Congenital diaphragmatic hernia: A 4-year experience in a single centre

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ABSTRACT

Background: This study aimed to evaluate congenital diaphragmatic hernia (CDH) patients in our department during a 4-year period. **Patients and Methods:** A retrospective study of 10 cases of CDH patients managed in the Neonatology and Pediatric Surgery Units of Goztepe Teaching Hospital from 2000 to 2004. **Results:** The mean birth weight of the patients was 2,600 g and the mean gestational age was 37.6 weeks. The mean admission time for the patients was 10.4 h. The mortality rate was 50%, comprising two deaths before operation and one during operation. The surviving infants followed for 1 year are free of any symptoms and complications. **Conclusion:** Conventional ventilation is an other treatment modality, especially in order to gain time before surgical operation for newborns with CDH.

Key words: Children, congenital diaphragmatic hernia, management

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INTRODUCTION

Congenital diaphragmatic hernia (CDH), one of the most common major congenital anomalies worldwide, occurs in about 1 per 2,000 to 1 per 4,000 live births.[1] Mortality from CDH has remained high, ranging from 20% to 60%,[2] despite advances in medicine. Hidden mortality arising from *in utero* deaths, termination of pregnancy and severely affected infants who die before transfer to a centre continues to underscore true outcomes.[3,4]

The present management of CDH employs sophisticated equipments and technologies. This in no little measure has resulted in higher survival rates in infants suffering from CDH compared with earlier results.[5] Sophisticated equipments and technologies like extracorporeal membrane oxygenation (ECMO) and high-frequency oscillatory ventilation (HFOV) are lacking in many centres. The present study evaluates the outcome of CDH patients managed in the absence of sophisticated modern armentaria in a resource-limited setting.

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PATIENTS AND METHODS

This was a retrospective study of newborn infants with CDH in the Neonatology and Pediatric Surgery Units of Goztepe Teaching Hospital, Turkey, from 2000 to 2004.

RESULTS

A total of 10 cases were admitted during a 4-year period. The demographic and clinical features of cases are identified in Table 1. The mean birth weight of the patients was 2,600 g and the mean gestational age was 37.6 weeks. The mean admission time for the patients was 10.4 h.

The patients presented with respiratory distress to the intensive care. Apart from one that was diagnosed antenatally, the rest were diagnosed by chest X-rays after clinical suspicion based on a history of severe respiratory difficulty since birth.

After diagnosis, the infants were put on a ventilator with an intermittent mandatory rate of 60/min, to obtain respiratory alkalosis of pH 7.45–7.50. Two infants were given surfactant to overcome hypoxemia. Majority of the infants underwent repair within 3 days of age. Direct closure of the defect was the preferred surgical repair.

The mortality rate was 50%, comprising two deaths before operation and one during operation; this latter patient who died during the operation had diaphragmatic agenesis. Two other infants died during the postoperation period due to sepsis.

Five infants were discharged within 10 days of surgical repair. They have been followed-up for 1 year without any residual symptom or complication.

Table 1: Demographic features of the cases
Range/n Mean

	Male/ female (n)	B. weight (g)	G. age (weeks)	Antenatal diagnosis	Admission h	Operation h	Surfactant	Respiratory acidosis	Exitus	Agenesis
Range	3/7	1,500–3,700	34–40	2	1–72	24–72	2	8	5	1
Mean		2,600	37.6		10.4	51				

DISCUSSION

The aim of treatment of CDH is to overcome pulmonory hypertension, hypoxemia and persistent foetal circulation associated with this condition. The present management of CDH in many developed centres employs the use of ECMO, HFOV, inhaled nitric oxide (NO), surfactant and delayed surgery,[4] although ECMO and NO have not proved to have a benefit or change the outcome.[6]

Our centre, like many others, lacks these sophisticated equipment and technologies,[7] except for conventional ventilation, which was introduced only in 2000. The first principle of treating these patients is to overcome hypoxia and stabilise them, preferrably in respiratory alkolosis, until surgery is carried out. Studies showed that a delayed operation, performed after 48 h of birth, has a better outcome than if performed earlier.[8] This was also our experience in the present sudy, where the patients were stabilised for an average period of 51 h before the procedure.

The mortality rate in the present study was 50%, which is in agreement with other reports.[3] The mortality rate in CDH has not changed over the past years.[9] Three deaths were due to unstable pathophysiology, which might be due to severe pulmonary hypoplasia and diaphragmatic agenesis. This was not confirmed because we did not perform autopsies. Although the rate of associated anomaly is high in the literature,[4] surviving babies in the present report were without anomalies. There was no morbidity related to the respiratory system. *In utero* deaths, termination of pregnancy and severely affected infants who die before transfer to a center continue to 'hide' the true incidence and outcome of CHD.[4,5]

Because only two patients were diagnosed antenataly, true incidence and outcome of CDH newborns in Turkey is not known. Thus, we think that in Turkey this hidden mortality is high and that data are missing. We strongly suspect that mortality from CHD is very high in Istanbul, and most of the physicians take for granted that death in these children is inevitable. Because there is lack of randomised controlled trials to provide evidence-based management guidelines, we hope to establish a CDH study group with obstetricians, paediatric surgeons and neonatologists all over Turkey and to establish centres for managing CDH patients.[10]

Before the construction of an NICU in our hospital, the CDH patients were lost without proper diagnosis. We suppose that this is the outlook in most teching hospitals lacking an NICU to take care of CDH babies. To us, the outcome of mangement of CHD in this report was good, considering that ours was a newly constitued intensive care unit. The implication is that these patients could survive without sophisticated gadgets.

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