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Congenital Diaphragmatic Hernia : Is Preoperative Stabilization Beneficial ?

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Abstract

Background/Propose: **Congenital diaphragmatic hernia (CDH) is one of the very high risk conditions in pediatric surgery. It has a high mortality rate in neonates with presenting symptoms shortly after birth. Results of treatment are not yet satisfactory. The purposes of this study are to review our experience with CDH and to evaluate whether preoperative stabilization with delayed surgical repair would be more beneficial than the immediate operation.**

Materials and Methods: **Medical records of infants with CDH admitted to the Queen Sirikit National Institute of Child Health during the period of 1992-2001 were reviewed. The study emplasized the outcomes of treatment, especially in neonates with presenting symptoms shortly after birth.**

Results: **There were 165 patients, male to female ratio was 1.4:1. Twenty-nine patients developed respiratory distress later than 24 hours after birth and 28 of them (96.6%) survived. The remaining 136 infants had respiratory distress within 24 hours after birth were managed in various approaches. From 1992-1994, 44 of the 136 patients underwent immediate surgical correction and 12 of 44 patients (27.7%) survived . From 1995 -1997, 48 patients were treated by either immediate or partially delayed operation . Only 12 of the 48 patients (25%) survived. From 1998-2001, 44 patients were managed under the concept of preoperative stabilization with delayed operative correction. Twenty-six of the 44 patients (59.1%) survived.**

Conclusion: **Preoperative stabilization with delayed operative correction yielded improved survival for the treatment of CDH at our institute. Permissive hypercapnia is one of the most technique for achieving preoperative stabilization with high frequency oscillatory ventilation for stand-by use if permissive hypercarbia is ineffictive.**

Neonates born with a congenital posterolateral (Bochdalek) diaphragmatic hernia represent a significant clinical challenge. The treatment of this entity remains a difficult problem, however a variety of recent developments in respiratory support and therapy for these infants have resulted in an improved survival rate. Original concept of management was the emergency repair of the diaphragm in order to rapidly achieving the return of gastrointestinal contents into the abdomen. But this approach could not help to increase the survival rate to any significant degree. During the past 20 years, there has been a radical change in the surgical management of infants with congenital diaphragma-tic hernia (CDH). The concept

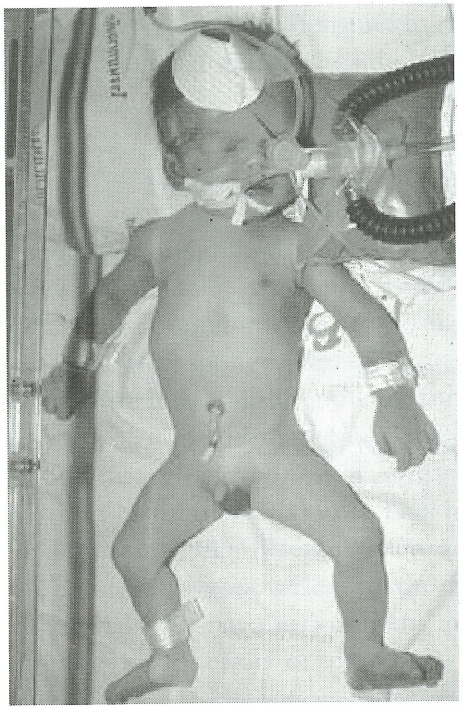

Fig. 1 Photograph showing an infant respiratory distress at birth required immediate respiratory support.

of treatment has changed from immediate or emergency surgery to delayed surgery since 1984.[1-8]

In 1996, CDH management strategy in the Department of Surgery, Queen Sirikit National Institute of Child Health (formerly the Children's Hospital) has been adjusted from emergency to delayed surgical repair in some cases and finally to the preoperative stabilization approach and delayed surgical repair in every case since 1998. The present study was undertaken to review our experience with CDH management in the past 10 years and allowed us to evaluate whether preoperative stabilization with delayed surgical repair would be more beneficial than the immediate operation.[9]

Materials and Methods

A retrospective review of the medical records of the CDH patients treated at the Department of Surgery, Queen Sirikit National Institute of Child Health during the 10-year period between January 1992 and December 2001 was carried out. The data were collected from existing medical records for the modes and outcomes of treatments in various concepts in each period of time. The studies emphasized the outcomes of treatment in CDH patients who developed presenting symptoms within 24 hours after birth (Figure 1). They were divided into 3 groups according to the periods of study.

Group I, between 1992-1994, our strategy of management was immediate repair within 6 hours of admission. Group II, during the second period between 1995-1997, immediate surgery remained the treatment in most cases, however, preoperative stabilization with delayed surgical repair was applied in some cases. Immediate or delayed surgery was based on individual surgeon's decision. A program of aggressive alkalosis was performed by both hyperventilation and chemical means. Infants who were treated with stabilization were intubated on admission and given intravenous pancuronium bromide (Pavulon) 0.1-0.2 mg/kg and Fentanyl 10 microgram/kg every 4 hours. They were mechanically ventilated at the rates ranging from 40-150 breaths per minute (bpm) with short inspiratory times. Peak inspiratory pressure (PIP) was maintained between 15-40 mmH_2O. This program was set up in order to maintain pH above 7.4, the pCO_2 below 60 mm Hg, and oxygen saturation greater than 90 per cent. Delayed operative repair was performed after the patient conditions were approaching the set-up criteria. Group III, the third period between 1998-2001, all patients underwent preoperative stabilization and delayed surgery. The strategy was to employ either hyperventilation towards alkalosis (high-pressure ventilation) or permissive hypercapnia (pressure-limited ventilation).[8] For permissive hypercapnia, PIP was limited between 15-30 cmH_2O and PEEP at 5 cmH_2O. Ventilator setting was maintained at the preductal oxygen saturation of 90 per cent or greater. High-frequency oscillatory ventilation (HFOV) was used as stand-by therapy if conventional mandatory mechanical (CMV) had failed. HFOV with inhaled nitric oxide (iNO) was tried in some cases who failed to improve with CMV. The duration of stabilization ranged from 1-7 days.

Results

One hundred and sixty-five CDH patients were treated between 1992-2001. Male to female ratio was 96:69 (1.4:1). Of the 136 infants who developed the respiratory distress shortly after birth, there were 44

cases in Group I (1992-1994) 48 cases in Group II (1995-1997) and 44 cases in Group III (1998-2001). Of the 165 cases, 136 infants developed respiratory distress with 24 hours after birth. The survival rate of these 136 cases was 36.8 per cent. The 29 remainders presented the symtoms later than 24 hours after delivery. Their survival rate was 96.6 per cent (Table 1).

Infants in Group I (44 cases) underwent immediate surgery within 6 hours (from admission to surgery) with twelve survivors (27.3%).

Group II (48 cases) managed as either immediate or delayed surgical repair had only 12 survivors (25%). Sixteen of the 48 cases (33.3%) died preoperatively. Five of the 32-operated cases did not require intercostal drainage (ICD) and 3 of them (60%) survived.

Group III (44 cases) all were treated with preoperative stabilization and delayed operative repair. Twenty-six of the 44 patients (59.1%) survived, while 13 patients (29.5%) died preoperatively (Table 2). Seventeen of the 31-operated patients received no ICD and 16 of them survived. The 14 remaining operated patients received ICD and 10 of them survived. There was no statistical sighnificant difference between the survivals of with and without ICD placement. HFOV with iNO were used in 5 patients with 3 of them succumbed during stabilization and 2 patients died postoperatively from other complications.

Factors presumed to influence survival were shown in Table 3. The initial pH below 7.0, the pCO_2 over 60 mmHg and contralateral pneumothorax were associated with survival rates in 10.0, 31.8 and 41.7 per cent, respectively. No ICD placement were noted in 22 patients of the 107-operated patients and 19 of them (86.4%) survived.

Of the 29 patients who died during preoperative

Table 1 Results of the treatment of CDH in 165 patients

Onset of Symptoms	No.	Survival	
		No.	%
1. Within 24 hrs.	136	50	36.8
1.1 0-6 hrs.	*131*	*45*	*34.4*
1.2 6-24 hrs.	*5*	*5*	*100.0*
2. After 24 hrs.	29	28	96.6
2.1 1 day-1 month	*8*	*7*	*87.5*
2.2 over 1 month	*21*	*21*	*100.0*

Table 2 Comparison of the survival rate in various period of time in 136 cases with presenting symptoms within 24 hours

Modes of Treatment	Total	Survival	
		No.	%
Immediate surgery (1992-1994)	44	12	27.3
Immediate + partially delayed surgery (1995-1997)	48	12	25.0
Delayed surgery (1998-2001)	44	26	59.1

Table 3 Factors presumed to influence survival

Information	Total	Survival	
		No.	%
1. pH < 7.0 at admission	20	2	10.0
2. pCO_2 > 60 at admission	66	21	31.8
3. Contralateral pneumothorax	12	5	41.7
4. Operation without ICD	22	19	86.4

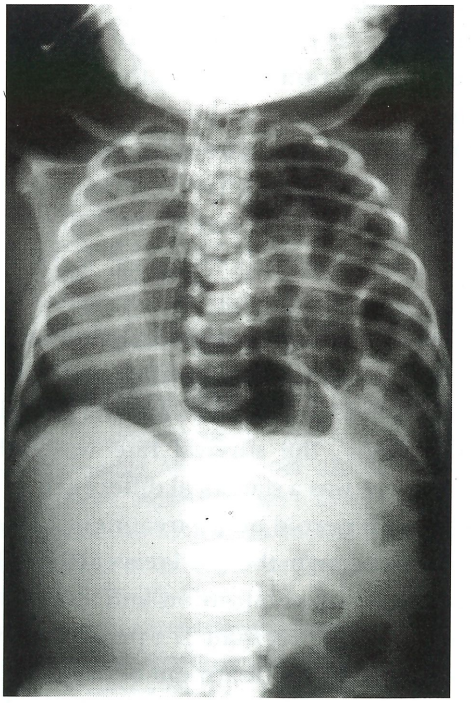

Fig. 2 A chest film of an infant showing the typical findings of gastric and intestinal herniation into the left plural cavity, left lung collapse and shifting of the heart to the right was seen in cases who died within 6 hours after birth and admission.

Table 4 Correlation between timing from admission to operation with the survivals

Duration	Total	Survival	
		No.	%
Operation	107	50	46.7
within 6 hours	49	12	24.5
6-24 hours	25	16	64.0
1-3 days	24	14	58.3
3-7 days	9	8	88.9

stabilization (1992-1995), most of them succumbed within 6 hours of admission due to gastric and intestinal herniation into the left pleural cavity (Figure 2). Five of 13 patients, who died within 6 hours, had lung hypoplasia and complex congenital heart diseases found at autopsy. Six patients died between 1-7 days during stabilization due to pneumonia and septicemia.

Of the 107-operated patients, only 25 per cent of CDH patients who underwent immediate surgery survived. If stabilization was maintained over 1-7 days, the survival rate increased to 58.3-88.9 per cent (Table 4).

Discussion

Despite the development of modernized intensive care units, increasing experience of CDH treatment, and the introduction of novel therapeutic options such as extracorporeal membrane oxygenation (ECMO), HFOV and iNO, overall survival rate reported in the literature was still in the range of 50-60 per cent.[10,11] Overall survival rate of the 136 patients with presenting symptoms shortly after birth from our institute was even lower than 50 per cent. Our data of 13 deaths within 6 hours after admission out of the 29 patients suggested that CDH infants in our instutute were in critically ill condition on referral and admission. From this study, the survival rate was rather low. However, there was a statistical difference among the survival of each peroid of the treatment. The survival rate of 59.1 per cent in the last 4 years (1998-2001) with full application of stabilization and delayed surgery was significantly higher than 27.3 per cent survival rate of immediate surgical treatment between 1992-1994.

Thus, in our opinion preoperative stabilization with delayed surgery is the best modality for the management of critically ill CDH patients.

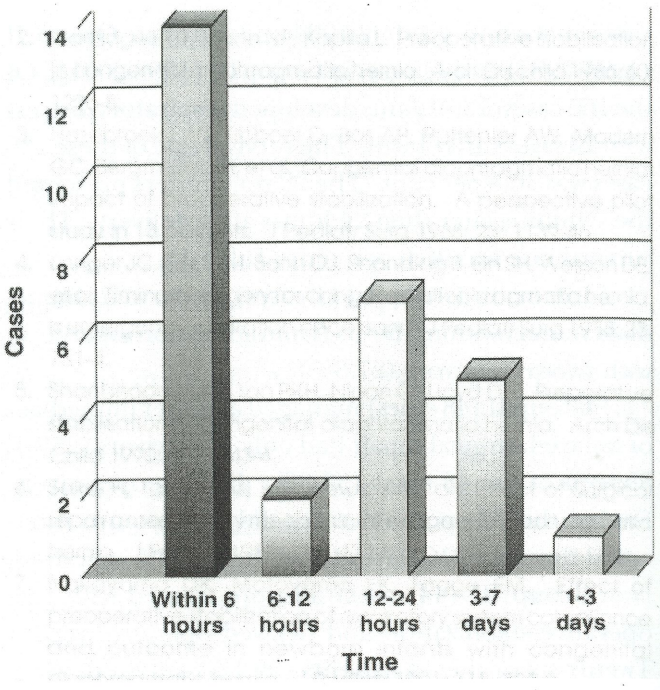

Fig. 3 Analysis of 29 non-operated-dead cases : correlation between duration of stabilization with number of deaths.

According to Ladd and Gross reports,[12-14] it was traditional for CDH to be repaired as an emergency procedure. The rationale of this approach is that returning the abdominal viscera into the abdomen allows expansion of the lungs resulting in improved ventilation. However, it has never been proved that emergency repair of CDH is beneficial.[8] Many infants deteriorate with increasing hypoxia and die after a brief transient period of about 18 hours. Disadvantage of early operation is that the surgical stress superimposed on an early unstable infants with CDH can aggravate pulmonary hypertension. It was therefore suggested that delayed operation preceded by a period of stabilization might be beneficial because of the decreased risk of pulmonary hypertension.[5-7]

Cartridge et al[5] reported an improved survival from 12.5 per cent after immediate surgery to 52.9 per cent after stabilization for 4-6 hours before surgery. Vacanti et al[4] proposed in 1984 that hyperventilation and alkalinization appeared to be a worthy strategy to reduce persistent pulmonary hypertension in the neonates (PPHN). In the contrary, many authors cautioned that hyperventilation and alkalinization may be the causes of barotrauma and right-to-left shunt.[8,15,16] In 1985, Wung et al[8] introduced the technique of pressure-limited ventilation or permissive hypercapnia.

When this strategy was used to minimize barotrauma and right-to-left shunt, survival of the critically ill CDH was increased over 80-94 per cent.[8,15-17]

ECMO has been used as a method of improving oxygenation in the presence of right-to-left shunt, reducing pulmonary hypertension and minimizing barotrauma caused by CMV.[18-20] Many reports indicated that early stabilization, delayed surgical repair and ECMO improved survival in high-risk CDH.[16,21-23] However, complications of ECMO including intracranial hemorrhage, cerebral palsy, cognitive disability, hearing loss, convulsion, blindness and delayed speech development have been reported.[24-28] We have no experience of using ECMO for CDH at our institute.

HFOV and iNO have been used to reduce pulmonary hypertension and improve oxygenation. Our 5 patients treated with this technique did not survive. Finer et al[29] suggested that iNO improved oxygenation and decreased the need for ECMO in hypoxic infants from any conditions other than CDH.

A varieties of new therapeutic options for CDH include surfactant therapy,[30] intratracheal pulmonary ventilation.[31] Partial liquid ventilation[32-34] and fetal surgery for CDH correction[35-38] have been under investigation for future clinical application.

Conclusion

This study of a 10-year experience during the changing strategy of management in infants with CDH at our institute provides data to support that preoperative stabilization with permissive hypercapnia is the appropriate strategy. CMV with limited pressure should be set up at the PIP 15-25 cmH_2O, PEEP 3-5 cmH_2O and IMV 60 bpm. The main objective of stabilization is to maintain the pH at 7.3 or over, preductal oxygenation over 90 per cent and pCO_2 around 60 mmHg. If this approach fails, HFOV with low pressure is another option.

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