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Simplification of Ventilatory Management and Elective Surgical Repair in Congenital Diaphragmatic Hernia

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Abstract

Background : **Congenital diaphragmatic hernia (CDH) presented in the first 12 hours of life still carries a high mortality rate in the range of 50 percent. Its concept of management and ventilatory support has evolved considerably and more sophisticatedly. Simplification of ventilatory management with comparable outcome is justified and should be sought for.**

Materials and Methods : **The author's plan of ventilatory management was to ventilate the patients with low peak inspiratory pressure, not exceeding 20 cm H_2O, in keeping of a neutral pH; delayed elective surgical repair without a chest tube; and minimal handling of the infant. Retrospective data collection was carried out from the records of all infants with CDH treated by the author over a 10-year period (1992-2001) at the Department of Surgery, the Children's Hospital.**

Results : **Twenty-two patients were treated. Sixteen had respiratory distress within the first 12 hours of life, eleven of whom survived (68%). The remaining six who had later presentation survived (100%).**

Conclusion : **The ventilatory management of CDH can be simplified and employed with comparable good results.**

Congenital diaphragmatic hernia (CDH) is one of the most challenging conditions in pediatric surgery. Its concept of management has evolved considerably over the latter half of the past century in response to the high mortality rate of newborn infants with CDH presented in the first 12 hours of life.

The timing of surgery, type of ventilatory support and its strategy, the necessity of a chest tube following repair, are the areas of controversy and are still evolving. Survival outcome as the ultimate indicator of the success of treatment, together with the patient's quality of life, should be considered with respect to cost-effectiveness for each sophisticated modality. The wisdom is to simplify the management, accomplished with less struggle, but not less care; and to yield comparable, if not better, results.

The purpose of this study was to (1) analyze treatment outcomes in the author's experience and; (2) advocate the simplification of ventilatory management in CDH.

Materials and Methods

The Children's Hospital, Bangkok, is one of the largest tertiary care pediatric referral centers in Thailand. On average, 15-16 neonates with CDH are admitted annually. No infant was referred after surgical repair.

Before 1993, surgical repair at the Children's Hospital was performed on an emergency basis. Post operative ventilatory care was directed towards alkalosis. Patients were hyperventilated to keep pCO_2 below 40 mmHg. Since 1993, sur-gical repair has been performed on a delayed basis after the so-called stabilization period, the duration of which was arbitrary. The ventilator strategy differed among the surgical staff. High-frequency oscillatory ventilation (HFOV) was available in 1996 and was used in certain cases refractory to conventional mechanical ventilation (CMV). Extracorporeal membrane oxygenation (ECMO) has not yet clinically been applied at the Children's Hospital.

The author's plan of ventilator management was to minimize pulmonary barotrauma from hyperventilation by using the lowest peak inspiratory pressure (PIP) as possible ($\leq$ 20 cmH_2O) in keeping blood pH relatively neutral (7.29-7.45), irrespective of pCO_2 and pO_2 levels. The pO_2 level was not taken into consideration since a specimen for blood gas analysis was normally taken from capillary blood.

Oxygen saturation was monitored by pulse oxymeter. HFOV was not employed except in some patients who required PIP > 20 cmH_2O. When satisfactory pH in the neutral or near neutral range could be maintained for a certain period of time, usually one or two days, surgical repair was then carried out electively. Chest tube was usually not placed following repair. Ventilatory support was continued postoperatively, using the same setting objective until the infants showed adequate spontaneous respiration. The patients were left relatively undisturbed in order to minimize their exertion and thus oxygen consumption.

A database was collected retrospectively from the records of all infants with CDH treated by the author over a 10-year period from 1992 to 2001. Survival data were compared by chi-square test.

Results

Twenty-four CDH patients were treated by the author between 1992 and 2001. Two died without surgery and were excluded. Sixteen patients had respiratory distress within the first 12 hours of life. Six patients had later presentation. Yearly distribution and survival outcome of the 22 infants wre shown in Figure 1.

Of the 16 patients who had respiratory distress within the first 12 hours of life, ten were male and six were female. Birth weights ranged from 1,470 to 3,900 grams (mean 2,439 grams). Two patients had a right-

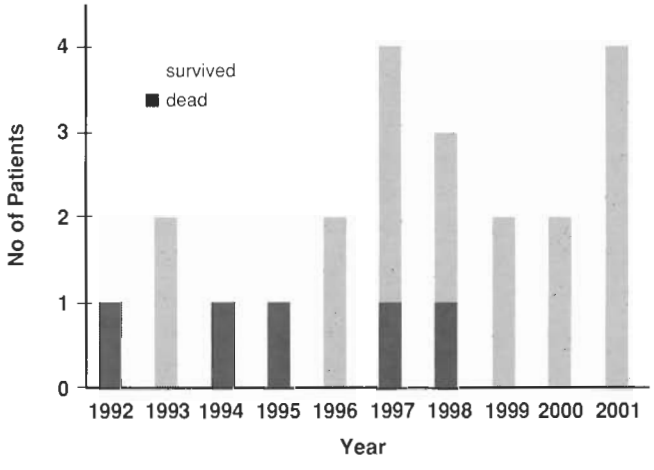

Fig. 1 Yearly distribution and survival outcome of the 22 patients with CDH.

Table 1 Presence of sac, stomach, liver versus survival (< 12-hour presentation group)

	Total	Survivors	p Value
Defect with sac	4	3	p > 0.05
without sac	12	8	
Stomach herniated	8	6	p > 0.05
not herniated	8	5	
Liver herniated	5	3	p > 0.05
not herniated	11	8	

sided defect while the remaining fourteen had a left sided one. The presence of a hernial sac, gastric herniation, or liver herniation had no influences on survival (Table 1).

Preoperative CMV settings with PIP ≤ 20 cmH_2O and a rate of ≤ 60 per minute were applied to 12 patients and the number of patients with acidosis (pH < 7.29) was reduced from eight to two. Sodium bicarbonate was given to nine patients who had concomitant metabolic acidosis. Three patients required PIP > 20 cmH_2O and rate > 60 per minute, two of them were eventually switched to HFOV and succumbed after surgical repair. Record of CMV settings was not available in one patient.

Surgical repair was carried out electively in all except one who was treated in 1992 before the concept of delayed surgery has been adopted in the department. Preoperative stabilization periods varied from one to six days; i.e., one day in 8 patients, two days in 2, three 4 days in 3, four days in one, and six days in one patient. Prosthesis was required in 3 patients, each with a very large defect. Chest tubes were placed in only two patients; one preoperatively for bilateral pneumothorax, the other following surgical repair. Of the remaining 14 patients, in whom no chest tube was placed, the trap air in the ipsilateral pleural cavity were completely reabsorbed within a few days (Figures 2 and 3).

Postoperative CMV settings were relatively similar to those set preoperatively. Ventilatory support was needed after surgical repair for 1-23 days; namely, 1-7 days in 9 patients, 8-14 days in 2, and > 14 days in 5. Malrotation of intestine was the most common associated anomaly. However, the record of this anomaly was incomplete. Associated anomalies, apart from malrotation of intestine, were detected in 7 infants as shown in Table 2.

Eleven patients survived (68%). Their length of stay was 9-54 days (mean 32 days). Follow-up time varied from 2 months to 3 years with satisfactory long term outcome. Five patients died, three of them succumbed to respiratory insufficiency, one to sepsis,

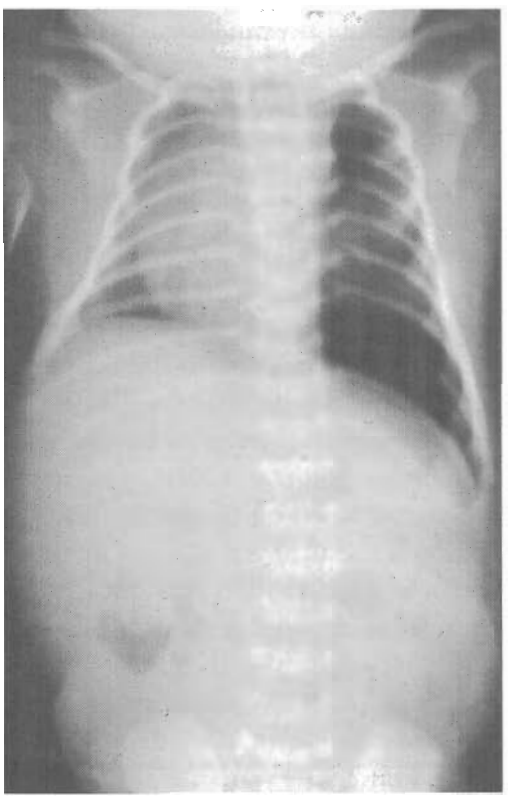

Fig. 2 Chest film : immediately after surgical repair of left-sided CDH without a chest tube. Note the pneumothorax of the left pleural cavity.

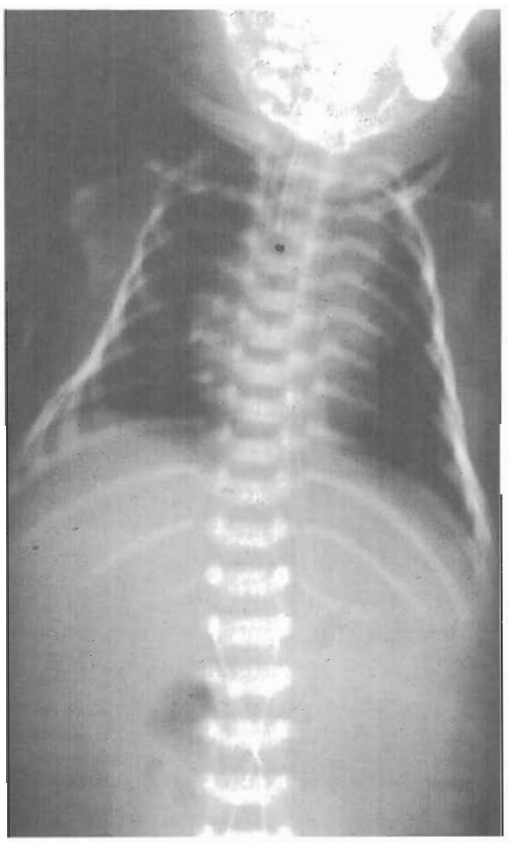

Fig. 3 Chest film : same patient as in Figure 2, at one day after surgical repair of left-sided CDH. Note the decrease of pneumothorax and restitution of the mediastinum.

Table 2 Associated anomalies other than malrotation of intestine in 7 infants (< 12- hour presentation group)*

Anomaly	No. of Patients
Congenital heart disease	2
Undescended testis	3
Pinna deformity	2
Vertebrae	1
Trisomy 13	1

**Some patients had more than one anomaly*

and the other died from cerebral anoxia resulted from cardio-pulmonary arrest during transfer to this hospital despite subsequent successful preoperative stabilization and surgical repair.

Of the 6 patients presented later than 12 hours of life, one was male and five were female. Birth weights ranged from 2,100 to 3,400 grams (mean 2,780 grams). Four patients had a defect with sac, two on the right side, and two on the left. The other two had a left-sided defect without sac. Only these latter two patients needed preoperative CMV, one with PIP 20 cmH_2O and rate 40 per minute; the other with PIP 15 cmH_2O and rate 75 per minute. Both had the support extended for two days postoperatively. No chest tube was placed in all patients except one. Anomalies of the vertebrae and ribs were noted in one infant. All six patients survived. Length of stay was 4-11 days (mean 7 days). Follow-up time varied from 2 months to 3 years with good results.

Discussion

In 1848, Bochdalek, professor of anatomy in Prague, described CDH occurring through the posterolateral defect that now bears his name. The first attempt at repair in an infant was documented by Bettman and Hess[1] in 1929. Successful surgical treatment was rare until 1940, when Ladd and Gross[2] reported 19 cases with 12 survivors. By 1951, Gross and colleagues[3] had repaired 63 infants with 55 survivors (87%), most of whom presented in the first 24 hours of life. Since then, there have been no published series with comparable survival outcome despite advances in neonatal intensive care. CDH presented within the first 12 hours of life still carries a mortality of over 50 per cent. What the intervening fifty years have taught us is that CDH is not merely a surgical problem, with a straightforward surgical solution, but a physiological problem resulted from abnormalities in the development of the pulmonary parenchyma,[4] pulmonary vasculature,[5-8] and surfactant system.[9] Furthermore, the rationale of emergency repair was challenged by the study of Sakai et al[10] which indicated that respiratory compliance actually fell, rather than improving, after surgery. Thus, the initial conservative approach was adopted and are now widely accepted.[11-13]

Regarding the ventilatory support, it had always been directed towards alkalization of the patients by hyperventilation, sometimes unavoidably with high pressure, leading to pulmonary barotrauma. Hence, the value of this aggressive strategy is questioned. Injury to the hypoplastic lungs by hyperventilation outweighs its benefit, if any. The landmark paper by Wung et al[14] has challenged the belief that hyperventilation was beneficial in persistent pulmonary hypertension of the newborn (PPHN). Those authors reported 100 per cent survival in a series of infants with severe PPHN, in which pressure-limited ventilation was used and hypercarbia was ignored, now commonly known as permissive hypercapnia.

HFOV had been advocated on the basis that its gas delivery mechanism would result in less pulmonary barotrauma.[15,16] Some investigators claimed that HFOV improved survival in CDH patients,[17] while others indicated otherwise that HFOV had not significantly improved overall survival, despite its proven ability to reduce $PaCO_2$ and elevate pH.[18] Moreover, although HFOV has been used to treat neonates for almost two decades, doubts remain as to its efficacy and potential for serious adverse side effects.[19]

Since the early 1980s, with the introduction of ECMO for the management of respiratory failure in the newborn, there has been an increasing tendency to use it to support infants with CDH who deteriorate. This approach is based on the presumption that the deterioration is due to abnormality of the pulmonary vascular bed and right to left shunting. Many series enthusiastically reported improved survival with ECMO.[20-27] However, the technology has its own morbidity, particularly the effect on the developing central nervous system of the infants.[28,29] Besides, recent series from centers with a large number of CDH patients indicated that survival was not significantly increased with ECMO.[30-34] Over the past 5 to 7 years, not surprisingly, fewer and fewer babies are treated

with this therapy, and ECMO is considered by many to reflect a bad outcome in neonates.[35]

The other areas of experimentation are inhaled nitric oxide[36-38] and liquid ventilation.[39,40] Both are preliminary and yet to be conclusive.

All these costly modalities have offered no significant survival advantage over CMV and may cause more complications.

The preoperative ventilator strategy in this study is to use low PIP, not exceeding 20 cmH_2O; since PIP,[41] rather than mean airway pressure,[42] closely correlates with pCO_2, hence is the determinant of ventilation, and if high, of pulmonary barotrauma. However the pH, not pCO_2 nor pO_2, is considered as responsive indicator since it is the summation of acid-base balance. Thus, the concept of permissive hypercapnia is applied. Surgical repair is deferred and to be performed in an elective, unhurried environment. Chest tube is not placed following repair of CDH because the trap air in the pleural cavity has no adverse effect on the hypoplastic lung, which, any how, will not expand soon. The air is proven to be readily reabsorbed in a few days. Some investigators even suggested that a chest tube would lead to loss of back pressure preventing the hypoplastic lung from rapid expansion and eventual barotrauma.[43,44] Without a chest tube, the need for tube care is obviated, and the infant is less disturbed. The survival outcomes of 68 per cent in the < 12-hour presentation group and of 100 per cent in the > 12-hour presentation group are comparable to those of large tertiary care pediatric centers with sophisticated technology and strategy.[18,34]

In conclusion, the management of CDH can be simplified as (1) preoperative CMV support with PIP $\leq$ 20 cmH_2O to keep a neutral blood pH and maintaining them for a few days, (2) delayed elective surgical repair without a chest tube, and (3) minimal handling of the infant in order to lessen its exertion and oxygen consumption. These are proven to yield comparable outcome.

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