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The price of success in the management of congenital diaphragmatic hernia: is improved survival accompanied by an increase in long-term morbidity?

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Abstract

Background: The impact of "gentle ventilation" (GV) strategies on morbidity and mortality of patients with congenital diaphragmatic hernia (CDH) in our institution has not been determined. This study reviews the primary and secondary outcomes of our patients with CDH treated with the GV approach.

Method: We performed a retrospective chart review of respiratory, neurologic, nutritional, and musculoskeletal morbidities in patients with CDH treated at a single institution between 1985 and 1989 with conventional ventilation (CV) compared with those treated from 1996 to 2000 with GV.

Results: There were 77 CV-treated and 66 GV-treated patients with CDH, with 51% survival in the CV cohort compared with 80% in the GV group (χ^2, $P < .05$). At 3-year follow-up, we found no statistically significant differences in the frequency of respiratory (38% of CV patients, 50% of GV patients), neurologic (29% of CV patients, 34% of GV patients), or musculoskeletal morbidity (46% of CV vs 29% of GV-treated patients). There was a difference in nutritional morbidity as indicated by the increased frequency of gastrostomy tube use in the GV-treated patients (34%) compared with the CV patients (8%; χ^2, $P < .05$).

Conclusion: The implementation of GV techniques has significantly decreased mortality in infants with CDH. This has been associated with a documented increase in nutritional morbidity among survivors.

The management of congenital diaphragmatic hernias (CDHs) has evolved over the past 50 years since the first reports of successful surgical outcomes by Gross [1] in 1953. It is now recognized that restoration of the normal chest and abdominal anatomies and the surgical repair of the diaphragmatic defect play only a minor role in the overall management of these infants, as the underlying pulmonary hypoplasia and the accompanying physiologic derangements often dictate the outcomes in these patients [2]. The

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surgical literature has reported progressive improvements in the mortality rates of patients with CDH through early stabilization of the neonate, recognition of the role of conventional mechanical ventilation in producing lung injury [3], and the role for delayed surgical repair [4]. The use of high-frequency oscillatory ventilation (HFOV) or the early use of extracorporeal membrane oxygenation (ECMO) has been reported to be beneficial in minimizing lung injury and improving overall survival among patients with CDH in multiple centers [5,6].

Our institution had previously reported our experience with HFOV and permissive hypercapnea to stabilize patients with CDH before surgical repair [7]. Over the past decade, this form of ventilation has been widely applied to patients with CDH admitted to our hospital who develop respiratory failure. In this report, we have updated our review of the outcomes of our patients with CDH during 2 representative periods of CDH management, from 1985 to 1989, during which conventional mechanical ventilation techniques were still widely used in patients with CDH, and from 1996 to 2000, during which gentle ventilation (GV) became the standard of practice. For the purposes of this study, GV is defined as the use of permissive hypercapnea, early transition to HFOV to minimize airway pressure, and liberal use of inhaled nitric oxide for the treatment of pulmonary hypertension. We postulated that there may be an increase in survival among patients with CDH during the GV era and that this enhanced survival might be associated with increased morbidity, particularly among those infants who may not have otherwise survived during the pre-GV era.

1. Methods

A retrospective chart review was performed of patients with CDH treated at the Hospital for Sick Children, a tertiary care pediatric hospital with a referral population of more than 5 million. All patients with CDH were initially admitted to the intensive care unit and stabilized before operative repair. Between 1985 and 1989, patients with CDH were managed with conventional ventilation (CV) or transitioned to HFOV for respiratory failure, and surgery was performed within the first 24 hours of life. In comparison, patients treated between 1996 and 2000 were managed in the intensive care unit with CV or GV, and surgical repair was delayed until patient was stable on CV. Extracorporeal membrane oxygenation was only used for patients who demonstrated adequate oxygenation and ventilation initially, and who subsequently failed to maintain oxygenation and/or cardiorespiratory stability on both CV and GV. When indicated, nitric oxide was used to supplement other supports in the treatment of pulmonary hypertension associated with CDH.

After hospital discharge, patients with CDH were followed in the general surgery clinic at 1 month, then 3 months up to 1 year postoperatively. For patients with CDH with ongoing respiratory symptoms, the respirology team was included in the patient's follow-up and management. The clinic records detailing respiratory, neurologic, nutritional, and musculoskeletal morbidities for each patient identified through the general surgery and respirology clinic visits were assessed. If no active issues were identified, each patient would be seen annually at the respirology clinic with chest x-ray until 16 years of age before discharge if stable. Pulmonary function tests were performed after 7 years of age.

When indicated, nutritional, neurologic, and cardiovascular investigations would be added to the clinical assessments of patients with CDH. Patients demonstrating growth retardation and poor weight gain (less than 25th percentile for age) were assessed for gastrostomy tube (G-tube) feed supplementation and followed by a clinical dietician. Patients with gastroesophageal reflux disease (GERD) by clinical and radiologic assessment and the medications or procedures to treat GERD were identified from the clinic notes. Neurologic morbidities as indicated by the need for specialized education classes, hearing devices, and delayed speech or motor development requiring speech, occupational, and physical therapy were determined from the patient records and by clinical referrals. Musculoskeletal deformities were identified in the follow-up clinics by clinical assessment as well as contribution to restrictive lung disease in the adolescent through pulmonary function tests. Outcomes between the CV and GV groups were compared using χ^2 test. The study was approved by the research ethics board at the Hospital for Sick Children (REB no. 1503).

2. Results

In the CV period from 1985 to 1989, 77 patients with CDH were admitted to our institution compared with 66 patients over the period from 1996 to 2000 (Table 1). Of the 77 patients in the CV group, only 11 patients (15%) were ventilated using HFOV, whereas 81% were ventilated using CV and 3 patients (4%) required no intubation before operative repair. None of the patients with CDH were managed by ECMO during this period. In contrast, 24 of the 66 patients with CDH treated in the GV group (36%) were ventilated using HFOV, compared with 35% using CV and 25% not requiring preoperative ventilatory support.

Table 1 Mortality and treatment strategies among patients with CDH in the CV and GV periods

Treatment strategies	CV (n = 77)	GV (n = 66)
Survival* (%)	51	80
HFOV-treated (%)	15	36
ECMO-treated (%)	0	4
% of Survivors with ≥3-y follow-up postrepair	61	69

* $P < .05$, χ^2 test.