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Experience with the Management of Congenital Diaphragmatic Hernia at the University College Hospital, Ibadan

VO Adegboye*, SI Omokhodion+, O Ogunkunle+, MO Obajimi*+, AI Brimmo*, OA Adebo**

Summary

Adegboye VO,Omokhodion SI , Ogunkunle O , Obajimi MO, Brimmo AI, Adebo OA. Experience with the Management of Congenital Diaphragmatic Hernia at the University College Hospital, Ibadan. *Nigerian Journal of Paediatrics* **2002;29:40**. A retrospective review of 44 cases of congenital diaphragmatic hernia (CDH) treated at the Cardiothoracic Surgical Unit, University College Hospital (UCH), Ibadan, between August 1976 and May 1999, was carried out. Thirty (68.2 per cent) of the 44 were males and 14 (13.8 per cent) were females. The patients were analysed in three groups: 31 patients who underwent surgery were analysed as two groups: inborn (born within UCH), 10 patients, and outborn (born elsewhere), 21 patients; the third group consisted of 13 patients who were not operated upon. All the inborn patients and 18 (85.7 per cent) of the outborn had left-sided defects; the remaining three of the outborn had right-sided defects. Eleven (84.6 per cent) of those who didn't undergo surgery, had left-sided defects while the remaining 15.4 per cent had bilateral defects. The three groups had comparable gestational ages (inborn vs outborn, $p > 0.05$; inborn vs unoperated, $p > 0.05$; outborn vs unoperated, $p > 0.05$) and birth weights (inborn vs outborn, $p > 0.05$; inborn vs unoperated, $p > 0.05$; outborn vs unoperated, $p > 0.05$). Those who did not undergo surgery had lower Apgar scores and higher incidence of associated congenital malformations than the patients who underwent surgical repairs. The differences in the mean of 5 minute Apgar scores among the three groups were not statistically significant ($p > 0.05$). Age at surgery was higher in the outborn (mean 55.5 ± 42.2 hours) than the inborn (mean 9.2 ± 3.9 hours) patients. The difference between the mean age at surgery was statistically significant ($p < 0.05$). Majority of those requiring operation (24 patients, 77.4 per cent) had repair by thoracotomy, while the remaining seven (22.6 per cent) had repair by the abdominal approach. Most of the patients had primary repair while two required diaphragmatic patches with dacron. Sixty per cent of the inborn patients required post operative ventilation compared with only 23.8 per cent of the outborn. Survival was 71.4 per cent for the outborn and 40 per cent for the inborn. It is concluded that CDH is a disease with a range of severity. The higher survival rate among the more mature outborns suggest a natural selection of those with minimum respiratory impairment.

Keywords: Congenital, Diaphragmatic hernia.

Introduction

THE incidence of congenital diaphragmatic hernia (CDH) varies from 1 in 2,000 to 1 in 5,000 live births.[1,2] This incidence is greater in stillbirths and abortions.[3] There is no sexual, racial or geographical predilection, and most maternal factors show no consistent relationship.[3] It was generally considered a fatal disease in the early 20th century during which time, watchful waiting was the management approach. The encouraging report by Ladd and Gross[4] on the surgical correction of CDH in the neonate was a landmark in the management of the disease. Immediate postnatal repair of the hernia was previously the standard care.[5-7] However, several reports in the 1980s demonstrated that survival was not different when repair was delayed by a period of preoperative stabilization.[8-10] To further improve survival in a subset of the patients, extracorporeal membrane oxygenation (ECMO),[11,12] exogenous surfactant,[13] inhaled nitric oxide,[14,15] and various ventilation techniques[16,17] have been used in the

University College Hospital, Ibadan

Cardiothoracic Unit of the Department of Surgery
*Senior Lecturer
**Professor
Department of Paediatrics
+ Senior Lecturer
Department of Radiology
*+ Senior Lecturer

Correspondence : VO Adegboye

perioperative care of the infant with CDH. This communication reports a retrospective study of our experience with 31 patients surgically treated between August 1976 and May 1999.

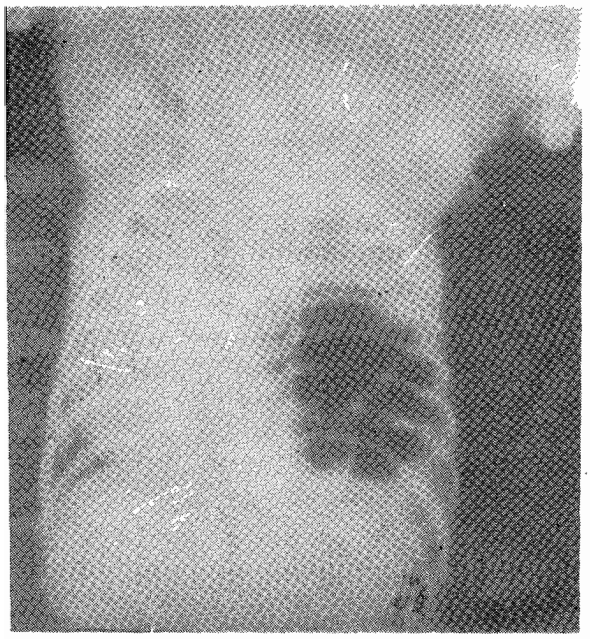

Fig. 1 (a) Preoperative posteroanterior (PA) view of a chest radiograph showing a left sided congenital diaphragmatic hernia (CDH) as a large tubular lucency in most of the left hemithorax.

Patients and Methods

The Unit's record of the cardiothoracic surgical service of the University College Hospital, Ibadan, indicated that 64 patients with CDH were referred to the Unit between August 1976 and May 1999. Retrospective analysis of the

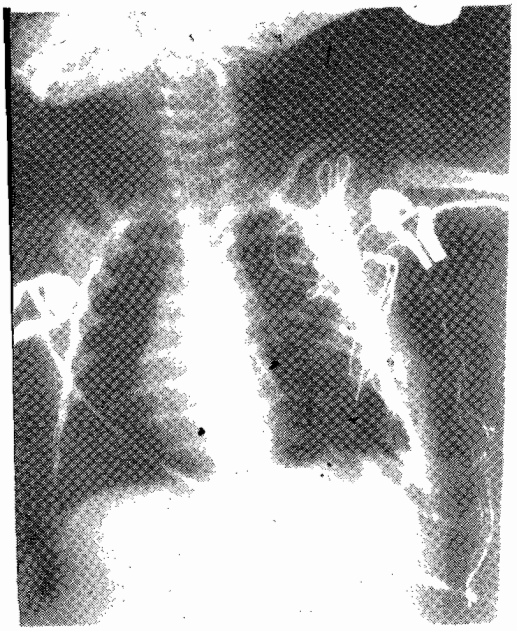

Fig. 1 (b) Postoperative PA view of the same patient.

Table I

Characteristics of 44 Patients with Congenital Diaphragmatic Hernia

	Number of Cases	*Mean Gestational Age (wks)**	*Mean Birthweights (kg)***	*Mean Apgar Scores*$^{+}$		*Mean Age at Surgery (hrs.)*$^{++}$
				1 min	*5 min*	
Operated						
Inborn						
	10	38.0± 2.5	3.1 ± 0.6	4.1 ± 2.6	7.2 ± 1.5	9.2 ± 3.9
		(34 – 42)	(2.2 – 4.0)	(0 – 8)	(5 – 9)	(4 – 17)
Outborn	21	38.8 ± 3.0	3.1 ± 0.4	5.9 ± 1.2	8.1 ± 1.2	55.5 ± 4.22
		(35 – 45)	(2.5 – 3.8)	(1 – 8)	(6 – 9)	(24 – 1300)
Unoperated	13	34.7 ± 4.4	2.9 ± 0.7	2.5 ± .1	5.7 ± 2	-
		(28 – 41)	(1.5 – 3.6)	(0 – 6)	(2 – 9)	

Means ± SD; Ranges in parentheses

* Inborn vs outborn t = -0.25, P > 0.05
Inborn vs unoperated t = 0.216, P > 0.05
Outborn vs unoperated t = 0.76, P>0.05

** Inborn vs outborn t = 0, P>0.05
Inborn vs unoperated t = 0.216, P>0.05
Outborn vs unoperated t = 0.24, P>0.05

$^{++}$ Inborn vs outborn t = 29, P<0.05

$^{+}$ P> 0.05

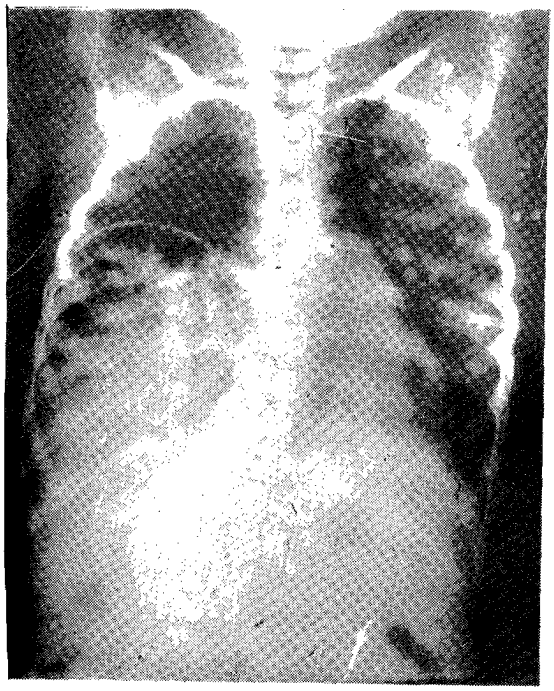

Fig. 2(a) Preoperative anteroposterior (AP) view of a chest radiograph showing the CDH as a non-homogenous opacity in the right lung base.

medical records were possible in 44 patients; incomplete information prevented adequate evaluation of 20 others. Information obtained included gestational ages, birth weights, Apgar scores at birth, ages at surgery and sources of referral. Other clinical information obtained included the mode of preoperative care, surgical approach in those who underwent repairs, and the outcome of management.

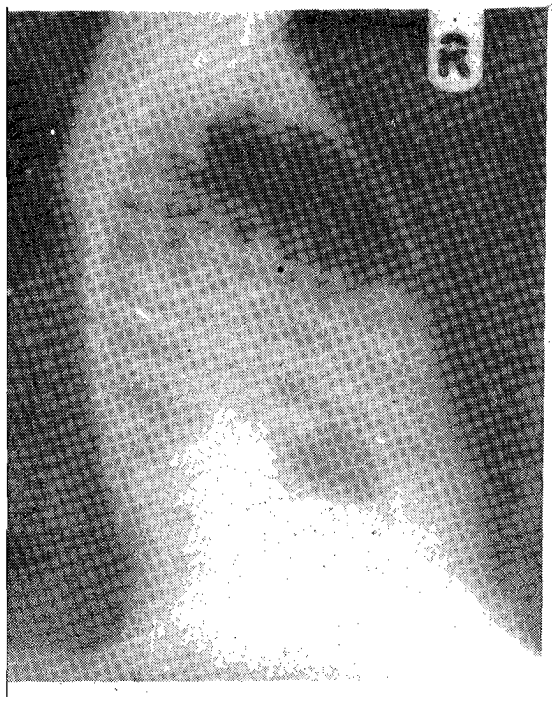

Fig 2 (b) Preoperative right lateral view of the same radiograph showing bowel loops in the thorax.

All referrals came through the department of paediatrics of the hospital. The patients were classified as inborn if delivered at the hospital, or outborn when delivered elsewhere. The management of CDH at the hospital consisted of early repair with postoperative support using conventional positive pressure ventilation.

Statistical analysis

Student's unpaired 't' test was used to compare mean gestational ages, birth weights, Apgar scores and ages at surgery among inborns and outborns who had surgery and those who did not. The differences between the groups were considered significant when $P < 0.05$.

Results

Thirty-nine (88.6 per cent) of the 44 had left sided defects (Fig 1a), three (6.8 per cent) had right sided (Fig 2a) and two (4.5 per cent) had bilateral defects. Thirty (68.2 per cent) of the patients were males and 14(31.8 per cent) females. Table I summarizes some of the clinical data of the patients. The 31 patients who underwent surgery, were grouped as inborn (10 patients) or outborn (21 patients) while the 13 who had no surgical intervention were included in the survival analysis.

Twenty-eight (90.3 per cent) of the 31 who had surgery, had left sided defects; this consisted of all inborn patients and 18 (85.4 per cent) of the outborn. The remaining three outborn (14.3 per cent) patients had right sided defects. Similarly, a majority (11 patients, 84.6 per cent) of those not operated upon, had left sided defects, and the remaining two (15.4 per cent) had bilateral defects. The three groups had comparable gestational ages and

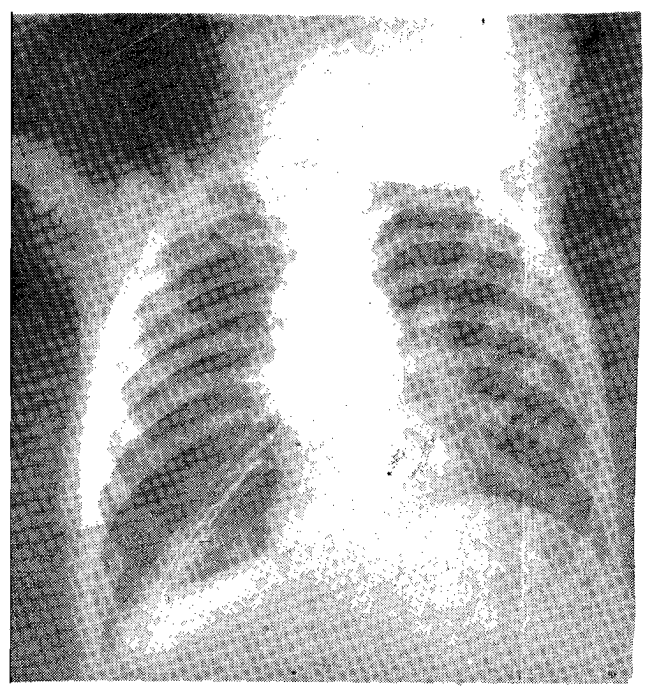

Fig 2 (c) Postoperative posteroanterior (PA) view of the same patient.

Table II

Congenital Diaphragmatic Hernia and Associated Malformations in 44 Patients

	Operated		*Not operated*
	Inborn (n=10)	*Outborn (n=21)*	
Diaphragmatic defects			
Mild/moderate	10	19	11
Severe (by size or bilateral defects)	-	2	2
Cardiac			
Truncus arteriosus	-	-	1
Tetralogy of Fallot	-	-	1
Ventricular septal defect	-	-	1
Genito-urinary			
Hydronephrosis	-	-	1
Renal cyst	-	-	1
Skeletal			
Hip dislocation	-	1	-
Anterior abdominal wall defect	1	2	-
Gastrointestinal tract			
Duodenal atresia	-	1	2
Imperforate anus	-	-	1
Volvulus / intussusception	1	1	1
Number of associated congenital malformations	2	5	9
Incidence of associated congenital malformations (%)	20	23.8	69.2

Table III

Operative Procedures and Outcome in 31 Cases of CDH

	Inborn n = 10		*Outborn n = 21*	
	Right	*Left*	*Right*	*Left*
Side of defect	-	10	3	18
Approach				
Thoracotomy	-	7	3	14
Laparotomy	-	3	-	4
Other procedures				
Abdominal wall stretching	4	-	-	9
Malrotation procedure	1	-	-	-
Colostomy	-	-	-	1
Diaphragmatic patch	-	-	-	2
Survival	4 (40%)		15 (71.4%)	
Mortality	6 (60%)		6 (28.6%)	

birth weights (P>0.05; Table I), and although the Apgar scores at one and five minutes were lower in the unoperated, the differences in the means of the five minute Apgar scores among the three groups were not significant (P>0.05). The mean age at surgery was significantly higher for the outborn than the inborn (P<0.05; Table I).

The incidence and pattern of associated congenital malformations were comparable for the operated groups but lower than in the unoperated as summarized in Table II. Thoracotomy was the commonest operative approach (24 patients, 77.4 per cent) but in seven patients (22.6 per cent), repair was through the abdomen because of associated abdominal symptoms. Malrotation procedures were rarely used (one patient, 3.2 per cent; Table III). Diaphragmatic patches with dacron were used in two patients because it was impossible to achieve a good repair without tension.

Majority of the outborn patients (16 of 21, 76.2 per cent) did not require mechanical ventilatory support while most (six of 10) of the inborn needed such support in the early postoperative period. Postoperative complications included pneumothorax from barotraumas; this was related to the treatment of delayed re-expansion of ipsilateral lung in two inborn patients, and empyema

thoracis with septicaemia in three patients (one inborn and two outborn).

Four of the inborn group (40 per cent) and 15 (71.5 per cent) from the outborn group survived and were discharged to follow up. The overall survival rate was 64.5 per cent. The period of follow up ranged from six months to five years. There was no long-term follow up in this series. The commonest complaints at follow-up were lack of weight gain, recurrent respiratory infections and easy fatigability in three of the inborn and eight of the outborn. The three inborn patients who had recurrent respiratory infections had mechanical ventilation for at least 48 hours, while the eight outborn with the same complication presented with bronchopneumonia. Follow-up chest radiographs were essentially normal among those followed up. No lung function tests were done.

Discussion

This review shows that congenital diaphragmatic hernia (CDH) is associated with a clinical spectrum of respiratory problems. The stage of pulmonary development at which visceral herniation occurs can explain the clinical spectrum of respiratory problems preoperatively in this disease.[18-20] This hypothesis was used to divide patients with CDH into four groups.[21] The first group experiences visceral herniation early in the course of bronchial branching resulting in bilateral pulmonary hypoplasia with uniformly fatal outcome. Herniation that occurs during the stage of distal bronchial branching results in unilateral hypoplasia. In this second group, there is a delicate balance between pulmonary vascular and ductal resistances. In the third and favourable group, herniation occurs late in gestation and the respiratory distress follows air swallowing at delivery. In this situation, there is compression of an otherwise adequate lung. In the fourth group, herniation is a postnatal event; this group also has excellent prognosis.

All the inborn patients who underwent surgery, most of the unoperated and the minority in the outborn group in the present study were symptomatic early. This partly explains the differences in mortality among the three groups. The subset of patients who are symptomatic early in life is the focus of interest of all current survival interventions.[11-17] These were described as having "critical" neonatal diaphragmatic hernia by Reynolds *et al.*[22]

An observation from our policy of early surgery was that the patients who survived surgery were older at the time of surgery than non-survivors and their survival probably indicates well developed lungs bilaterally. The outborn survivors of surgery most likely represent those whose herniation occurred late in gestation or postnatally; hence the increased survival in spite of minimal postoperative ventilatory support. Most of the non-survivors had an initial early postoperative stable respiratory and haemodynamic status which was most often followed by a deterioration in their cardio-respiratory status. Two previous series from our unit reported eight patients with CDH seen between 1970 and 1973, and another case simulating pectus carinatum (1975) in a two-year old boy.[23,24] The mortality among six of the eight patients who had surgical repair was 50 per cent. The remaining two patients died pre-operatively, of associated congenital malformations. The surgical survivors were older than one week at surgery. The operative mortality occurred among those that were symptomatic immediately at birth. This pattern seems to corroborate the finding in the present study that the operative survivors in our catchment area have less severe pulmonary hypoplasia and fewer associated congenital malformations.

The notion that CDH might not be a surgical emergency was first proposed by Sakai *et al*[25] and later confirmed by Nakayama *et al.*[26] They showed that immediate surgical repair of CDH was followed by a decrease in respiratory compliance. This alteration is currently accepted as caused by the usual postoperative alterations in water and sodium balance.[27] The main essence of delayed repair is the stabilization of the cardio-respiratory and haemodynamics status of the patients.[17,22,27-29] However, what constitutes an appropriate period of stabilization remains unclear.[28,29] Nio *et al*[29] carried out a prospective randomized trial of delayed versus immediate repair in patients who became symptomatic within the first 12 hours of life. Their results show neither an advantage for emergency surgery nor a clear advantage for a long delay in a clinically stable infant. Infants who are clinically stable and can be operated on, under conditions of satisfactory supportive care may undergo early repair. Those who are unstable early in their course or have evidence of persistent intermittent pulmonary hypertension can be placed on ECMO in the perioperative period before repair. A study comparable with ours showed a survival of 100 per cent among minimally distressed babies and of 54 per cent among the "critical" CDH cases.[30] Corresponding values in our study were 71.4 per cent and 40 per cent, respectively. Improvement in survival in both groups is achievable with improved early cardio-respiratory resuscitation. Among the "critical" CDH, survival rates of between 75 and 81 per cent have been achieved with delayed surgical repair, early postnatal high frequency oscillatory ventilation and in selected patients, ECMO.[17,31] Of prognostic importance is the complexity of associated congenital anomalies of other systems.[7,32] The presence of associated cardiac defects and an absence of the diaphragm have been associated with higher mortality.[32,33] A higher incidence of associated complex congenital anomalies in the group that did not undergo surgery in this series, agrees with this, although no definite pattern of these congenital malformation can be drawn.

While we, like other groups,[34,35] used the thoracic approach in the majority of our patients, the choice between subcostal abdominal and thoracic approach has not been a major outcome determinant in the literature.[7] The two patients who had patch repair in our series died

of respiratory failure. Technically and pathophysiologically, the need to use a patch portends a large defect, severe hernia and higher mortality than primary repair.[7] Tissue oedema and the desire to minimize bleeding by avoiding a dissection of the posterior rim of the diaphragm may occasionally make primary repair difficult.[36] In such instances, there may be some benefit in attempting to recreate a dome shape to the diaphragm rather than a tight closure.[37] The fact that all the mortalities in those operated upon, occurred with left sided defects suggest that the left sided hernia was more frequently associated with severe pulmonary dysfunction than right sided defects although pooled data[7] showed no difference in mortality rates of babies with right or left sided defects.

Our follow up finding of recurrent respiratory infections has not been a common occurrence in the literature. However, many workers[20,1] have reported some respiratory abnormalities such as decreases in total lung capacity and vital capacity, as well as reduced residual volume in preoperative patients.[38] In a long-term postoperative follow-up of some patients, their weights and heights were average for age.[39] Loss to follow up has prevented meaningful long-term postoperative data. However, the few patients we reviewed were comparable to their peers in weight, height and in exercise tolerance.

We conclude that CDH is a disease with a range of severity which depends on the stage of airway development at which herniation occur. The higher survival rate among the more mature outborns suggest natural selection of those with minimal respiratory impairment. Those with more severe pulmonary pathology diagnosed very early in life (i.e. <12 hours) in our institution will require improved and aggressive preoperative cardiopulmonary stabilization followed by appropriate postoperative care.

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