Congenital Diaphragmatic Hernia: Antenatal Diagnosis and Successful Repair In Preterm Neonate Case Report

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ABSTRACT

Most of the congenital diaphragmatic hernias observed in the left hemi diaphragm. The pathogenesis of congenital diaphragmatic hernias is not completely understood; but abnormal development of the diaphragm at 6-10 weeks' supposed to be the cause in most of the cases. Although congenital diaphragmatic hernias usually occurs sporadically, environmental exposures have been implicated. Improvement of the antenatal imaging techniques has allowed early diagnosis and evaluation of the associated anomalies. In antenatally diagnosed congenital diaphragmatic hernias cases, multidisciplinary team management and delivery at tertiary centers with proper facilities may be provided to optimize the outcome.

A 39-year-old woman, with the history of one previous cesarean section, was admitted to the hospital at 25 weeks' gestation, because of placenta previa and her antenatal ultrasound showed fetal congenital diaphragmatic hernias with polyhydramnios. During hospitalization, she was monitored for hemoglobin levels, consumptive coagulopathy, and fetal well-being. Betamethasone was given to accelerate the fetal lung maturity, and magnesium sulphate for fetal neuro-protection. Due to an attack of heavy antepartum hemorrhage, delivery occurred at the gestational age of 28 weeks + 6 days. The delivered female newborn was admitted to the neonatal intensive care unit because of respiratory distress and prematurity, and received surfactant, dobutamine and intravenous antibiotics. Chest examination of the studied neonate showed diminished air entry on the left side, and chest X-ray showed stomach shadow in the left hemi thorax. After exclusion of congenital heart diseases and intracranial hemorrhage, the baby was successfully operated on the postpartum 7th day, and discharged from the neonatal intensive care unit 60 days after the operation.

Two months after discharge from the neonatal intensive care unit, examination of the studied neonate was successfully operated on the postpartum 7th day showed normal growth parameters and appropriate motor and sensory development for her age.

Antenatal diagnosis of congenital diaphragmatic hernias allows multidisciplinary team management and delivery at tertiary center with the proper facilities to optimize the outcome.

Keywords: Congenital diaphragmatic hernia, Prenatal diagnosis, Repair, Premature

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Introduction

Congenital diaphragmatic hernia (CDH) is a rare disorder which involves the left hemi diaphragm in 75% of the cases; 15% are right-sided and 10% bilateral (1). The pathogenesis is not completely understood; but abnormal development of the diaphragm at 6-10 weeks' gestation supposed to be the cause in most of the cases. Although most cases occur sporadically, environmental exposures have been implicated, including smoking, alcohol, vitamin A deficiency, thalidomide, and anticonvulsants (2-5).

With the improvement of the antenatal imaging techniques, early diagnosis of CDH has been possible as well as the evaluation of the associated anomalies and syndromes. Associated anomalies, most commonly of renal, gastrointestinal, cardiovascular and central nervous systems, found in 40%-60% of live-born infants with CDH (6,7).

Antenatal diagnosis of CDH allows multidisciplinary team management involving an obstetrician, neonatologist, and pe-

diatric surgeon, and delivery at tertiary center with the proper facilities to optimize the outcome (8,9).

This report represents a case of CDH diagnosed antenatally, to highlight the importance of early diagnosis, and the good outcome when managed through multidisciplinary team approach in a tertiary center.

Case Report

A 39-year-old woman, with the history of one previous cesarean section, was admitted to the hospital at 25 weeks' gestation, after a mild attack of antepartum hemorrhage due to placenta previa. Her antenatal ultrasound showed fetal CDH (Bochdalek hernia) and polyhydramnios, but no other associated anomalies. During hospitalization, she was monitored for hemoglobin levels, consumptive coagulopathy via platelet count, prothrombin, bleeding and clotting times, and fetal well-being and informed consent was obtained. Hemoglobin measurements as well as tests for consumptive coagulopathy were performed especially during active bleeding. Fetal well-being was evaluated by fetal movement counts, daily fetal heart rate (FHR) recordings, and weekly trans-abdominal ultrasonographic examinations to measure amniotic fluid volume, fetal growth and umbilical artery Doppler indices (10,11). Betamethasone was given to accelerate the fetal lung maturity, and magnesium sulphate for fetal neuro-protection the day after admission (12,13).

Due to an attack of heavy antepartum hemorrhage, delivery occurred at the gestational age of 28 weeks + 6 days. The delivered female newborn was 960 grams, with APGAR scores of 5, 6, and 8, at 1, 5 and 10 minutes respectively. She was admitted to the neonatal intensive care unit (NICU) because of respiratory distress and prematurity, placed on continuous mandatory ventilation (CMV) and received surfactant after the delivery, and monitored by oxygen saturation and blood gases. Intravenous (IV) fluids given were monitored by the input/output fluid chart. Additionally, she received dobutamine, and IV antibiotics according to the results of the cultures performed as a part of neonatal sepsis screening protocol.

On admission to NICU, chest examination of the studied neonate has revealed diminished air entry on the left side, with displacement of the heart to the right side; and chest X-ray has shown the stomach shadow in the left hemi thorax, pushing the heart to the right side of the chest (Figure 1).

Echocardiographic examination performed two days after NICU admission, revealed a small atrial septal defect (ASD) and patent ductus arteriosus (PDA), which were considered normal for a premature baby, and the cranial ultrasound showed no intracranial hemorrhage (ICH).

The studied neonate was successfully operated on the postpartum 7th day when her cardio-pulmonary functions were stable, and the diaphragmatic defect identified (Figure 2) was repaired after reduction of the contents to the abdominal cavity using interrupted stitches (9,14). (Figure 3).

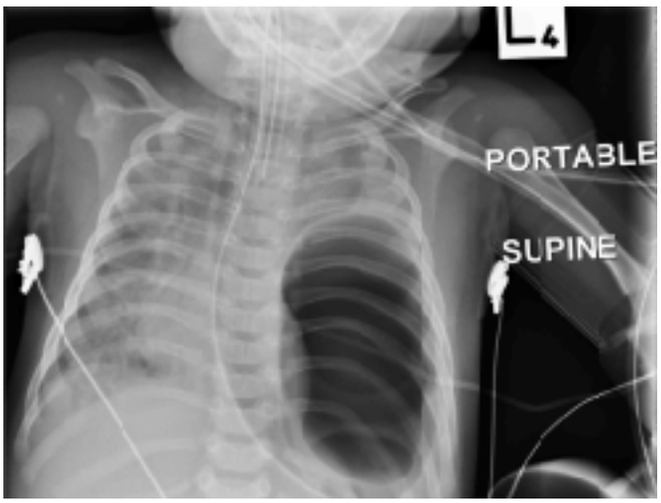

Figure 1: Chest X ray of the studied neonate before surgery showing the stomach in the chest, pushing the heart to the right side.

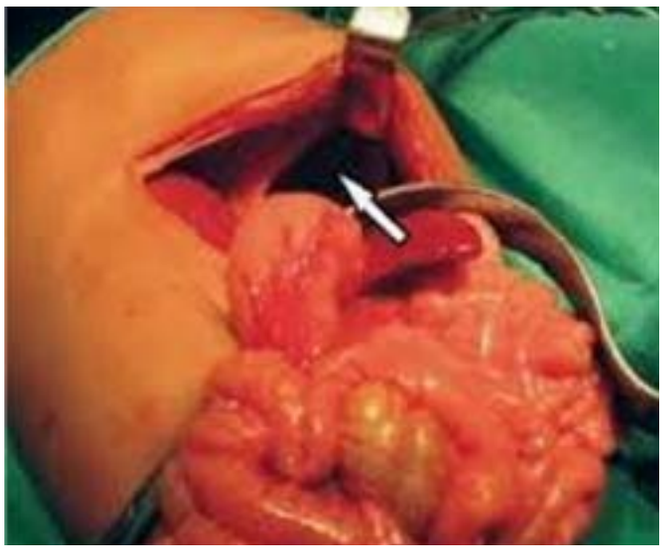

Figure 2: Intra-operative image shows the congenital diaphragmatic hernia on the left side after reduction of its contents

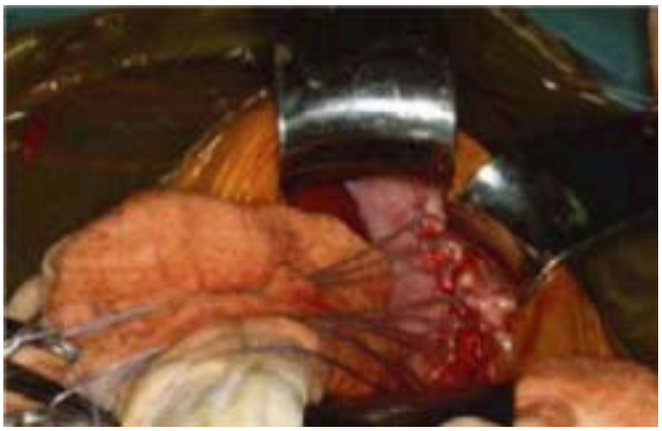

Figure 3: Intra-operative image shows the repaired congenital diaphragmatic hernia on the left side, using interrupted stitches

On the postoperative 3rd day, the studied neonate shifted to synchronized intermittent-mandatory ventilation (SIMV), and dobutamine infusion was completely stopped. On the 12th day, she shifted to pressure support ventilation (PSV), oral feeding started through orogastric tube, and the rates of IV fluids and total parenteral nutrition (TPN) were reduced. On the 21st day, she was extubated and placed on nasal continuous positive airway pressure (CPAP).

Forty days after the operation, her weight increased to 1421 grams, oral feeding rate was increased, IV fluids and TPN were stopped, and nasal CPAP was shifted to heating, ventilation and air conditioning (HVNC).

The small ASD and PDA were closed spontaneously, and she was 2120 grams and on complete oral feeding and room air when discharged from the NICU 60 days after the operation for out-patient follow-up (Figure 4).

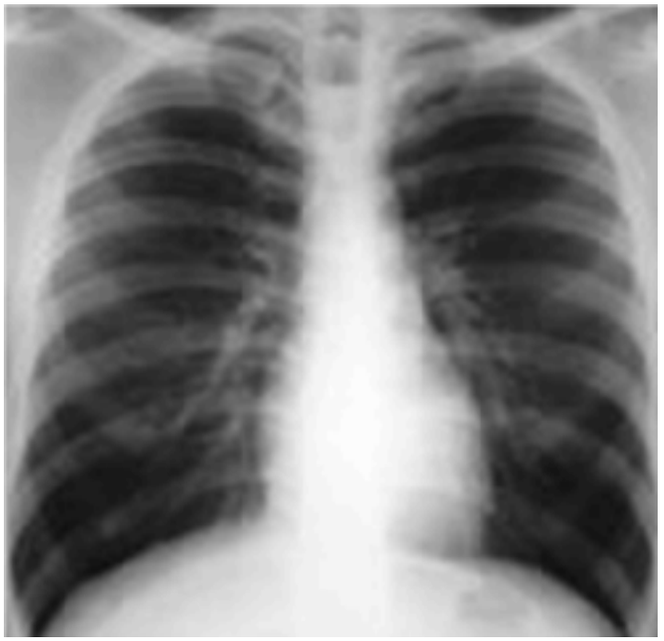

Figure 4: Chest X-ray of studied neonate before discharge from the hospital

Two months after discharge from the NICU, examination of the studied neonate at the out-patient department showed normal growth parameters and appropriate motor and sensory development for her age.

This case report was approved by the hospital institute ethical committee and informed consent was taken from the parents of the baby.

Discussion

The incidence of the CDH ranges between 1:2000 and 1:5000 live births (15). The mortality rate in neonates with CDH remains high with an overall survival rate of around 60% (15,16).

Congenital diaphragmatic hernia is one of the major challenges of perinatal medicine and surgery, and antenatal diagnosis allows multidisciplinary team management to optimize the outcome (9).

Infants with CDH usually present with respiratory distress due to pulmonary hypoplasia, which require endotracheal intubation, and pressor support (17).

Surgical management of the CDH involves reduction of the herniated abdominal content, and repair of the diaphragmatic defect. Researchers agree in that, repair of the diaphragmatic defect should be delayed until the cardio-pulmonary functions of the newborn are stable (9,14).

Traditionally, the repair performed via an open surgical approach, using either a patch repair or a muscle flap; the minimally invasive techniques may be associated with a higher rate of hernia recurrence (18). In the current case report, CDH was diagnosed antenatally at 25 weeks' gestation and was managed by a multidisciplinary team. Delivery was planned in a tertiary center with intensive care facilities. Immediately after the delivery, the studied neonate was admitted to the NICU because of respiratory distress, prematurity and low birthweight, as the gestational age was 28 weeks+6 days at delivery and the birthweight 960 grams; then operated successfully on the 7th postpartum day, after stabilization of cardio-pulmonary functions. She was discharged from the NICU 60 days after the operation when she was 2120 grams, and two months later out-patient examination showed normal growth parameters and appropriate motor and sensory development for her age.

Tsao et al, have emphasized that the outcomes in preterm infants with CDH are worse compared to term infants, the survival rate being around 50% in preterm infants after surgical correction (19).

A systematic review of controlled trials showed that the neonatal survival improved in infants diagnosed prenatally and delivered at a tertiary center with intensive care facility (20).

Mortality was higher in the presence of additional anomalies, iatrogenic lung injury, pulmonary hypoplasia, and pulmonary hypertension (19,20).

Tovar, has notified that, the best hospital reported survival rate was 80% in CDH, but it remained around 50% in population-based studies (9).

Taking the current case into consideration, it may be concluded that, antenatal diagnosis of CDH allows multidisciplinary team approach and delivery at tertiary center with proper facilities to optimize the outcome.

Antenatal diagnosis of CDH allows multidisciplinary team approach and delivery at tertiary center with proper facilities to optimize the outcome.

✍: *Ethical approval: The case report approved by the institute ethical committee of Ahmadi hospital and informed consent taken from the studied woman.*
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