Congenital Diaphragmatic Hernia

Background:

CDH is a defect in the diaphragm that is associated with a variable amount of intraabdominal organ extrusion. Incidence is about 1/3000 births. It is an early gestational event with incomplete closure of the diaphragm leading to bilateral pulmonary hypoplasia, pulmonary hypertension, and increased airway reactivity. The most common and largest diaphragmatic defect occurs through the left posterolateral pleuroperitoneal canal (foramen of Bochdalek) and accounts for 75% of all cases. The remainder occurs in the anterior foramen of Morgagni and paraesophageal locations. These infants can have profound hypoxemia, which causes right-to-left shunting through the ductus arteriosus. A Vicious cycle is set in place in which the already elevated pulmonary vascular resistance is further exacerbated by the severe arterial hypoxemia, hypercarbia, and acidosis. The ductus arteriosus remains patent and fetal circulation persists.

Pre-operative Management:

Current management is aimed at medically stabilizing the cardiorespiratory status of the newborn (correct hypoxia, acidosis, and cardiovascular stability) before surgical repair. The specific timing of the surgery is not as important as stabilization of the neonate's condition. Specific goal of preoperative medical management include achievement of a preductal oxygen saturation of at least 90% and correction of metabolic acidosis. The neonates are usually intubated shortly after birth and when mechanically ventilated the settings should be as low as possible to allow for moderate permissive hypercarbia to minimize ventilator-induced lung injury. Dopamine and milrinone may be needed to maintain hemodynamic stability. Surgery should be delayed until pulmonary vascular resistance and ventilation can be maintained with low peak inspiratory pressures and FiO2. Lung protective ventilation strategies include: Target SaO2 > 85%, permissive hypercapnia (PaCO2 <65 mmHg, pH >7.25), PIP < 25 cmH2O, PEEP 3-5 mmHg, and RR < 65. Inhaled nitric oxide and ECMO may be necessary.

Anesthesia Management:

If mechanical ventilation has not already been initiated, a rapid-sequence tracheal inbutaion will be performed. Induction medications appropriate for RSI in critically ill newborn include propofol/opiate and succinylcholine. Atropine may be added to prevent bradycardiac following administration of sux. Anesthesia can be maintained with low dose volatile anesthetic (if patient tolerates), opiates, and muscle relaxants. Two pulse oximeters (preductal and postducal locations) are useful to monitor the degree of shunting. Placement of preductal arterial cannula (right radial artery) is

recommended for monitoring blood pressure and acid-base status. Venous access should be avoided in the lower extremities because compression of the inferior vena cava may occur following reduction of the hernia. Nitrous oxide should be avoided because its diffusion into loops of intestine may result in distention and compression of functional lung tissue. After the abdominal contents are returned to the abdomen, an attempt to inflate the hypoplastic lung is not recommended. Expansion of the lung is unlikely and excessive airway pressures can damage the contralateral lung. Because these neonates have an underdeveloped abdominal cavity, abdominal closure may lead to increased intraabdominal pressures and compression of the inferior vena cava. Placement of a pulse oximeter to the lower extremity may indicate abdominal compartment syndrome and circulatory compromise.

Reference:

Hines, Roberta L. *Stoelting's Anesthesia and Co-Existing Disease.* Philadelphia: Elvsevier Saunders, 2012. p594-596.