



LEFT CONGENITAL DIAPHRAGMATIC HERNIA A rare case report

¹Dr. Ankush Maheshwary, ²Dr. Arvinder Singh, ³Dr. Karandeep Singh Bhatti
¹MBBS Medical Officer, ²MBBS MD Pediatrics Senior Resident, ³MBBS Medical Officer
¹Mrs. Khushbir Kalra Memorial Hospital, Amritsar, India

Abstract- Congenital diaphragmatic hernia (CDH) is a condition where abdominal content protrudes through a defect in the diaphragm which occurs as a result of incomplete fusion of pleuroperitoneal membrane during fetal development. CDH is normally diagnosed prenatally which has proven to improve the prognosis, but failure to do so can lead to serious cardiorespiratory implications in newborns. Early management after birth includes bowel decompression and prevention of development of pulmonary hypertension along with maintaining adequate oxygenation. Surgery is the only definitive treatment. We present this case report of a 4 hour old male child with severe respiratory distress who was referred from another facility requiring urgent care.

Keywords- Congenital diaphragmatic hernia, diagnosis, management, prognosis

I. INTRODUCTION

The incidence of congenital diaphragmatic hernia is 1 in 2500 births ^[1]. Majority of cases have an isolated diaphragmatic defect but it can be associated with cardiac, Gastrointestinal or Genitourinary anomalies with chromosomal aneuploidy such as trisomies ^[2]. Varied degree of pulmonary compression by intrathoracic bowel protrusion, causes decreased pulmonary branches, limiting multiplication of alveoli, pulmonary artery hypertrophy and decreased functional lung mass affecting ipsilateral lung more but both lungs are involved eventually. These newborns are predisposed to develop Persistent pulmonary hypertension creating an intracardiac right-to-left shunt and eventually hypoxia and acidosis ^[3]. Due to the progress made in fetal ultrasonography, in 60% of cases the diagnosis of CDH occurs prior to delivery ^[1], often associated with a history of polyhydramnios. Those that are missed present with cyanosis, grunting, tachypnea soon after birth. CDH is rarely seen in adults with a little over 100 cases reported, often diagnosed incidentally on imaging ^[3]. The prognosis of late presenting CDH patients is usually better than in those cases which were diagnosed prenatally or shortly after birth because management of these patients is different and they have a better overall quality of life ^[1].

There are three basic types of congenital diaphragmatic hernia ^[4] -

- 1) Posterolateral or bochdalek's hernia (occurring approximately at 6 weeks during fetal development)
- 2) Anterior morgagni's hernia
- 3) Hiatus hernia

II. CASE REPORT

A sick looking term neonate presents 4 hours after birth in the pediatrics emergency with significant respiratory distress requiring mechanical ventilation. He was born via normal vaginal delivery, had a birth weight of 1675 gms with apgar of 5/7 in first and fifth minute. Mother had discharge of clear fluid per vaginum for 2 days prior to delivery (likely prolonged rupture of membrane).

On physical examination the child had chest retractions and audible grunting. Chest appeared barrel shaped and abdomen was scaphoid (fig. 1), palpation and auscultation of abdomen were within normal limits. Point of maximum impulse was displaced and bowel sounds were heard on left side of chest. Neurological examination was also normal. Fontanelle were open and anus was patent. Genital examination was normal and no congenital gross malformations were present.



Figure 1: scaphoid abdomen



Figure 2: chest x-ray PA view with displaced mediastinum; trachea (white arrow); heart (red arrow); cystic lucency on depicting bowel (blue arrow)

Modified Downe's score calculated to be 4/10

Grunting- 2

Cyanosis- 0

Respiratory rate- 1

Air entry- 0

Retractions- 1

Mother (G3P3) had an uneventful 1st and 2nd trimester and no history of substance abuse or similar complaints were found in previous childbirth. Chest x-ray was performed immediately and it showed significant displacement of mediastinum to the right with bowel protrusion and left lung hypoplasia (fig. 2).

After bowel decompression, the child was placed on CPAP in PICU, adequate antibiotics and IV fluids initiated. Although no murmurs were audible but a suspicion of congenital cardiac defect was definitely ruled out after doing an echocardiography. Regular pulse oximetry for early detection of hypoxemia done and child was kept under observation. Later he was sent for pediatric surgical consultation and repair after insuring stabilization on 2nd day of hospitalization.

III. DISCUSSION

Severe respiratory failure is the major cause of neonatal death and among the causes of severe respiratory failure in newborn; CDH remains most life threatening^[5]. Defect in posterolateral location is most common (70-75%) and majority are on the left side (85%) and less frequently on the right side (13%) or bilateral (2%)^[2].

Infants with CDH require mechanical ventilation and sometimes extracorporeal membrane oxygenation (ECMO) in the neonatal period. Hernia repair is typically performed through the thoracic or abdominal route. Small diaphragmatic defects are usually repaired by primary repair with non-absorbable sutures. For large defects, prosthetic patches or tissue-engineered grafts are used to avoid causing excessive tension after repair. With the development of surgical techniques, operations of CDH occur through minimally invasive techniques such as laparoscopy or thoracoscopy^[6]. Even after successful surgery many children require long term oxygen supplementation and diuretic therapy. Major respiratory complications include tracheobronchomalacia, pneumothorax, and secondary lung infection (especially viral pneumonia) that could precipitate terminal respiratory failure even months after surgery^[1].

IV. CONCLUSION

Congenital diaphragmatic hernia if diagnosed prenatally with the use of morphological ultrasonography has a better overall prognosis. After birth the neonate should be managed in a well equipped institution, with surgery being the only definitive treatment available and early repair ensures prevention of persistent pulmonary hypertension. It is imperative for physicians to maintain low threshold while diagnosing a case on clinical grounds even with an inconclusive chest x-ray. We presented the case of a newborn baby with CDH who was referred from another institution due to lack of infrastructure, prompt mechanical ventilation and imaging ensured favorable outcome after operative repair.

REFERENCES

- [1] Topor L, P. T. (2015, Jan-Feb). *Left congenital diaphragmatic hernia -- case report*. Retrieved March 2020, from PubMed: <https://www.ncbi.nlm.nih.gov/pubmed/25800323>
- [2] Praveen Kumar Chandrasekharan, M. R. (2017). Congenital Diaphragmatic hernia – a review. *Maternal health, Neonatology and perinatology* , 3:6.
- [3] Lava JR, Hettwer GA, Reginatto CJ, et al. Congenital diaphragmatic Bochdaleck hernia: case report. *Int Arch Med*. 2012;5(1):30. Published 2012 Oct 30. doi:10.1186/1755-7682-5-30
- [4] *Pediatric Congenital Diaphragmatic Hernia*. (2014, april 25). Retrieved march 2020, from medscape: <https://emedicine.medscape.com/article/978118-overview>
- [5] Greer, J., Babiuk, R. & Thebaud, B. Etiology of Congenital Diaphragmatic Hernia: The Retinoid Hypothesis. *Pediatr Res* **53**, 726–730 (2003). <https://doi.org/10.1203/01.PDR.0000062660.12769.E6>
- [6] Gujar A, Rodrigues DD, Patil K, Tambe U, Sinha S, Bhushan A. Rare case report - congenital diaphragmatic hernia presentation in adult. *Indian J Surg*. 2013;75(Suppl 1):44–46. doi:10.1007/s12262-011-0321-7