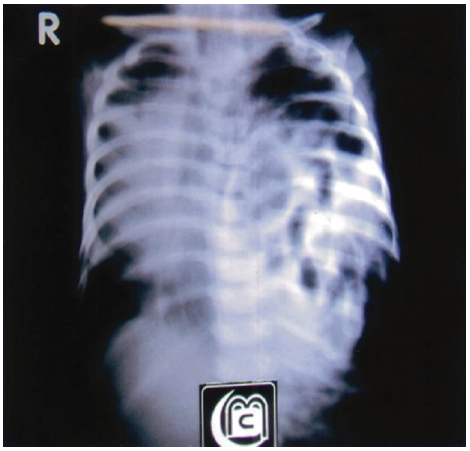

Fig. 1. Plain X-ray of the abdomen and chest showing intrathoracic intestine and mediastinal shifting.

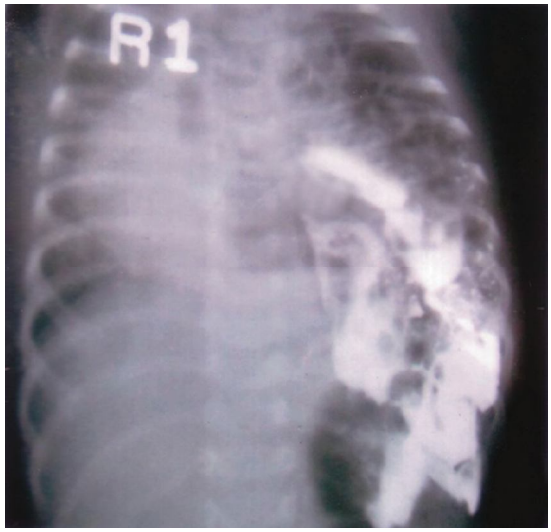

Fig. 2. Contrast gastrointestinal tract X-ray showing intrathoracic intestine.

Discussion

The perioperative management of CDH had shown many advances over the recent years. During 1980s, CDH was considered as an emergency condition requiring early surgical repair, but this was associated with more deterioration of respiratory compliance after surgical repair.[22] Since early 1990s, the management strategies have been changed depending on emergent treatment of disturbed physiology rather than the disturbed anatomy. This change included preoperative stabilization with delayed surgical repair.[23] Medical management for stabilization of patient include inhaled nitric oxide, high frequency oscillatory ventilation, and extracorporeal membrane oxygenation (ECMO) and use of exogenous surfactant.[24-28] Most important preoperative determinant is respiratory stability. The baby should be acyanotic with or without assisted ventilation. CDH is associated with a clinical spectrum of respiratory problems. The stage of pulmonary development at which visceral herniation occurs can explain the clinical spectrum of respiratory problems preoperatively in this disease.[29-31] Predictors of a worse prognosis include earlier presentation and severity of the disease.

A plain X-ray of the thorax and abdomen may present the position of the herniated viscera. Blood gases and pH status reflect the efficiency of gas exchange and other derived indexes are helpful in refining the assessment.[32,33] Ultrasonography of the heart is necessary for ruling-out associated malformations, for measuring the right-to-left shunt and for estimating the severity of pulmonary hypertension.[34-36] In this study diagnosis was confirmed in all cases by plain X-ray abdomen and thorax (Fig. 1). Atypical presentation may create diagnostic confusion and needs special investigation. Late presenting case in this study was also diagnosed by upper gastrointestinal tract contrast series (Fig. 2). All of the cases were Bochdalek's type because we were selectively dealing with left sided CDH. Right sided CDH repair is technically difficult due to presence of liver under right dome. The diaphragmatic defect may be covered by membrane in 10-15% cases. A case of CDH with membrane was also detected. The choice of peroperative surgical approach either thoracic or abdominal depends on surgeon but per-abdominal approach is preferable. In the present study, approach was per-abdominal through left subcostal incision. The diaphragmatic defect may be closed directly or by prosthesis, and in this study, defect was directly by interrupted prolene suture.

The incidence of associated congenital anomalies in infants with CDH ranges from rare to as high as 56%.[37] Cardiac anomalies are the most common, followed by genitourinary, gastrointestinal, central nervous system, skeletal and chromosomal abnormalities. We found 3 (17.7%) cases where anomaly was cleft lip and palate, undescended testes and hypospadias. Due to limitation of advanced facilities, selective cases without major anomalies were dealt with in this study. Studies showed that the presence of major congenital heart disease, such as hypoplastic left heart syndrome in infants with CDH, is associated with a very high mortality.[37,38]

Post operatively assisted respiration should be provided according to the need. These may vary from oxygen inhalation to artificial ventilation to ECMO. Survival rate in this study was 100% and only one baby needed overnight artificial respiration with ETT and AMBU after a transient attack of cyanosis. Oxygen inhalation with traditional nasal tube was given for 24 to 48 hours post operatively in all other cases. The use of nasal CPAP (5-8 cm H_2O) has significantly reduced the

need for long-term ventilation in many babies with CDH. The increasing survival of 'borderline babies' has seen in more babies needing long term CPAP and oxygen.[39]

All babies should have X-ray of the chest and abdomen (anterior-posterior and lateral view) immediate postoperative period. Postoperative X-ray showed one case of pneumothorax required chest drain for 5 days in this study. Tolerating enteral feeding can be problematic. Gastro-oesophageal reflux is frequently present and often treated with thickened feeds and proton pump inhibitors (both of uncertain value and in the case of thickening agents it is potentially harmful). Continuous feeds (24-30 Kcal) can be a useful strategy to ensure adequate caloric intake. Infant positioning, prone with slight head up, can also be helpful. No other complication in postoperative period was found.

Conclusion

CDH has a range of severity which depends on the stage of airway development at which herniation occurs. The higher survival rate among the more mature babies suggests natural selection of those with minimal respiratory impairment. It is presently accepted that it should be undertaken only after cardio-respiratory functions are stable. Good prognosis is expected after repair of the defect on a stable patient even in non intensive care setup.

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Suggestion for citation of the above:
Ali MB, Saha AK, Hossain SM, Ahmed SFU, Maruf AA. Study on management of congenital diaphragmatic hernia. Mediscope 2016;3(1):16-21.