Duchenne Muscular Dystrophy (Pseudohypertrophic Muscular Dystrophy)

Stephanie Black

Risk

- X-linked recessive; 1:3500 live male births; few known cases in females.
- Often undiagnosed until age 3–5 y; periop complications can occur before diagnosis.
- Deterioration through puberty to death usually before age 25 y.
- Periop risks may be present in female carriers.

Perioperative Risks

- Respiratory failure, prolonged mechanical ventilation
- Cardiac failure (CHF or arrhythmias)
- Hyperkalemia and rhabdomyolysis

Worry About

- Poor cardiac contractility, dilated cardiomyopathy, cardiac arrhythmias, pulm Htn from sleep apnea, MVP
- Poor respiratory function, restrictive lung disease from scoliosis, chronic pneumonia
- Aspiration risk from gastroparesis and dysphagia
- Possible hyperkalemic arrest with succinylcholine and volatile agents
- Associated with malignant hyperthermia-like syndrome unresponsive to dantrolene

Overview

- Most boys die from pneumonia, but heart failure is usually present by adolescence.
- Gradual onset of muscle wasting, replaced by fat/fibrosis, causing pseudohypertrophy.
- Hyperkalemic response to depolarizing NMBs may develop years before the onset of DMD symptoms; the prediagnosis infant may present with only mild gross motor delay.
- Increased sensitivity to nondepolarizing NM blockers.
- Use of Ca^{2+}-channel blocker (e.g., verapamil) may prolong or even cause NMB.
- Resting tachycardia common; cardiac involvement in 70% of cases, cardiac debilitation usually late.

Etiology

- X-linked recessive disease causing absence of dystrophin, destabilizing the sarcolemma
- Muscles (including myocardium) gradually replaced with fat and connective tissue

Usual Treatment

- Corticosteroids may increase strength and delay progression.
- Spinal rodding and fusion for scoliosis may increase comfort and ease of wheelchair use; pulmonary deterioration continues, and life may be only minimally prolonged.
- Tendon releases for contractures.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Conduction: heart block, SVT, prolonged QT Cardiomyopathy: Ventricular dilation or fibrosis, pHTN	Tachycardia CHF Sx: Orthopnea, DOE	Opening snap (MVP) CHF: crackles, JVD, edema	ECG ECHO Cardiac MRI
RESP	Decreased volume and flows Restrictive lung disease Sleep apnea Recurrent pneumonia/aspiration	SOB Snoring, apneic spells	Hypoxia Crackles Poor inspiratory effort	PFTs CXR SpO_2 on RA, polysomnogram
GI	Dysmotility, gastric dilatation, paralytic ileus			
GU	Bladder paralysis, impotence			
CNS	Decreased IQ		Mental status exam	
MS	Scoliosis, kyphosis Contractures Muscle destruction Macroglossia Poor IV access	Gross motor delay Progressive weakness Exercise intolerance	Pseudohypertrophy of calves, wheelchair dependence	Spine films Elevated CK levels (20–100× normal)

Key References: Birnkrant DJ, Panitch HB, Benditt JO, et al.: American College of Chest Physicians consensus statement on the respiratory and related management of patients with Duchenne muscular dystrophy undergoing anesthesia or sedation, *Chest* 132(6):1977–1986, 2007; Hayes J, Veyckemans F, Bissonnette B: Duchenne muscular dystrophy: an old anesthesia problem revisited, *Paediatr Anaesth* 18(2):100–106, 2008.

Perioperative Implications

Preoperative Preparation

- ECHO, ECG, and pulm function tests preop.
- Avoid or limit preprocedure sedation.

Monitoring

- Consider invasive cardiac monitoring based on EF and surgical procedure.
- Nerve stimulator.

Induction

- Succinylcholine contraindicated because of hyperkalemia risk.
- Limit volatile anesthetic exposure secondary to MH-like response.
- Avoid depressants of cardiac contractility; minimize arrhythmogenic medications.
- Consider regional technique to avoid the risks of GA.

Maintenance

- Variable response to NM blockers; titrate to effect.
- Consider using a "nontriggering" technique (MH precautions, TIVA, "clean" machine).
- Optimize regional or neuraxial blocks to minimize GA exposure.
- Avoid hypoxemia, large fluid shifts, and anemia to prevent uncompensated cardiomyopathy.

Emergence

- Potential for prolonged ventilator dependence greatest when vital capacity <30% of predicted.
- For GA cases, consider extubating directly to BIPAP and/or CPAP, weaning later.
- Outpatient surgery may be inadvisable due to late respiratory depression (cause unclear).
- Avoid postop shivering, which may cause rhabdomyolysis.

Anticipated Problems/Concerns

- Respiratory failure.
- Cardiomyopathy, CHF.
- Supraventricular tachydysrhythmias.
- Rhabdomyolysis, hyperkalemia, and cardiac arrest in response to succinylcholine and volatile agents have been described in boys years before clinical signs of DMD present.
- MH-like picture may be unresponsive to dantrolene.

Duodenal Atresia

Lynne G. Maxwell

Risk

- Incidence 1:5000–10,000 live births
- Male to female incidence is equal
- Trisomy 21 in 20–30%
- 45% are premature infants of pregnancy complicated by polyhydramnios
- Incidence of polyhydramnios 32–81%
- Mortality 3–5%; due not to duodenal atresia but to associated CHD or prematurity

Perioperative Risks

- Hypoxemia associated with immature lungs
- Hypoxemia due to CHD, persistent fetal circulation (pulm Htn)

Worry About

- Ventilation problems associated with prematurity.
- Other associated anomalies in 50% of cases: esophageal atresia (7%), other intestinal atresias, renal anomalies (5%), malrotation of the gut (30%), volvulus, imperforate anus (3%), annular pancreas (25%).

- CHD associated with trisomy 21 (ASD, VSD, AV canal).
- Aspiration on induction of anesthesia secondary to bowel obstruction.
- May be associated with cystic fibrosis.
- Late presentation can be associated with dehydration, hypovolemia, and hypochloremic alkalosis.

Overview

- Frequently premature infant of pregnancy complicated by polyhydramnios.
- Polyhydramnios may occur in the absence of premature birth.
- Diagnosis frequently made by prenatal ultrasound, allowing for parental counseling and planning for early repair; may not be detected until 28–32 wk gestation because of delay in development of proximal duodenal dilation.
- Vomiting after birth: May be copious and bile stained. If obstruction is proximal to ampulla of Vater, emesis is nonbilious.
- Flat abdomen.
- Dx is made by "double bubble" on abdominal x-ray (dilated stomach and proximal duodenum).
- Obstruction may be caused by partial obstruction: stenosis with perforated web or diaphragm rather than atresia; Dx may be delayed.
- Coexisting distal intestinal atresia is rare (<1%).

Etiology

- Unknown in sporadic cases
- More common in trisomy 21

Usual Treatment

- Fluid resuscitation with correction of any electrolyte abnormalities should precede surgery (especially in cases of prolonged vomiting).
- Surgical repair (duodenoduodenostomy) is curative; associated malrotation should be ruled out.
- Surgical technique may be open laparotomy or laparoscopic.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	CHD—ASD, VSD, AV canal Persistent fetal circulation	Trisomy 21	Murmur Cyanosis	ECHO CXR Pulse oximetry
RESP	Respiratory distress syndrome of prematurity	Polyhydramnios Gestational age <36 wk	Tachypnea Retractions Flaring Grunting	CXR Pulse oximetry
GI	Duodenal obstruction Associated esophageal atresia	Bilious vomiting No gas in abdomen	Scaphoid abdomen	Abdominal x-ray Unable to pass OG tube
RENAL	Structural anomalies		Palpation of kidneys	Abdominal ultrasound

Key References: Aguayo P, Ostlie DJ: Duodenal and intestinal atresia and stenosis. In Holcomb III GW, Murphy JP, Ostlie DJ, editors: *Ashcraft's pediatric surgery,* ed 6, Philadelphia, PA, 2014, Elsevier, pp 414–429; Olsen M, Avery N, Khurana S, et al.: Pneumoperitoneum for neonatal laparoscopy: how safe is it? *Paediatr Anaesth* 23(5):457–459, 2013.

Perioperative Implications

Preoperative Preparation

- OG tube to decompress stomach and reduce gastric contents
- IV catheter placement with hydration (20 mL/kg NS) if Dx delayed beyond 24–48 h; correct electrolyte abnormalities.
- Surfactant for premature infants with significant lung disease.
- Confirm intramuscular vitamin K given as part of normal newborn care.

Monitoring

- Arterial monitoring for ABGs, electrolyte, and Hgb determination only in premature infants with significant lung disease, those with CHD, or those with extreme dehydration due to protracted vomiting; otherwise noninvasive BP sufficient as minimal blood loss expected.
- Temperature.
- Urinary catheter (small feeding tube) may be helpful in assessing adequacy of fluid resuscitation.
- Pulse oximetry, ECG, and end-tidal carbon dioxide and gas monitoring; preductal and postductal pulse oximetry in pts with congenital heart disease or persistent fetal circulation.

Anesthetic Technique

- Suction OG tube with infant supine and in left and right decubitus positions prior to induction and intubation.
- Awake intubation after preoxygenation only for actively vomiting, volume-depleted infants with abnormal airway anatomy.
- Rapid-sequence induction after preoxygenation for normovolemic pts with normal airway anatomy.
- Avoid N_2O to prevent intestinal distention.
- Nondepolarizing muscle relaxant helpful for surgical exposure.
- Second peripheral IV after induction.

Airway

- Precautions to prevent aspiration
- Abnormal airway anatomy unlikely

Preinduction/Induction

- Pt may be hypovolemic due to vomiting and/or poor feeding.
- Correct dehydration, hypochloremic alkalosis (failure to do so can shift oxyhemoglobin dissociation curve to left and reduce O_2 delivery to tissues).
- Debubble IV lines to prevent paradoxical air embolism.
- Type-specific blood available for transfusion (rarely needed).

Maintenance

- Mechanical pressure ventilation with rate 20–25, PIP 20–25, PEEP 2–5 to achieve adequate ventilation (tidal volume 8–10 mL/kg).
- Air/O_2 mixture to achieve O_2 saturation 92–96%, although some use 100% O_2 to provide reserve; data on retinopathy of prematurity due to operative exposure to 100% O_2 is not conclusive, and surgical retraction may restrict ventilation and cause atelectasis, which can cause desaturation.
- Surgical retraction/pressure on liver may decrease venous return and cause hypotension.
- Hemorrhage and/or air or carbon dioxide embolus has been reported prior to or after trochar insertion when laparoscopic technique is used; this may result in CV collapse, requiring CPR. Resuscitation drugs should be available.
- Cease insufflation of abdomen and evacuate gas from abdomen, left-side down/Trendelenburg position, 100% O_2, fluid administration, epinephrine bolus, cardiac compression if no cardiac output.
- Fentanyl or remifentanil/vecuronium or vecuronium/isoflurane or sevoflurane for premature infants.
- In full-term infants who may be immediately extubatable, consider caudal catheter threaded to low thoracic position. Dose with bupivacaine 0.25% (or ropivacaine 0.2%) with 1:200,000 epinephrine 0.5–0.75 mL/kg followed by continuous epidural infusion of 0.1% bupivacaine or ropivacaine at 0.2 mL/kg/h for postop pain relief.

Extubation

- May require postop ventilation if pt premature or has CHD.
- Full-term infants with effective epidural anesthetic and no or low-dose opioid administration may be extubated if effective spontaneous ventilation.

Anticipated Problems/Concerns

- Prematurity/respiratory distress syndrome/apnea.
- CHD.
- Hemorrhage, air, or gas embolus may occur at start of laparoscopic procedure.
- Risk of aspiration may continue postop; leave OG or NG tube in place.
- Later risk of GE reflux higher than normal (17%).
- Adequate fluid replacement.
- Other associated anomalies.