Analytical Retrospect of the First Successful Thoraco-laparoplasty For Congenital Diaphragmatic Hernia In a 1-day Old Newborn In Rwanda: A case review

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ABSTRACT

Successful diaphragmatic surgeries with abdominal approaches have been reported nowadays in most developed and fully equipped surgical centers. Few reports exist in developing countries due to the rarity of the disease, and insufficiency of well-equipped centers for its accurate diagnosis and management.

The following analytical retrospect describes a 1-day old newborn with congenital diaphragmatic hernia (CDH) received after 24hours of life and treated successfully at King Faisal Hospital Rwanda (KFH), with hernioplasty through a thoraco-abdominal approach. The newborn recovered perfectly well after surgery without complications. The discussion, herein, extends on the rationale and new practical concepts in the combination of both medical and surgical therapies, highlights some updated anticipatory preoperative and postoperative measures for a better overall outcome on such major and complex disease.

Keywords: Congenital - Diaphragmatic hernia - Thoraco-laparoplasty - Surgery - Rwanda

INTRODUCTION

CDH is a rare malformation. Its incidence is approximately 1 per 2,000 - 4,000 live births worldwide [1, 2, 3], with an overall survival of 56.3-90% in developed countries [4,5,6] and a widely distributed mortality ranging [1]: < 25% (developed countries) to > 75% (developing countries) [4,7,3,8]. The sex-ratio (M:F) remains high 3-8:1 [1], with a significant number recorded in developing countries due to the attention paid to male children (male dominated societies) [5]. Few articles exist in developing countries, and no report has yet been published in Rwanda or elsewhere in the Eastern African region.

Diaphragmatic hernias are caused by a defect or the absence (Partial/complete) of closure of pleuroperitoneal apertures which results in the protrusion of abdominal organs in the pleural cavity. The expansion of abdominal contents in the chest depends on the level and location of the defect, and other associated thoracoabdominal malformations. The disease is classified into two [5,9]: 1.Bochdalek hernia in which there is posterolateral diaphragmatic defect; and 2. Morgagni hernia where the diaphragmatic defect is located anteromedial on either side of the septum transversum and the thoracic wall.

For the past 30 years, great progress has been made in developed countries due to the improvement of prenatal versus postnatal diagnostic modalities, and the adequate optimization with proper management of the disease. Many authors stipulate that the high predictive risk factors of mortality are: time of prenatal diagnosis, low APGAR level at birth, and the higher mean of oxygen index (OI) - a measure of the level of ventilator support - ; whereas the low predictive risk factors remain the nature and characteristics of the defect, the persistent pulmonary hypertension of newborns (PPHN), and the type of therapy provided [1]. Prenatally detected cases (30%) [1] have a significantly reduced survival to surgery, an overall low survival and lower Apgar scores at one minute (p-value < 0.04); what predetermine in fact the overall mortality of CDH in early infancy, and explain the early presentation and the severe symptoms both identified as the predictors of mortality, of greater degree or longer duration of visceral herniation, therefore an increased association to severity of pulmonary hypoplasia [4].

In fact, it has been reported the early hospital presentation of right-sided CDH compared to left-sided, even though left-sided presents more frequently (75%) [5,1]; with a greater incidence of isolated CDH 76% [1,10]. No statistical significant difference in gestational age at birth and birth weight has been observed to influence the overall mortality or its prediction [4]. The hospital stay has been reported to be 10–30 days (mean 12 days); and it has been observed that in developing countries, most patients present late after 24h of life explained mostly by the origin of patients from remote areas [5].

CASE PRESENTATION

A 01 day-old newborn boy referred from the Rwanda Military Hospital (RMH) with acute respiratory distress and high suspicion of dextrocardia. Admitted in the Neonatology intensive care unit at KFH, the patient was immediately stabilized and put on respiratory support while a multidisciplinary team - made of pediatricians, general surgeons, a thoracic surgeon, and an intensivist - was convened for surgical and intensive care treatment. They reported a term delivery with Cesarean section due to incomplete fetal engagement and distress (APGAR 6/10-8/10). The pregnancy went on without major concern, without chronic or infectious conditions, with no reported risk factors for fetal malformation, and all prenatal visits were observed regularly and correctly; although, they reported long delivery and rupture of placental membranes for 15 hours with no antibiotics coverage.

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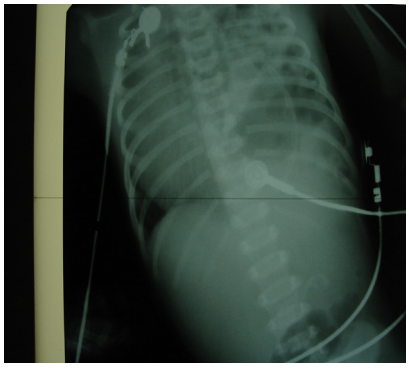
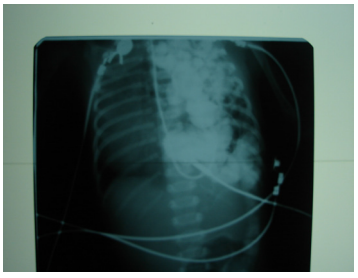
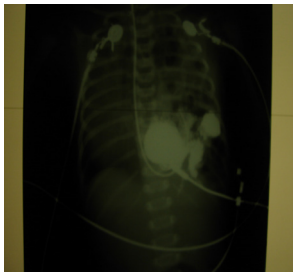
At arrival, the newborn had polypnea (RR=80), moderate cyanosis central (SaO2=85%), nasal flares, paradoxal thoracoabdominal movements, moderate pectus excavatum, right-sided cardiac impulse and the chest auscultation revealed heart sounds in the right hemithorax. We noted bilateral and symmetrical chest expansion, even though decreased right breath sounds were reported, and completely abolished on the left lung with dull left basal hemithorax. The Full blood count (FBC) showed white blood cells WBC=22.100/mm³; (PNN=20.4%), 66% lymphocytes, Hb=14.6g/dl, Platelets= 266.106/ml, a normal CRP and serum electrolytes panel.
The chest radiography with anteroposterior view in supine position showed multiple air infiltrations with features of aero enteric images, right mediastinal deviation, and right pulmonary parenchymal collapse.

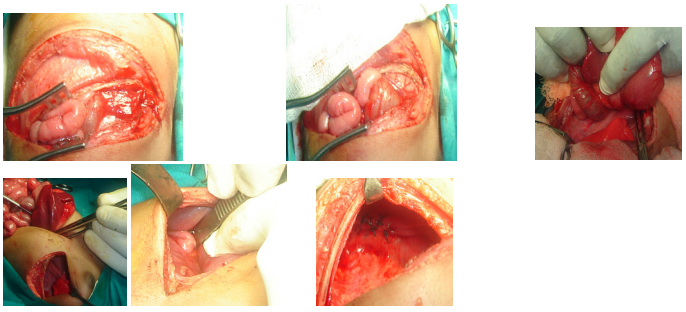


The cardiac ultrasound showed normal heart shape, size and function. The barium meal highlighted the protrusion of contrasted enteric loops into the left thoracic cavity; hence the confirmation of left congenital diaphragmatic hernia.

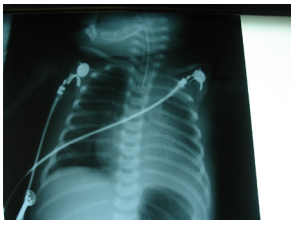


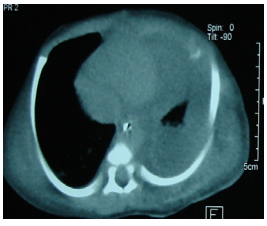


The thoracic surgeon was called 48 hours after the admission into NICU (at 72 hours of life), and after discussion of the overall optimization status within the treating team, the decision to operate was taken. The preliminary surgical approach was thoracotomy which failed to reduce the hernia into the abdomen, and an extended midline abdominal incision was therefore made to approach the abdominal organs protruding into the thorax, and were finally reduced back in the abdomen. Intraoperatively, the stomach, bowels and upper part of spleen were found in the thorax. An advanced left pulmonary hypoplasia and low degree right pulmonary hypoplasia were noted, but neither secondary organ malformation nor other subsequent injury damage was reported.



After surgery, the newborn was readmitted in NICU; the respiratory was good on mechanical ventilation for the first 2 days, thereafter was extubated and kept on oxygen face mask. On the 3rd day postoperatively, he passed flatus and stools. Soft meal nutrition was started through nasogastric tube, and we noted intolerance to feed. We decided to make a 3days-window for further bowel maturity, and continued with small amount of 5cc every 3hours which were progressively tolerated. The newborn developed facial and peripheral edema; and the serum albumin level was noted low. We implemented an intravenous human albumin infusion every 3 days that improved him clinically with a noticeable progressive decrease of edema.
Multiple chest radiographies and computed tomography showed in postoperative left pulmonary hypoplasia, recentralization of the mediastinum, and a good right lung ampliation. Postoperatively, we nebulized the patient with epinephrine every 3 hours coupled with chest physiotherapy aiming to reopen the left lung collapsed alveoli.





The newborn was discharged one month later, in a good clinical status, stable with vigorous movements, and adequate developmental milestones regarded the age; although oxygen nasal prolongs had to be continued at the nearest district hospital until complete recovery and good saturation on room air. An appointment after one month was planned to evaluate the pulmonary maturity and function for eventual wean off continuous oxygen support; and the patient showed up 4 weeks after the discharge back to the referring hospital in good general status.

DISCUSSION

Embryologically, the diaphragmatic development (4-12th week of gestation) results from the fusion of the septum transversum and the posterior mesenchymal proliferation; and herein approximately, the diaphragmatic hernia forms between the 6-8th week of gestation. The fusion of those

2 primitive remnants makes the central of the diaphragm; separating therefore the organs into 2 main different cavities. Though, 2 large apertures remain posteriorly and form pleuriperitoneal canules (Bochdalek foramen) which will be covered by secondary pleural and peritoneal serosa. In fact, the closure of the pleuroperitoneal canules occurs on the 8th week, starting from the right side than the left side; what explains the high frequency of left congenital hernias [11,12,13].

The pathophysiology of inadvertent presence of abdominal organs in the thoracic cavity results into 3 main consequences [11,14] : 1.Homolateral predominant pulmonary hypoplasia characterized by the reduction of the number of total alveoli; 2. Increase of pulmonary resistance (with reduction of vascular bedsites) which results in pulmonary arterial hypertension with persistence of a shunt effect; and 3. Abnormal organ attachment with irregular organ rotations. Thereof, the main complaint is acute respiratory distress (Polypnea, dyspnea, cyanosis) refractory to oxygen support which makes suspect primary neonatal infection, cardiac congenital malformation and/or congenital diaphragmatic hernia [13,15,16,17].

The clinical findings are mostly decreased chest expansion and air entry on the affected side with a scaphoid abdomen. The pulmonary auscultation shows absent breath sounds homolateral to the hernia with eventual digestive borborygmi. The diagnosis is on imaging modalities which include radiographic investigations (82.6% confirmatory) [5] showing a hemithorax with multiple aero colic images features of enteric organs in the chest, a contralateral mediastinal shift (67%) [1,13,16,17] with equal incidence rate if we consider the side of the defect in the thorax; but on the other hand, the oral contrast study confirms 17.4% of the remaining unclear diagnoses [5].

A protocol of gentle ventilation to avoid barotraumas, and inhaled nitric oxide (iNO) or intravenous magnesium sulphate has been widely recommended [1], but few hospitals are well equipped to implement them. The rationale behind that protocol is to provide adequate range of hemodynamic and respiratory stability of the newborn, safe surgical environment for definitive care, and decrease the postoperative complications. Stabilization meant settled pulse and respiratory rate without any chest retraction or evidence of cyanosis, Oxygen saturation of more than 88% on pulse oximetry, and arterial blood gas levels of $pCO2 < 60$ and $pO2 > 40$ [5]. Herein, Surgery is performed when the infants' vital signs, general condition and blood gases are well stabilized for at least 24 hours. However, the optimal timing of surgery is still not universally accepted. Rozmiarek et al reported that cardiac defects such as Ventricular or atrial; renal failure and initial blood gases were significant factors that influence the survival rate; and not the timing of surgery [18]; therefore delayed surgery allows improving the postoperative conditions which influence mostly the mortality within the stabilization of hemodynamic functions prior to surgery [1]. The management of CDH has actually changed from being seen as a dire emergency to nearly elective situation where adequate preoperative preparation and stabilization of the patient are paramount and undertaken before any plan for theater [5]. Actually, delayed surgery has become a widely accepted strategy for treatment of CDH; and Sakai et al reported the worsening of cardiopulmonary function following early surgical repair [19].

Surgical techniques include laparotomic approaches via the subcostal incision both for left- or right-sided hernia, which are the preferred procedures [9,20,14,21,17] then reduction of herniated contents and closure of the defect using mesh repair with polypropylene sutures [5]. In fact, any subsequent condition or other treatable malformation is corrected at the same time. Nowadays, thoracoscopic surgery and other techniques are done in highly-equipped centers for repair of CDH with good results [22], but we don't have quite good experience with them in our setting. The prevention of the development of pulmonary hypertension by preventing patient agitation and avoiding hypothermia or hypoglycemia along with maintaining the intra-vascular volume status remain the paramount measure for the preoperative optimization in all newborn patients as an alternative treatment in case adequate highly effective protocols are not available. On the other hand, postoperative chest tube placement would remain a random surgical decision prior to surgery to eliminate selection bias. It has not been recognized as a positive effective factor for survival [5], therefore the risk of mortality increases [23], related infections (empyema) increase [5]; and recent reports claim good results without using the chest tube [24,25].

In fact, regarding the discussion above, the patient herein reported presented the same chest radiological findings, a normal cardiac ultrasound, and the barium meal confirmed the diagnosis of left diaphragmatic hernia. The multidisciplinary therapeutic approach, in the neonatal setting, was oriented mostly on the respiratory support due to the imminent life threatening distress. Even though, we were not able to use the most top technologies and highly recommended protocols, we proceeded with an endotracheal intubation and adequate ventilation support, a nasogastric tube insertion to decompress the stomach, sedation and curarization, close hemodynamic and ventilator monitoring, acidosis correction, and a complete malformation investigations panel [8,26,17]. The patient was admitted in the neonatology intensive unit, was put on mechanical ventilation with a PEEP=5mmHg, Pi=15mmHg for adequate respiratory support, and a complete septic workup was without particular concern. The surgery was performed on day-3 of life after respiratory stabilization, and the approach was through a laparotomy incision with hernioplasty after reduction of the eviscerated abdominal content.

Early most common postoperative complications usually include wound dehiscence, ileus, conjugated hyperbilirubinemia, pneumothorax requiring drainage, and Acinetobacter sepsis [4]; but the overall most common complications of CDH remains PPHN and pulmonary hypoplasia; and on long term, few patients develop chronic lung disease [1].

The mortality remains high within the first 72 hours of life (30-50%); and nearly 100% when symptoms start within 12 hours of life; which is explained by the low APGAR score [4], Severe PPHN [1] , and eventual gross bowel ischemia with gangrene of herniated viscera [27].

The high survival rate reported in well-equipped settings note that the mortality is not due to the disease itself but the early stabilization and adequate management. Some reports agreed that a higher proportion of infants who develop PPHN die, but this has never been statistically proven to be significant; though, significance is retrospectively noted in reportedly dead patients with higher OI (reflecting more severe PPHN) than the survivors ($p < 0.04$); therefore associating the mortality to severe PPHN rather than simple PPHN [1].

In developing countries, it remains unclear which strategy or combination of strategies offers the best survival rates. Most of the reported studies on CDH were non-randomized

and the number of patients was small. Nowadays, from the limited evidences available, better outcomes are reported in infants delivered at tertiary centers with surgical repair delayed until hemodynamic and respiratory stability are achieved, and with the judicious use of non-aggressive mechanical ventilation and permissive hypercapnia.
Reports from developed countries describe improved survival with high frequency ventilation, inhaled nitric oxide (iNO) and extracorporeal membrane oxygenation (ECMO), apart from delayed surgical repair [28, 29]. When the diagnosis is made prenatally, the mortality remains high 100% than when done postnatally (75%) with an overall mortality of 75% before surgery, and 15% after surgery. The worse prognosis was reported in fetuses with CDH associated with liver herniation in the hemithorax compared to similarly-affected fetuses without liver herniation [30].
Coincidently, the relatively increased survival rate, in developing countries, is attributed to delayed arrival rather than highly performed preoperative management [5]. Many of the neonates born with CDH might die undiagnosed in our remote areas; what makes late presentation more common, and even in very poor clinical condition with an obvious need of stabilization before definitive surgical management.

CONCLUSION

Congenital diaphragmatic hernia remains a rare pathology and more rarely diagnosed in settings with limited resources. The diagnosis is highly suspected with respiratory distress of the newborn associated with deviation of the mediastinum. Barium meal remains the gold standard investigational test. The cardiopulmonary ultrasound is essential in the exclusion of associated congenital malformations. Surgery should be discussed as soon as the diagnosis is confirmed to prevent complications related to delay of respiratory support and implement early correction of pulmonary hypoplasia. Cardiothoracic surgeries are actually performed in our setting, and the prognosis could be promising for an upgraded setting confronting more and more simple or complex congenital diaphragmatic hernias.

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