

CONGENITAL DIAPHRAGMATIC HERNIA: THE IMPACT OF PREOPERATIVE STABILIZATION ON OUTCOME

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Despite the fact that it is diagnosed antenatally, and despite the availability of improved intensive care, congenital diaphragmatic hernia (CDH) is still associated with high mortality.¹⁻³ To overcome this problem, a variety of techniques have been proposed to decrease the pulmonary vascular resistance, and correct the hypoxia, acidosis and hypotension. These include *in utero* surgical repair,⁴ introduction of extracorporeal membrane oxygenation,^{5,6} and an extended period of preoperative stabilization.⁷⁻¹² Several investigators have proposed extended preoperative medical stabilization followed by delayed repair, but the results of such an approach in terms of overall outcome are controversial.¹⁰⁻¹² This report represents our experience with the management of 47 patients with congenital diaphragmatic hernia and analyzes the effect of preoperative stabilization on outcome.

Patients and Methods

This is a retrospective study of all patients admitted since 1982 to Tawam Hospital, Al-Ain, United Arab Emirates, with the diagnosis of congenital diaphragmatic hernia. The medical records of these patients were reviewed for age at presentation, sex, gestational age, birth weight, whether born at Tawam Hospital or referred from another hospital, associated anomalies, type and site of hernia, contents of the hernia, preoperative and postoperative management, blood gas parameters and outcome. The blood gases, as well as the alveolar arterial O₂ gradient ($AaDO_2 = [173 \times FiO_2] - [PaCO_2/0.8] - PaO_2$) were analyzed in terms of outcome. The effect of preoperative stabilization, as well as changes in blood gas parameters, were also analyzed using Student's *t*-test. In the stabilized group, surgery was delayed in an attempt to achieve a pH of more than 7.2, PO₂ of greater than 50 torr, and PCO₂ of less than 50 torr.

After evaluation at the Intensive Care Unit, endotracheal intubation was performed when indicated, and the patients were ventilated, using positive pressure ventilation (PPV) to start with, and positive end expiratory pressure (PEEP). Chest and abdominal x-rays were obtained, and respiratory support, as well as the ventilatory settings, were adjusted according to the blood gas estimates, which were obtained via umbilical and/or radial arterial catheters. Bowel decompression was achieved by nasogastric aspiration. All ventilated infants were paralyzed, and volume expanders as well as dopamine were used when indicated to maintain their blood pressure. Talozoline, surfactant, nitric oxide and dexamethasone were not used in any of our patients, and metabolic acidosis was corrected with bicarbonate therapy.

All operations were performed by a transverse upper abdominal incision. After reducing the contents, primary closure of the diaphragmatic defect was performed and prosthetic graft was used whenever primary repair without tension could not be achieved.

Results

From 1982 to 1997, a total of 47 cases of congenital diaphragmatic hernia were admitted to our hospital. There were three cases of Morgagni's hernia and two cases of eventration of diaphragm. These were excluded from the analysis. The remaining 42 patients had congenital posterolateral diaphragmatic hernia. Eighteen were born at our hospital, while the remaining 24 were referrals from other hospitals. There were 24 males and 18 females, and 34 were full-term infants. The mean gestational age of the remaining eight patients was 35 weeks (32-37 weeks). Thirty-two had left-sided hernia and 10 had right-sided hernia. The mean birth weight was 2.96 kg (1.58-4.6 kg). All patients presented within six hours of birth, except four, who presented at the ages of 4 days, 10 days, 4 months and 1½ years, respectively. All four infants survived, but were excluded from further analysis.

Of the remaining 38 patients who presented within six hours of birth, one was a conjoined twin with left posterolateral diaphragmatic hernia in both twins. They required emergency separation because of respiratory

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TABLE 1. *Associated anomalies.*

Patient #	Anomalies
1	Down syndrome, Fallot's tetralogy
2	Atrial septal defect, hemangioma abdominal wall
3	Bilateral undescended testes, hypospadias
4	Bilateral undescended testes, obstructive uropathy
5	Myelomeningocele with hydrocephalus
6	Bilateral undescended testes
7	Unilateral hair lip and cleft palate, syndactyly right hand, short rib syndrome, deformed low-set ears, narrow chest, short upper arm

TABLE 2. *Clinical features and outcome.*

Clinical feature	Non-survivors	Survivors	P-value
Birth weight	2.9±0.55 (1.58-3.4)	3±0.57 (1.8-4.6)	0.4182*
Gestation	38.7±2.9 (32-42)	39.4±1.8 (33-40)	0.4352*
Preoperative stabilization (hours)	6.45±6.9 (2-25)	17±11.5 (2-48)	0.0111**
Duration of ventilation (days)	2±1.9 (0.25-6)	12.2±16.9 (1-64)	0.0564†

*Not significant; **significant; †marginally significant; all values are mean±SD.

TABLE 3. *Preoperative blood gas and alveolar-arterial O₂ gradient in relation to outcome.*

Blood gas result	Non-survivors	Survivors	P-value
pH	7.2±0.14 (6.98-7.44)	7.37±1.89 (7.19-7.51)	0.0005
pO ₂	51.3±16 (32-74)	114.8±61.5 (44-245)	0.0037
pCO ₂	57.5±23.2 (34-109.6)	37.1±7.8 (20.6-52)	0.0010
AaDO ₂	521.3±89.13 (326.6-623.5)	304.22±174.6 (38.8-619)	0.0006

All values are mean±SD and range.

distress. Intraoperatively, both were found to have left posterolateral diaphragmatic hernia. They shared the same liver and were also attached at the pericardium anteriorly. They were separated and both hernias repaired, but soon after, both twins died. Excluding malrotation, other associated anomalies were seen in seven patients (18.4%) (Table 1).

Four of our patients were not operated on and died prior to surgery. Of the remaining 34 patients, 12 died, including the conjoined twin, thus giving a postoperative mortality of 35.3% and an overall mortality of 42%. The clinical features of the survivors compared to the non-survivors are shown in Table 2. The immediate preoperative blood gas parameters and the alveolar-arterial O₂ gradient of the survivors were compared to the non-survivors (Table 3). Twelve of our patients were operated on within four hours of birth, and eight of them died (66.7%). The remaining 22 patients were stabilized preoperatively. The duration of preoperative stabilization ranged from 8 hours to 48 hours (mean 18.4 hours), and only four of them died (18.2%). The effect of preoperative stabilization in terms of outcome and changes in blood gas parameters, comparing the initial blood gas parameters to those immediately prior to surgery, are shown in Table 4. Comparing the site of the hernia,

three out of the seven right-sided hernia patients died (42.9%), and nine out of 27 left-sided hernia patients died (33.3%). The contents of the hernia were variable, and included stomach, liver, spleen, and small and large intestines, but of interest was the presence of the liver in nine of the 12 non-survivors (75%), but in only nine of the 22 survivors (40.1%). The stomach on the other hand, was present in the hernia of three of the non-survivors (25%), and 11 of the survivors (50%).

Discussion

The management of newborns with congenital diaphragmatic hernia has changed over the years, from an extreme surgical emergency where newborns were taken immediately to the operating room with severe hypercarbia and acidosis to reduce the contents and allow expansion of the compressed lung, to one of extended preoperative stabilization followed by delayed repair.⁸⁻¹³ In addition, the newer forms of treatment, such as extracorporeal membrane oxygenation, high-frequency ventilation and the use of surfactant and nitric oxide,^{5,6,8,13,14} are all attempts at decreasing the mortality, as congenital diaphragmatic hernia presenting within the first six hours of life is associated with a mortality of 40%-50%.²⁻⁴ This high mortality is attributed to several factors. Although associated anomalies may contribute to mortality, this is mainly due to hypoxia secondary to lung hypoplasia. Emergency repair, however, does not correct the already pre-existing pulmonary hypoplasia, but may on the contrary worsen the respiratory status postoperatively.^{15,16} If associated anomalies are excluded in patients with congenital diaphragmatic hernia, the main determinant factors of outcome are the age at presentation (<6 hours) and the preoperative blood gases.^{2,3,17,18}

Several investigators have proposed extended preoperative medical stabilization, followed by delayed repair. The aim of the extended period of preoperative stabilization is to decrease the pulmonary vascular resistance, improve and stabilize the patient in terms of adequate ventilation (that is, improving oxygenation, eliminating the hypercarbia and acidosis), and improve cardiovascular stability. The results of such an approach in terms of overall outcome are, however, controversial, but a definite fact is that by adopting such a policy, the survival rate would not be worse, but on the contrary may be better.⁸⁻¹⁰ Our overall mortality was 42%, and the postoperative mortality was 35.3%, but in terms of mortality in relation to preoperative stabilization and timing of surgery, eight of the 12 patients operated on early died (66.7%), while only four (18.2%) of the remaining 22 who were operated on after a period of stabilization died. We did not evaluate other factors such as birth weight and gestational age (Table 2), which were of significance in terms of prognosis, but preoperative stabilization was found to be of significance. The effect of this is reflected in the

TABLE 4. The effect of preoperative stabilization in terms of changes in blood gas parameters initially and immediately prior to surgery, and the relation to outcome.

Blood gas parameters	Initial values			Immediate preoperative values		
	Non-survivors	Survivors	P-value	Non-survivors	Survivors	P-value
pH	7±0.13 (6.83-7.3)	7.2±0.15 (6.9-7.45)	0.0028	7.2±0.14 (6.98-7.44)	7.37±1.89 (7.19-7.51)	0.0005
pO ₂	73±34.4 (37-152)	73±34.4 (37-152)	0.1316 ^a	51.3±16 (32-74)	114.8±61.5 (44-245)	0.0037
pCO ₂	56.3±13.4 (25.7-79)	56.3±13.4 (25.7-79)	0.0028	57.5±23.2 (34-109.6)	37.1±7.8 (20.6-52)	0.0010

^aNot significant; all values are mean±SD and range

significant improvement in blood gas parameters (Tables 3 and 4).

A difficult question is whether delaying surgery and extending the period of stabilization is the crucial factor in improving outcome, or whether this was the natural course of these patients. Our study was retrospective and the patients were not randomized, but in congenital diaphragmatic hernia, it is difficult to fairly randomize the patients in a prospective study, as congenital diaphragmatic hernia represents a heterogeneous group of patients and the degree of pulmonary hypoplasia in them is variable. In a prospective randomized trial of delayed versus immediate repair of congenital diaphragmatic hernia, Nio et al.¹¹ found no difference in a clinically stable infant. In a retrospective study, on the other hand, Breaux et al.⁸ reported an improved survival in patients with congenital diaphragmatic hernia, utilizing a strategy of delayed repair, but in their study, the use of extracorporeal membrane oxygenation (ECMO) in some of their patients may have contributed to the improved survival. Wilson et al.,¹² on the other hand, found no improvement in survival of high-risk patients with congenital diaphragmatic hernia undergoing delayed repair and preoperative ECMO. As well, Langer et al.¹⁸ did not find any significant difference in outcome between a group of 31 patients undergoing operation at a mean of 4.1 hours after admission, and a later group of 30 infants repaired at a mean of 24.4 hours following admission.

Although our overall mortality is high, the successful survival of all 22 patients except four in the stabilized group calls for the adoption of such a policy. ECMO is not available in our hospital, and we feel that it does have a place in the management of some of these patients. Until the emergence of other interventional measures that are likely to improve the overall survival, a policy of preoperative stabilization and delayed repair is to be advocated.

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