Maintenance
- Protect end organs, especially heart, renal, and CNS, because they are often compromised by DM.

Extubation
- Awake. May require mechanical ventilation and ICU admission if pH less than 7.2, compromised mental status, and/or high risk of aspiration.

Adjuvants
- Same as for diabetes

Postoperative Period
- Potential for hypoglycemic injury from rapid increase in insulin sensitivity when surgical cause of DKA corrected.
- Subsequent medical management should be continued by physician with expertise in diabetes.

Anticipated Problems/Concerns
- Hemodynamic instability from combined volume deficiency, acidosis, and pre-existing CV disease
- CNS dysfunction from metabolic and lyte abnormalities (hypokalemia, hypophosphatemia), both early and late

Diaphragmatic Hernia (Congenital)

N. James Halliday | Jibin V. Samuel

Risk
- Occurs in approximately 1 in 2500–5000 births; 12–25% have associated anomalies, in particular cardiac (20%), chromosomal (5–16%), and neurologic.
- Parents who have one child with isolated defect have 2% chance with next child.
- Usually left sided (90%) due to defect in foramen of Bochdalek and are more common in boys. Morgagni hernias (2–5%) located anterior are more common in girls. Remainder through esophageal hiatus.

Perioperative Risks
- 30–60% mortality despite improved diagnosis and management
- Degree of pulm hypoplasia and associated CNS and CV malformations affect mortality
- Timing of diagnosis associated with the prognosis

Worry About
- Hypoxemia and acidosis
- Pulm Htn and CHD
- Shock
- Tension pneumothorax

Overview
- Classified by site of herniation
  - Posterolateral defects (Bochdalek) are left sided (largest and associated with greatest degree of pulm hypoplasia). Morgagni hernias rare; parasternal, less symptomatic, and therefore diagnosed at later age
- Between 4–9 wk of age the pleuroperitoneal membrane forms with the left closing after the right. In Bochdalek and Morgagni, normal development of the diaphragm and digestive tract does not occur
- Degree of lung hypoplasia determined by time of defect during fetal development and amount of abdominal contents in chest. Although ipsilateral lung is most affected, both lungs are abnormal and result in decreased numbers and function of alveoli; hypoplastic lung with smaller pulm artery and decreased arterial branching causes high vascular resistance

Usual Treatment
- Initial treatment involves determining the severity of associated congenital anomalies and degree of illness.
- Goal is semielective surgery when pt is medically stable.
- Posterolateral defects require surgical repair (does not resolve the pulmonary dysfunction).
- Small defects closed primarily; larger defects use artificial diaphragm, which contributes to postop resp failure.
- In most cases, abdomen is closed primarily after correction but a silastic pouch may be used with increased intra-abdominal pressures.
- Fetal surgery has been accomplished in those severely affected with increased degree of pulm hypoplasia. FETO can be done to trigger lung growth. The Cochrane group reviewed two studies looking at a total of 97 women who underwent FETO and found insufficient data indicating improvement in perinatal mortality.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Mediastinal shift, associated ASD, VSD, coarctation, tetralogy of Fallot (23%)	Displaced cardiac impulse	CV exam	ECHO
RESP	Resp distress, pulm Htn	Decreased breath sounds on affected side Prominent ipsilateral chest	Pulm exam	CXR, ABG
GI	Malrotation, atresia (20%)	Scaphoid abdomen	Abdominal exam	
GU	Hypospadias		Inspection	
CNS	Spina bifida, hydrocephalus, anencephaly (28%)		Inspection and neurologic exam	US, CT
METAB	Acidosis, hypoxemia, hypercarbia			ABG

Key References: Aggarwal S, Stockmann P, Klein MD, et al.: Echocardiographic measures of ventricular function and pulmonary artery size: prognostic markers of congenital diaphragmatic hernia? *J Perinatol* 31(8):561–566, 2011; Crivell RM, Andersen C, Dodd JM: Prenatal treatments for babies with congenital diaphragmatic hernia, *Cochrane Database Syst Rev* 11:CD008925, 2015.

Perioperative Implications

Perioperative Management
- ECMO provides temporary support until perinatal circulation matures and less sensitive to vasoconstrictive stimuli (1–2 wk).

Preoperative Preparation
- Avoid triggers for pulm vasoconstriction.
- Goals include a PaO_2 >80, $PaCO_2$ 25–30, normal or elevated pH, and normothermia (hypothermia increases O_2 consumption).
- For pulm Htn, can also use nitric oxide 20–80 ppm, sildenafil.
- Avoid gaseous distension of stomach with early placement of NG tube.
- With compromised neonates, ET intubation, sedation, paralysis, and ECMO may be required if conventional or high-frequency ventilation fails.
- All neonates with resp distress require invasive monitoring using preductal right radial a-line. If severe, consider preductal and postductal A-line and pulse oximetry.
- IV access best in upper extremities to avoid possible IVC obstruction from increased intra-abdominal pressure.
- Watch for pneumothorax (sudden deterioration in BP or oxygenation); consider prophylactic contralateral chest tube; equipment needed should be available.

Anesthetic Technique
- Opioids well tolerated; inhaled halogenated anesthetics may cause significant hypotension; avoid N_2O, which distends gas-filled intestines.
- Avoid peak pressures more than 25 cm H_2O.
- High frequency, low tidal volume preferred.
- Continue nitric oxide if given preop.
- Lung mechanics change during surgery; may require hand ventilation.

Postoperative Period
- Continued muscle relaxation/opioids and ventilatory support.
- Once stable, assess need for continued resp support.
- If A-aDO_2 gradient >400 mm Hg or if cardiopulmonary deterioration, continue resp assistance.
- Persistent hypoxemia while on high FIO_2 suggests persistent pulm Htn with R-to-L shunting.
- Minimize ET suctioning, correct metabolic acidosis.
- Deliver adequate nutrition.
- High degree of neurologic problems, whether or not infants placed on ECMO; seizures, developmental delay, and hearing loss in 20–30%, but pulm outcomes are usually good.