Preoperative stabilisation in congenital diaphragmatic hernia

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Abstract
Between January 1983 and November 1986, 26 newborn infants with congenital diaphragmatic hernia were treated by early operation at a mean of 7 hours of age. A further 23 infants admitted between December 1986 and December 1989 were stabilised for a mean period of 40 hours before operation. There was no significant difference in survival between the two groups. Delayed operation is not detrimental to infants with congenital diaphragmatic hernia.

It is traditional for congenital diaphragmatic hernia to be repaired as an emergency procedure. The rationale of this approach is that removing the abdominal viscera from the chest allows expansion of the lungs with improved ventilation.¹ It has never been proved, however, that emergency repair of congenital diaphragmatic hernia is beneficial.

A disadvantage of early operation is that the surgical stress superimposed on an already unstable patient with congenital diaphragmatic hernia can precipitate or aggravate pulmonary hypertension. It was therefore suggested that delayed operation preceded by a period of stabilisation might be beneficial because of the decreased risk of pulmonary hypertension.¹⁻³ Cartlidge *et al* reported improved survival after stabilisation for four to 16 hours before surgery,² but this has not been confirmed by others.^{1,3} The purpose of this study was to assess the outcome in a group of patients who had delayed operation with preoperative stabilisation compared with that in patients who had conventional emergency repair of the congenital diaphragmatic hernia.

Patients and methods
All patients with congenital diaphragmatic

hernia admitted to Royal Liverpool Children's Hospital, Alder Hey between January 1983 and December 1989 were studied. The case notes were reviewed and gestational age, age at the onset of symptoms, age at operation, duration of preoperative stabilisation (if any), survival, and necropsy data on those who died were extracted.

Group 1 comprised the patients admitted between January 1983 and November 1986 all of whom underwent conventional emergency repair of congenital diaphragmatic hernia. The patients in group 2 were admitted between December 1986 and December 1989; these were stabilised before operation by paralysis with pancuronium, mechanical ventilation, intravenous infusion of crystalloids and colloids, and monitoring of blood gas tensions and ventilation. Pharmacological support, which included tolazoline, dopamine, and dobutamine, was used as indicated. Infants who did not improve clinically during the period of stabilisation were not operated on. After operation the supportive measures mentioned above were continued as indicated for patients in both groups. The differences between the two groups were compared with the Student's *t* test and χ^2 test; probabilities of <0.05 were considered significant.

Results
There were 49 patients: 26 in group 1 and 23 in group 2. The patients in the two groups were similar in terms of birth weight, maturity, side of defect, age at presentation, and incidence of associated anomalies (table).

Five patients died before operation in group 1 (19%) and seven in group 2 (30%); this was not significant. The remaining 37 patients underwent surgical repair of the diaphragmatic hernia and the results are summarised in the table. The mean age at operation in group 1 was 7 hours, and in group 2 was 47 hours. The survival rates of 54% and 48%, respectively, were not significantly different.

Necropsies were carried out on 15 infants, in nine of whom total lung weights were available. The mean (SD) total lung weight was 28 (17.07) g in group 1 (n=4) and 17 (8.56) g in group 2 (n=5); this was not significant. Normal total lung weight for a newborn is 60 (15) g.⁴

Discussion
It has long been appreciated that there can be deterioration even after the congenital diaphragmatic hernia has been repaired. Meticulous cardiopulmonary support of the neonate is

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Comparison of two groups of newborn infants with congenital diaphragmatic hernia. Values are expressed as number (%) unless otherwise stated

	Group 1 (early operation) (n=26)	Group 2 (delayed operation) (n=23)
Presentation:		
<6 hours	25 (96)	21 (91)
6–12 hours	1 (4)	2 (9)
Gestational age <38 weeks	3 (12)	3 (13)
Mean (range) birth weight (g)	2760 (947–4000)	2877 (1000–4100)
Associated major anomaly	2 (8)	3 (13)
Right sided hernia	3 (12)	4 (17)
Age at operation (hours):		
Mean (SD)	7 (3)	47 (40)*
Range	4–18	12–168
Total No who survived	14 (54)	11 (48)

*p<0.001.

critical and may improve survival in some cases.^{5,6} Pulmonary hypoplasia and persistent pulmonary hypertension are two important factors that determine the outcome in neonates with diaphragmatic hernias.^{1,2} Additional factors include associated congenital and chromosomal abnormalities.⁷ There is no treatment for pulmonary hypoplasia at present, apart from early prenatal repair of fetal diaphragmatic hernia which remains highly experimental, and neonatal lung transplant (which may be an option for the future). Any improvement in survival rates, therefore, should take account of factors that are likely to moderate pulmonary hypertension. Langham *et al* reported from the extracorporeal membrane oxygenation (ECMO) central registry that survival in critically ill neonates with a predicted 100% mortality was 58% when ECMO was used.⁸ ECMO improves survival mainly by resting the lungs and, possibly, reducing the risk of pulmonary hypertension. ECMO is not yet available in the United Kingdom. The appreciably lower total lung weight in most of our patients who died, particularly in group 2, indicates that pulmonary hypoplasia was the main cause of death.

Cartlidge *et al* reported that survival improved from 12.5% after early operation (done as soon as possible after birth) to 52.9% after stabilisation for four to 16 hours before operation.² This relatively short stabilisation period may not be adequate in haemodynamically labile patients and it is difficult to explain the impact of such a policy on survival rates. Furthermore, a survival rate of 12.5% after early operation is much lower than others have reported.^{3,6} Langer *et al* reported 61 neonates from Toronto and found a mortality of 58% in the early surgery group (n=31) and 50% in the delayed surgery group (n=30).³ From these results, which were not significantly different, they concluded that diaphragmatic hernia repair should be done only when the patient is satisfactorily stabilised. Our findings of 54% and 48% survival in the early and delayed surgery groups, respectively, corroborate the observations of Langer *et al*.

There are a number of possible reasons for

the similar survival rates in our two groups of patients. Firstly, the mean lung weights of infants who died did not differ significantly between the two groups, probably because of the small numbers involved; they were, however, lower in group 2, implying that these were higher risk patients than group 1. Secondly, prenatal ultrasonography has led to earlier diagnosis and transfer to neonatal surgical centres of infants with a poor chance of survival.^{5,9} This may have been a factor affecting improved survival in group 2, but we do not have adequate data to confirm this.

In summary, in patients with congenital diaphragmatic hernia, overall survival rates after preoperative stabilisation and delayed surgery are at least comparable with the results of early operation. It is possible that continued evaluation of preoperative stabilisation and delayed surgery may bring about an improvement in survival and also avoid operations on those who are not likely to survive.

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