Congenital Diaphragmatic Hernia : Don't Haste Surgery

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The management of congenital diaphragmatic hernia (CDH) in 12 neonates who had the onset of symptoms in the first 24 hours of birth was studied prospectively. The aim of study was to assess the efficacy of prior stabilization by conventional medical therapy and determine the optimum time for successful surgery. Arterial blood gas analysis showed pH below 7 in 5(41.7%) neonates, between 7 and 7.2 in 3 (25%) and more than 7.2 in 4 (33.3%) cases. Arterial pO2 was less than 40 mmHg in 4(33.3%) cases, between 40 and 80 mmHg in 6(50%) and more than 80mmHg in 2(16.6%) cases. The best pH obtained during stabilization period was 7 to 7.2 in 3(25%) and more than 7.2 in 9(75%) cases whereas best possible pO2 obtained was between 70-80mmHg in 2(16.6%), cases and above 80mmHg in 10 (83.3%) cases. Mean time period for prior stabilization was 36.2 hours. Mortality rate was 41.7% in neonates stabilized preoperatively whereas it was 93.4% where earlier policy of urgent surgery was adopted. The authors recommend that prior stabilization followed by delayed surgery results in a survival rate superior to that associated with early operation in neonates with CDH.

Key Words: Diaphragmatic Hernia, Pulmonary Hypertension, Congenital.

The treatment of CDH remains a challenging problem in neonates who are symptomatic at birth. It has undergone a revolutionary change from previous urgent surgery to the present practice of initial stabilization and delayed surgery. Persistent pulmonary hypertension (PPH), responsible for poor cardiovascular and pulmonary status is the major cause of gloomy outcome in these unfortunate babies [1]. Hypoxaemia due to PPH results in metabolic acidosis which itself aggravates PPH by causing pulmonary vasoconstriction. Pulmonary hypertension (PH) can be brought under control by interrupting this vicious cycle. It can be achieved by conventional medical therapy comprising of pharmacological agents and prolonged ventilatory support. Pulmonary vasodilators, inotropes and sodium bicarbonate are the drugs used to stabilize the cardiovascular and respiratory status in these neonates. However, controversy still continues regarding the duration of stabilization and optimum timing of surgery [2]. Other therapeutic measures used for stabilization of cardiovascular and respiratory status are extra corporeal membrane oxygenation (ECMO) and high frequency oscillatory ventilation (HFOV) [5,6]. Recent development for the management of poor risk cases of CDH are fetal surgery [7] and lung transplant [8].

The results of emergency surgery in our unit for patients of CDH symptomatic at birth were disappointingly poor. Facility of ECMO and NFOV are not available so the authors decided to study the effects of prior stabilization by conventional medical therapy on the outcome of surgery in all cases of CDH who were symptomatic on the first day of life.

Patients And Methods

The study was carried out at the department of paediatric Surgery Mayo Hospital Lahore from May 1991 to April 1992. A total number of 12 neonates were studied . All had the onset of symptoms in the first 24 hours of birth. Thirteen neonates with CDH who developed symptoms after 24 hours of birth were not included in the study. Detailed history was taken at the time of admission and special enquiry was made about respiratory distress, cyanosis at birth and the treatment received for them. A thorough clinical examination especially of respiratory and cardiovascular system was done on each patient.

Investigations carried out were complete blood picture, urine analysis, plain x-ray of chest and abdomen, serum electrolytes and arterial blood gases. After confirmation of the diagnosis following preoperative stabilizing measures were taken.

- All the neonates with CDH were ventilated mechanically after paralyzing them with pancuronium. Morphine was used to sedate them. Ventilatory settings were adjusted to induce respiratory alkalosis so as to minimize pulmonary vasoconstriction. Attempt was made to keep pH higher than 7.45 and pO2 as high as 100 mm Hg.
- Arterial lines were placed in 6 patients whereas transcutaneous electrodes were used as and when required to determine arterial pO2 and PCO2.
- Tolazoline 1-2 mg/kg/hour or chlorpromazine in a bolus dose of 1-2mg/kg was used for refractory hypoxemia.
- Dopamine or Dobutamine was used as inotrope.

Patients were subjected to surgery when the best possible condition as assessed by pH and pO2 was achieved. Transabdominal approach was adopted for surgery. The defect in diaphragm was closed with interrupted mattress silk sutures. No prosthetic material was needed as primary closure was feasible in all the cases. Postoperatively the patients were ventilated and stabilizing agents were used as and when required. Parenteral nutrition was started on 3rd postoperative day and continued till the patient was able to accept oral feeds. Patients were weaned off the ventilator when

improvement in ventilatory parameters was noticed.

Results

In a retrospective review of 2 years prior to the start of the study, records of all the cases of CDH presenting within the first 24 hours of life were examined. Urgent surgery without proper preoperative stabilization of the patients was performed in all 15 cases of CDH during that period with single (6.66%) survivor. Out of 12 cases of present series, adopting the protocol of delayed surgery preceded by a period of stabilization, 7(58.3%) survived. Out of 12 neonates there were 7 males and 5 females. Age at the time of presentation of patients is shown in Table I.

Only 1(8.3%) patient was premature, remaining 11(91.7%) were full term babies, All the patients had left sided CDH. Presenting symptoms are shown in table II, Arterial pH and pO2 at the time of admission prior to any stabilization are shown in table III and Iv respectively. Arterial pH and pO2 after stabilization a shown in table V & VI respectively. Mean interval between the admission and surgery was 36+2 hours. Cause of death was sepsis in 2 patients , disseminated intravascular coagulation in 1 and respiratory failure in 2 patients. One patient was admitted twice for adhesive bowel disease. There was no evidence of retrolental fibroplasia.

Discussion

The primary focus of therapy for CDH has been rapid reduction of hernia and repair of the defect in the diaphragm [9]. The rationale of early surgery was to remove herniated viscera from the chest as early as possible [10] . However even after apparently successful surgery for CDH, children continued to die of respiratory failure. This was found to be due to continuing hypoxemia and metabolic acidosis of PPH [11]. The anatomically abnormal pulmonary vascular bed in neonates with CDH is capable of showing exaggerated response to factors which produce pulmonary vasoconstriction resulting in progressive hypoxia [12].

Survival of patients with CDH symptomatic at birth has improved in past years with the introduction of a protocol of initial stabilization with conventional medical management followed by delayed surgical repair. Stabilization consisted of alkalinization of the patient's pH with a combination of hyperventilation and bicarbonate administration. During this period of stabilization. muscularized arterioles of already hyperplastic pulmonary circulation undergo vasodilatation [2,13]. Once this reactivity of pulmonary circulation subsides recurrence of pulmonary hypertension becomes very unlikely provided the precipitating factors for pulmonary vasoconstriction are avoided. Surgical repair of hernia in the presence of acidosis and hypoxia resulting from worsened respiratory mechanics could be one of the factors [15]. After paralysis and positive pressure ventilation hernia partially reduces making it seem unlikely that bowel exerts any significant pressure on the ipsilateral lung [14]. All children of CDH have some degree of bilateral pulmonary hypoplasia which seems to be a direct cause of respiratory and haemodynamic instability. [16] Children with CDH rarely improve after emergency operation , in fact many of them are significantly worse postoperatively [17] which is thought to be due to delayed onset of persistent fetal circulation . All these observations weaken the rationale for emergency repair of CDH. Most of the postoperative deaths in neonates with CDH are related to hypoxemia due to pulmonary vasoconstriction combined with myocardial failure. High pulmonary vascular resistance in CDH causes right to left shunting through intrapulmonary channels, foramen ovale or patent ductus arteriosus [18]. Pulmonary hypertension can be controlled with pulmonary vasodilators (morphine, tolazoline and chlorpromazine)[1,19]. Hyperventilation produces respiratory alkalosis which indirectly brings down the pulmonary vascular resistance. After prolonged ventilation intestines collapse and surgery can be performed easily because there is less difficulty in closing the abdomen [20].

Several authors have reported equal or improved results with delayed surgery without the availability of ECMO. Cartlidge et al [21] found a significant improvement in the survival from 12.5% with early repair to 52.5% in neonates stabilized preoperatively for 4 to 16 hours. Sigalet et al reported 100% survival without ECMO in 24 patients after stabilization and elective repair at an average age of 4.4 + 2.8 days. However studies presented by Nio et al [23] and Langer et al [14] did not show significant difference in survival rates of neonates undergoing delayed surgery from those having early operations. A survival rate of 38% with conventional medical stabilization of up to 48 hours in neonates who had the best post ductal pO2 of less than 100 mmHg and 93% with best Post ductal pO2 of more than 100 mmHg was reported by Pusic et al [24]. .In our series we could never obtain pO2 of 100 mmHg during stabilization period and a survival rate of 58% is comparable to that reported by Hazebroek et al [20] of 38% after a mean stabilization period of 10.5 hours.

We consider that the key to survival lies in the management of dangerous combination of acidosis and hypoxia. If acidosis is controlled, hypoxia is tolerated remarkably well. We attempted to minimize acidosis by enhancing peripheral and renal perfusion with low doses of dopamine (2-10 micro gram/kg/min) and limited hypoxia by prolonged ventilation. We recommend that all the neonates of CDH with impaired respiratory functions should be ventilated for a minimum of 30 to 36 hours or even longer. Return to full respiratory support for a further period may be needed postoperatively. Extra corporeal membrane oxygenation support plays important role in therapy protocol for neonates unresponsive to conventional treatment of stabilization [25] although Wilson et al found that overall survival of neonates with high risk CDH did not improve even with ECMO [26].

As far as the impact of predictive values of arterial blood gases on the survival of neonates with CDH is

concerned , we focus our attention on pH and pO2 as advocated by Biox-Ochoa [27]. The maintenance of pCO2 was not strictly regulated because prolonged hypocapnea leads to decreased cerebral circulation which might have dangerous effects like seizures or decreased intelligence later in life [28]. We find that 30 to 40 hours was a safe period for ventilation if patients behaved well throughout with little or no support of drugs. This study is significant in our set-up where ECMO is not available. Results obtained are encouraging. Increased awareness of the disease among doctors, early referral and prompt management will go a long way in improving the outlook of neonates with CDH.

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