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Right Sided Congenital Diaphragmatic Hernia

Alaoui Othmane[1,2]; Mahmoudi Abdelhalim[1,2]; Khattala Khalid[1,2]; Bouabdallah Youssef[1,2]*

[1]Department of Pediatric surgery, Hassan II University Hospital, Fez, Morocco.

[2]Sidi Mohamed Ben Abdallah University of Fez; Faculty of Medicine and Pharmacy and Dental of Fez, Morocco.

***Corresponding Author(s): Othmane Alaoui**

Department of Pediatric Surgery, Hassan II University Hospital, Morocco.

Tel: 00212676905942;

Email: othmanechirped@gmail.com

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Abbreviations: CDH: Congenital Diaphragmatic Hernia; LHR: Lung To Head Ratio; HFOV: High Frequency Oscillatory Ventilation; CT Scan: Computed Tomography Scan; ECMO: Extracorporeal Membrane Oxygenation.

Abstract

Congenital diaphragmatic hernia is a rare condition that affects approximately 1 in 2500-5000 births and has been classified as posterolateral, anterior or central.

The posterolateral defect occurs on the left side (85%), although it may involve the right side (13%) or be bilateral (2%). Equivalent survival has been reported for left-sided versus right-sided congenital diaphragmatic hernias, although the latter have been associated with a high incidence of pulmonary complications.

We report the evolution after surgical treatment of right diaphragmatic hernia in one case.

Introduction

Embryologically, the diaphragm develops between the 8th and 12th week of gestation. The septum transversum separates the thoracic cavity from the abdominal cavity, and muscle fibers migrate into this membrane. Left-sided anomalies are more common because of the late closure of this membrane. The loss of lung development may be explained by the loss of space to develop, the diagnosis of CDH can be made before birth by ultrasound in 90% of cases. Fetal ultrasound findings include polyhydramnios, intestinal loops in the chest, echogenic chest mass.

Two distinct features were used for risk stratification:

1) A low Lung-to-Head Ratio (LHR).

2) A hepatic hernia in the chest.

Initial postnatal treatment is aimed at resuscitating and stabilizing the infant in cardiopulmonary distress. Pulmonary hypertension and associated cardiac abnormalities are assessed by echocardiography. The timing of surgery is based on the clinical judgment and discretion of the surgeon.

Case report

A male neonate with a gestational age of 10 days was delivered by simple vaginal delivery, weighed 3.8 kg, was born with faint crying, dark in color, intubated and connected to a ventilator (HFO ventilation was instituted with a mean pressure of 13, FiO2 100%,. Chest radiograph **(Figure1)**, and CT scan **(Figure2)** confirmed a right congenital diaphragmatic hernia. Ultrasound revealed pulmonary hypertension. Other examinations were normal. The newborn was taken to surgery after stabilization.

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Surgery was performed by thoracotomy at the level of the 5th right intercostal space. The entire liver was intra-thoracic with the intestinal loops. The liver was twisted at 90 degrees in the thoracic cavity. The stomach and spleen were found in the abdomen. Only a thin rim of the diaphragm on the anterior aspect was present, and the posterior rim was absent **(Figure 3)**. The right lung was very small.

The liver could be lowered into the abdomen with difficulty, and the intestinal loops were reduced. Diaphragm repair was performed with 2-plane with non-absorbable sutures through the anterior border and pleural flap **(Figure 4)**.

The abdomen was closed primarily without much tension. After surgery, the baby was returned to the neonatal intensive care unit and maintained on high-frequency oscillatory ventilation. The baby was placed on nasogastric feeding on the fifth postoperative day, first at half strength and then at full strength. The chest radiograph on postoperative day four showed expansion of the right lung **(Figure 5)**, and the baby was weaned from ventilation after two weeks. The baby made a remarkable recovery and was discharged from the hospital on postoperative day twenty-nine; he is being followed up.

The evolution is favorable with 2 years of hindsight, without aggravation or appearance of respiratory disorders.

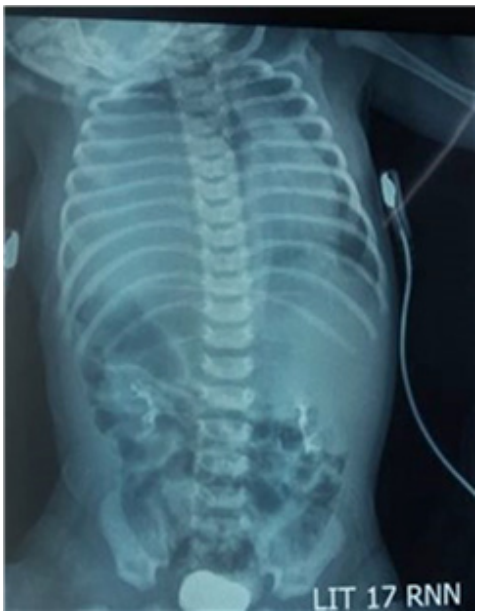

Figure 1: Homogeneous pulmonary opacity (shriveling the lung at the top and pushing back the trachea on the left) with image of colon filling the base on the right.

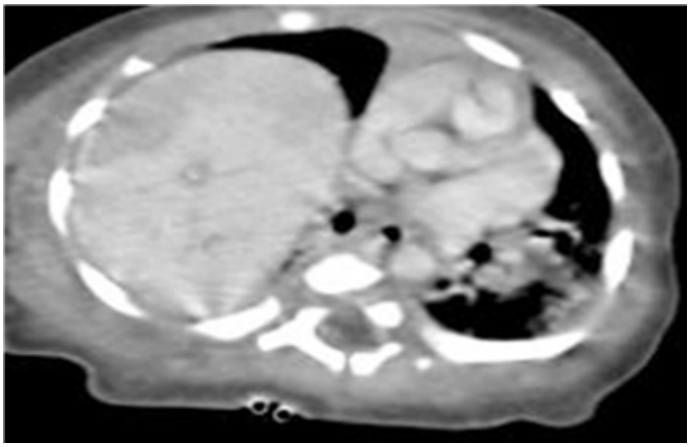

Figure 2: CT scan of chest showing liver at the level of heart.

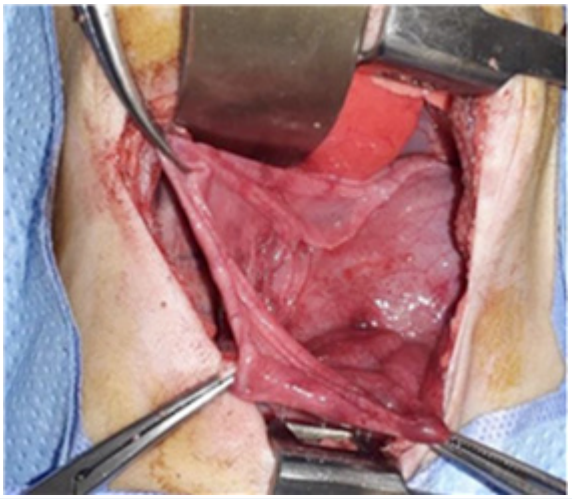

Figure 3

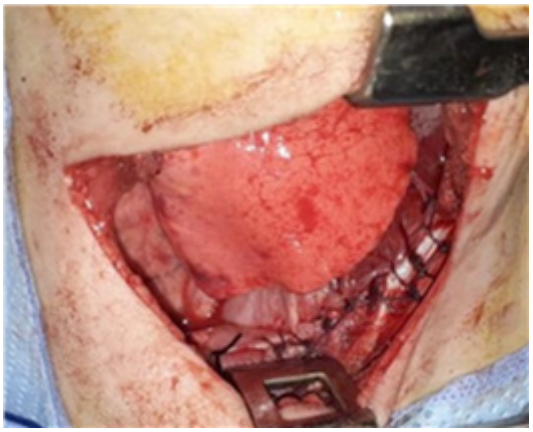

Figure 4

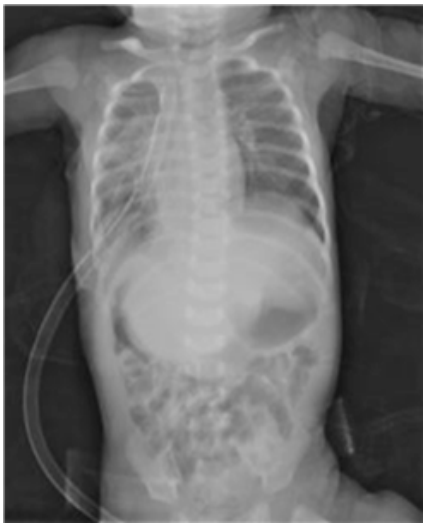

Figure 5: Chest X-ray on postoperative day four.

Discussion

Right-sided congenital diaphragmatic hernia (CDH) have been associated with higher morbidity and mortality than left-sided defects [1-3]. Whether the side of the anomaly has prognostic value remains controversial, as other groups have reported no difference [3] or better survival [4] for those who were managed before birth.

It is clear that the size of the anomaly is inversely correlated with survival and is the strongest predictor of outcome, both in terms of survival and morbidity [5,6].

The surgical repair was difficult in this right sided CDH because the size of the defect. Was more than four centimeters. Only a thin rim of diaphragm was present anteriorly and the posterior rim was absent completely. The reduction of liver posed a difficult problem. Liver replacement in the abdomen can be complicated by kinking of hepatic veins causing profound hypotension. Potential anatomic anomalies such as possible hepatopulmonary fusion [7,8] anomalous venous drainage uniquely associated with right sided defects. Survival based on liver herniation alone is 43% as compared to 93% survival without liver herniation [9]. The series published by Fischer et al. [10] has shown the survival rate (right CDH 55% to left CDH 77%) ECMO requirement (right CDH 40% Vs left CDH 15%) prosthetic material in R CDH Vs L CDH (76% Vs 41%) and abdominal wall (38% Vs 19%) repairs. These data support that right side CDH carries a high mortality and morbidity. The repair of a CDH may be as variable as clinical management. A thoracic approach is our attitude. While other surgeons prefer a transverse incision on the ipsilateral side, the type of repair is dependent on the size of the defect. If the defect is small, a tension free primary surgical closure should be performed with non-absorbable sutures. If the defect is wide primary closure may be attempted by one of the patch methods (**Prosthesis, Muscle Flap** [11,12,13]**, Bioactive Material** [14]).

Conclusion

Preoperative physiologic stabilization and subsequent elective repair have become the cornerstone of CDH management as in our case. The introduction of a treatment protocol, using high-frequency oscillatory ventilation from birth, a vasodilator, and delayed surgical repair after stabilization, improved the survival of this neonate with CDH. Success in this difficult case is optimized by close cooperation between the neonatologist, anesthesiologist, and pediatric surgeon. Although the hospital stay was prolonged, the survival outcome is excellent in this high-risk case.

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