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Original Research Article

Congenital Diaphragmatic Hernia Treatment and Outcome Analysis: A 7-Years Experience in a Northern Part of Karnataka in Single Center

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ABSTRACT

Background: This study aimed to retrospectively analyze of Treatment and Outcome in congenital diaphragmatic hernia (CDH) patients in our department during a 7 year period.
Patients and Methods: A retrospective study of of all CDH infants managed in our Neonatal and Pediatric Surgery department between July 2009 and July 2016. Prenatal and maternal as well as perinatal and neonatal data were collected, including outcome parameters.
Results: Total 108babies were studied. The mean birth weight of the patients was 2,200 g and the mean gestational age was 36 weeks. The mean admission time for the patients was 72 hours. Major congenital anomaly was present in 11%. The mortality rate was 25%. Prenatal diagnosis babies, delay in diagnosis due to late referral, earlier gestational age at birth, lower birth weight, who required pre operative ventilator support and major congenital anomalies mainly cardiac anomalies were associated with high mortality rate.
Conclusion: Our data suggest a higher survival rate. We speculate that today's cases of congenital diaphragmatic hernia are probably milder than in the past due to earlier and more detailed prenatal diagnosis and subsequent termination of pregnancies for the more severe forms of the disorder.

Key words: Congenital diaphragmatic hernia, prenatal diagnosis, congenital anomalies.

INTRODUCTION

Congenital diaphragmatic hernia (CDH), one of the most common and serious congenital anomalies in the neonatal intensive care unit (NICU), occur in approximately 1 of every 2500 live-born neonates. [1] A mortality rate has remained high ranging from 20-60% despite advances in medicine. The prognosis of CDH is related to pulmonary hypoplasia as well as primary abnormality of airway branching, combined with the coexistence of other major malformations. [2] The epidemiology and outcomes of CDH are well described in developed countries, [2] but few data exist for developing countries, particularly with respect to risk factors and outcomes such as morbidity and mortality. New modes of preoperative treatment have led to an incremental increase in survival rate. The present management of CDH employs sophisticated equipments and technologies. This sophisticated equipments and technologies like extracorporeal membrane oxygenation (ECMO) and high-frequency oscillatory ventilation (HFOV) are lacking in many centres. The current study retrospectively reviewed our institution's experience in the management and surgical outcome of CDH over the last 7 years.

MATERIALS AND METHODS

This study was conducted at the SDM Hospital Sattur, Dharwad. This hospital is a tertiary referral centre with neonatal surgery services. Detailed information was obtained from review of maternal and infant medical records of 108 CDH patients, admitted to the NICU between July 2009 and July 2016. The objectives of this study were to document the true incidence, management, and outcomes. Children who were diagnosed over the age of 1 month and Infants with diaphragmatic eventration were excluded from the study. Our institute adopted a standardized approach to newborn infants with CDH. This strategy includes planned delivery at term or near term, immediate intubation if requires, nasogastric tube (active suction), sedation and gentle ventilation strategy. Surgical repair was delayed as required until the infant was hemodynamically stable. The main data analyzed included demographics, age at presentation, birth weight, sex, Apgar scores, resuscitation at delivery, inborn or outborn, clinical features, timing of detection (prenatal, postnatal), presence of associated malformations, preoperative respiratory management, timing of surgical repair, side of defect(left or right), site and size of the diaphragmatic defect, condition of lung (hypoplasia or normal) , description of the abdominal organ herniation, operative procedures carried out, acute postoperative complications, profiles of postoperative respiratory care, durations of mechanical ventilation, the outcome of treatment, follow-up, duration of follow up, and hospital stay. The study was approved by the ethics committee.

RESULTS

A total of 108 patients were enrolled in the study. 96 patients underwent diaphragmatic repair. The demographic and clinical features of cases are given in Table - 1. The mean birth weight of the patients was 2,200 g and the mean gestational age was 36 weeks. The mean admission time for the patients was 72hrs.Onlyeighteen (17%) cases were diagnosed prenatally. The ultrasound intrauterine diagnosis of CDH was earlier in 24 weeks. Prenatally diagnosed infants were delivered in our center. Postnatal diagnosis occurred in 83%(n-96) cases. The patients presented with respiratory distress to the NICU. Apart from eighteen there were diagnosed antenatal, the rest were diagnosed by chest X-rays after clinical suspicion based on a history of severe respiratory difficulty since birth. Mean gestational age at diagnosis 3 days. 33% (n-36) babies were inborn and 67% (n-72) referred babies from outside, 58% (n-63) babies were male and 42% (n-35) were female. Eighteen (17%) were born preterm (less than 37 weeks of gestation) and 36 (33%) were of low birth weight (less than 2500 g). The gestational age at birth and the birth weight were both associated strongly with survival rates. Major congenital anomaly was present in 11 %(n-12) cases and minor congenital anomalies being present in 16% (n-48) cases. These anomalies included significant cardiovascular anomalies (*n-15*), genitourinary anomalies (*n*-12), musculoskeletal anomalies (*n-9*), gastrointestinal anomalies (*n-9*), and dysmorphic features (n-*6*). The presence of an additional major anomaly was associated with a poor survival rate. Majority of the infants underwent repair within 6 days of age. In preoperative respiratory management 31 % of patients required preoperative mechanical ventilation and stabilisation, mean stabilisation period was 72 hrs.Total 96 infants underwent surgical repair of CDH, mean age at surgery 96 hrs of after birth, 94%(n-90) were left side defect and only 6%(n-6) were right side, Mean size of diaphragmatic defect was 4cm, 50 % of lung have hypoplasia at time of surgery, 80% having stomach as content followed by 70% small bowel, 60% have colon, and 20% of having spleen as content of hernia, all defect closed with non absorbable suture material. 61% (n-66) patients required postoperative mechanical

ventilation support. Mean duration of post operative mechanical ventilation was 3 days. Post operatively total parenteral nutrition given to all patients till they tolerate enteric feed. Eighteen patients developed pneumonia improved with treatment. Nine patients developed superficial wound infection improved with conservative treatment. The mortality rate was 25%, comprising four deaths before operation and five other infants died during the postoperative period. Eighteen one infants were discharged within 15 days of surgical repair. The total survival rate of CDH was 75% (27/108). Twelve of the patients did not undergo surgery, and all died. The overall operative mortality was 16% (15/96). The majority of deaths occurred within the first 7 days of life. Factors like [Table -2] prenatal diagnosis babies, delay in diagnosis due to late referral, earlier gestational age at birth, lower birth weight, who required pre operative ventilator support and major congenital anomalies mainly cardiac anomalis were associated with high mortality rate. All the survivors are alive and well, with weight and height appropriate for their ages over a follow-up period of 3 months-7 years. There was five recurrence of hernia among the survivors which was repaired.

Table 1. Characteristics of infants with CDH

Characteristic	*n*(%)
Gender	
Male	63(58)
Female	36(42)
Gestation age at birth	
Less than 37 weeks	18(17)
more than 37 weeks	90(83)
Birth Wt	
Less than 2500gms	36(33)
More than 2500gms	84(67)
Side of hernia	
Left	90(94)
Right	6(6)
Diagnosis	
Prenatal	18(17)
Postnatal	90(83)
Hospital of birth	
Inborn	36(33)
Outborn	72(67)
Other congenital anomaly	
Major	12(11)
Minor	18(16)

Table 2. Predictors of factors in infants with CDH

Characteristic	Survivors (n)	Nonsurvivors (n)
Prenatal diagnosis	9(27)	9(9)
Other major anomaly	3(27)	3(9)
Preoperative mechanical ventilator support	18(27)	15(9)
Gestational age at birth Less than 37 weeks	6(27)	12(9)
Birth weight Less than 2500gms	21(27)	15(9)

DISCUSSION

Congenital diaphragmatic hernia is one of the most common congenital disorders, making it a common anomaly seen by a pediatric surgeon. There is no gender, racial or geographical predilection. 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. [3] The prenatal detection rate for CDH varies enormously in published studies, from 10% to 79%. [4] Studies of cases with a prenatal diagnosis frequently report mortality rates of 75%, [5,6] which is similar to our study. The proportion of infants with coexisting major anomalies is which reported rates of 37% to 47%. [7,8] Most other studies [2,9] found associated major anomalies to be important predictors of mortality rates. In our study incidence of coexisting major anomalies is low, but like other studies it is associated with high mortality rates.

The aim of treatment of CDH is to overcome pulmonary 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. [3] CDH is no longer believed to require immediate surgery, since the primary problem after birth is not the herniation of abdominal viscera into the chest but rather severe pulmonary hypoplasia associated with pulmonary hypertension. [10] As a result, most centers practice delayed surgical repair of CDH following a period of preoperative stabilization. [11, 12] Our centre also following the same. Our centre, like many others, lacks these sophisticated equipment

and technologies like ECMO, HFOV, inhaled nitric oxide (NO), except for conventional ventilation. The first principle of treating these patients is to overcome hypoxia and stabilize them, preferably in respiratory alkalosis, 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. [12,13]

Our overall and the post-surgery mortality compared to that reported in the literature. [1,14] The mortality rate in the present study was 25%, which is in agreement with other reports. [15] Prenatal mortality (known also as "hidden mortality") should also be mentioned since spontaneous abortions are common in fetuses with diaphragmatic hernia, but its prevalence was not available for the present study. The overall survive rate of CDH among patients in NICU ranges between 21% and 83%. [4,16] In our study, we found a total survive rate of CDH in newborns to be 75%. The survive rate of CDH in NICU appears, therefore, to be similar in comparison to recent studies in high-risk CDH patients, [16,17] but to be lower in comparison to some other studies in isolated CDH patients. [18,19] Low gestational age at birth, low birth weight, [18,19] to be associated with higher mortality rates in CDH, our finding also similar to these studies. Associated major congenital anomalies mainly cardiac are considered to be poor prognostic factors by many authors, [7,8] we found similar comparison in our study.Studies of cases with a prenatal diagnosis frequently report mortality rates of 75% [5,6] similar to our study.

Today we probably deal with milder forms of CDH, due to earlier and more detailed prenatal diagnosis and subsequent termination of pregnancies for the more severe forms or severe forms not survive till they reach hospital. True incidence and outcome of CDH newborns in our setup is not known. Thus, we think that in our setup this hidden mortality is high and that data are missing. We strongly suspect that mortality from CHD is very high, 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 state and to establish centres for managing CDH patients.

CONCLUSIONS

CDH presents with a wide spectrum of disease that can make it difficult to accurately predict outcome. CDH is a challenging medical situation for which a prompt postdelivery multidisciplinary treatment approach should be taken. The overall mortality rate for this condition remains high, despite increased prenatal detection, transfer to tertiary institutions for delivery, and advances in neonatal care, We summarized data from the last 3 years of infants with CDH referred to a our institute, study to determine survive rates, risk factors, and outcomes of CDH in a NICU. To us, the outcome of management of CHD in this report was good, considering that ours was a newly constituted neonatal intensive care unit. The implication is that these patients could survive without sophisticated gadgets and technologies.

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