Impact of a current treatment protocol on outcome of high-risk congenital diaphragmatic hernia.

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BACKGROUND: There is considerable debate regarding the optimal management of congenital diaphragmatic hernia (CDH) in high-risk infants (those cases presenting with respiratory distress within 2 hours of birth or those diagnosed prenatally). The aim of this study was to analyze patient outcomes using a new treatment protocol for CDH in a tertiary care non-extracorporeal membrane oxygenation (ECMO) neonatal unit. METHODS: The records of 78 consecutive neonates with CDH presenting to Bambino Gesù Children's Hospital from 1996 to 2001 were analyzed retrospectively. Of these infants, 70 high-risk patients were identified (prenatal diagnosis or respiratory distress requiring intubation and assisted ventilation within 2 hours after birth), regardless of associated anomalies, medical condition on presentation, or degree of pulmonary hypoplasia. A prenatal diagnosis was made in 46 of 70 (66%) patients. Associated lethal malformations were present in 6 of the children (8.5%). The patients were placed in 3 historical groups: group 1, 19 patients from 1996 to 1997, group 2, 22 patients from 1998 to 1999, and group 3, 29 patients from 2000 to 2001. In the first 2 groups, a new protocol was introduced using inhaled nitric oxide (iNO) and high-frequency oxygen ventilation (HFOV). In the third group, gentle ventilation and permissive hypercarbia were also used routinely. Mortality and severe morbidity--defined as O2 requirement at discharge, need for a tracheostomy, neurologic impairment, or bilateral hearing loss-were evaluated when the patients were at 6 months old. Univariate analysis was performed. RESULTS: The 3 groups were comparable with respect to predictive risk factors such as side of hernia, prenatal diagnosis, polyhydramnios, stomach and liver in the thorax, associated lethal malformations, and patch. Overall survival rate significantly increased from 47% (9 of 19) in group 1 and 50% (11 of 22) in group 2 to 90% (26 of 29) in group 3 (P =.02). None of the 19 patients in group 1 had severe morbidity compared with 2 of 22 (9%) patients in group 2 and 2 of 29 (7%) patients in group 3. Hearing loss was observed in 4 patients. Mortality rate and preoperative pneumothorax significantly decreased in group 3 compared with groups 1 and 2 (P =.03 and P =.00, respectively). CONCLUSIONS: (1) The application of new treatment protocol for CDH, using gentle ventilation and permissive hypercarbia, produced a significant increase in survival with concomitant decrease in morbidity. (2) The rate of pneumothorax was significantly decreased by the introduction of permissive hypercarbia and gentle ventilation. (3) As more infants survive CDH without the use of ECMO, severe long-term sequelae of CDH can be recognized in these children