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Original Article

Clinical profile of congenital diaphragmatic hernia and their short-term outcome in a tertiary care neonatal unit: A retrospective study

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ABSTRACT

Background: Recent developments in the antenatal diagnosis, surgical techniques, and neonatal intensive care had widely increased survival rates in neonates with congenital diaphragmatic hernia (CDH) in the western world. In developing countries, however, high mortality in neonates with CDH still continues to be a challenge. **Objective:** The aim of this study is to study the clinical profile of neonates with CDH and to analyze the various factors affecting mortality. **Materials and Methods:** In this retrospective study, 148 babies with a diagnosis of diaphragmatic hernia admitted to a tertiary care neonatal unit in South India, from the year 2010 to 2015, were reviewed. **Results:** The total survival rate was 58.1%, and the operative survival rate was 85.1%. Prenatal diagnosis was made in only 7 cases, and of these, 5 (71.42%) survived. Higher mortality was associated with age at admission <24 h, low Apgar score, early onset of respiratory distress, right-sided CDH, presence of persistent pulmonary hypertension of newborn, and high FiO_2 requirement at the time of admission, during stabilization and surgery ($p<0.01$). Significantly higher mortality was also observed among babies who required positive-pressure ventilation during transport, required inotropes during hospital course ($p<0.01$), and had low PaO_2, high PCO_2, and high oxygenation index ($p<0.01$). **Conclusion:** Babies admitted to hospital within 24 h of age and who manifested early had a poor outcome indicating severe disease. Antenatal diagnosis of this condition should improve to prevent delay in stabilization and poor outcome.

Key words: *Congenital diaphragmatic hernia, Outcome, Pulmonary hypertension*

Congenital diaphragmatic hernia (CDH) remains one of the most difficult problems of the perinatology and neonatal surgery. Western literature showing 90% of survival rate is not reflected in our population. Initial stabilization of the neonate with advanced ventilatory techniques and optimal management of associated pulmonary hypertension by the neonatologist has significantly lowered the operative mortality in developed countries. However, the mortality rate in developing countries is still 40-50% [1]. Several predictors of mortality have been suggested, including polyhydramnios, early prenatal diagnosis, intrathoracic stomach, intrathoracic liver, presence of associated anomalies, right-sided hernia, and low Apgar scores [2].

Hence, this study was carried out to analyze the clinical profile of neonates with CDH and determines the risk factors associated with mortality in these neonates.

MATERIALS AND METHODS

This observational retrospective study was conducted after getting the Institutional Ethics Committees approval. Medical records of 148 neonates who were admitted to medical and surgical newborn units of a tertiary care neonatal center in South India and diagnosed to have CDH between 2010 and 2015 were reviewed. They were analyzed for a place of referral, mode of transport, age at diagnosis (Antenatal age [gestational age] and postnatal age), presenting features, associated anomalies, peroperative findings, short-term outcomes, survival/death and complications, and duration of hospital stay.

Statistical Analysis

Data were analyzed using SPSS version 20. All categorical data were summarized using frequency and percentages, and all continuous data were described using mean and standard deviation or median and interquartile range based on the distribution. To check the association of different clinical parameters (risk factors) on the survival of the CDH baby, independent sample t-test or Mann-Whitney U-test was applied for the continuous measurements after checking normality assumption, and Chi-square test or Fisher's exact test was applied for the categorical observations. p value was considered statistically significant at 5% level of significance for all comparisons.

RESULTS

Case records of 148 patients with a diagnosis CDH were reviewed. The mean age at admission, birth weight, sex, and

mode of delivery among survivors and non-survivors is shown in Table 1. The median age at admission to the unit was 2 days. 94 (63.51%) babies underwent hernia repair at a median of 2 days after admission. Operative survival rate was 85.1% (80/94). In this study, 69% (102/148) of the babies were referred from government hospitals. Only 7 (4.7%) babies out of the 148 were antenatally diagnosed and 5 (71.4%) of these survived.

Higher mortality was associated with age at admission <24 h, low Apgar score, early onset of respiratory distress, presence of persistent pulmonary hypertension of newborn (PPHN), and high FiO_2 requirement at the time of admission, during stabilization and surgery ($p<0.01$). Significantly higher mortality was also observed among babies who required positive-pressure ventilation during transport, required inotropes during hospital course ($p<0.01$), low PaO_2 (46.29±10.05) mm Hg, high PCO_2 (47.06±6.78) mm Hg, and high oxygenation index (46.56±8.97) (Tables 1 and 2). 9 (6%) babies had right-sided CDH, out of which seven babies underwent surgery and two of them survived. 53 (62.35%) out of 85 babies who presented before 24 h of age expired, and they contributed to 85% of non-survivors. Only 9 (14%) of these babies who were admitted after 24 h died. The predominant clinical features were respiratory distress and decreased breath sounds on the affected side.

The most common associated congenital anomalies found on surgical exploration were malrotation and pulmonary hypoplasia. 94 (63.51%) babies underwent surgery, and 3.7% developed post-operative complications such as wound infection. Median post-operative days after which feed were started were four. 54 (36.48%) died without surgery because of poor respiratory status on arrival.

Table 1: Baseline characteristics: Comparison between survivors and non-survivors

Variables	Survivor, *n* (%) *n*=86	Non-survivor *n* (%), *n*=62	p
Gestational age*	38.19 (1.41)	37.43 (2.18)	0.017
Birth weight*	2752.73 (327.17)	2611.45 (385.8)	0.011
Sex			
Males	62 (72.09)	44 (70.97)	0.881
Females	24 (27.91)	18 (29.03)	
Age at admission			
<24 h	32 (37.21)	53 (85.48)	<0.01
2-7 days	40 (46.51)	9 (14.52)	
>8 days	14 (16.28)	0 (0)	
Apgar*			
1 min	7.14 (0.77)	5.82 (1.6)	<0.01
5 min	8.35 (0.68)	7.16 (1.31)	<0.01
Onset of symptoms			
<24 h	47 (54.6)	57 (91.94)	
>24 h	39 (45.35)	5 (8.06)	
Mode of delivery			
Vaginal	36 (41.86)	20 (32.26)	
LSCS	50 (58.14)	42 (67.74)	
Antenatal diagnosis			
Yes	5 (71.42)	2 (28.57)	
No	81 (57.44)	60 (42.55)	
Mode of transport			
Ambulance	63 (73.26)	45 (72.58)	0.927
Others	23 (26.74)	17 (27.42)	
Respiratory support during transport			
Hood O_2	74 (86.05)	18 (29.03)	<0.001
BTV	5 (5.81)	30 (48.39)	
No support	7 (8.14)	14 (22.58)	

***Mean (SD), LSCS: Lower segment Cesarean section, BTV : Bag and tube ventilation, p<0.05**

DISCUSSION

CDH symptomatic at birth is traditionally considered a surgical emergency, and correction of the defect is carried out as early as possible [3]. However, poor pulmonary functions contribute to a poor outcome, and hence, the need for prior stabilization with ventilator support and management of pulmonary hypertension [1]. The most common presentation in this study was respiratory distress and decreased breath sounds on the affected side. 57% of the babies had presented within 24 h life. The previous study by Jain et al. documented respiratory distress and decreased breath sound on the affected side as the most common presentation, whereas only 15% of them presented within 24 h of life.

The mortality was 62% in babies who presented early and 14% in those who presented after 24 h. This could be due to severe lung hypoplasia and associated severe PPHN leading to early manifestations. As per the recent studies, the overall survival rate ranged from 21% to 83% [4]. In this study, the overall survival rate was 58.1%, and the operative survival rate was 85.1%. The higher survival in babies who were operated highlights the need for stabilization, control of PPHN, and early surgery. Krishna and Mitra studied 7 children (more than 48 h of age) and found that 57% survived [9]. Those babies, who presented within 24 h, with low Apgar score, required PPV during transport, high FiO_2 requirement at admission and at the time of surgery, high oxygenation index, requirement of inotropes during hospital stay, and presence of PPHN had a poor outcome.

Ruza et al. reported that babies requiring low FiO_2 and achieving higher PaO_2 had better survival [4,6]. In this study, high $PaCO_2$ (>45 mm Hg), low PaO_2 (<40 mm Hg), and high pulmonary pressure were significantly associated with higher mortality. Babies with higher PCO_2 (>60 mm Hg) indicate poor pulmonary condition mostly with pulmonary hypoplasia [7]. Availability of high-frequency oscillatory ventilation, inhaled nitric oxide, and extracorporeal membrane oxygenation could have yielded a better outcome. Pre-operative management of pulmonary hypertension and hypoplasia is critical factors which determine the ultimate outcome [8]. In this study, only 7 (4.7%) babies out of the 148 were diagnosed antenatally, and 5 (71.4%) of these survived. This better survival could probably be due to the use of appropriate resuscitation methods soon after birth,

Table 2: Clinical parameters: Comparison between survivors and non-survivors

Variables	Survivors mean (SD) (*n*=86)	Non-survivor mean (SD) (*n*=62)	p[£]
FiO_2 at the time of admission	41.3 (19.27)	98.31 (7.84)	<0.001
FiO_2 during hospital course	44.02 (20)	93.87 (15.9)	<0.001
SpO_2	93.58 (4.01)	80.66 (11.29)	<0.001
PaO_2	58.29 (9.73)	46.29 (10.05)	<0.001
PCO_2	38.47 (5.56)	47.06 (6.78)	<0.001
Pulmonary pressure	16.66 (8.31)	43.11 (15.38)	<0.001
Oxygenation index	16.44 (3.02)	46.56 (8.97)	<0.001
FiO_2 requirement at the time of surgery	36.56 (12.31)	99.19 (3.75)	<0.001
Inotropes			
Yes	35 (40.70)	62 (100)	<0.001
No	51 (59.30)	0 (0)	

[£]t-test, p<0.05

prior stabilization of babies before transport and early transport. It reflects that skill of antenatal diagnosis of CDH during fetal ultrasound examination should be upgraded for the better management of these babies.

CONCLUSION

This study revealed that babies presented within 24 h had severe disease (lung hypoplasia) and those who required high FiO_2, PPV, high oxygenation index, and requirement of inotropes support had severe PPHN, and hence, had a poor outcome. Babies who were fit to undergo surgery had higher survival rate. Follow-up those babies who survived after surgery should be carried out in future to analyze the morbidity and any long-term complications.

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