Left Posterolateral Congenital Diaphragmatic Hernia in Neonatal Revelation Form: A Case in Togo, A Developing Country

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Abstract

Congenital diaphragmatic hernia is a serious malformation with a permanent high mortality despite the considerable work it raised in respiratory and hemodynamic stabilization. The mortality is higher in developing countries where deep lack of resuscitation conditions is habitual. We hereby report a case of neonatal revealed CDH in a newborn admitted to the SOTH three days after his birth for respiratory distress and cyanosis. The boy had Apgar score of 8-9-8. His birth weight was 3700 g. There was no antenatal ultrasonography. About four hours after his birth he presented respiratory distress syndrome with cyanosis. He was managed for “neonatal infection and dextrocadia” until thoraco-abdominal x-ray was performed four days later. The x-ray showed stomach and colonic lights in the left hemi thorax displacing the mediastinum on the right. On admission, the baby was fully conscious but had major respiratory distress with cyanosis more remarked on extremities and on the lips. The left hemi thorax was bulging and less moving than the right side. The heart sounds were displaced to the right side. Neither ultrasonography of heart, blood gases nor pH status was done in emergency. After one day of resuscitation the baby underwent laparotomy. Herniated organs were stomach, spleen, transverse colon and the left lobe of the liver. There was no hernia sac. The left lung was seriously hypoplastic. It was made a freshening of the banks with direct closure of diaphragmatic defect by interrupted sutures with no absorbable suture. There was no intestinal malrotation. The newborn was kept oxygen depending during five days in resuscitation department. Postoperative heart ultrasonography was normal. The outcome was favorable after a follow up of six months.

Keywords: Bochdalek hernia, Congenital diaphragmatic Hernia, Children, Togo

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Introduction

Defined as the presence of an orifice in the diaphragm more often left and posterolateral that permits the herniation of abdominal contents into the thorax, Congenital Diaphragmatic Hernia (CDH) is a rare condition that occurs in less than 1-5 on 10,000 births [1,2]. It is a serious malformation. The mortality relative to only hospital statistics runs from 10 to 30% despite the important improvement of the management achieved during last years. When cases relative to terminations of pregnancy, spontaneous abortions, stillborns, pre-hospital and/or preoperative deaths and surgical mortality are considered, real mortality is between 50% and 60% [3-5]. In developed countries, it is currently accepted that a policy of "delayed" surgery associated with "gentle ventilation" and occasionally Extra-Corporeal Membrane Oxygenation (ECMO) support yields the best results recorded [6]. Conditions for this management especially resuscitation facilities are not often optimal in sub-saharan African countries [7-9]. We hereby report a neonatal revelation form of left posterolateral CDH managed in a newborn at Sylvanus Olympio Teaching Hospital (SOTH) in Lomé, in order to present the hard conditions of this management and the results.

Case Report

Figure 1: Thoraco-abdominal face x-ray of the newborn: notice stomach and colonic lights in the left hemi thorax displacing the mediastinum on the right, and fading of the left diaphragmatic cupola.

Figure 2: Left diaphragmatic copula defect seeing during laparotomy after reduction of herniated organs.

Figure 3: Closure of diaphragmatic defect by interrupted sutures with no absorbable suture.

Figure 4: Postoperative thoraco-abdominal face x-ray (7 days after operation): notice acceptable right lung re-expansion and repositioning of heart and the mediastinum.
It was a male newborn admitted to the SOTH three days after birth for respiratory distress and cyanosis. The boy was born at date in a district health center situated 7 km from SOTH. He cried at birth and had Apgar score of 8-9-8. His birth’s weight was 3700 g. There was no antenatal ultrasonography. About four hours after birth, he presented respiratory distress syndrome and cyanosis. There was a right deviation of the heart at auscultation. Oxygenotherapy was started and he was kept fasting. An antero-posterior thoraco-abdominal X-ray was wanted. Because of poor financial conditions of the parents, the X-ray was performed four days after birth. The X-ray (Figure 1) showed digestive images in the left hemithorax with right deviation of the mediastinum and left diaphragmatic cupula non observable. The right lung was compressed by the heart and the mediastinum. The diagnostic of left CDH was retained and the baby was transferred to SOTH the same day for management. The examination at admission noted respiratory distress with cyanosis more intense at the ends and on the lips. There was also insufficient oxygenation (83%). The respiratory frequency was 80 cycles per minute and the cardiac one was 148 per minute. The left hemithorax was globular and less mobile than the right side. Vesicular murmurs were absent on the left side and replaced by digestive bowel sounds. Heart sounds were moved to the right. The abdomen was excavated. Neither ultrasonography of heart nor blood gases and pH status were done in emergency. After one day of resuscitation including intubation, mechanic ventilation in pure oxygen with a ventilator frequency of 40-60 cycles per minute, nasogastric intubation, monitoring of vital parameters (cardiac frequency, saturation in oxygen, blood pressure) and taking a venous for analgesia and sedation; the baby underwent surgery. A left subcostal incision was used. Herniated organs were stomach, spleen, transverse colon and the liver left lobe. The diaphragmatic defect was left and posterolateral. After reduction of herniated organs, we noticed that the diaphragmatic defect was oval and measured 6 cm over 3 (Figure 2). There was no hernia sac. The left lung was seriously hypolpastic. There was no intestinal malrotation. We performed excision of the defect’s edges, followed by direct closure with non absorbable suture (Figure 3). The newborn was kept intubated for 24 hours but he remained dependent on oxygen for four to five days in resuscitation department. Postoperative thoraco-abdominal X-ray (7 days after operation) showed acceptable right lung re-expansion and repositioning of the heart and the mediastinum (Figure 4). Postoperative heart ultrasonography was normal. The outcome was favorable with a follow up of six months.

**Discussion**

DCH is a rare malformation. It occurs in less than 1-5 on 10.000 births [1,2,10]. It is a serious condition with a high mortality rate despite major improvements in resuscitation [3-5] which are routinely available in developed countries. In fact, CDH was described many years ago [11,12] but survival after repair was not achieved until the 20th century. Pioneers of pediatric surgery [13] reported amazingly low mortalities; nowadays the severity of the condition surfaces when abortions, stillbirths and pre-hospital deaths are considered, adding a “hidden mortality” to operative and postoperative demises [14]. The pathophysiology of lung insufficiency and persistent pulmonary hypertension that threaten survival are currently better understood and a new policy of “delayed” surgery coupled with “gentle ventilation” and occasionally extra-corporal membrane oxygenation (ECMO) are adopted [6]; but the results remain disappointing since mortalities near 50% are still reported when all deaths are taken into account in population-based series [15].

In developing countries especially sub-Saharan African countries, resuscitation conditions are limited and important diagnosis problems are noticed [8]. Then, after birth, respiratory distresses are often considered as manifestations of neonatal infection and managed like that, making the diagnosis delayed [8]. Precious time is then lost, sometimes with useless dangerous treatment.
Elsewhere, the lack of antenatal diagnosis contributes also to the poor conditions of management. In our case, there was no antenatal ultrasonography and the baby was born in a district health center without pediatric department and managed for “respiratory distress and dextrocardia” until the X-ray was realized. Medical continuous training for patricians could help to fulfill this gap. The thoraco-abdominal X-ray was hardly done four days after the birth. Radiological examination must be done quickly. A thoraco-abdominal radiogram is often enough to make the diagnosis, showing intrathoracic multiple digestive images associated with right deviation of the heart and mediastinum and reduced digestive pneumatization in the abdomen. In the pediatric surgery of SOTH, neither heart ultrasonography nor blood gases were available. It was the same for ECMO. These are the conditions of management of CDH in several developing countries. The diagnosis of CDH may be suspected during pregnancy or done after birth when a newborn presents with acute respiratory distress. Although the simple thoraco-abdominal X-ray is sufficient to the diagnosis, the most important problem in our conditions is the way to have respiratory and hemodynamic stabilization; in fact these are essential before any surgery [9,10]. Without blood gases, pH status and ultrasonography of the heart, we did blind resuscitation in our case till surgery. Our preoperative control parameters were tegument coloration, blood oxygen saturation and respiratory frequency. Our patient underwent surgery with oxygen saturation of 83% despite nasal oxygenation. We didn’t know about the seriousness of the pulmonary hypertension in this case because of not having heart ultrasonography in emergency. It is known that main factors of mortality are the severity of pulmonary hypoplasia and the presence of pulmonary hypertension [10,16,17]. Other associated malformations can worsen the prognosis. They are very common in stillbirths (85%) and would range from 20% to 50% among alive newborns. There were no associated malformations in our case; it was a major favorable factor in that situation. Surgical management is usually performed by laparotomy by a subcostal incision ipsilateral to the hernia. Herniated viscera are reduced, and the diaphragmatic gap is visualized and lung volume assessed. The closure of the diaphragmatic defect is performed either by direct suture or by using prosthetic material [16]; the choice of the procedure depends on the size of the orifice and the existence of a posterior diaphragmatic rim, which is the pledge of solidity. This surgery can be made by laparoscopic approach especially in older children with a delayed presentation [17-20]. The question that remains discussed is the establishment of a diaphragmatic prothesis. Some authors believe that it improves lung compliance in case of large defect when others consider that there is a risk of recurrence of the hernia. In order to succeed the management, whenever prenatal diagnosis is made, it is advisable to direct the expectant mother to a tertiary perinatal center in which all the necessary obstetric, neonatal and surgical skills are concentrated [21]. In our case, the baby was born in a district health center and wasted there precious time even before the diagnosis. Surgical repair of CDH used to be in the past a vital emergency. It is actually accepted that it should be undertaken only after stabilization of cardio-respiratory functions. A policy of “delayed” surgery coupled with gentle ventilation and occasionally ECMO support yields the best results recorded [6]. Several medications are used to treat the pulmonary hypertension: tolazoline [22-24] and prostacyclins [25,26] were tried first, but they did not produce good results. Prostaglandin 1E is occasionally used [27]. Inhaled nitric oxide, a well known smooth muscle relaxant is in turn widely used [28-31] with variable results, although there is no strong evidence of its benefits [32]. Inhibitors of phosphodiesterase like sildenafil, known for their vasodilator action, are currently used in some cases [33], but again, there is only anecdotal evidence of their benefits. These medications are coupled with inotropic agents, like dobutamine (up to 20 mg/kg/min), dopamine (<10 mg/kg/min) [34] and also, sometimes with peripheral vasoconstrictors, like adrenaline at low doses, aimed at reducing the shunting by increasing pressure in the
systemic circulation. It is rare to see sub-saharan African countries publications talk about these medications in the management of CDH.

Although many questions are expected to be resolved, we have first in our context to make available early diagnosis and optimal resuscitation conditions which are already achieved in developed countries.

**Conclusion**

Although the evolution was favorable in our case, it is important to retain that the mortality of CDH even in developed countries remains very high despite the improvement of resuscitation conditions. Developing countries are still characterized by a deep lack of required conditions as far as the management of CDH is concerned, from the antenatal diagnosis to the postoperative follow-up. The improvement of the management of lung hypoplasia is necessary for developing countries; but before that, we have to focus on early diagnosis. Retraining doctors, pediatricians, obstetricians and mid-wives and sensitilizing expectant mothers can help to achieve this goal.

**References**