Congenital diaphragmatic hernia (CDH) is a cardiopulmonary anomaly that causes severe respiratory disorder. The incidence of CDH is estimated to be 1 in 600 to 18,000 births, with an average of 1 in 5,000 live births [1]. The hallmarks of CDH include hypoxia, a scaphoid abdomen, and evidence of bowel in the thorax by auscultation or radiography. Although surgical interventions and medical care have been developed over many years, the mortality rate from CDH remains in the range of 40% to 50% because of severe lung underdevelopment in these patients [2].

Early in fetal development, the pleuropertoneal cavity is a single compartment. If there is delayed or incomplete closure of the diaphragm, or if the gut returns early and thus prevents normal closure of the diaphragm, a diaphragmatic hernia will develop. Left or right posterolateral foramen of Bochdalek or the anterior foramen of Morgagni are the three possible diaphragmatic defects. Due to the later closure of the left side of the diaphragm than the right side, higher incidences of left diaphragmatic hernia are anticipated (90%) [2].

The anesthetic technique chosen depends on the size of the defect and the anticipated postoperative respiratory status. In general, anesthesia is maintained with low concentrations of volatile agents or opioids, muscle relaxants, and air as tolerated.

Thus far, very few articles about total intravenous anesthesia (TIVA) for CDH repair, and its relationship to lung dysfunction in CDH have been published. Our case provides valuable experience for further investigation.

CASE PRESENTATION

This female newborn was a full-term baby with a gestational age of 37 weeks and a body weight of 2,680 g. The mother was 34 years old with no evidence of gestational diabetes mellitus, eclampsia, or thyroid disease during pregnancy. She had no history of smoking, alcohol or drug use during pregnancy. The venereal disease research laboratories test and hepatitis B markers were all negative. The mother had...
received regular outpatient follow-up and the prenatal examination was normal.

However, after cesarean section delivery, the baby had an abnormal Apgar score of 7/8. Grunting and generalized cyanosis were noted despite the use of oxygen.

Neither meconium stain nor delay of initial crying was noted. The baby was admitted to our neonatal intensive care unit (NICU) and underwent endotracheal intubation. Chest roentgenography showed that bowel gas had accumulated in the left thorax, and bowel sounds were noted in the left thorax (Figure). Arterial blood gas data showed hypoxemia and respiratory acidosis. Under the impression of CDH, high-frequency oscillatory ventilation (HFOV) with inhaled nitric oxide (iNO) was used. The initial settings for HFOV were as follows: inspiratory/expiratory (I/E) ratio, 1:1; frequency, 15 times/second; mean airway pressure, 12 cmH₂O; fractional oxygen concentration in inspired gas (FiO₂), 100%; amplitude, 66 cmH₂O; stroke volume, 23 mL; and NO concentration, 20 ppm. After HFOV with iNO, the pH and partial pressure of carbon dioxide in arterial blood returned to normal. The partial pressure of oxygen in arterial blood (PaO₂) was more than 90%, but hyponatremia was noted. Two days later, surgical intervention was scheduled.

HFOV with iNO was used during surgery. Anesthesia was induced with ketamine (10 mg), rocuronium (1.5 mg), and fentanyl (3 µg). The maintenance hourly dosages were ketamine at 5 mg, rocuronium at 0.7 mg, and fentanyl at 3 µg by continuous intravenous infusion. No inhalational anesthetics were used. Hemodynamics were stable during surgery. Left thoracotomy was performed to repair the diaphragmatic defect. Complete repair of the diaphragmatic defect and return of the gut, stomach, and liver to the abdomen were confirmed by radiography. A small chest tube was placed into the left thorax. Blood gas analysis showed normal pH and PaO₂, but hyponatremia, metabolic acidosis, and hypocapnia were noted. Sodium bicarbonate 4.17 mEq was given according to the blood gas data. The process of surgery was smooth and the baby was returned to the NICU after surgery.

HFOV with iNO was used continuously until the day after surgery, then the ventilator was set to the conventional mechanical ventilation mode. The settings were as follows: I/E ratio, 1:2.4; rate, 35 times/minute; peak inspiratory pressure, 14 cmH₂O; peak end-expiratory pressure, 4 cmH₂O; and FiO₂, 40%. iNO was stopped at the same time. The circulation was improved after the operation. Unfortunately, right upper lobe pneumonia was noted 3 days later.

To wash out the accumulated CO₂, the ventilator was changed to the HFOV mode again until the next day. Two days later, the baby started to receive nasogastric tube feeding instead of total parenteral nutrition. She recovered and was discharged 1 month later.

**DISCUSSION**

In this case, we considered that TIVA was a more favorable technique than inhalational anesthesia for repair of the diaphragmatic defect because the three factors that affect inhalational anesthetic uptake (solubility in the blood, alveolar blood flow, and the partial pressure difference between alveolar gas and mixed venous blood [3]) are greatly affected by CDH. Since the embryologic diaphragm and lung develop at the same time, the degree of pulmonary hypoplasia is related to the timing of herniation of abdominal organs into the pleural cavity. The earlier the herniation, the more severe the pulmonary hypoplasia [2]. Based on the anatomic location of the defects in the diaphragm, the CDH...
is classified as eventration, diaphragmatic hernia, or absent diaphragm [1]. In this case, total compression of the left lung by the abdominal organs was observed on chest roentgenogram. The patient presented with the clinical signs of respiratory dysfunction (hypercapnia, respiratory acidosis, and hypoxemia) caused by the decrease in the number of alveoli and bronchial generations, and abnormal pulmonary vasculature [4,5]. Therefore, the uptake of inhalational anesthetics might be difficult with the increased intrapulmonary shunting in this baby. Also, bronchoconstriction may be easily induced because of irritation by inhalational anesthetics and the bronchial hyperreactivity of pulmonary hypoplasia [6], and the cardiovascular system might be compromised by the inhalational anesthetics. Therefore, TIVA rather than volatile anesthetic agents was chosen because anesthetics are more readily taken up via the intravenous route (peripheral or central venous catheter) and rapidly reach therapeutic plasma levels.

In neonates and infants, stroke volume is relatively fixed by a noncompliant and poorly developed left ventricle. The cardiac output is therefore very dependent on heart rate. Patients with severe pulmonary hypoplasia usually have fixed right-to-left shunting at the level of the ductus or patent foramen caused by persistent pulmonary hypertension (PPH) [7]. For maintenance of systemic vascular resistance and heart rate, ketamine seems an appropriate anesthetic agent because of its central stimulation of the sympathetic nervous system and inhibition of the reuptake of norepinephrine [8]. At the same time, we used fentanyl to stabilize the patient’s cardiovascular system and for analgesia.

In the past, when it was believed that the herniated contents caused lung collapse and respiratory failure, CDH was considered a surgical emergency. However, it is now clear that CDH is associated with pulmonary hypoplasia, abnormal pulmonary vascular reactivity, and pulmonary immaturity [9]. The pulmonary hypertension as well as pulmonary hypoplasia play major roles in respiratory failure, and lung compression by the herniated viscera is therefore considered a minor factor [10]. Thus, delayed surgery and preoperative stabilization to reverse the PPH are widely employed [11,12]. The time it takes to stabilize the condition varies from 24 to 48 hours in infants with only mild pulmonary hypertension and hypoplasia, to 7 to 10 days in neonates with severe pulmonary hypertension and hypoplasia [13]. Traditionally, conventional mechanical ventilation is used to hyperventilate the neonate with a low tidal volume and high respiratory rate (60–120/minute) to pH 7.55–7.60. Positive airway pressure during mechanical ventilation should not exceed 25 to 30 cmH2O to reduce the risk of barotrauma [4,7]. In recent years, HFOV seems to be a more favorable pre- and postoperative technique in CDH [14]. Cacciari and colleagues compared HFOV with conventional mechanical ventilation in CDH and demonstrated that HFOV treatment was a more valuable technique for preoperative stabilization and for intra- and postoperative respiratory treatment in newborns with CDH [15]. Bouchut et al confirmed that CDH can be safely managed using HFOV during anesthesia for the repair procedure [16].

NO is a strong vasodilator. The purpose of using iNO is to reduce the PPH without any systemic side effects. Many studies recommend the use of iNO for preoperative stabilization and postoperative treatment [17–20]. However, the effects of iNO on neonates with CDH remains controversial [21]. Although some infants show an improvement in oxygenation, most recently, the National Inhaled Nitric Oxide Study group found that iNO therapy did not reduce the need for extracorporeal membrane oxygenation (ECMO) or mortality in infants with CDH based on a randomized, double-masked, controlled, multicenter study [22]. Another study by Christou et al suggested that HFOV enhances the effectiveness of iNO in infants with PPH to reduce the need for ECMO [23]. The effect of iNO is still controversial, so further investigations to evaluate the benefit of using iNO in CDH must be undertaken.

**Conclusion**

From the relatively smooth surgical outcome in our case, we believe that the combination of HFOV with iNO and TIVA is a successful anesthetic technique for the surgical repair of CDH.

**References**


全靜脈注射麻醉應用於
先天性橫膈膜疝氣修補術 — 病例報告

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先天性橫膈膜疝氣是一種會導致嚴重心肺功能異常的先天性心肺畸形。傳統上，我們使用吸入性麻醉藥物合併人工呼吸器、鴉片類藥物以及肌肉鬆弛劑來維持手術所需要的重要性。在這個病例上，我們使用了全靜脈注射麻醉以及高頻震盪呼吸器合併一氧化氮來維持手術所需要的重要性。手術後，這名病人情況良好，並且於一個月後康復出院。

關鍵詞：先天性橫膈膜疝氣，全靜脈注射麻醉，高頻震盪呼吸器，吸入性一氧化氮
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