The Japanese Approach to Congenital Diaphragmatic Hernia

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Introduction

Previously the outcome of antenatally diagnosed congenital diaphragmatic hernia (CDH) was thought to be poor as Adzik reported a 70% mortality rate in 38 cases. However, recent data published by the CDH Study Group on more than 400 infants did not reveal any association between prenatal diagnosis and survival. Similarly, in Japan, over 50% of the CDH cases are now antenatally diagnosed with improvements in prenatal ultrasound, and many centers including our institution have reported satisfactory outcomes in this patient population.

The Maternal and Perinatal Center at the Tokyo Women's Medical University, located in the center of Tokyo, serves as one of the tertiary neonatal centers in central Tokyo. All the CDH cases we have experienced for the past 5 years were diagnosed antenatally by obstetricians between the 21st and 36th week gestation, and mothers were referred to us for further evaluation and treatment with the wish that they would like to continue with their pregnancy. Although our center is a referral center for perinatal care of high-risk pregnancies and critically ill newborns, we do not provide extracorporeal membrane oxygenation (ECMO) therapy. Therefore, we need to maximize lung protective strategies from the birth of neonates with CDH to avoid the use of ECMO. To achieve this goal, we have developed a new management strategy for antenatally diagnosed CDH including: (1) delivery of a "depressed baby" (no breathing and swallowing) by cesarean section under general anesthesia; (2) exclusive use of high-frequency oscillatory ventilation (HFOV) in the NICU prior to surgery; (3) delayed surgery following short-term stabilization. We herein report our results (survival and long-term morbidity) of 12 consecutive patients who followed the above management protocol.

Prenatal management

Once a prenatal diagnosis of CDH was confirmed and the parents expressed their wish to continue their pregnancy, mothers were transported to our institution. They received genetic counseling including fetal karyotyping, and repeated ultrasound examination to determine the following: (1) the lung-thorax transverse area ratio (LT ratio), which is calculated by a simple ratio of right and left lung area to thorax area in a cardiac four-chamber view, to assess the severity of the pulmonary hypoplasia; (2) the presence of other congenital abnormalities, particularly those affecting the cardiovascular system, the central nervous system, and airway structures.

Serial measurements of LT ratio are also important because it changes with the progress of pregnancy. LT ratio of less than 0.25 indicates that the fetus has a severely hypoplastic lung and requires close attention to determine the timing of delivery.

Management during delivery

Antenatal diagnosis of CDH has not been thought to mandate the necessity for cesarean section for the delivery of CDH babies. Although they are routinely intubated on birth in the delivery room and are immediately transferred to the NICU, some neonates have already developed pneumothorax and/or intestinal distention on admission to the NICU. These complications are postulated to be attributed to the struggle or gasp during the initial resuscitation, bucking after intubation, and inadvertent high peak inspiratory pressure during transport. Since the neonates with CDH have pulmonary hypoplasia and a fixed number of alveoli, a single pneumothorax requiring a chest tube or mediastinal shift due to intestinal distension compressing the contralateral lung will result in a lung that is less capable of performing its function than it was prior to injury.

To circumvent these problems, we have attempted to provide transplacental anesthesia to deliver an antenatally diagnosed CDH baby depressed sufficiently enough to allow us to intubate the baby without breathing or swallowing effort. Our anesthetic management for cesarean section is, therefore, a general endotracheal anesthesia consisting of fentanyl (at least 10 µg/kg before the incision), propofol (target effect site concentration is 4 µg /ml), and muscle relaxant (vecuronium 0.3mg/kg for induction). Although the number of our cases is small, this anesthetic technique successfully prevented the baby from breathing and swallowing at birth.

Postnatal management

Regardless of the clinical presentation, high-frequency oscillatory ventilation (HFOV) was initiated on admission to the NICU, and was exclusively used before surgery. The patient received extensive medical therapies to achieve preoperative stabilization, which included: infusion of sedatives and muscle relaxants; inotropic support and volume infusion to maintain stable circulation; administration of pulmonary vasodilators such as prostaglandin E_1, or inhaled nitric oxide; administration of surfactant to improve oxygenation. Preoperative stabilization was defined by the following criteria: (1) normal hemodynamic variables with minimal circulatory support, (2) no signs of persistent pulmonary hypertension (PPHN) evidenced by no difference in pre-/post-ductal oxygen saturation; (3) adequate oxygenation and ventilation by minimal setting of HFOV ($F_1O_2 < 0.3$, mean airway pressure< 8 cmH$_2$O, peak to peak pressure level < 50 cmH$_2$O). Surgical correction was attempted only when the preoperative stabilization was achieved.

Preliminary results

From 1996 through 2001, we treated 12 patients following the above strategy and all completed a two-year follow-up observation period. All twelve neonates were delivered at median 37th week (range 36-39) of gestational age by cesarean section under uneventful general anesthesia. The median birth weight was 2.7 kg (range 1.9-3.3). Ten out of the 12 patients had LT ratio of less than 0.2, indicating a significant pulmonary hypoplasia. Eleven out of the 12 patients were delivered depressed sufficiently enough for smooth resuscitation and transfer to the NICU. Two patients were excluded from further analysis because

extensive medical support was withdrawn: one due to chromosomal abnormality (18 trisomy), and the other due to associated complex cardiac anomalies. Of the remaining 10 patients, 9 patients achieved preoperative stabilization to undergo successful surgical repair, and all survived. The remaining patient, the only baby that failed to be depressed during delivery (the mother received thiopental, succinylcholine, nitrous oxide, and sevoflurane for general anesthesia), showed pneumothorax and mediastinal shift on initial chest X-ray evaluation in the NICU and developed PPHN refractory to medical therapies; this patient eventually expired before surgical intervention. Of the 9 survivors, eight patients were discharged home without pulmonary complications and remained free of pulmonary morbidity during the two-year follow-up. Three developed postoperative gastroesophageal reflux and were successfully treated with medication. For the eight "fine" CDH survivors, the median time (postnatal age) to surgical repair, extubation, and discharge were 4 days (range 3-8), 18 days (range 10-25), and 33 days (range 25-67), respectively. The remaining patient, born with LT ratio of less than 0.1, experienced severe pulmonary morbidity including prolonged preoperative stabilization (60 days) and postoperative severe bronchopulmonary dysplasia, which led to failure of weaning off the ventilator and tracheostomy.

Summary

Iatrogenic barotrauma from harsh ventilation is now recognized to be an important factor for a high morbidity and mortality of CDH. Such recognition led to the hypothesis that transplacental anesthesia might smooth out the initial resuscitation before admission to the NICU, thereby improving the prognosis of antenatally diagnosed CDH. In this small case series, our CDH management achieved a high survival rate (90%) without the use of ECMO therapy, and all the CDH survivors were free from pulmonary complications.

References:

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