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# Delayed Surgery for Congenital Diaphragmatic Hernia

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**Infants with congenital diaphragmatic hernia (CDH) operated on at 12 hours of age have poor prognosis. Development of severe hypoxemia due to hypoplastic lung and pulmonary vascular constriction is the major prognostic factor. Infants who show reasonable gas exchange at first but develop severe hypoxemia a few hours following surgery (honeymoon period) may have benefited from delayed radical surgery. During the waiting period, the patient should be kept on a minimal handling and stabilizing protocol.**

**In our institute, 24 CDH patients were treated from 1970 to 1995. These patients were divided into two groups : group I (n = 11) was the first stage (1970~1984), group II (n = 13) was the second stage (1985~1995). The total survival rate of these two groups did not improve. However our experience with the last six cases using this stabilizing protocol between 1989 and 1995 suggests that delayed repair surgery for CDH improved the survival rate. The survival rate changed from 33.3% in group I to 44.4% in group II. We report our cases, including clinical status, surgical findings and treatments.**

**Key words :** congenital diaphragmatic hernia, delayed surgery

## Introduction

Congenital diaphragmatic hernia (CDH) has one of the worst prognoses of any disease, in spite of remarkable improvements in most outcomes of the major neonatal due to progress in perioperative management. The physiological mechanisms of this disease have been clarified recently, and delayed operation has been chosen in some institutes. The present study was undertaken to compare the outcome of emergency operation with delayed operation for CDH in our department.

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## Materials and Methods

From 1970 to 1995, 24 patients were referred to our institute for surgical treatment of CDH. We reviewed their charts for antenatal conditions, birth weights, gestational ages, symptoms, the presented time of the symptoms, preoperative conditions and management. The first antenatal diagnosed case was transferred to our department in 1983. Until that time, CDH patients had always been operated on just after admission. However now we are treating the earlier cases of CDH, especially those expressed during the first 12 hours of life, according to the new protocol, in which the principle of management is delayed operation (Table 1). If the patients did not develop respiratory symptoms, such as tachypnea, mild to moderate dyspnea, retractions or cyanosis, within the first 12 hours after birth, early operations were performed. On the other hand, if symptoms developed within the first 12 hours, we treated the patient according to the stabilizing protocol and planning of delayed operation. For this study, patients were divided into two groups : group I was treated before 1984, and group II was treated from 1985 to 1996. In 1989, we started the new management using the stabilizing protocol for CDH patients who presented symptoms within the first 12 hours.

### *Stabilizing Protocol*

We treated patients in whom symptoms developed early according to the stabilizing protocol : (1) Minimum handling. (2) Sedation and muscle relaxant : At the induction, pancuronium 0.02~0.3 mg/kg and morphine chloride 0.05~0.2 mg/kg were injected in two shots, then maintenance doses of pancronium 0.03~0.1 mg/kg/hr and morphine chloride 0.1~0.2 mg/kg/hr were given. (3) Mechanical ventilation : either conventional ventilation (IMV) or high frequency oscillatory ventilation (HFO). At the start of HFO, parameters were set at $FiO_2$ 1.0, frequency 15Hz, mean airway pressure (MAP) 12 cm $H_2O$ and amplitude 20. (4) Gastrointestinal drainage using 8 Fr nasogastric sump tube. (5) Less water in the first 48 hours

of life, 40~60 ml/kg/day. (6) Control of blood pressure and urination with dopamine 3~10 μ g/kg/min and dobtamine 3~10 μ g/kg/min. (7) Monitoring of body temperature, electrocardiogram, blood pressure, urine volume, percutaneous gas pressure, and pulse oxy-meter (upper and lower extremities). (8) If persistent fetal circulation (PFC) occurred, at first we tried hyperventilation, venous injection of sodium bicarbonate, increase of sedative drugs and posture change. (9) If the patent fetal circulation could not be controlled, we used lipo-PGE1 (0.002~0.005 μ g/kg/min) and Imidalin (1~2 mg/kg/min; bolus injection and 0.25~2.0 mg/kg/min, continuous dose) starting with volume expander preparing for the decrease of blood pressure.

## Results

A total of 24 patients were studied 11 in group I and 13 in group II. The results are summarized in Table 1. In group I, the age at the time of diagnosis varied, and total survival rate was 72.7%. In group II, the age at the time of diagnosis was lower than that of the former group : prenatal = 5, within the first 12 hours = 4, 12 to 24 hours = 1 and after 24 hours = 3. Total survival rate was 61.5%, lower than that of group I. However, among the patients who developed symptoms within the first 12 hours of life, the survival rate of group II (44.4%) was better than that of group I (33.3%).

Data on the eight patients who died are summarized in Table 2. In all these cases, symptoms appeared before 6 hours, except for in one case (case 1). Two cases were right sided, and all defects were completely closed without any materials. Six patients needed preoperative respiratory support, which consisted of intratracheal intubation only in one patient, intermittent mechanical ventilation in four patients and high frequency osccillatory ventilation in one patient. Most of these patients died because of hypoplastic lung and postoperative induced PFC, but some of them could have survived if they had been treated by the delayed operation method.

The six cases that were treated by the stabilizing protocol and delayed operation are summarized in Table 3. Four of them were diagnosed by prenatal fetal ultrasonography. The stabilizing period was two to nine days. Five of these patients are alive and showing a good postoperative course. Unfortunately, ipsilateral pneumothorax occurred during the stabilizing period in one patient, who died 5 hours after birth. In this case, the mean airway pressure was too high to maintain better oxygenation, and the hypoplastic lung was damaged.

**Table 1.** Prognosis of Congenital Diaphragmatic Hernia

| Age st time of diagnosis | 1970−1984 alive | dead | (servival rate) | 1985−1995 alive | dead | (servival rate) | Total alive | dead | (servival rate) |
|---|---|---|---|---|---|---|---|---|---|
| prenatal | 0 | 1 | ( 0 %) | 4 | 1 | ( 80 %) | 4 | 2 | (66.7%) |
| <12 hrs | 1 | 1 | ( 50 %) | 0 | 4 | ( 0 %) | 1 | 5 | (16.7%) |
| 12−24 hrs | 1 | 1 | ( 50 %) | 1 | 0 | (100 %) | 2 | 1 | (66.7%) |
| 24 hrs≦ | 6 | 0 | (100 %) | 3 | 0 | (100 %) | 9 | 0 | (100 %) |
| | 8 | 3 | (72.7%) | 8 | 5 | (61.5%) | 8 | 3 | (72.7%) |

**Table 2.** Summary of Patients who Died of Congenital Diaphragmatic Hernia

| Case | Age Sex | BW (GA) | Symptoms | Preop. management (associated anomaly) | Defect (Organs) | Postop. course | Survival period |
|---|---|---|---|---|---|---|---|
| 1 | 15 hrs M | 2,020 g (36 w) | cyanosis dyspnea | IMV (6 hrs) | 5 x 4.5 cm (St·Sp·L·G) | IMV sepsis·DIC | 29 day |
| 2 | 5 hrs M | 2,020 g (37 w6d) | cyanosis | Oxygenation (ARM) | 3 x 4 cm : C (St·Sp·L) | IMV colostomy | 3 day |
| 3 | Pre.D. F | 2,860 g (36 w6d) | cyanosis abd. distension | intubation·$O_2$ (Duodenal Atresia) | Rt, 3 x 4 cm (L·St) | IMV | 4 hrs |
| 4 | 4 hr M | 2,897 g (37 w4d) | cyanosis retractive resp. | Oxygenation (multiple hemangioma) | 4 x 5 cm (St·Sp·L·G) | IMV DIC·cardiac failure | 83 days |
| 5 | at birth M | 1,680 g (38 w4d) | dyspnea | IMV | Rt, 3 x 3 cm (L·St) | IMV 18-trisomy | 3 days |
| 6 | 3 hrs M | 3,400 g (39 w) | dyspnea cyanosis | IMV | 4 x 3 cm (St·Sp·G·L) | sirculatory disturbance | 3 hrs |
| 7 | Pre.D. M | 3,660 g (41 w) | cyanosis | IMV | 5 x4 cm (St·Sp·G) | progressive acidosis | 9 hrs |
| 8 | at birth M | 2,740 g (38 w) | resp. arrest | IMY→HFO | NO | contralateral pneumothorax | 26 hrs |

IMV : Intermittent mechanical ventilation, HFO : High frequency oscillatory ventilation, ARM Ano-rectal malformation
Herniated organs ; St = stomach, Sp = spleen, L = liver, G - amall and/or large intestine
Case 1−3 : Goupe I , Case 4−8 : Groupe II

**Table 3.** Cases with delayed operation : Congenital Diaphragmatic Hernia

| Case | Sex | Time or Dx | Bw(g)/GA | Delivery | Apgar | Stabilizing period | Prognosis |
|---|---|---|---|---|---|---|---|
| 1 | ♂ | GA20w2d | 2,570 g/39w5d | vaginal | 6→7 | 4 days | alive |
| 2 | ♀ | GA20w1d | 3,100 g/39w3d | C/S | 4→7 | 7 days | alive |
| 3 | ♂ | GA27w5d | 3,180 g/39w3d | vaginal | 3→4 | 2 days | alive |
| 4 | ♂ | GA28w | 3,180 g/41w3d | C/S | 6→6 | 9 days | alive |
| 5 | ♂ | 15 hrs | 3,340 g/40w | vaginal | 8→9 | 3 days | alive |
| 6 | ♀ | 2 hrs | 2,648 g/41w | vaginal | 9→10 | 5 days | dead |

C/S : Caesarean section, Apgar Score (1 min.→5 min.)

## Discussion

In 1953, Gross[1] reported the first successful surgery of a case of CDH in which symptoms were expressed in the first 24 hours. At that time the most important physiological problem related to CDH was thought to be herniated organs depressing the lung, and until 1979, an earlier surgery was considered the best treatment. In the 1980s, it was suspected that the prognostic factor was the hypoplastic lung. Consequently, Miyasaka et al[2], proposed delayed operation for CDH, preventing PFC. They believed that emergency operation during the early stage was unwise because fetal circulation is shifting and unstable during this period.

Atkinson et al[3], reported the results of a multi-center trial, in which symptoms were expressed in the first 12 hours and needed intratracheal intubation. There was no difference between the emergency operation cases and the delayed operation cases, and they reported that delayed operation improved patient survival rate with or without extracorporeal membranous oxygenation (ECMO).

Patients with CDH that does not express any symptoms within 12 hours after birth generally have acceptable lung function and a survival rate close to 100% after operation. On the other hand, high-risk patients presenting respiratory distress within 12 hours after birth may have significant pulmonary hypoplasia, and there is a high mortality rate among such patients.

Several new techniques have been used in the treatment of CDH during the last decade. HFO has been successfully used in neonates who have severe respiratory distress and pulmonary hypertension. ECMO has been used preoperatively, perioperatively, and postoperatively in high-risk patients when conventional treatment has failed. A significant increase in survival rate when using ECMO has been shown in several reports[4,5,7]. An exception, however, are neonates with pulmonary hypoplasia caused by CDH or some other cause, in which HFO seems to be less successful[4,5]. Nevertheless HFO as early intervention, rather than rescue therapy, may be advantageous, since this avoids iatrogenic trauma induced by conventional ventilation. In our experience, one patient died during the stabilizing period because of iatrogenic barotrauma that led to contralateral pneumothorax. ECMO shoud be considered in case as such as this.

Nitric oxide (NO) is normally synthesized in endothelial cells. As a potent vasodilator, it is a principal regulator of vascular tone. Inhaled NO acts as a selective vasodilator of the pulmonary vascular bed because it is rapidly metabolized before it reaches the systemic circulation[6]. Its success rate may, however, be lower in CDH patients than in neonates in general.

The present investigation suggests that in our department the use of preoperative stabilization with HFO and/or conventional ventilation and delayed surgery improved the recovery rates of CDH patients whose symptoms were expressed in the first 12 hours. However, further investigation is needed to determine the clear parameters that can be used with blood gas data. Also, other methods, such as ECMO or NO inhalation, should be used in the more critical cases to improve the survival rate of the neonates with CDH in which symptoms are expressed within the first 12 hours.

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