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Challenges in the management of early versus late presenting congenital diaphragmatic hernia in a poor resource setting

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ABSTRACT

Background: Despite the advances in management, congenital diaphragmatic hernia (CDH) has continued to pose a significant challenge to paediatric surgeons. This is amplified in a setting like ours where there is a dearth of facilities to cope with the problem of CDH. This study was undertaken to highlight the peculiarities of the management of CDH in a poor resource setting. **Methods:** All confirmed cases of CDH were prospectively documented from 2003 till date. **Results:** Seven children were treated from 2003 till date. The diaphragmatic defect was on the left side in six (83.8%) and on the right side in one (17.7%). All the patients had primary closure of the defect without patch via an abdominal approach. The three patients presenting at birth died while the remaining four patients survived. **Conclusion:** With inadequate neonatal intensive care facilities, the severe early presenting CDH has a dismal prognosis. In contrast, the late presenting CDH poses more diagnostic challenges; but once identified and appropriate treatment instituted, it has an excellent prognosis. We recommend that physicians should include CDH in the differential diagnosis of patients with birth asphyxia and in patients with chronic respiratory symptoms with failure to thrive.

Key words: Congenital diaphragmatic hernia, outcome, poor resource setting

INTRODUCTION

Congenital diaphragmatic hernia (CDH) has continued to pose a significant challenge to paediatric surgeons. Despite advances in prenatal diagnosis, transport, intensive care unit (ICU) care and new treatment modalities, overall mortality rate has continued to remain high.[1,2] In poor resource settings like ours, the challenges for the management of CDH are more as there are no adequate facilities for prenatal diagnosis, transport and neonatal intensive care. These result in most of the severe cases of CDH being missed (hidden mortality) or they die in the early postnatal period and just a select group of late presenting CDH with less severe pulmonary hypoplasia survives, as is evidenced by earlier reports on CDH from Nigeria.[3-6]

We report the experience with the management of seven cases of CDH in our hospital over a 6-year period.

MATERIALS AND METHODS

This is a prospective observational study of all cases of confirmed CDH presenting at this hospital from December 2003 till date. The neonatal cases were admitted into the special care baby unit which has limited facilities for neonatal intensive care. A neonatal ventilator became available only in 2007.

Records were entered into a proforma and included age at presentation, birth weight, sex, Apgar scores, and clinical features, results of radiological investigations and presence of associated malformations. Others included the operative findings, operative procedures carried out, the outcome of treatment, follow-up and duration of follow-up.

RESULTS

There were seven children comprising five boys and two girls. The median age at presentation was 10 days with a range from few minutes after birth to 7 months. In three patients, the diagnosis was made few minutes after birth, two patients presented within the neonatal period on day 10 and 14, while in two patients the presentation was after the neonatal period [Table 1]. In six of the seven patients, the defect was located on the left and it was on the right in the seventh patient. All

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Table 1: Patients' characteristics, clinical presentation, associated malformations and investigations

Case/year	Age/sex	Birth weight	Delivery mode	Apgar scores (5 minutes)	Clinical features	Associated malformations	Investigations and results
1/2003	4 months/male	—	SVD	Unknown	Fever, cough, dyspnoea, failure to thrive, intercostals recession	Down's syndrome	Chest X-ray: presence of bowel loops in left hemithorax
2/2004	1 hour/male	3.25	SVD	4	Dyspnoea, cyanosis, scaphoid abdomen, subcostal recession	Undescended testis	Chest X-ray: presence of bowel loops in left hemithorax
3/2005	2 hours/male	2.70	SVD	3	Dyspnoea, cyanosis, scaphoid abdomen	Unknown	Chest X-ray: presence of bowel loops in left hemithorax
4/2006	12 days/male	2.55	CS	5	Dyspnoea, cyanosis, bulging left hemithorax	Nil	Chest X-ray: presence of bowel loops in left hemithorax
5/2006	2 hours/female	—	SVD	4	Dyspnoea, cyanosis, subcostal recession	Nil	Chest X-ray: presence of bowel loops in left hemithorax
6/2008	7 months/ female	3.70	SVD	Unknown	Dyspnoea, failure to thrive, pectus excavatum	Pectus excavatum, midgut malrotation	Chest X-ray: ill-defined diaphragmatic silhouette on the right; diagnosis confirmed by barium enema
7/2009	10 days	—	SVD	Unknown	Fever, dyspnoea, anorexia	Nil	Chest X-ray: presence of bowel loops in left hemithorax

CS – caesarian section; SVD – spontaneous vaginal delivery

the patients presenting at birth had severe respiratory distress associated with cyanosis and had 5 minutes Apgar scores of between 4 and 5. The older patients presented with recurrent difficulty in breathing, cough and failure to thrive. Details are shown in Table 1. There were four associated malformations in three of the seven (43%) patients. These were Down's syndrome (1), undescended testis which was also a content of the hernia in the chest (1) and pectus excavatum and midgut malrotation (1).

The diagnosis was confirmed in six of the seven cases with plain chest radiography that revealed the presence of bowel loops in the left hemithorax with shifting of the mediastinum to the right in five patients [Figure 1]. In the sixth patient (case 6), the bowel loops were on the right with shifting of the mediastinum to the left. She also had a barium enema confirming the findings on chest radiography [Figure 2].

Six of the seven patients had reduction of the herniated viscera [Figure 3] and primary repair of the defect using non-absorbable Prolene 0 sutures via a laporatomy. The seventh patient (case 4) died before surgery while he was being stabilised. The parents refused consent for a postmortem examination. In one patient (case 6), a modified Ladd's procedure was also carried out to correct the associated midgut malrotation [Table 2].

All the three patients who presented immediately after birth died. The remaining patients who presented at 10 and 12 days and all those who presented outside the neonatal period survived. Four of the seven patients survived, thus giving a survival rate of 57%. All the survivors are alive and well, with weight and height appropriate for their ages over a follow-up period of 5 months–6 years. There was also no recurrence of hernia among the survivours. Details are given in Table 2.

DISCUSSION

CDH is rare with an incidence of 1:2000 pregnancies[7] the lower incidence is because of the "hidden mortality" of the severe forms of CDH who die inutero.[8,9] There is no gender, racial or geographical predilection.[7] The literature on CDH in Nigeria consists of mostly single case reports, or as in our experience, relatively small series spanning over a long period of time.[3-6] We see an average of one case of CDH per year. This relative rarity of CDH in our practice may be accounted for by the absence of antenatal diagnosis and the loss of most of the severe form of CDH in the immediate postnatal period. In Nigeria, antenatal care is not routine and many children are born at home unsupervised or at best supervised by traditional birth attendants (TBAs). Coupled with early feeding, a patient with severe CDH will likely die. However, all the patients in our series had supervised antenatal care and delivery in hospital and 80% had antenatal ultrasonography (USS) but CDH was missed despite the presence of polyhydramnios in

two of the mothers. Even when delivery takes place in the hospital, cases with CDH may be lumped together and managed as birth asphyxia and may succumb. Since postmortem examination is not routine, CDH in still births and early neonatal deaths is also missed. The relative rarity of CDH in our settings may be explained by the above factors.

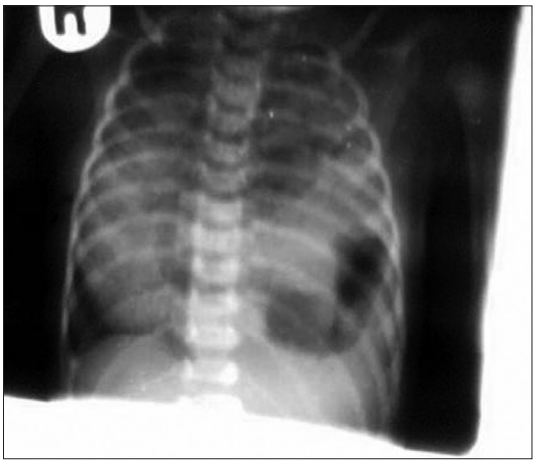


Figure 1: Plain chest X-ray showing the presence of bowel loops in the left hemithorax with shifting of the mediastinum to the right

The spectrum of clinical presentation of patients with CDH varies widely. This is determined by the severity of pulmonary hypoplasia and the presence of persistent pulmonary hypertension (PPHN). Elsewhere, most cases of CDH are diagnosed antenatally or at birth.[10] This permits counselling of parents regarding prognosis and preparing the health care team towards eventual delivery of the CDH patient. Facilities for skilled resuscitation including extracorporeal membrane oxygenation (ECMO) are also available in such centres to provide optimal conditions for the surgical procedure.[10,11] However, there still remains a subset of severe CDH patients with liver up in the chest and a prenatal sonographic lung to head ratio (LHR) of less than 1, who do badly with standard postnatal care is considered for antenatal intervention. Antenatal intervention for CDH has gone through the following phases: open surgical repair, open surgical tracheal occlusion (TO), endoscopic external TO and endoscopic

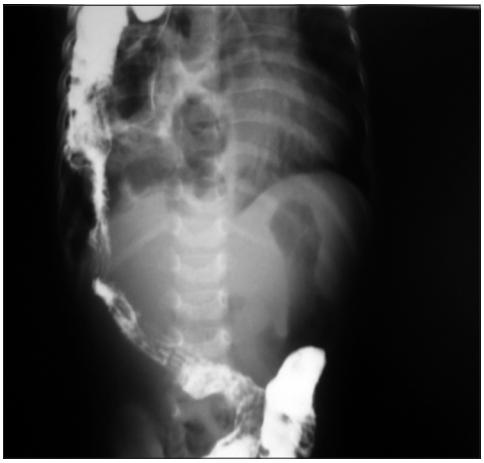


Figure 2: Barium enema with the colon inside the right hemithorax with shifting of the mediastinum to the left

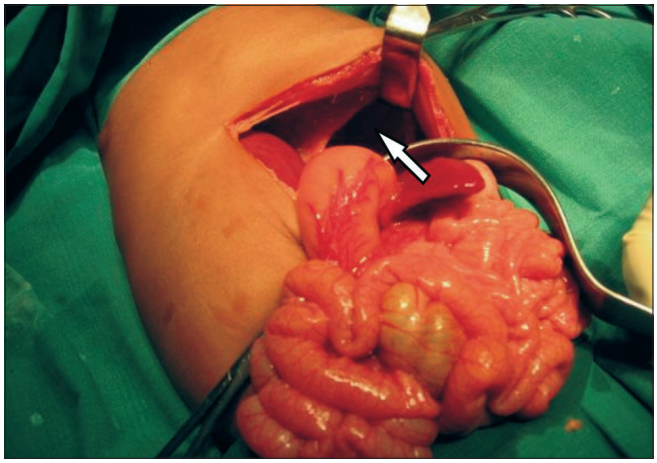


Figure 3: Intraoperative findings after reduction of hernia contents from the left hemithorax; arrow inside the diagphragmatic defect

Table 2: Operative findings, surgical procedures and outcome of treatment in seven cases of CDH

Patients	Operative findings	Contents of hernia	Surgical procedure	Outcome/follow-up
Case 1	Left posterolateral (10 × 6 cm)	Whole colon and small intestine, liver, spleen	Reduction and closure	Alive and well at 6 years follow-up
Case 2	Left posterolateral (10 × 8 cm)	Stomach, small and large intestines, spleen, left kidney, left testis	Reduction and closure	Died
Case 3	Left posterolateral	Unknown;died preoperatively	Nil	Died
Case 4	Left posterolateral (6 × 4 cm)	Transverse and ascending colon, small intestine, left kidney	Reduction and closure	Alive and well at 3-years follow-up
Case 5	Left posterolateral (8 × 6 cm)	Small intestine, stomach, spleen	Reduction and closure	Died
Case 6	Right posterolateral (3 × 4 cm)	Small intestine, ascending, transverse to proximal descending colon, midgut malrotation	Reduction and closure, modified Ladd's procedure	Alive and well at 20-months follow-up
Case 7	Left posterolateral (5 × 4 cm)	Stomach, transverse colon	Reduction and closure	Alive and well at 6-months follow-up

endoluminal TO. Randomised control trials in the United States, however, have not shown any advantage of these over standard postnatal interventions and therefore have been abandoned.[12] The application of antenatal minimally invasive TO in some European centres for the CDH-induced lung hypoplasia holds some promise, and with improvements, will determine its feature role.[13] None of our patients had antenatal diagnosis of CDH even though antenatal USS was done in five of the mothers but the diagnosis was made only during the postnatal period in all the patients. In Nigeria, some of the earlier reports have shown that all the patients presented outside the neonatal period.[5] In our practice, there are limited facilities for adequate resuscitation and stabilisation of patients with CDH presenting at birth; therefore, only a select group of good prognosis CDH with milder form of pulmonary hypoplasia present. The severe forms probably succumb early in the neonatal period. Our group of CDH who were diagnosed at birth presented with the classical symptoms and signs of CDH and was easily identified. On the other hand, the two patients who presented outside the neonatal period had repeated treatment for bronchopneumonia in outlying hospitals before presenting to us and the diagnosis was delayed in both of them. In a large series from multicentre studies, the clinical presentation of late presenting CDH was found to be acute or chronic respiratory or gastrointestinal symptoms or may even be completely asymptomatic. They also found predominance of gastrointestinal symptoms with left sided defects.[14] Plain chest radiograph was enough to make the diagnosis of CDH in six of our seven patients. However, the value of plain radiography has been questioned by some workers especially in the late presenting CDH.[15,16] Radiographs of CDH have been misinterpreted as pleural effusion, pneumothorax with inappropriate insertion of a chest tube into the herniated viscus in the chest and sometimes with consequences of gastrointestinal perforation.[5,14,17] Radiographs following insertion of nasogastric tube have helped in improving the diagnosis in such difficult cases.[17]

The management of CDH has shifted from emergency surgery to delayed surgery. This allows for stabilisation of patients with CDH associated with severe lung hypoplasia and PPHN. Sakai *et al,*[18] have shown worsening of cardiopulmonary function following early surgical repair. The period of preoperative stabilisation also leads to the avoidance of operative procedures in those with severe pulmonary hypoplasia, who might be expected to die. This subset of patients who have severe CDH is the focus of interest of all the current survival interventions. However, the optimal management for all CDH remains unknown because of the wide spectrum of the disease. But the most important factor that has improved postnatal care is the introduction of gentle ventilation like the high frequency oscillation ventilation (HFOV) which provides effective ventilation with protection against barotraumas.[19] The other therapies include the use of inhaled nitric oxide (iNO) and magnesium sulphate to counteract PPHN, administration of surfactant, corticosteroids *in utero* and novel therapies like the use of liquid ventilation with perfluorocarbons for patients under ECMO,[20] and lung transplantation.[21]

We favour the abdominal approach for the repair of CDH because visualisation of the defect is excellent, reduction of the herniated viscera is easier and associated abdominal pathology like the midgut malrotation in one of our patients can be more conveniently tackled. In an instance, difficult reduction of the herniated viscera via a thoracotomy necessitated widening of the diaphragmatic defect.[22] Also, in cases of increased intrabdominal pressure, reduction of viscera may be impossible because stretching and abdominal patching cannot be performed through a thoracotomy. Elsewhere, both laparoscopic and thoracoscopic techniques have been used to repair CDH. But in the newborn these techniques are associated with high incidence of technical and physiological complications and, therefore, their applications are more preferable in the older child.[23]

The overall survival in our series was 57%. This was achieved in the late presenting cases with less severe hypoplasia and is similar with the experience of other workers in similar circumstances.[4,5] In a poor resource setting like ours, only the group consisting of late presenting CDH with less severe pulmonary hypoplasia survives.[4-6] The severe forms of CDH presenting at birth are likely to die as in our experience.

In conclusion, the diagnosis of the classical CDH at birth is straightforward but this poses enormous challenges with the initial management in a poor resource setting like ours and until the standard of our neonatal intensive care facilities improves, we may continue to record this dismal outcome of management of the early presenting CDH as is in our experience. On the other hand, the main challenge with the late presenting CDH is diagnostic, and once the correct diagnosis is made and appropriate treatment offered, the prognosis is excellent. A misdiagnosis may be associated with a high morbidity and mortality in this otherwise good prognosis group. A high index of suspicion is required especially in

a patient with a recurrent history of difficulty in breathing, failure to thrive and indeterminate chest X-ray findings.

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