

Successful Management of a Delayed Left Congenital Diaphragmatic Hernia in a 24 Days Baby Boy with Severe Respiratory and Hemodynamic Distress

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Abstract

Congenital diaphragmatic hernia (CDH) is characterized by the absence of all or part of a diaphragmatic dome. It represents 8% of all congenital malformations, with an incidence of 1 in 2500 births in the United States. While many cases are discovered prenatally or during the immediate postnatal period, 5 to 25% of CDH can be late diagnosed. It is associated with varying degrees of pulmonary and pulmonary arterial hypertension leading to a significant cause of morbidity and mortality. CDH has been rarely reported in Africa and never in Cameroon where there is a profound lack of appropriate infrastructures to help manage these children. We are reporting the case of 24 days newborn presenting with severe respiratory distress and hemodynamic compromise diagnosed with a left diaphragmatic hernia with subsequent emergent surgical repair.

Keywords: Congenital diaphragmatic hernia; Pulmonary hypoplasia; Pulmonary arterial hypertension; Respiratory distress

Introduction

Congenital diaphragmatic hernia (CDH) is characterized by a partial or complete absence of the diaphragmatic dome [1]. It represents 8% of all congenital malformations, with an incidence of 1 in 2500 births in the United States [2]. While many cases are discovered prenatally or during the immediate postnatal period, 5 to 25% of CDH can be diagnosed long after birth [2]. CDH is associated with varying degrees of pulmonary hypoplasia (PH) due to chronic compression of the herniated abdominal organs thru the diaphragmatic defect and pulmonary arterial hypertension (PAH) [3-5]. If late-onset forms have a good prognosis [6], early forms, with less favorable prognosis, require medical techniques often invasive to stabilize the patients before a reconstructive surgery can be undertaken in a second time [5,7-10]. The disease is uncommonly reported in Africa and no case has been reported in Cameroon. We are reporting the case of a full term newborn whose left posterolateral diaphragmatic hernia was found three weeks after birth.

Case Report

A newborn of 24 days was referred to us from a peripheral healthcare facility for management of a congenital diaphragmatic hernia. The child was born from healthy parents, the pregnancy and the delivery were normal with a birth weight of 2,550 g, the Apgar score was 9 at one minute and 10 at five minutes after birth. There was no noticeable abnormality during the first two weeks of life.

The parents consulted one week earlier at a peripheral health center for fever in the newborn. Two days after admission, he started presenting a progressive respiratory distress. Laboratory tests showed a significant anemia, leukocytosis predominantly neutrophilic and moderate thrombocytopenia. A chest x-ray (Figure 1) showed an elevation of the left diaphragmatic dome and the presence of digestive structures above the diaphragm. A chest Computed tomography (Figure 2) showed herniation of bowel loops filling the left chest with a complete collapse of the left lung and a shift of the mediastinum to the right. The diagnosis of a CDH was made and the child transferred to us from a distance of 250 km. He was transported in a simple ambulance, on nasal oxygen with no possibilities of intubation.

On arrival, the child was in severe respiratory distress and significant

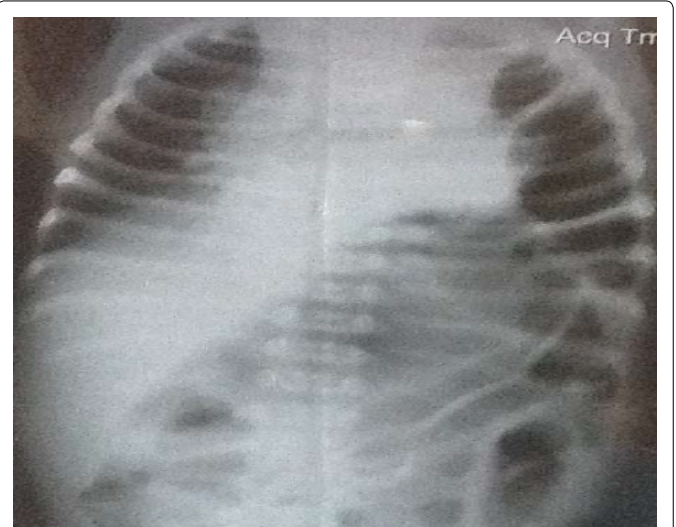


Figure 1: Thorax x-ray showing elevated left diaphragmatic dome and digestive loops above the diaphragm.

hemodynamic collapse (severe bradycardia, cyanosis, hypoxemia). The whole team (Surgeons, anesthesiologist, pediatricians, and nurses) was waiting for him. He was immediately taken to the operating room where he arrested on the table. An emergent laparotomy was then performed with reduction of the herniated organs (colon, stomach, spleen, and small bowel) in the abdomen. Concomitantly, the child was intubated

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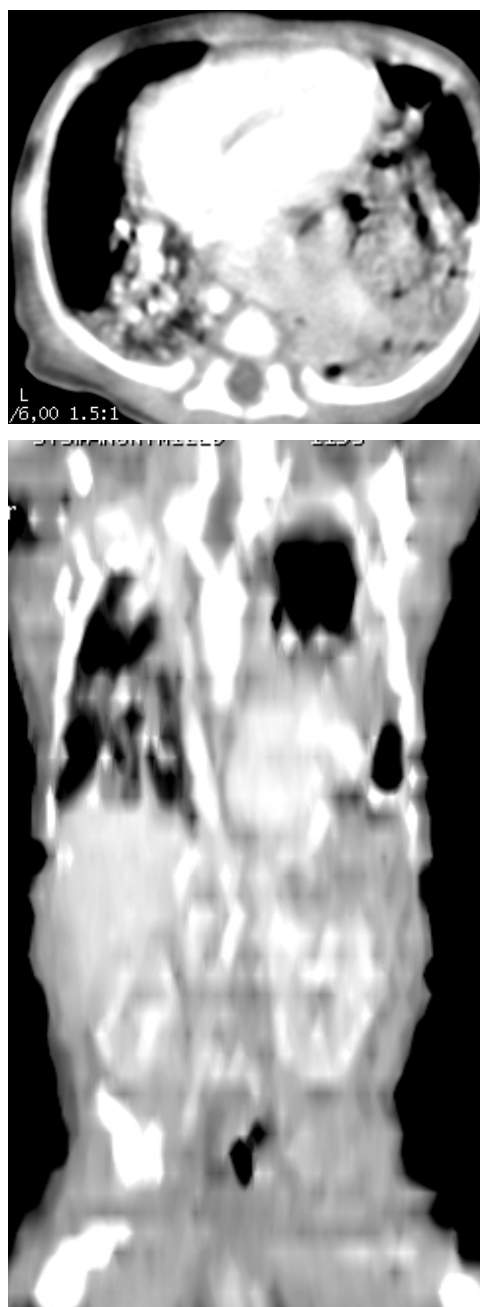


Figure 2: Thoracic TDM showing intrathoracic protusion of bowel loops filling the left chest with a complete collapse of the left lung, and a shift of the mediastinum to the right.

and resuscitative measures carried out. These manoeuvres were successful with return of the heartbeat and the disappearance of the signs of shock. The congenital diaphragmatic hernia, of 7 cm diameter on the left side was closed without problems with a non-absorbable suture. Because of the short live cardiac arrest and the absence of a pediatric respirator, he was kept intubated and ventilated manually for 8 hours after surgery (until the next morning). Breast milk feeding was started as soon as he was fully awake meaning in the afternoon. The postoperative course was uneventful. The child was referred back to his doctors a week after admission. He was seen one month later for follow up in our surgical clinic, he was doing well and the control chest X-ray at one month did not show any significant abnormality (Figure 3).



Figure 3: Postoperative chest x-ray one month after the surgery showing a left diaphragmatic dome at normal height and good ventilation of the left lung.

Discussion

CDH is characterized by the persistence of an orifice in the diaphragm. This orifice is located in 84% of cases on the left posterolateral region of the diaphragm, in 12% of the cases on the right posterolateral region and in 2% of the cases, on both sides [11]. The size of this orifice is variable from a few millimetres to a complete agenesis of the diaphragmatic dome. CDH appears very early during pregnancy because the development of the diaphragm takes place before 6 weeks of gestation. There are many theories on its formation (bowel interposition, anomaly of innervation by the phrenic nerves, anomaly at the level of pleuropulmonary membranes...) [11].

The persistence of this diaphragmatic orifice will disrupt the process of reintegration and joining of the intestine [6]. There are two major forms: in early forms, abdominal organs develop in the chest while in late forms, the viscera will secondarily migrate into the chest [1]. Our case is a late form. Although we have found a 7cm orifice, it seems difficult to explain why the child became symptomatic only 2 weeks after birth. The stomach, the spleen, most of the right and left colon and small bowel were in the chest resulting in an associated malrotation.

This permanent presence of the abdominal viscera in the chest will interfere with the development of the lung. The herniated abdominal organs exert a mechanical stress: inspiratory decrease in activity of the diaphragm in utero, increase in lung fluid leak through the larynx, less distension of the lung tissue by decrease in lung expansion space [11]. This constant compression of the lung will result in pulmonary hypoplasia. There is also an increase in the number of type II pneumocytes creating a morphologic (thicker alveolar septa) but also a functional impairment of the alveolo-capillary barrier [11]. The induced pulmonary hypoplasia will result in hypoxemia and acidosis due to bronchial dysplasia and the reduction of the alveolar exchange's surface [6].

Besides pulmonary hypoplasia, CDH results in proliferation of the vascular smooth muscle cells with thickening of the media of the pulmonary arteries [11]. This is the cause of PAH which can also be related to the reduction of the vascular bed due to the PH [12]. PAH causes a right to left shunt which will increase the hypoxia and the acidosis which in turn will accentuate the PAH creating a vicious cycle [6]. The CDH is always associated with varying degrees of PH and PAH that influence the prognosis of this disease [3-5].

Despite improved medical and surgical means of care of patients with CDH, mortality related to this disease remains high [3-5,8,13,14]. Although surgery is the definitive treatment of this malformation, its implementation at an early stage is increasingly refuted by several authors who prefer secondary repair to obtain a respiratory, circulatory and metabolic stabilization [5,7-10]. If in the late-onset forms, this stabilization is usually easily obtained by conventional methods (oxygen administration with or without tracheal intubation, spontaneous or low stress assisted ventilation, correction of acidosis), in severe forms revealed at birth, other techniques (extracorporeal membrane oxygenation, high frequency oscillatory ventilation, inhaled nitric oxide) are used to stabilize these patients [5,6,8,9].

In our environment, there is a profound lack of appropriate materials and infrastructures; there is no pediatric ventilator available, children are ventilated manually [15]. There are no possibilities to use ECMO. Conventional mechanical ventilation techniques with high pressure and hyperventilation run the risk of barotrauma related to bronchial or alveolar rupture [16]. Alveolar over distension induced by mechanical ventilation leads to visible lesions (mainly pneumothorax) and invisible lesions which are associated with a pulmonary and systemic inflammatory response [16]. This systemic inflammation may lead to organic failure which is responsible of an elevated death rate [16]. The barotrauma is frequent in patients with acute respiratory distress syndrome [17], with the values up to 60% in some old series [18]. In addition, mask ventilation promotes distension of the herniated viscera, thus increasing respiratory distress [10]. Emergency surgery, in precarious respiratory and cardiovascular conditions remains the only alternative in these patients. However, immediate intubation of children with CDH avoiding mask ventilation will minimize the herniation of the viscera in the chest [10].

In our case, the severe respiratory distress and hemodynamic compromise was aggravated by the poor conditions in the ambulance during the transport, and the child was saved in extremis, on arrival because the whole team was expecting him. We have reported the importance of the multidisciplinary management of the patient in critical conditions [19]. As we have reported for the fetus, newborns may have a stronger autoregulation which enable them to have a better recovery in face of a cardiac arrest [19]. We believed that the child extubation was delayed because of the metabolic acidosis following the cardiac arrest and the deleterious respiratory conditions that prevailed few days before he was referred to us. This type of pathology is a major challenge in our country where there is a lack of proper material and infrastructure, affected children do not have a proper environment to survive, and this must be the reason why the disease seems to be rare: the patient died before a diagnosis can even be made.

Conclusion

CDH is a rare disease with dismal prognosis correlated to the degree of PAH and bronchopulmonary dysplasia and hypoplasia. Its management uses heavy medical techniques in order to stabilize these patients for a secondary surgical repair. As shown in this case, this attitude should be reconsidered in some specific environment. Good

prognosis was possible in this child because the whole team was there on arrival.

A consent was taken from the parents who have accepted that the case be reported to the scientific community, because they believe that their experience worth to be known.

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