IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 24, Issue 2 Ser. 1 (February. 2025), PP 51-55
www.iosrjournals.org

Twenty Years Experience With Clinical Outcome Of Congenital Diaphragmatic Hernia: A Single Centre Study

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Abstract
Purpose: *To analyze twenty years of experience with Congenital Diaphragmatic Hernia (CDH) at a single center.*
Methods: *This study included 81 CDH cases admitted to the NICU and Pediatric ward from 2004 to 2023. Clinical records, operative findings, and outcomes were prospectively recorded in an EXCEL database and analyzed retrospectively. Patients were divided into four subgroups based on age at hospital presentation: Group A (inborn < 24 hours), Group B (outborn admitted within 24 hours), Group C (1-30 days), and Group D (>30 days). Outcomes at one year post-operation, including deaths and cases of leaving against medical advice, were recorded.*
Results: *CDH occurred on the left side in 69.13% of cases, right side in 29.6%, and bilaterally in 1.23%. Uncontrolled persistent pulmonary hypertension was present in 71.6% of deaths. All deceased patients required high-frequency oscillatory ventilation and inotropic support. At one year, 2.4% of patients in Groups A, B, and C had a Developmental Quotient of 70-85, indicating mild developmental delay on the Development Assessment Scale for Indian Infants (DASII). At one year, 97.5% of babies who underwent CDH surgery were alive and thriving.*
Conclusion: *This study from an Indian university teaching center shows CDH outcomes comparable to landmark studies from the Western world.*
Keywords: *Congenital Diaphragmatic hernia, Mortality and Morbidity, Neurodevelopmental Outcome*

Date of Submission: 27-01-2025 Date of Acceptance: 07-02-2025

I. Introduction

Congenital diaphragmatic hernia (CDH) is characterised by defect in the development of diaphragm which leads to protrusion of abdominal content in the thoracic cavity. Its incidence varies across the population from 0.8 to 5/10,000 live birth [1]. It accounts for 8% of all major congenital anomalies [2]. Advancement in the management of CDH over the past decades like delayed surgical repair, gentle ventilation, optimisation of pulmonary artery pressure before surgery, less muscle relaxation and sedation; has led to improvement in post-natal outcome of CDH. Western literature has reported 70- 90% survival rate of CDH [3]. However, mortality reported from developing country continues to be high in the range of 40-50% without any follow up experience [4]. This study was done to depict 20 years of clinical experience and long-term outcome of CDH from a single tertiary care teaching hospital.

II. Materials And Methods

This was a retrospective observational study carried out in the division of Pediatric surgery and Neonatology /Pediatrics of a teaching hospital and tertiary referral centre, New Delhi, India. All patients with diagnosis of CDH admitted in NICU and Pediatric ward between the period of 2004 to 2023 were included. Recruited patients were divided into 4 subgroups according to the age of presentation in hospital. Group A included inborn babies with antenatal diagnosis of CDH. Group B included babies transferred/referred from outside hospital within 24 hours of birth. Group C included neonates between 1 to 30 days of life. Group D included patients who presented after 1 month of life with diagnosis of CDH.

The clinical records, operative findings and clinical outcome of patients admitted in the NICU and Paediatric ward was tabulated and recorded in EXCEL database. Follow up of all patient was done in Pediatric surgery and Pediatric outpatient at least every 3 monthly from the date of discharge till 1 year. Following that they have been followed annually and whenever there were symptoms. Neurodevelopmental assessment of Group A, B and C patients were done using DASII (Developmental assessment scale for Indian infants) scale at 1 year of age. The clinical data was entered prospectively with retrospective analysis. All deaths, leave against the medical advice and clinical outcome at 1 year of post-operative day were recorded. All the patients were given medical management as per standard NICU and PICU protocol (i.e. invasive monitoring, 2D echocardiography, mechanical ventilation, blood products, intravenous antibiotics, parenteral and enteral nutrition).

III. Results

Total of 81 patients with CDH were included in the study during the period of 20 years. Group A, B, C, and D included 11, 23, 20, and 27 patients respectively. Age group of Patients included were from newborns to 10 years of age. Out of 81 patients, seven patients succumbed to the illness. Five patients died pre surgery (Group A-1, Group B- 4) and 2 patients died post-surgery (Group B-1 and Group C-1). Presurgical death were related to disease process itself. One Postsurgical death from Group B was due to Acinetobacter sepsis on post op day 8. Another post-surgical death from Group C was 1 year after surgery due to ARDS (Acute respiratory distress syndrome) caused by HINI pneumonia. Thus, overall survival rate was 91.3 %. Those who went for surgery after initial stabilisation (n= 76), only two deaths were recorded in the first year. The post operative survival on day 365 is 97.5%, with three babies who needed further laparotomy for adhesive obstruction, infected goretex sheet with adhesion and growth failure due to severe gastroesophageal reflux. One further baby is awaiting surgery for severe pectus excavatum.

Demographics:

CDH was most common among male (Male- 81.5%, female-18.5%). Amongst Group A, B, and C 90% were of >37weeks gestation and 10% were between 28 to 36 weeks of gestation. Mean Birth weight for group A, B and C were 2.8 ± 0.6 Kg, 3.75 ±5.08 and 2.73 ± 0.41 Kg respectively. For group D there was incomplete record of gestational age, APGAR score and birth weight and APGAR score of <5 was seen in 5 out of 7 patients who died. Right side CDH cases were 29.6%, left side 69.13% and Bilateral 1.23%. (Table 1)

Table 1- Demographics

		Group A	Group B	Group C	Group D
Total patients		11 (13.5%)	23 (28.3%)	20 (24.6%)	27 (33.3%)
Sex	Male	10 (90.9%)	18 (78.2%)	17 (85%)	21 (77.7%)
	Female	1 (9.09%)	5 (21.7%)	3 (15%)	6 (22.2%)
Gestation (weeks)	>37	11 (100%)	19 (82.6%)	18 (90%)	
	28-36	0	4 (17.3%)	2 (10%)	
Delivery	NVD	1 (9.09%)	3 (13%)	8 (40%)	
	LSCS	10 (90.9%)	20 (87%)	12 (60%)	
Hernia side	Right	1 (9.09%)	1 (4.3%)	6 (30%)	16 (59.2%)
	left	10 (90.9%)	22 (95.6%)	14 (70%)	10 (37.03%)
	Bilateral	0	0	0	1 (3.7%)
Apgar score	<5	1 (9.09%)	5 (21.7%)	2 (10%)	
	>5	10 (90.9%)	18 (78.2%)	18 (90%)	

Medical Management:

Persistent pulmonary hypertension (PPHN) was present in 11 of 81 patients (Group A-2, group B-8, Group C-1 and Group D-0). High frequency oscillatory ventilation was required for 10 patients (Group A-2, group B-7, Group C-1 and Group D-0). Five out of seven patients who died had PPHN and required HFOV support. Inotropic support were required in 12 patients out which seven succumbed to illness. (Table 2).

Table 2- Medical management

		Group A	Group B	Group C	Group D
Ventilation	Conventional	9 (81.81%)	16 (6905%)	19(95%)	27 (100%)
	HFOV	2 (18.18%)	7 (30.43%)	1 (5%)	0
PPHN		2 (18.18%)	8 (34.78%)	1 (5%)	0
Inotropes		5 (45.45%)	14 (60.8%)	3 (15%)	0

Surgical management: Depicted in Table 3.

Table 3- Surgical management

Type of repair	Group A	Group B	Group C	Group D
Open primary repair	10 (90.9%)	21 (91.3%)	18 (90%)	19 (70.37%)
Patch repair	1 (9.09%)	2 (8.6%)	2 (10%)	1 (3.7%)
Laparoscopic/Robotic repair	0	0	0	7 (25.9%)

Associated Anomalies:

Malrotation (7.4%) was the most common anomaly associated followed by Patent ductus arteriosus (6.17%) and bowel volvulus (3.7%). Other associated anomalies were Exomphalos, Dextrocardia, Atrial Septal Defect, Ventricular Septal Defect and Right Pelvi-ureteric junction obstruction.

Post Operative complications:

Immediate (48 to 96hours) post operative complications were Wound Dehiscence (6%), Pneumothorax (4.9%), Pleural effusion (3.7%) and Chylothorax (1.2%). Long term (>1 month) post operative complications were Gastroesophageal reflux disease (7.4%), Adhesive intestinal obstruction (4.9%) & severe pectus excavatum (1.2%).

Neurodevelopmental Outcome: At 1 year of age, in Group A, B and C 2.4% had Developmental Quotient between 70-85 indicating mild developmental delay (motor delay) on Development Assessment Scale for Indian Infants (DASII).

IV. Discussion

Advancement in the field of neonatal and pediatric surgery has led to decrease in the CDH mortality. Recent guidelines encourage delayed surgery only after pulmonary pressures get stabilised [5]. In our study delayed surgical approach was followed in the treatment of neonatal CDH cases.

CDH survival rate in developed countries is upto 90%. However, developing countries still lags behind with the mortality being 40-50%. In our study overall survival rate was 91.3 % and post operative survival of 97.5% after one year of follow up post-surgery. This survival rates are easily comparable to the survival rate reported in any of the western literatures. Table 4 compares mortality rate of our study with the study done on this subject in India.

Table 4- Comparison of survival rate of our study with Indian studies.

Author / year	N= total number of patients	Duration/follow up	Survival rate
Jain et al/ /2002 [12]	N=86 Age at presentation **group1**-<24 hours-13 **group2**->24hrs to 30 days-34 **group3**->30 days-39	1993–2000 No follow up	The survival rate was 0%, 87.5% and 97.4% in Group 1, 2 and 3 respectively.
M molugan et al/2017 [4]	N=148	2010-2015 No follow up	**Survival rate-** 85%
Aihole et al/2018 [13]	N=83	2005- 2017 No follow up	**Survival rate –** 84.3%
Kamal nain rattan et al/2019 [16]	**N**=42	2001-2016 No follow up	**Survival rate-**69%
Jayasree et al/ /2019 [19]	**N**=120	2013- 2018 No follow up	**Survival rate-** 73.4%
Current study/2023	N=81	2004-2023 1 year follow up	**Survival rate-** 97.5%

Male preponderance was seen in our study similar to the various studies done on this subject [6,7]. Right side CDH was the most common presentation in the group D patients and it was associated with good outcome as opposed to the poor outcome mentioned with right side CDH in most of the literatures [8]. The reason could be late presentation and the smaller size of defect. Major associated congenital anomalies carry the poor prognosis [9,10]. Malrotation of the gut was the most common associated congenital anomalies with CDH in group A, B and C. Mortality was also higher in this subset of patients.

Low APGAR score (<5) was associated with increased risk of mortality [11,4,10]. Five out of seven patients in our study who died had APGAR score <5 at 1 minute of life. Age of presentation is also good predictor of mortality in CDH. Studies done on this aspect published that late presentation is associated with good survival

rate and early age of presentation is associated with poor prognosis [12,9]. This was also seen in our study, as there was 100% survival in group D patients whereas group A, B and C accounts for all the mortality seen in this study.

In various literature uncontrolled Persistent Pulmonary Hypertension (PPHN) was associated with increased risk of mortality [13,14,15]. This result was consistent with our study as six out of seven who died had uncontrolled PPHN. Requirement of HFOV and inotropes is also associated with poor prognosis [16]. In our study, seven out of twenty-two who required inotropic support succumbed to the illness and seven out of ten patients who required HFOV support died.

Most common long term post operative complication as reported in a study done by Wan Yee teo et al, Alshaima Algamdi et al and Vageriya NL et al was gastroesophageal reflux disease. In our cohorts similar result was found. Patch closure of the large defect in CDH is associated with recurrence of CDH [17]. In our study 2 out of 6 patient (33.3%) of patch closure had recurrence. Primary repair and robotic closure didn't have any recurrence of CDH.

CDH is associated with good neurodevelopmental outcome provided the initial delivery and ventilation has been of high quality. Studies have reported gross motor delay, expressive and receptive language delay and hearing disability [18,10]. In our study developmental assessment was done using DASII scale and we found mild gross motor delay in 2.4 % of cases.

There are paucity of data on long term (1 year) follow up of CDH in Indian literature. After extensive literature search we found only one Indian study done by Vageriya NL et al, who has followed up 42 patients for 8 months [18]. Our study is the only Indian study who has followed up 81 patients for 1 year with improving result and evaluated long term post op complication along with the neurodevelopmental outcome. This will help in counselling the outcome of the disease to the family of the patients and will boost the comprehensive management of CDH in our country.

V. Conclusion

This is the first Indian study reporting comparable results of medical and surgical management of congenital diaphragmatic hernia with the best published results from the western units. This study establishes that the short term and long term outcome of this complex condition requires team work, excellent intensive care support and surgical experience on a platform where there is seamless integration of these services. This study establishes the maturation of Indian neonatology and neonatal surgery to international standards hitherto unreported from any centre in our country.

WHAT IS ALREADY KNOWN?

Mortality rate of Congenital Diaphragmatic hernia in developing countries is very high. However, in western centres survival rates are better.

WHAT THIS STUDY ADDS?

There is a paucity of studies on one year post-operative outcome of CDH in India. Survival rates comparable to western centres in neonates with severe CDH can be achieved in India.

References

[1] Chandrasekharan PK, Rawat M, Madappa R, Rothstein DH, Lakshminrusimha S. Congenital Diaphragmatic Hernia–A Review. Maternal Health, Neonatology And Perinatology. 2017 Dec;3:1-6.

[2] Aly H, Bianco-Batlles D, Mohamed MA, Hammad TA. Mortality In Infants With Congenital Diaphragmatic Hernia: A Study Of The United States National Database. Journal Of Perinatology. 2010 Aug;30(8):553-7.

[3] Balayla J, Abenhaim HA. Incidence, Predictors And Outcomes Of Congenital Diaphragmatic Hernia: A Population-Based Study Of 32 Million Births In The United States. The Journal Of Maternal-Fetal & Neonatal Medicine. 2014 Sep 1;27(14):1438-44.

[4] Molugan M, Kamalarathnam CN, Muthukumaran J. Clinical Profile Of Congenital Diaphragmatic Hernia And Their Short-Term Outcome In A Tertiary Care Neonatal Unit: A Retrospective Study. Indian Journal Of Child Health. 2017 Sep 26:435-7.

[5] Puligandla PS, Grabowski J, Austin M, Hedrick H, Renaud E, Arnold M, Williams RF, Graziano K, Dasgupta R, Mckee M, Lopez ME. Management Of Congenital Diaphragmatic Hernia: A Systematic Review From The APSA Outcomes And Evidence Based Practice Committee. Journal Of Pediatric Surgery. 2015 Nov 1;50(11):1958-70.

[6] O'Mahony E, Stewart M, Sampson A, East C, Palma-Dias R. Perinatal Outcome Of Congenital Diaphragmatic Hernia In An Australian Tertiary Hospital. Australian And New Zealand Journal Of Obstetrics And Gynaecology. 2012 Apr;52(2):189-94.
[7] Long AM, Bunch KJ, Knight M, Kurinczuk JJ, Losty PD. Early Population-Based Outcomes Of Infants Born With Congenital Diaphragmatic Hernia. Archives Of Disease In Childhood-Fetal And Neonatal Edition. 2018 Nov 1;103(6):F517-22.
[8] Bidiwala A, Bishara J, Burjonrappa S. Right Sided Congenital Diaphragmatic Hernia: Diagnostic, Prognostic And Therapeutic Implications. Inincisional And Congenital Diaphragmatic Hernia (CDH): Risk Factors, Management And Outcomes 2016 Jan 1 (Pp. 113-130).
[9] Wang Y, Honeyford K, Aylin P, Bottle A, Giuliani S. One-Year Outcomes For Congenital Diaphragmatic Hernia. BJS Open. 2019 Jun;3(3):305-13.
[10] Khachane Y, Halliday R, Thomas G, Maheshwari R, Browning Carmo K. Outcomes For Infants With Congenital Diaphragmatic Hernia: A Single Campus Review With Low Extracorporeal Membrane Oxygenation Utilisation. Journal Of Paediatrics And Child Health. 2022 Jan;58(1):90-6.
[11] Teo WY, Sriram B, Alim AA, Ruan X, Rajadurai VS. A Single-Center Observational Study On Congenital Diaphragmatic Hernia: Outcome, Predictors Of Mortality And Experience From A Tertiary Perinatal Center In Singapore. Pediatrics & Neonatology. 2020 Aug 1;61(4):385-92.
[12] Jain A, Singh V, Sharma M. Congenital Diaphragmatic Hernia: Our Experience-A Brief Review. Indian Journal Of Anaesthesia. 2002 Nov 1;46 (6):426-9.
[13] Aihole JS, Gowdra A, Javaregowda D, Jadhav V, Babu MN, Sahadev R. A Clinical Study On Congenital Diaphragmatic Hernia In Neonates: Our Institutional Experience. Journal Of Indian Association Of Pediatric Surgeons. 2018 Jul;23 (3):131.
[14] Mohseni-Bod H, Bohn D. Pulmonary Hypertension In Congenital Diaphragmatic Hernia. Inseminars In Pediatric Surgery 2007 May 1 (Vol. 16, No. 2, Pp. 126-133). WB Saunders.
[15] Suda K, Bigras JL, Bohn D, Hornberger LK, Mccrindle BW. Echocardiographic Predictors Of Outcome In Newborns With Congenital Diaphragmatic Hernia. Pediatrics. 2000 May 1;105(5):1106-9.
[16] Rattan KN, Dalal P, Singh J. Clinical Profile And Outcome Of Neonates With Congenital Diaphragmatic Hernia: A 16-Year Experience From A Developing Country. Proceedings Of Singapore Healthcare. 2019 Mar;28(1):74-8.
[17] Macchini F, Raffaeli G, Amodeo I, Ichino M, Encinas JL, Martinez L, Wessel L, Cavallaro G. Recurrence Of Congenital Diaphragmatic Hernia: Risk Factors, Management, And Future Perspectives. Frontiers In Pediatrics. 2022 Feb 9;10:823180.
[18] Vageriya NL, Shah R, Mane SB, Daginawala T, More P. Evaluation Of Long Term Outcome Of Congenital Diaphragmatic Hernia Survivors: A Single Centre 8 Years' Experience In A Developing Nation With Limited Facilities. International Surgery Journal. 2020 Apr 23;7(5):1366-72.
[19] Chandramati J, Nair LS, Menon SM, Prabhu A, Abraham M, Viswanathan N, Ponthenkandath S. Outcome Of Congenital Diaphragmatic Hernia: A Single Center Experience. Journal Of Neonatal Surgery. 2020 May 6;8(4):29.