INTRODUCTION
Congenital diaphragmatic hernia (CDH) occurs in 1 of every 2000 - 3000 live births.1 Conventional ventilatory techniques are now being replaced with ventilatory techniques utilizing the concepts of permissive hypercapnia and high frequency oscillation ventilation.2,3 Despite optimized ventilatory management and supportive care, pulmonary hypertension persists in a subset of these patients. Although, inhaled nitric oxide (iNO) results in a reduction in pulmonary hypertension and improvement in oxygenation without any systemic side effects, its benefit in the infant with CDH remains controversial.3,4

In this report, the authors present the successful management of postoperative pulmonary hypertension by iNO in a newborn with CDH.

CASE REPORT
A male infant weighing 3000 grams delivered by caesarean section due to fetal distress at 39 gestational weeks. At first examination, he was cyanotic, tachypneic and had intercostal retractions. Auscultation of the lungs revealed poor air entry on the left, with a shift of cardiac sounds over the right chest. He was intubated and admitted to NICU because of severe respiratory distress.

The first chest X-ray of the patient showed intestinal loops in the left hemithorax with mediastinal shift to right (Figure 1). Therefore, a diagnosis of CDH was confirmed. Conventional pressure-controlled mechanical ventilation was initiated. Echocardiography showed patent foramen ovale, a small patent ductus arteriosus, and cardiac dextroposition.

The patient was operated on the second day of life. On laparotomy, there was a large (5 cm) posterolateral defect in the left hemidiaphragm with the spleen and intestines. There was also malrotation of the gut that was corrected. After reducing the viscera and repairing the diaphragmatic defect the patient was readmitted to NICU for postoperative care. While he was on conventional ventilation, his oxygen saturation was gradually decreased. Persistent carbon dioxide retention was detected on arterial blood gas analyses. Echocardiography showed that there was severe pulmonary hypertension (mean pulmonary artery pressure, mPAP = 68 mmHg). The ventilator mode was changed to high frequency oscillation (HFO) with an initial mean airway pressure of 12 cmH2O, a ∆P of 28 cmH2O, an oscillation frequency of 8 Hz and 100% oxygen together with 20 p.p.m. iNO. Subsequently, pulmonary hypertension was gradually reduced and dose of iNO was progressively decreased with no rebounds in mPAP and stopped 8 days from the start. The patient was discharged on the 26th day of surgery with full enteral feeding in good health.

DISCUSSION
CDH is a very serious congenital defect associated with high mortality rate. The syndrome is characterized by migration of abdominal viscera to thoracic cavity a variable degree of pulmonary hypoplasia associated with pulmonary hypertension, decreased cardiac output and impaired oxygenation.

The old management strategy of immediate surgery is now replaced by the principle of physiologic stabilization
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and delayed surgery. In this case, the patient was hemodynamically stable on conventional ventilation with a good blood gases condition before operation. However, pulmonary hypertension developed at early postoperative period.

Extracorporeal membrane oxygenation (ECMO) has been shown to salvage some of the most severely affected neonates with CDH complicated by pulmonary hypertension. Further insights into the pathophysiology of CDH and the introduction of less invasive therapeutic techniques such as HFO, iNO, surfactant, and perfluorocarbon liquid ventilation may replace the need for ECMO. However, the ideal treatment remains elusive.

Endogenous iNO is an important modulator of vascular tone in the pulmonary circulation. It reduces pulmonary hypertension, with improvement in oxygenation but no change in the systemic vascular resistance. However, its benefit in the infant with CDH remains controversial.

Neonates with CDH may have a poor response to iNO because of lung hypoplasia and abnormalities on the pathway NO-cGMP. Potential benefits should be expected by combined therapy with iNO and agents that act on the vascular metabolism pathway of the lung, such as sildenafil and prostacyclins. In this case, ventilatory mode was changed to HFO and iNO was started as soon as pulmonary hypertension was detected. The patient was successfully treated by iNO without need of combined therapy.

It is well-known that reduction in the pulmonary vascular bed with pulmonary hypoplasia associated with CDH is the major cause of treatment failure. As in this case, iNO may be useful in the management of pulmonary hypertension developed at early postoperative period.

REFERENCES


Figure 1: Chest X-ray showing intestinal loops in the left hemithorax with mediastinal shift to right.