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Case Report

Complicated Case of Congenital Diaphragmatic Hernia and Effects of Mechanical Ventilation on the Survival and Management Options

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Abstract

Congenital diaphragmatic hernia (CDH) remains one of the most challenging conditions to treat within the pediatric surgical and medical communities. Mortality remains high inspite of modern treatment modalities including ECMO, mechanical ventilation and surfactant therapy. Lung hypoplasia, dysmaturity and persistent pulmonary hypertension of the newborn adds to the complexity of the problem. Early versus late presentation has different outcome in terms of morbidity and mortality. Availability of mechanical ventilation has changed the prognosis of congenital diaphragmatic hernia (CDH) patients significantly. CDH patients on ventilator have limited options for transport of these sick neonates to Operation Theater and better medical facilities. Management strategies in a case of ventilator dependent neonate with CDH are described and morbidity and mortality associated with this condition is discussed.

Keywords: *CDH (Congenital Diaphragmatic Hernia); Ventilation; Management Options*

Introduction

Congenital diaphragmatic hernia (CDH) is a fascinating and challenging birth defect that usually results in tragic loss for the child and family. Frequently diagnosed ultrasonographically at 20 weeks gestation, the diagnosis can overwhelm affected parents who never knew the condition existed and seldom can understand it adequately. Affected newborns face myriad potential issues including pulmonary insufficiency, pulmonary hypertension, hemodynamic instability, associated cardiac defects, eventual feeding difficulties and significant risk of dying or of surviving with substantial morbidity. Well-meaning but often inadequately informed physicians and caregivers too frequently view the situation as either hopeless or requiring dramatic but completely unproven prenatal interventions. However, an improvement in understanding of CDH physiology has led to dramatic improvements in CDH survival in many centers. Some however argue that improvement in survival is nil, but this is, in large part due to continued significant rates of pregnancy termination with this diagnosis in developed countries and rapid adoption of adjuvant CDH treatments and slower penetration of fundamental changes in CDH therapy.

Case Report

A three days old male infant born to G3P3A0D0 mother by Spontaneous vaginal delivery after term pregnancy was admitted to neonatal intensive care unit of a private hospital for respiratory difficulty just after birth. Initially baby was managed in incubator with O_2 by

head box and on aggravation of respiratory distress and development of cyanosis (blue discoloration) of face and peripheries 'baby was intubated and put on mechanical ventilation. As baby stabilized and cyanosis disappeared, detailed physical examination revealed bulging chest and scaphoid abdomen. On suspicion of a potential abnormality initial chest and abdominal radiography was performed that disclosed presence of intestinal loop shadows inside left side of chest, so a diagnosis of left diaphragmatic hernia was made. A call was sent to pediatric surgery service for operative management of patient. At this time baby was ventilator dependent on SIMV mode with absent cyanosis and O_2 saturation in Rt. Radial artery of > 95% and in Rt. Leg of 90% with a difference of preductal and postductal saturation of approximately 5%. Abdomen was scaphoid with a buldging chest that revealed intestinal shadows on supine chest x-ray and kinking of NG tube in chest confirming a diagnosis of Lt. sided Congenital diaphragmatic hernia.

Because of ventilatory dependence of patient surgical repair of CDH was planned in the NICU with consent of parents and NICU consultant.

After appropriate IV line maintenance and medication, abdomen was opened with Lt. subcostal oblique incision and it was found that all of stomach, small intestine and greater part of large intestine went into chest up to splenic flexure and spleen, left kidney was residing in Left side of thoracic cavity through a left Bochdalack's defect. Left lung size was small and it could not be inflated. All intestinal contents were reduced through the defect that was repaired using interrupted 2/0 silk sutures. On trying to close the abdomen after placing intestinal contents in abdominal cavity, patient became unstable with bradycardia and low O_2 saturation, so reduced contents were placed in a silo bag (made of ordinary transfusion bag) sewed to abdominal wound margins with continuous vicryl 3/0 sutures. Post operatively patient remained stable and one unit of fresh blood was transfused. SIMV mode mechanical ventilation was continued to maintain O_2 saturation above 90%.

On 2nd post-operative day patient passed small amount of meconium and partial Total Parenteral Nutrition was started. After some 36 hours, on 2nd postoperative day greenish patches could be seen over small intestine along with small amount of greenish fluid and sludge in the silo bag. A diluted gastrograffin upper GI series was carried out that revealed distal small intestinal perforation. So, a second look laparotomy was planned and carried out in the same settings, that revealed a single small tiny perforation some 14 cm proximal to Ileocecal valve. Immediately an ileostomy was fashioned and anchored on the side wall of silo bag and abdomen was closed after saline lavage. Post-operatively SIMV mode ventilation was continued along with a diagnosis of Necrotizing enterocolitis and CDH. Later patient developed septicemia with development of sclerema. Broad spectrum antibiotics imipenem, fortum and metronidazole in appropriate dosages were continued to address persistent pulmonary hypertension of the newborn because of pulmonary hypoplasia, I.V. $MgSO_4$ was also started in the treatment from 3rd post-operative day onwards. A central line was needed to be secured that was done through umbilical vein.

As Patient was unable to maintain O_2 saturation above 90% so referral for better management to a tertiary care facility with better equipment and specialized services was planned. Patient was referred for High Frequency Oscillatory Ventilation to the Children`s Hospital Lahore. In the meantime patient developed worsening sclerema and DIC for which antibiotics were continued along with fresh frozen plasma. But patient could not recover and succumbed to septicemia and DIC on 4th postoperative day.

Discussion

Epidemiology and etiology

Although cause of CDH is not known, it has been associated with abnormalities on every human chromosome and overt chromosomal abnormalities are seen in 10 - 40% of affected patients reviewed by Witter's and Howe. When CDH occurs with problems of chromosome number as in Turner syndrome (monosomy X), Down syndrome (trisomy 21), Edward syndrome (trisomy 18) and Patau syndrome (trisomy 13), CDH frequently occurs in Pallister Killian syndrome (tetrasomy 12 P). It can present syndromally based on single gene abnormality as in Densy Drash syndrome (WTl), Spondylocostal Dysostosis (DLL3) and Neonatal Marfan syndrome (FBNl). Most case as of

CDH however occur as isolated events. The risk of having a second affected child after a first non-syndromic presentation is estimated at 2%, a 50 fold increase from estimated baseline risk of 0.04% (1: 2500).

Pathophysiology

Babies born with CDH face some fundamental physiological problems. Herniation of the abdominal contents into the chest of developing fetus compresses developing lungs. With larger defects and earlier herniations having greater effects on developing lungs resulting in pulmonary hypoplasia with attendant persistent pulmonary hypertension of the newborn that affects the gas exchange what is main function of developing lungs in fetus.

Markers of severity

One parameter used to assess severity of CDH is LHR (fetal lung to head ratio) either prenatally using ultrasound or MRI, or postnatally using chest radiography. Variable results have been found using LHR from 0.62 to 1.86 with mean LHR of 1.0 to 1.4 culminating in a favorable outcome. Another valid method to estimate severity of CDH during prenatal and postnatal evaluations is to evaluate the extent of herniated abdominal contents. In Left CDH, herniation of bowel alone with stomach and liver in abdomen correlates with good lung development and adequate diaphragm for primary closure. Patients thus affected should do well with minimal mortality and need for extracorporeal membrane oxygenation (ECMO). Patients having bowel and stomach in the chest but liver in abdomen have intermediate risk and patients who have bowel, stomach spleen and liver in chest have more sever CDH and are at significant risk of morbidity/mortality and need for ECMO. These patients have insufficient native diaphragm for closure and require a large patch. Patients at highest risk are those, who has large amounts of liver in the chest and has correspondingly low LHR. Those patients having Rt. Sided CDH and have major part of liver in the chest evidenced by gallbladder on ultrasonography pose a challenge and likely will require ECMO. Another important anatomic defect correlated with prognosis is presence/absence of CDH "sac". When present, CDH pathophysiologically is less sever. It appears that sac holds abdominal contents back a bit or possibly herniation occurred later in gestation. The presence of sac at surgery correlates with less sever pulmonary hypoplasia predicting improved prognosis.

Fetal surgical intervention

Original concept is to repair CDH in the fetus and return the fetus to the womb for further lung growth and development and deliver the repaired fetus electively near term. In a study performed only 5 out of 21 repaired fetuses could survive. Several techniques were applied for hypoplastic lung of CDH to grow including tracheal ligation, occluding tracheal clips and removable intra tracheal balloons placed by fetoscopy. Open fetal tracheal ligation was associated with poor survival, so fetoscopic tracheal ligation was attempted with better results. Recurrent laryngeal nerve injury was found to be the complication of the procedure.

Postnatal care

Surfactant

Evidence is lacking regarding better prognosis after use of exogenous surfactant in CDH patients. Same holds true for development of chronic lung disease and use of ECMO.

Pulmonary hypertension

Pulmonary hypertension in CDH has two components, fixed and reactive. The fixed component is due to small pulmonary vascular bed- an effect of pulmonary hypoplasia. This fixed component is slow to resolve requiring weeks, moths and even years. The reactive component is due to changing resistance of pulmonary arterioles in CDH. This may be due to changes in pulmonary compliance, loss of

lung volume and resultant hypoxic vasoconstriction. Maintaining appropriate lung volume while minimizing risk of barotrauma is most appropriate therapy. Historically demonstration of hyperventilation and alkalosis effectively decrease pulmonary vascular resistance and can reverse the direction of shunting in neonatal pulmonary hypertension syndromes including CDH. This is a roller-coaster modal of lung failure because of recurrent ductal shunting requiring further increase in ventilator settings and development of a vicious circle.

ECMO

ECMO evolved to rescue infants from pulmonary hypertension and ventilator induced lung injury. Hyperventilation was sometimes profoundly successful in reversing ductal shunting and providing relief from desaturation but at an unacceptable cost. Initially ECMO was called upon as a rescue therapy when patient developed pulmonary hypertension and lung injury after corrective surgery being frequently performed in first 24 hours. ECMO use was found to be specially advantageous in those infants having predicted mortality of 80%, but not in patients with less sever disease.

Recently timing of ECMO supporting CDH has changed. This reveals a trend towards preoperative stabilization with ECMO rather than postoperative rescue therapy. Now venovenous ECMO is being institutionalized commonly than veno arterial ECMO as former is less associated with seizures and cerebral infarctions. The survival was comparable in both groups. Venovenous ECMO can be further converted into veno arterial ECMO as need arises. Some of the centers, however are using ECMO sparingly while having good number of surviving CDH patients.

Inhaled nitric oxide

The best hope of a powerful non-toxic agent to control pulmonary hypertension in infants who have CDH has been nitric oxide. Though treatment with nitric oxide can have rapid and sometimes a dramatic effect on oxygenation, the effect is transient. Accordingly inhaled nitric oxide has not been shown to improve survival or decrease the need for ECMO. Nitric oxide combined with High frequency oscillatory ventilation (HFOV) was found to be associated with good results in CDH patients.

Sildenafil

A specific phosphodiestrase-5 inhibitor was demonstrated to be more effective at improvements in pulmonary hypertension. Evidence is lacking regarding its use in CDH.

Gentle ventilation

The development of neonatal ventilation strategy pioneered by Wung and associates is the single most significant advance in the management of CDH. This strategy significantly limits inflation pressures allowing tolerance of hypercapnia and relative post-ductal hypoxemia and eliminates hyperventilation. Most problematic in the era of hyperventilation was the concept that pulmonary vascular resistance could normalize in the presence of hypercapnia and respiratory acidosis. Detailed ventilation analysis showed that pneumothorax rates decreased from 43% to 2% and survival improved from 50% to 89% in patients treated with strict lung protective strategy that avoided hyperventilation and limit infusion pressures to less than 25cm of water. ECMO was used to support infants who had critical deficiencies of oxygen delivery. High survival rates in CDH also have been reported by some investigators using HFOV, but others reported no improvements compared with conventional ventilators. Same concept of tolerance to hypercapnia and relative post ductal hypoxemia and elimination of hyperventilation can be applied to HFOV as well. The important conceptual breakthrough was that ventilatory support must be as gentle and non-toxic as possible to maximize CDH survival.

Surgical repair

Surgical repair is an important part of patient care and primary issue is when to repair. Pulmonary gas exchange often improves in 24 hours after birth and respiratory system compliance also improves with preoperative stabilization though there is no evidence that delayed repair is harmful. There is also no convincing evidence that such delay improves survival or decreases risk of pulmonary hypertension.

Most surgeons prefer repair through an open sub-costal approach. Patients having mild CDH have enough muscle for primary repair but those having more sever CDH especially with liver in the chest do not have enough native diaphragm to close the defect. Such patients require patch to close the defect that is a marker of severity of CDH and these patients are at increased risk of gastro esophageal reflux. As patch material does not grow with the patient, there is risk of future diaphragmatic herniation recurrence.

Outcomes

An improved understanding of CDH pathophysiology and expanded application of lung protective treatment strategies is resulting in an increased number of CDH survivors. Infants surviving CDH are at increased risk of brain injury, neurodevelopmental disability, hearing loss, feeding difficulties, gastro esophageal reflux, lung disease scoliosis, pectus excavatum and recurrence of diaphragmatic hernia. Some of these outcomes are anatomic and are unavoidable, others reflect potential toxic effects of treatment strategies and might be avoided or eliminated in future. Though most infants having CDH survive without major neurological sequelae, newborns with sever CDH are at an increased risk of periods of hypoxemia, poor perfusion and need for ECMO. Such infants are at high risk for hypoxic-ischemic brain injury and other neurological effects. A review of 31 CDH patients who required ECMO showed that 35% had CNS abnormalities on CT scan manifested primarily as enlarged ventricles, focal and diffuse brain atrophy and intra cranial hemorrhage. At 2 years these patients have mild cognitive and physical delay. In three separate series 44 - 55% of CDH survivors required hearing amplification due to hearing loss [1-15].

Conclusion

Congenital diaphragmatic hernia is not an uncommon disorder in Pakistan and early presenters usually present with respiratory distress and mechanical ventilation is required for management and stabilization of these sick neonates. Pediatric surgeons are required to accomplish and facilitate surgical management of these patients.

Management options for patients as our index case are limited because of patient dependence on mechanical ventilator and transportation to operation theater is either not possible as equipment and devices are fixed or is difficult to manage. The concept of carrying out surgical procedures in NICU (Neonatal intensive care unit) is a novel approach to manage these situations as patient is already intubated and maintained on mechanical ventilation and need not to be transferred to Operation Theater.

As disinfection and aseptic techniques are practiced and maintained in NICU, the environment provides an alternative to (though not ideal) operation theater. Moreover, NICU like operation theater has restricted entry of personnel. The surgical team, anesthesia availability and necessary equipment can be arranged in NICU and surgery can successfully be performed there.

Our index case had two more unique features, need of abdominal space (silo like) to avoid abdominal compartment syndrome as preoperatively patient developed O2 desaturation and tachycardia and development of NEC (necrotizing enterocolitis) postoperatively. Both issues were managed successfully, first by using ordinary blood bag in place of silo preoperatively to manage raised intra-abdominal pressure causing tachycardia and desaturation and second by construction of ileostomy on the side wall of bag as patient developed NEC postoperatively in second successful surgical procedure in NICU.

The last and most lethal issue to be tackled is sepsis consequent upon immune compromised status of the patient, poor nutrition, defects in maintenance of aseptic environment and technique to avoid infection related developments. We managed these issues with maintenance of asepsis in NICU and introduction of partial total parenteral nutrition and institution of antibiotics to fight killer organisms responsible for infection and its consequences.

Our index case developed sepsis and NEC postoperatively and died on 4th postoperative day.

We recommend that ideal place for surgery of neonates is operation theater, but as the need arises it can successfully be performed in NICU as it provides suitable alternative for the compelling situation provided asepsis can be maintained and surgical and anesthesia personnel along with necessary equipment and devices can be arranged and managed in NICU.

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