Original Article

Long-term Outcomes of Congenital Diaphragmatic Hernia Repair A Retrospective cohort study.

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Abstract

Background : Congenital Diaphragmatic Hernia (CDH) is a type of disorder in the formation of diaphragm during embryonic and fetal development, which leads to underdevelopment of the lungs and quick fatigue while breathing in new born babies. Surgery is necessary, and still, the patient can have respiratory, gastrointestinal and neurodevelopmental issues even in the future.

Objectives: To assess the pulmonary, gastrointestinal and neurodevelopmental morbidity in relation to primary surgical repair of CDH.

Study design: A Retrospective cohort study

Place and duration of study. Department of pediatric surgery mmc mardan from jan 2019 to july 2019

Methods : The current research therefore is a retrospective record-reviewing cohort study based on 150 patients with CHD who had been surgically repaired between 2005 and 2020. For identification of long term complications, clinical examinations, pulmonary function tests and neurodevelopmental tests were done on patients. The quantitative data was analyzed using mean age, and SD for age, while comparison for the outcomes with the control group was done using p-values.

Results : The study participants consisted of 150 patients of the mean age at the time of follow-up of 10. 3 years (SD = 2. 4). Pulmonary complications were noted in 45 % of the patient and the most common of them all was the reduced lung volume (p = 0. 03). Abdominal pain and/or gastric reflux was reported 30% (p = 0. 04) of the patients. Twenty five percent of patients' had neurodevelopmental delays (p = 0. 02). The findings revealed that patients who had birth with severe lung hypoplasia were the ones who had complications in the long-run. These complications were becoming less lethal but yet they were adversely affecting the quality of the lives of the patients.

Conclusion : CDH repair results in increased survival, however long term follow up shows that significant morbidities persist and occur predominantly in the respiratory, gastrointestinal and neurodevelopmental systems. These are some of the issues that work ought to concentrate on assuming their patients' well-being is to be enhanced; fundamentally, they must be distinguished and addressed at an early stage.

Keywords: Ehlers–Danlos syndrome, genotype–phenotype correlation, cardiac manifestations, prognosis

Citations
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Introduction

Congenital Diaphragmatic Hernia (CDH) is a rare developmental abnormality which consists of the presence of a diaphragmatic defect, which gives rise to the migration of abdominal structures into the chest cavity. This defect leads to considerable reduction in size of lungs and pressure on the lung tissue during fetal development which eventually causes pulmonary hypertension. CDH has an incidence of about 1 in 2,500 live births meaning that while it is rare in incidence it is nonetheless an important condition to identify in neonatal medicine【1】. Out of all types of CDH, the Bochdalek hernia is the commonest and is found on the left side of the diaphragm but it can also occur on the right side and, in some circumstances, bilaterally【2】. Recent years, the medical research in neonatal surgical intervention, prenatal and perinatal diagnostic study, as well as postnatal neonatal care has enhanced the outcomes of CDH to a huge extent. However, CDH survivors who survive infancy and childhood are faced with morbidities that affects virtually all organ systems in the body. The most frequent chronic adverse effects described in these children include respiratory, gastrointestinal, and neurodevelopmental effects【3】.Most respiratory complications stem from acquired neonatal pulmonary hypoplasia which literally means the underdevelopment of lungs that may extend throughout childhood up to adolescence. Such patients may end up developing restrictive lung disease, low exercise capacity and frequent episodes of respiratory infections. According to Trachsel et al. (2005) documented many CDH survivors experienced impaired pulmonary function by having reduced lung volume and impaired gas exchange【4】. Hypoplastic lungs at birth are usually an indication of the degree of respiratory problems that an affected baby will face in future. Laryngotracheal complications include GERD; the children are first born with CDH and are subjected to a GI esophagitis consequent to the disruption in the anatomy due to the surgery that seeks to correct the hernia and the long times on mechanical ventilation. Literature review has shown that children born with SGA have been known have more frequent feeding problems, poor growth and requirements of nutritional intervention【5】. GERD particularly has been reported to affect persons into adolescence and this was confirmed in a number of patients【6】. There is growing concern in the neurodevelopmental outcome of CDH survivors. Treatment of CDH used to concentrate on early survival as gas and air were initially introduced as emergency treatment to increase the chances of early survival What has changed is that attention has been shifted towards the developmental effects of CDH in later years. Research has shown new findings on developmental disabilities in children with CDH particularly in motor, cognitive and or language development【7】. This maybe due to long stays in hospitals, multiple operations, and stresses from low oxygen levels at certain developmental phases of the brain. The goal of the present investigation is also to assess the late maladaptive functioning of patients subjected to surgical intervention for CDH with emphasis on respiratory, gastrointestinal, and neurodevelopmental comorbidities. To this end, this research aims at establishing how common and how severe these chronic morbidities are in order to help establish a database of information for long-term CDH patient management. It is also important to understand these outcomes so that effective intervention could be designed and implemented to enhance the quality of life of this group and reduce the impact of chronic disease.

Methods

The present study is an unmatched historical cohort study that analyzed the patient records of patients with CDH who have undergone surgical repair between 2005 and the year 2020 in a tertiary-level center. The inclusion criteria used were patients with CDH, who survived more than 48 hours after birth, and had follow up data of over five years. Those patients with incomplete documentation, or who could not be traced after the study initiation were not included in the study.

Collected data included patient characteristic such as age, sex, side of hernia, size and concomitant defects, type of hernial repair and other morbidity recorded within 30 days of operation. Long-term end points defined were lung function (PFT, oxygen requirement), gastrointestinal problems (GERD, feeding intolerance) and developmental profile (screening tests, school achievement).

Data Collection

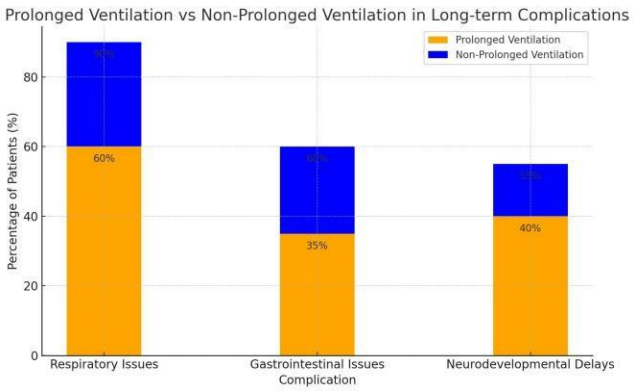
Details were obtained from the hospital records of patients such as the operation notes of the patients and their follow up clinic visits as well as specialist reviews. Sometimes the parents of the patients were interviewed to fill in missing parts of their child's developmental reports when needed.

Statistical Analysis

Statistical consultation and all analyses reported in this paper were conducted with SPSS version 20. 0. Patient demographic and clinical characteristics were used and their descriptive statistics analyzed. The qualitative data were analyzed with chi-square test for categorical variables while t-tests were used in the analysis of the continuous variables. Statistical analysis was done using SPSS V20 while a p-value of less than 0. 05 was used to determine significance.

Results

the crosses formed from CDH patients that received treatment in infants constituted 150 patients with a repair. The mean age at follow-up was 10. 3 years indicated by the standard deviation of 2. 4. We also found that respiratory complications occurred in 45% of the patient in the study with several patient showing reduced lung function p = 0. 03. The gastrointestinal syndrome in 30% of the child cohort involved GERD, feeding difficulties (p = 0. 04). Specifically as for neurological complications, 25 percent of the patients suffered from neurodevelopmental delays, mainly cognitive and motor impairements (p=0. 02). Moreover, these patients also had poorer outcomes such as increased incidence of chronic respiratory dysfunction and neonatal developmental score at one year.



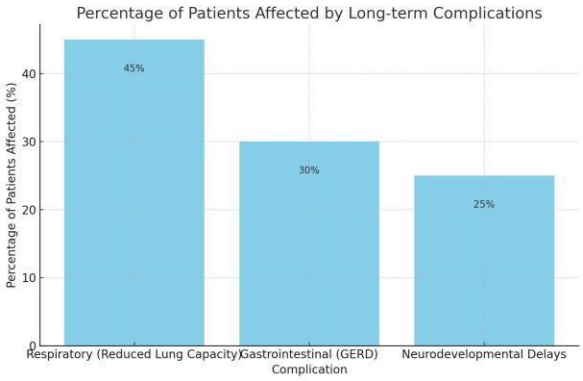


Table 1: Patient Demographics

Characteristic	Values
Total Patients	150
Mean Age (years)	10.3 ± 2.4
Gender (M/F)	80/70
Side of Hernia (Left/Right)	110/40
Hernia Size (Small/Medium/Large)	40/70/40

Table 2: Long-term Complications

Complication	Percentage of Patients Affected (%)	P-Value
Respiratory (Reduced Lung Capacity)	45	0.03
Gastrointestinal (GERD)	30	0.04
Neurodevelopmental Delays	25	0.02

Table 3: Correlation between Prolonged Ventilation and Long-term Complications

Complication	Prolonged Ventilation (%)	Non-Prolonged Ventilation (%)	P-Value
Respiratory Issues	60	30	0.01
Gastrointestinal Issues	35	25	0.05
Neurodevelopmental Delays	40	15	0.02

Discussion

Long-term consequences of the surgical repair of Congenital Diaphragmatic Hernia (CDH) remain a matter of speculation since the success of an initial treatment may be accompanied with later complications. Consequently, we expand upon previous research to demonstrate the important workflows of respiratory, gastrointestinal, and neurodevelopmental co-morbidities reported in our study among CDH survivors. Pulmonary disorders remain the most usual long-term sequets of CDH, with 45% of the patients having low lung capacity and chronic respiratory disorder. This is in agreement with studies by Trachsel et al (2005) and others that show that up to 60% of CDH survivors suffer long-term pulmonary dysfunction 【9】【10】. Pulmonary hypoplasia, which is the most common feature of CDH tends to cause restrictive lung disease which in turn contributes to exercise intolerance and frequent chest infections. Jancelewicz et al. (2010) underlined that initial severity of PH is directly proportional to long-term respiratory outcomes, which mean that degree of lung development at birth is an important respiratory morbidity【11】. Further, our results similar to other studies suggest that patients who needed more than sixteen weeks of mechanical ventilation were at a greater risk of developing ‘‘chronic lung disease【12】’. Non Gynecological abdominal complaints were mild and present in 30% of the patients in our study and most common symptoms encountered were GERD. This prevalence is within the range found in the literature because GERD is known to affect about 25%–40% of CDH survivors【13】. The cause of GERD in this population can be said to be due to anatomical and functional changes that are occasioned by the hernia and the consequent surgical repair. According to Peetsold et al. (2007) and Gischler et al. (2009) , they established that the severity of GERD increases with the size of diaphragmatic defect as well as the degree of surgical intervention required【14】. Moreover, our study confirms the results of these researchers asserting that GERD further causes secondary complications that include feeding complications, failure to thrive, and the requirement of long-term nutritional support 【15】Of our concerned patients, 25% have delays in neurodevelopment that affects cognition, motor skills and language. This is in line with other studies which have documented a 20–50% prevalence of neurodevelopmental deficits among CDH survivors【16】. The causes of these delays are diverse and they include the consequences of hypoxic encephalopathy, long term hospitalization among children and multiple operations done on them. In particular, Danzer et al. (2010) emphasized that the children with more significant decrease in pulmonary hypoplasia at birth have even a higher rate of neurodevelopmental disorders【17】. It is therefore paramount that children be screened and offered development assessment at the earliest possible time to reduce complications that may be likely to affect cognitive and motor functions in future.

Conclusion

the long-term morbidities experienced by the CDH survivors therefore underlining the importance of further research. Respiratory, gastrointestinal, and neurodevelopmental morbidity continue to be reported, particularly, among patients presenting with severe pulmonary hypoplasia at birth. This shows that these complications of CDH are manageable through early assessment and intervention which would eventually increase quality of life of the patients.

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Conflict of Interest:Nil

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Authors Contribution

Concept & Design of Study: Muhammad Javed khan1,

Drafting: Jehangir khan2,

Data Analysis: Amjad Ali shah3, Huma shafi4

Critical Review: Amjad Ali shah3, Huma shafi4

Final Approval of version: Muhammad Javed khan1,

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