Chapter 19: Anesthesia for Gastrointestinal Procedures

Kallol Chaudhuri

INTRODUCTION

FOCUS POINTS

1. Pyloric stenosis is a medical and not a surgical emergency managed first by volume repletion. Hypochloremic, hypokalemic metabolic alkalosis is the classic derangement and once corrected, pyloromyotomy is the surgical intervention of choice.
2. An international randomized multicenter trial did not find any strong evidence that infants exposed to general anesthesia for 1 hour during inguinal herniorrhaphy had any measurable neurocognitive or behavioral deficits at 5 years of age.
3. Insufflation of the peritoneum with CO_2 for laparoscopic procedures leads to increased intra-abdominal pressure (IAP) with sometimes clinically significant cardiorespiratory, neuro, and renal physiological changes.
4. Intussusception is the most common abdominal emergency in patients less than 2 years of age with classic presentation of abdominal pain, and bloody (currant jelly) stool.
5. Gastroschisis is usually an isolated abdominal defect, whereas omphaloceles are usually associated with a wide variety of abnormalities (cardiac defects, congenital diaphragmatic hernia, chromosomal anomalies).
6. Congenital diaphragmatic hernia (CDH) presents at birth with respiratory distress-tachypnea, grunting, use of accessory muscles, and cyanosis. Prognostic indicators include the degree of lung hypoplasia and the presence of pulmonary hypertension.
7. The classic presentation of appendicitis includes periumbilical abdominal pain with anorexia, nausea, and vomiting. Leukocytosis and fever may be present. Transabdominal ultrasound (US) is the most common diagnostic modality.
8. Hirschsprung disease or congenital intestinal agangliosis is the most common cause of intestinal obstruction in neonates. Neonates who fail to pass meconium within 48 hours of life should be assessed for this disorder.

A wide variety of gastrointestinal surgeries for various types of clinical disorders are performed in pediatric patients. Since the majority of disorders involve neonates and young infants, a general understanding of the embryological development would be helpful to appreciate the pathology and the clinical presentations of those disorders.

In this chapter, we will provide clinical presentations and treatment options, and brief descriptions of the anesthetic management of several pediatric diseases related to the gastrointestinal system: pyloric stenosis, inguinal hernia, umbilical hernia, laparoscopic surgery, intussusception, gastroschisis, omphalocele, congenital diaphragmatic hernia, appendicitis, aganglionic megacolon (Hirschsprung disease).

PYLORIC STENOSIS

Hypertrophic pyloric stenosis is the most common gastrointestinal surgical disorder in neonates. This disorder was first reported in 1717 by Patrick Blair, but a complete description with autopsy findings was then described by Harald Hirschsprung in 1887. The operative procedure of pyloromyotomy for pyloric stenosis was first described by Ramstedt in 1912.

The incidence of pyloric stenosis (PS) is 0.9 to 5.1 per 1000 live births[1] and is more common in first born male infants. It usually presents at 3 to 8 weeks of age. The primary pathology is segmental hypertrophy of circular muscle fibers of the pyloric region of the stomach, resulting in gastric outlet obstruction. No definite inheritance pattern is noted, but the possibility of polygenic mode of inheritance pattern influenced by environmental factors has been suggested. Erythromycin, a motilin antagonist,[2] and maternal smoking[3] have also been implicated to play a role in the development of PS.

Symptoms of PS begin in an otherwise healthy infant with nonbilious projectile vomiting that starts immediately following feeding. As vomiting continues, the progressive loss of gastric acid (hydrogen and chloride ion) and fluid leads to hypochloremic metabolic alkalosis. Loss of intravascular fluid initiates increased absorption of sodium in exchange for potassium at the renal tubules, stimulated by aldosterone. The classic metabolic abnormality is hypochloremic, hypokalemic metabolic alkalosis. Urine may be alkaline due to systemic alkalosis. However, due to persistent hypokalemia, H^+ is exchanged in renal tubules for Na^+ to preserve K^+. The loss of H^+ causes paradoxical acidic urine with a systemic alkalosis. At later stages, persistent dehydration may lead to poor tissue perfusion, lactic acidosis, and metabolic acidosis. The classic finding of “olive,” an olive-sized mass at the upper abdomen, is occasionally missing in PS patients. Confirmation of diagnosis is commonly done by ultrasound examination which has replaced the traditionally performed gastrointestinal barium study.

Management

Pyloric stenosis is a medical and not a surgical emergency. Replacement of intravascular volume and correction of electrolyte abnormalities (hypochloremia, hypokalemia, alkalosis) are essential prior to surgical correction. A tentative target of laboratory values includes serum bicarbonate >30 mEq/L, serum chloride >100 mEq/L, serum sodium >132 mEq/L, serum potassium >3.2 mEq/L, and serum pH 7.30 to 7.45.

Surgical correction of choice for PS patients is pyloromyotomy which includes incising the muscles of pylorus up to the layer of submucosa, leaving the mucosa intact. Currently, laparoscopic pyloromyotomy is preferred over open pyloromyotomy. The laparoscopic approach, compared to open approach, allows early initiation of postoperative feeding and shorter hospital stay.

Anesthetic Management

1. Standard ASA monitors should be placed prior to induction.
2. Patency of existing IV access should be confirmed.
3. A large orogastric catheter should be placed for decompression of stomach (four quadrant suctioning) prior to induction of anesthesia.
4. Glycopyrrolate (10 mcg/kg) may be considered to prevent reflex bradycardia during laryngoscopy.
5. Induction:

   Inhalation induction: Although these patients are at high risk for pulmonary aspiration, inhalation induction with oxygen and inhalation agent (sevoflurane) has been reported to be a safe technique.[4]

   Intravenous induction: Modified rapid sequence induction with low airway pressure ventilation is a popular method. The drugs used are propofol (2–3 mg/kg) or a combination of propofol (1–2 mg/kg) and ketamine (0.5–1 mg/kg). Ketamine by itself, at a dose of 1 to 2 mg/kg, has been shown to be an effective induction agent with minimum hemodynamic changes. Succinylcholine (2 mg/kg) or rocuronium (0.5–1 mg/kg) is commonly used as muscle relaxant. Rapid sequence induction with propofol (2.5–3 mg/kg) and remifentanil (2.5–3 mcg/kg) without any muscle relaxant was also used[5] for induction in pediatric patients, but hemodynamic changes, especially hypotension, are a concern in neonates.
6. Maintenance of anesthesia is achieved by using sevoflurane or desflurane in combination with oxygen and air. Nitrous oxide and remifentanil (2 mcg/kg/min) without any inhalation agent were also used without any significant hemodynamic alteration in open pyloromyotomy.[6] Nitrous is usually avoided in laparoscopic pyloromyotomy as it may cause distension of bowel. Intraoperative opioids (fentanyl) are also avoided because they may prolong emergence. Opioids can also increase the risk of postoperative apnea, a unique complication in these patients, thought to be related to persistent elevation of CSF pH, a determinant of respiratory drive.[7]

Relative intraoperative hypotension (systolic blood pressure <20% below baseline) was reported to be routine (in more than 70% patients less than 2

months old) and absolute intraoperative hypotension (mean arterial BP <35 mm Hg) was also reported to be common (12–21% in babies less than 2 months) during general anesthesia in patients undergoing laparoscopic pyloromyotomy.[8] This is of significant concern as these group of patients are most vulnerable to cerebral hypoperfusion.

Spinal anesthesia has been used successfully in laparoscopic pyloromyotomy. The advantages include avoidance of airway manipulations and related complications, avoidance of the debatable role of general anesthesia as neurotoxin in neonates, better hemodynamic profile with higher intraoperative blood pressure, and rapid recovery after surgery.[9,10] The technique was performed at L3-L4 level with a 22-gauge needle using hyperbaric bupivacaine (1 mg/kg).[9]

Postoperative analgesia is usually provided by

1. Infiltration of surgical wound by local anesthetic
2. Intravenous acetaminophen (15–20 mg/kg) preferable or rectal administration (30 mg/kg, variable absorption)

Opioids, such as morphine or fentanyl, are rarely needed for postoperative analgesia. Most patients tolerate oral feedings within a few hours after surgery.

INGUINAL HERNIA

Inguinal hernia is one of the commonest clinical conditions which requires surgical correction in pediatric patients. The overall incidence varies between 3.5% and 5% in term infants but is higher (13%) among the preterm infants.[11] It is more common in boys.

The majority of pediatric hernias are indirect in nature and result from failure of obliteration of the processus vaginalis. Processus vaginalis is a peritoneal diverticulum which precedes the testis in its journey to relocate from retroperitoneum to the scrotum at 25 to 33 weeks of gestation. The processus vaginalis normally fuses at 36th weeks of gestation and closes the internal ring. In cases where the processus vaginalis does not involute and remains patent, fluid or intraabdominal contents can migrate through the sac creating a hydrocele or indirect hernia, respectively.

The hallmark presentation for an inguinal hernia is a swelling at the groin or in the scrotum, the size of which increases with increased intraabdominal pressure (with crying, coughing). If the hernia becomes incarcerated, it presents with an irreducible tender erythematous swelling with symptoms of irritability, abdominal pain, and vomiting. The incarceration of intestines may lead to intestinal obstruction with worsening of previous symptoms along with abdominal distension, fever, and leukocytosis.

Because inguinal hernia usually does not resolve spontaneously and carries the risk of incarceration of bowel and testicular atrophy, all inguinal hernias in pediatric patients, especially in infants, are recommended to undergo surgical repair. However, the decision and timing of operative repair must be balanced against the overall medical condition of the child and potential anesthetic complications. Prematurity, which is associated in a number of these patients, can make the operative procedure more challenging. Early elective herniorrhaphy is also recommended in these patients; delaying the repair 1 week after diagnosis[12] or performing surgery beyond 40 weeks of conceptual age[13] significantly increases the risk of incarceration, strangulation, and intestinal obstruction.

In most cases, the surgical repair is carried out by laparoscopy; the advantages of laparoscopic repair include smaller incision and scar, shorter postoperative stay, and lesser need for postoperative analgesia. Laparoscopic approach also offers an option to assess the contralateral side without any significant injury to vas deferens and related blood vessels.

Anesthetic Management

Both general anesthesia and regional anesthesia have been used successfully for inguinal hernia repair.

General Anesthesia

General anesthesia with both endotracheal tube and laryngeal mask airway has been successfully used for repair of uncomplicated inguinal hernia. Endotracheal tubes are commonly used in laparoscopic procedures to provide adequate muscle relaxation and effective ventilation and in

complicated hernia repair. LMA (Proseal) has been successfully used in laparoscopic hernia repair of short duration, with average surgical time of 15 minutes and average total anesthesia time of half an hour.[14]

Premedication: Midazolam (oral 0.5 mg/kg; IV 0.05 mg/kg) as indicated

Standard ASA monitors

Induction: Inhalation induction with oxygen/nitrous/sevoflurane; intravenous induction with propofol (2–3 mg/kg) or combination of ketamine (0.5–1 mg/kg) and propofol (1–2 mg/kg) may consider supplementing with lidocaine (0.5 mg/kg) and fentanyl (1 mcg/kg) or remifentanil (2 mcg/kg)

Muscle relaxation with rocuronium (0.6–0.8 mg/kg)

Maintenance of anesthesia with oxygen/air/sevoflurane or isoflurane

Ondansetron (50–200 mcg/kg IV) and dexamethasone 0.15–1 mg/kg IV as antiemetics

May consider dexmedetomidine (0.5 mcg/kg IV) and/or ketorolac (1 mg/kg IV) for postoperative analgesia given prior to completion of surgery

Postoperative Analgesia

1. Local wound infiltration with local anesthetics (bupivacaine or lidocaine)
2. Acetaminophen (20 mg/kg IV)
3. Ilioinguinal nerve block with 0.2% ropivacaine[15]
4. Caudal block (prior to surgery): ropivacaine 0.2% at 1 mL/kg. Clonidine (1–2 mcg/kg) or dexmedetomidine (1–2 mcg/kg) is a common adjunct drug.

It is suggested that all formerly preterm infants of less than 60 weeks postconceptual age should be monitored until they are apnea free for 12 hours.

Awake Regional Anesthesia

Spinal anesthesia has been reported to be effective in a wide variety of pediatric surgeries including inguinal hernia repair, circumcision, pyloromyotomy, and orthopedic procedures. The technique was also found to be effective in infants undergoing inguinal hernia repair using laparoscopic approach.[16]

There are some distinct advantages of spinal anesthesia over general anesthesia in infants and neonates:

1. Decreased respiratory complications including hypoxemia (oxygen desaturation <90%).[17]
2. Better hemodynamic stability. Compared to sevoflurane anesthesia, the incidence of hypotension (MAP <35 mm Hg) was reported to be significantly lower in infants (less than 60 weeks of postconceptual age) who underwent inguinal hernia repair under spinal anesthesia.[18]
3. Decreased apnea in the first 30 minutes postoperatively in infants (60 weeks or younger postconceptual age) undergoing inguinal herniorrhaphy, though there was no difference between spinal and general anesthesia groups in the first 12 postoperative hours.[19]
4. Spinal anesthesia is usually performed in the sitting or lateral decubitus position at L4-L5 level using a 22- (1.5 in.) or 25-gauge spinal needle. Standard ASA monitors are placed, and IV access is confirmed. Currently recommended drugs for spinal anesthesia are isobaric bupivacaine (0.5%) or isobaric ropivacaine (0.5%) at 1 mg/kg. Bupivacaine 0.75% in 8.25% dextrose is also used at a dose of 0.75 to 1 mg/kg.

   The success rate of spinal anesthesia in neonates and infants primarily depends on the experience of the anesthesiologist; failure rate could be as high as 20%.[19] Total spinal anesthesia may occur especially during lifting the legs for placement of electrocautery pads in the back of the patient immediately after spinal. Respiratory support should be provided immediately with continuous monitoring of pulse, oxygen saturation, and blood pressure. Bradycardia and hypotension should be treated promptly. Post-dural-puncture headache is thought to be rare, but difficult to diagnose

in these group of patients.

Of note, although there have been concerns regarding neurotoxicity and long-term neurocognitive effects of younger patients undergoing general anesthesia, an international randomized multicenter trial did not find any strong evidence that infants exposed to general anesthesia for 1 hour during inguinal herniorrhaphy had any measurable neurocognitive or behavioral deficits at 5 years of age.[20]

UMBILICAL HERNIA

Umbilical hernia is the protrusion of bowel or omentum due to weakness or defective closure of the umbilical ring where umbilical vessels enter the umbilical cord. The condition is common in low–birth-weight infants and in African Americans. The hernia usually presents as a swelling during straining but does not cause any painful symptoms unless strangulated. The umbilical hernia may be associated with congenital hypothyroidism, Down syndrome, and mucopolysaccharide storage disease.

Majority of the umbilical hernias close spontaneously by 5 to 6 years of age. Surgery is indicated in cases of strangulation, in hernias causing symptoms, or ones that persist after 4 to 5 years of age.

Anesthetic Management

Usually provided by general anesthesia. Airway management with LMA can safely be provided in otherwise healthy child with a small to medium-sized uncomplicated hernia. For a large defect and in strangulated hernias, endotracheal intubation with inhalation or IV induction with propofol and rocuronium is recommended. Maintenance of anesthesia is usually provided with oxygen, air, and sevoflurane or desflurane.

LAPAROSCOPIC SURGERY

Minimally invasive approach surgeries, such as laparoscopic and robot-assisted laparoscopic procedures, have become increasingly popular in the last 10 to 15 years. The magnified image of the surgical field offered by advanced endoscopic instruments and improved laser technology allow the surgeons to perform precise dissection in several commonly performed pediatric surgeries. The advantages of minimally invasive surgeries include smaller surgical scars, less blood loss, fewer intraabdominal adhesions, shorter postoperative hospital stay, and decreased postoperative pain.

Pneumoperitoneum is created during laparoscopic surgery by insufflating carbon dioxide inside the peritoneal cavity, which allows better visualization. This process can potentially produce significant pathophysiological changes and complications:

- *Cardiovascular:* The increased intraabdominal pressure (IAP) causes a decrease in venous return, leading to a decrease in left ventricular preload, in cardiac output and in blood pressure. IAP may also cause significant reflex bradycardia by stimulation of the vagus nerve due to peritoneal stretching during needle placement.

  An increase in peripheral vascular resistance due to an increase in plasma norepinephrine during pneumoperitoneum may counterbalance the reduction in blood pressure.[21]

  Positioning of patient in the Trendelenburg position during robot-assisted surgery significantly increases cardiac filling pressures and cardiac workload.

  Due to the aforementioned negative cardiac effects, increased IAP may not be well tolerated by patients with hypovolemia, anemia, or significant cardiac disease.

  Pneumopericardium may occur when CO_2 passes via the membranous diaphragm or to the mediastinum via IVC. Serious injury to major vessels may happen due to accidental insertion of trocar into the vascular channels.

- *Respiratory:* Increased IAP from pneumoperitoneum pushes the diaphragm cephalad which increases peak inspiratory pressure, reduces compliance of the respiratory system (mainly chest wall compliance in neonates and infants), functional residual capacity (FRC), and total lung capacity. Loss of FRC may cause atelectasis and hypoxemia which get exaggerated in the head-down position. Depressed cardiac function during laparoscopy along with the pulmonary atelectasis increases *V*/*Q* mismatch. Pneumothorax may complicate the laparoscopy by entry of gas into

the pleural cavity during dissection around esophagus, a diaphragmatic defect, tear in the visceral peritoneum, or rupture of an emphysematous bulla.[22]

Higher solubility of CO_2 leads to higher absorption of the gas and hypercarbia; the solubility is age dependent, more in neonates due to the relatively larger peritoneal surface area.[23]

- *Renal:* Pneumoperitoneum may decrease urine output by reducing renal blood flow and glomerular filtration rate. Urine output may also be decreased by stimulation of ADH secretion, a result of the direct compression of renal parenchyma.[24] Oliguria may continue for several hours after surgery.[25]
- *Intracranial pressure:* Intracranial pressure may increase in laparoscopy. Head-down position, hypercapnia, and increased IAP are possible contributory factors. Increase in ICP with low cardiac output and increased intrathoracic pressure may compromise cerebral perfusion pressure.[26]
- *Carbon dioxide embolism:* This is a rare but serious complication of pneumoperitoneum, which can occur due to direct placement of needle in the vessels. CO_2 embolism may present as sudden onset of hypotension, hypoxemia, increased end-tidal CO_2, and arrhythmia, which can progress to pulmonary edema.

  Because increased IAP is related to these wide range of physiological changes and potential complications, it is suggested to keep the insufflation pressure low (8–12 mm Hg) in neonates and infants. Insufflation should begin at lowest flow rate, which may be increased as it attains the target pressure.[27] The IAP in older children should be limited to less than 15 mm Hg.

  Low-pressure pneumoperitoneum with IAPs not exceeding 5 mm Hg in infants and children did not reduce cardiac index.[28]

Anesthetic Management

There are several important issues to remember regarding anesthetic management of patients for minimally invasive surgeries. Since access to the patient gets limited after surgical draping and docking of robots (in robot-assisted cases), standard monitors should be properly secured. Placement of a precordial stethoscope over left chest is helpful in small babies to quickly diagnose endobronchial migration of ET tube. A second IV access should be considered in long procedures. As end-tidal CO_2 concentration may not represent the arterial pCO_2 accurately in very small children, placement of an arterial line may be considered in selected cases of long duration. Inaccurate end-tidal CO_2 may lead to incorrect ventilator strategies in small babies.[26] Proper positioning and padding are important for infants due to their high vulnerability to positional injuries. An orogastric tube is helpful for gastric decompression.

General anesthesia with placement of endotracheal tube is commonly employed in laparoscopic cases. The cuffed endotracheal tube should be properly secured at correct mid-tracheal point, since downward displacement during steep Trendelenburg position may cause hypoxia or bronchospasm. Inhalational agents with opioids (as indicated) are used for maintenance of anesthesia. Nitrous should be avoided to limit bowel distension. Muscle relaxation is necessary for effective ventilation and pneumoperitoneum. Proper monitoring of temperature and urine output is mandatory.

INTUSSUSCEPTION

Intussusception is the telescopic invagination of a proximal segment of intestine, usually in the ileocolic region (intussusceptum), into an adjacent segment. The incidence peaks in children between 4 and 7 months, higher in fall and winter, and is the most common abdominal emergency in patients less than 2 years of age. Incidence is approximately 1 to 4 per 1000 live births and more common in males. In 90% cases it is idiopathic, but a prior respiratory infection, viral gastroenteritis, intestinal lymphoid hyperplasia, and Henoch-Schonlein purpura have been associated with intussusception. In some patients, a lead point such as Meckel's diverticulum, intestinal polyp, appendix, lymphoma, or hemangioma are recognized. Interruption of venous drainage of the mesentery associated with intussusceptum leads to edema and mucosal bleeding and if not relieved may lead to strangulation, gangrene, and shock.

The classic presentation of intussusception in children is abdominal pain, palpable abdominal mass, and bloody (currant jelly) stool. Typically, the initial symptom is a paroxysm of abdominal pain with intermittent crying with flexed knees and hips. Progressively, the pain becomes more frequent, the child vomits, becomes lethargic, and bloody diarrhea ensues.

Ultrasound is commonly used for diagnostic purposes, which has sensitivity and specificity of 98%. The patient with typical presentation should begin receiving intravenous fluid boluses and transported to the radiology suite for emergent hydrostatic reduction under fluoroscopy, which is both diagnostic and therapeutic. Success rate of hydrostatic reduction is approximately 80%. The patient with suspected peritonitis, bowel perforation, shock, and multiple recurrences should be taken to the operating room for surgical reduction.

Anesthetic Management

1. Review patient's history, vital signs, associated medical issues, laboratory results (glucose, electrolytes, H/H)
2. Confirm patency of intravenous access, premedication if appropriate
3. In the operating room: placement of ASA monitors
4. Rapid sequence IV induction with propofol (2–3 mg/kg) or ketamine (1–2 mg/kg), fentanyl (1 mcg/kg) and rocuronium (1 mg/kg), and endotracheal intubation with appropriate sized cuffed tube
5. Placement of large intravenous catheter and arterial line if indicated
6. Placement of nasogastric tube to evacuate the stomach
7. Aggressive fluid replacement with normal saline, lactated ringers, 5% human albumin in patients with intravascular volume deficits (shock, peritonitis, perforation, etc.)
8. Appropriate antibiotics, antiemetics (ondansetron and dexamethasone)
9. Maintenance of anesthesia with air/oxygen/sevoflurane or desflurane with fentanyl boluses, if appropriate
10. Extubate when awake
11. Acetaminophen (20 mg/kg IV), morphine (0.05–0.1 mg/kg IV), and/or dexmedetomidine (0.5 mcg/kg IV) for postoperative analgesia

GASTROSCHISIS AND OMPHALOCELE

Gastroschisis and omphalocele are two most common congenital abdominal wall defects observed in pediatric patients.

Gastroschisis

Gastroschisis, where the intraperitoneal contents protrude through an inborn defect of the ventral abdominal wall, appears like a "ruptured physiological hernia" and was first accurately described by Bernstein in a left-sided case which he named "epigastro-schisis."[29] An increase in incidence of gastroschisis has been noted in recent years from 3.6 per 10,000 births during 1995 to 2005 to a current report of 4.9 per 10,000 births during 2006 to 2012.[30]

The defect in the abdominal wall in gastroschisis is a full thickness paraumbilical opening with evisceration of intestines without any covering membrane. It is approximately 3 to 5 cm in diameter and is commonly present to the right of the umbilical cord insertion. Herniated bowel usually contains jejunum or ileum, and often appears thickened, edematous, or covered with fibrinous exudate.

Maternal factors implicated in gastroschisis include younger age, use of vasoconstrictive agents (nicotine, cocaine), and maternal infections.[30] Gastroschisis is divided into simple and complex cases. Complex cases are associated with other pathologies, eg, intestinal atresia, volvulus, perforation, intestinal obstruction, and necrotizing enterocolitis. Neonatal mortality in complex cases is more than sevenfold higher than those with simple gastroschisis.[31] Pregnancy with gastroschisis babies also has a higher incidence of preterm birth, intrauterine growth retardation,[32] and

stillbirth.[33] Prenatal diagnosis of gastroschisis is commonly done by ultrasonography, usually diagnosed during the second trimester. It is important to differentiate gastroschisis from omphalocele, which is covered by a membranous sac. The umbilical cord is inserted into the membranous sac in omphalocele, unlike in gastroschisis where it is inserted into the abdominal wall.

Several etiological factors are proposed to explain the development of gastroschisis:

1. Result of failure of the body wall to close, primarily involving right lateral fold being unable to meet the left one in the midline
2. Vascular accident causing atresia of midgut either due to early involution of the right umbilical vein or intrauterine interruption of the right omphalomesenteric artery leading to infarction of the abdominal wall
3. Failure of the yolk sac to insert into the body stalk and thus forming an extra opening in the amniotic cavity through which intestine will develop

Embryological abnormalities are also thought to be perpetuated by some environmental factors, eg, tobacco smoking, exposure to pesticides, Chlamydia infection.[29]

Compared to spontaneous vaginal delivery, planned cesarean delivery (CD) did not show any decreased risk of complications related to bowel ischemia, bowel injury, and necrotizing enterocolitis.[34] Yet, the rate of CD is higher in the United States (60.9%) perhaps due to planned cesarean deliveries or unexpected fetal distress.[35]

Surgical treatment depends on the condition of the eviscerated bowel and the ability to reduce the eviscerated intestine inside the abdomen. Primary reduction and immediate closure without causing abnormally high abdominal pressure are helpful as they reduce the evaporation of fluid and exposure to infection. Staged reduction involves creating a silo to keep the bowel inside the bag and then gradually reducing the size of silo to allow the eviscerated bowel to enter into the abdominal cavity. The abdominal defect is closed with suture after completion of reduction. The complex gastroschisis cases, patients with atresia, perforation, volvulus are managed depending on the status of the intestine and overall clinical status of the patient.[36]

Omphalocele

Omphalocele, the other common congenital abdominal wall defect, presents with a midline protrusion of herniated intraabdominal organs through the umbilicus into the umbilical cord. Umbilical cord is inserted into the membrane which is composed of peritoneum, Wharton's jelly, and amnion. The herniated organs include midgut and occasionally spleen and gonads. The incidence of omphalocele is estimated to be 1.9 per 10,000 live births.[37] A fetus with omphalocele is at high risk for associated chromosomal abnormalities, intrauterine growth restriction (IUGR), premature delivery, and fetal death.[38] The infant mortality rate is reported to be around 28% and significantly higher in neonates born with chromosomal abnormalities.[37] The other factors associated with mortality are pulmonary hypertension and respiratory insufficiency at birth, large omphalocele containing 75% liver, and rupture of sac.[39]

Unlike gastroschisis, omphalocele is associated with a wide variety of malformations:

1. Beckwith-Wiedemann syndrome: omphalocele, macrosomia, and hypoglycemia
2. Pentalogy of Cantrell: omphalocele, anterior diaphragmatic hernia, sternal cleft, ectopia cordis, intracardiac defect (VSD, ASD, tetralogy of Fallot, etc.)
3. Chromosomal defect: trisomy 18, 13, 21. Chromosomal defects may occur up to 49% of fetuses diagnosed with omphalocele[40]
4. Congenital heart defect (ASD, PDA, VSD, tricuspid valve anomaly, VACTERL): 18% to 24% incidence of cardiac anomalies[41]
5. Genitourinary system: exstrophy of bladder or cloaca
6. Central nervous system defect: spina bifida, anencephaly
7. OEIS complex (omphalocele, exstrophy of bladder, imperforate anus, spinal defect)

8. Donnai-Barrow syndrome (omphalocele, congenital diaphragmatic hernia, corpus callosum agenesis)

Embryology

The embryo develops to a three-layered flat disc of ectoderm, mesoderm, and endoderm with amniotic cavity dorsally and yolk sac ventrally. Around the fourth to fifth week of gestation, due to increased growth of neural tube, the embryonic disc is folded toward the ventral surface and a fetal configuration develops. The four body folds (cranial, caudal, and two lateral) converge toward the umbilicus to form the umbilical ring which separates the developing body wall from the amnion. The gastrointestinal tract, which develops more rapidly in relation to the abdominal cavity, herniates through the umbilical ring into the umbilical cord. At around 10 to 12 weeks of gestation, the bowel returns to the abdominal cavity and fixes to its final position. The omphalocele probably develops due to malformations of the embryonic folding process which interferes with the physiological reentry of abdominal organs from the umbilical cord.[42]

Prenatal diagnosis of omphalocele is made reliably after the first trimester with ultrasonography and the detection of high maternal serum AFP levels and prenatal ultrasound. An omphalocele may be classified as small (<5 cm), giant (>5cm) with a sac containing liver, and ruptured. Karyotyping using chorionic villi sampling may be useful to rule out any associated major chromosomal abnormalities.[43]

Although controversy exists whether outcome is improved with CD,[44] surgical delivery is indicated in cases of giant omphalocele in order to prevent injury to the liver or disruption of the sac.

The small to medium-sized omphalocele in an otherwise stable neonate is closed by primary closure of the skin and fascia after reduction of the herniated organs and excision or inversion of the sac. In large omphaloceles, the ultimate goal is to provide complete reduction with closure of the defect without causing excessive intraabdominal pressure. The increased intraabdominal pressure may cause cardiorespiratory compromises such as the ones mentioned in minimally invasive procedures above. Intraabdominal hypertension (usually >20 mm Hg) can progress to abdominal compartment syndrome (ACS) with potentially devastating complications, eg, renal failure, gut ischemia, necrotizing enterocolitis, wound dehiscence, sepsis, and enterocutaneous fistula.

Central venous pressure monitoring has been reported as a useful guide to assess IAP; an increase of more than 4 mm Hg during placement of silo in delayed closure or fascia closure during primary repair correlated with a decrease in cardiac index.[45]

A giant omphalocele is surgically repaired with a staged closure initially placing a silicone plastic “silo” and gradually reduced it in size returning the herniated organs to the abdominal cavity over few days. Silastic silo closure is associated with wound infection and wound dehiscence. Staged closure has also been reported using intraabdominal tissue expander placement, component separation, and bipedicle skin flap technique. Escharotic therapy with topical application of silver sulfadiazine results in gradual epithelialization of the omphalocele sac. After completion of epithelialization, the ventral hernia is repaired using prosthetic mesh and skin flap.

Anesthetic Management of Gastroschisis and Omphalocele

Preoperative Management

1. Genetic evaluation to diagnose any coexisting chromosomal anomalies (eg, trisomy 13, 18, 21 in 30–40% cases).
2. Echocardiography for patients with omphaloceles to rule out congenital heart disease and pulmonary hypertension.
3. Abdominal ultrasound to look for any associated renal abnormality.
4. Blood sugar levels in premature patients and in patients with omphalocele and possible Beckwith-Wiedemann syndrome (12% cases).
5. Gastric decompression with NG tube in both gastroschisis and omphalocele to prevent intestinal distension.
6. Adequate intravenous access to ensure fluid resuscitation to replace evaporative fluid loss from herniated intestines.
7. Prevention of heat loss by increasing the ambient temperature. Also, the herniated bowel and omphalocele should be wrapped in warm saline soaked gauge to reduce evaporative loss. In case of gastroschisis, care should be taken to avoid kinking of the mesentery.

Intraoperative Management

1. Standard ASA monitors. Intraarterial catheter can be helpful to monitor continuous blood pressure, laboratory studies (glucose, electrolytes, arterial blood gases), fluid status. Pulse oximeter in right upper extremity (preductal) and in toes/feet (to assess lower extremity perfusion during closure of abdomen).
2. Central venous line to monitor the effect of increased abdominal pressure and as additional access for vasopressors and volume infusion.
3. Nasogastric tube placement and aspiration prior to induction.
4. Urinary catheter to measure urine output and intravesical pressure.
5. Temperature probe either rectal or esophageal.

Induction: Rapid sequence intravenous induction after adequate preoxygenation. Propofol (2–4 mg/kg) or ketamine (1–2 mg/kg) or a combination of lower doses of ketamine and propofol may be used for hypnosis. Rocuronium at 0.8 to 1 mg/kg produces intubating condition in 30 seconds. Glycopyrrolate (10 mcg/kg) to prevent vagal-mediated bradycardia during intubation and fentanyl (1 mcg/kg) may also be used as supplemental agents.

In patients with possible difficult airway (such as macroglossia in Beckwith-Wiedemann syndrome) a video laryngoscope (CMAC) with appropriate blade should be available.

Fluid management should be continued with D5 or D10 with 0.2% NaCl solution. In cases of neonatal hypoglycemia (serum glucose less than 40 mg/dL in term or preterm infant) D5 to D12.5W at 4 to 8 mg/kg/min should be used. For intravenous fluid replacement of surgical or evaporative/insensible fluid loss, replace with 5 to 15 mL/kg/h of lactated ringer's solution and/or normal saline, or 5% human albumin (50 mg/mL; 0.5 g/kg/dose).

Hypothermia (body temperature <36°C) should be prevented by increasing the OR temperature (78–80°F), by adding warming blanket on the operating table to reduce conductive heat loss, by placing overhead heat lamps to reduce radiant heat loss, and by transporting in a thermoneutral incubator or a bed with overhead heater.

Anesthesia is maintained with inhalation anesthesia (oxygen/sevoflurane or desflurane) and narcotics (fentanyl) boluses. Nitrous oxide should be avoided to reduce bowel distension.

Intraabdominal pressure is closely monitored during abdominal closure, and for serial approximation of Silo by intravesical pressure, intragastric pressure, or CVP. Other surrogate indicators of increased IAP include high ventilation pressure (peak pressure >30 mm Hg), high end-tidal pCO_2, significant drop in MAP not responsive to fluid boluses, and low pulse oximetry reading in lower extremities.

CONGENITAL DIAPHRAGMATIC HERNIA

Congenital diaphragmatic hernia (CDH) is a complex clinical syndrome represented by a development defect in the diaphragm and herniation of abdominal organs into the thoracic cavity, which can interfere with development of the lung. At birth, the pulmonary hypoplasia and pulmonary hypertension present a major threat to survival of the neonate. CDH commonly occurs in 1 in 3000 live births.[46] Majority of the hernia is left sided and the commonest defect is in posterolateral diaphragm (Bochdalek type). The other forms are anterior (Morgagni type) and central.

Approximately 30% to 40% of CDH cases are reported to have associated congenital malformations, eg, cardiovascular (VSD, ASD, tetralogy of Fallot), central nervous system abnormalities (neural tube defects, hydrocephalus), and limb abnormalities (polydactyly, syndactyly, reduction defects). Regarding associated chromosomal abnormalities, trisomy 18 is the most common one.[47]

Embryology

The diaphragm is developed from the following embryonic components: (1) septum transversum, which forms the central tendon of the diaphragm, and (2) pleuroperitoneal membrane: the primary source of the future diaphragm. Failure or defect in the fusion of the pleuroperitoneal membrane

with other components of the developing diaphragm about the 10th week of gestation is the main embryological pathology in the formation of CDH. As the right pleuroperitoneal canal closes before the left one, left-sided CDH is more common than the right-sided CDH.[48]

Herniation of abdominal contents in the lung cavity and shift of mediastinal structures to the contralateral side contribute to the pulmonary hypoplasia. An abnormal genetic expression, which regulates the development of both diaphragm and lungs may also be involved in causing pulmonary hypoplasia. Hypoplasia affects the total surface area of the lung involved with gas exchange. Developmental arrest of the pulmonary vasculature with increased thickness of the arterial wall decreases cross-sectional area of the pulmonary vascular bed and increases pulmonary vascular resistance.[49]

Diagnosis is usually made by ultrasonography during routine prenatal screening. The fluid-filled intestines with or without the liver, and displacement of the heart are seen in the thorax. Fetal lung-to-head ratio (LHR), as measured by ultrasound, helps to assess the lung hypoplasia. Lung size could also be assessed by MRI or 3D ultrasound. Herniation of the liver and stomach has been associated with poor outcome. Diagnosis in early pregnancy helps in prenatal counseling, referral to a tertiary center for advanced tests, fetal interventions, and termination of pregnancy, in selected cases.

Clinical Presentation and Management

Respiratory distress, characterized by tachypnea, grunting, use of accessory muscles, and cyanosis, is the cardinal feature of babies with CDH. Scaphoid abdomen may also be present. Auscultation of the chest may reveal bowel sounds on the left side of the chest, and rightward shift of the point of maximal cardiac impulse may be heard. A chest X-ray will confirm the diagnosis with bowel gas pattern in the chest and mediastinal shift.

Historically, CDH used to be considered a surgical emergency, and prompt surgical exploration to reduce the herniated organs back to the abdominal cavity was thought to be the primary treatment. Mortality though continued to remain high is primarily due to persistent pulmonary hypertension. Today, the key principle of management following birth is to optimize the respiratory status, deteriorated by the combination of lung hypoplasia, increased PVR, and right-to-left shunt. Initial management should include quick evaluation of the cardiorespiratory status, endotracheal intubation without prolonged mask and bag ventilation, establishment of IV access, and decompression of gut by nasogastric tube. Aggressive bag and mask ventilation should be avoided as it affects oxygenation by enlarging the stomach and compressing the lungs. Factors which worsen pulmonary hypertension, eg, hypoxia, acidosis, and hypothermia, should be avoided.

Proper ventilator strategies are important for newborns with CDH. Initial approach to resolve pulmonary hypertension and right to left shunt by inducing with aggressive hyperoxia and hyperventilation produced several significant complications, including barotrauma (pneumothoraces, pulmonary hemorrhage) and residual lung disease.[50]

The advent of the concept of “permissive hypercapnia” by gentle ventilation in the management of infant pulmonary hypertension reduced ventilator-induced lung injury and improved survival.

The current recommendations suggest keeping the ventilator mode as intermittent mandatory ventilation (IMV) with PIP <25 cm H_2O: PEEP 3 to 5 cm H_2O, keeping preductal SaO_2 >85% and pCO_2 <60 mm Hg. If the goals for oxygenation and ventilation are not accomplished by this method, high-frequency oscillatory ventilation (HFOV) may be used to optimize alveolar recruitment with mean airway pressure of 13 to 15 cm H_2O with a pressure delta ranging from 30 to 40 cm H_2O.[51] Use of HFOV as an early intervention strategy rather than rescue mode, keeping $PaCO_2$ around 40 to 55 mm Hg and pressure delta of 35 to 45 cm H_2O, has been reported to improve survival.[52]

The general medical management of persistent pulmonary hypertension in CDH includes administration of adequate intravascular volume, maintenance of hemoglobin and glucose at normal levels, and maintenance of normal temperature. Although high-frequency ventilation with inhaled nitric oxide (iNO) is considered for selected patients, iNO has not been shown to reduce risk of death or ECMO use. Use of other potential agents, eg, sildenafil, milrinone, prostaglandin analogues (prostacyclins, PGE1), and Bosentan, has not gained sufficient evidence for clinical use.

ECMO (Extracorporeal Membrane Oxygenation)

ECMO has been used in many centers to stabilize the infant prior to surgery. In ECMO, the blood is oxygenated outside the body, so the need for gas exchange in the lung is not needed. The following criteria have been considered by Scottish Congenital Diaphragmatic Hernia Network for ECMO.[53]

1. Inability to maintain preductal saturation >85% despite optimal ventilation and management of pulmonary hypertension
2. Arterial pCO_2 above target range with respiratory acidosis (pH 7.15) despite optimum ventilation
3. PIP consistently above 25 cm H_2O to achieve ventilator goals or failure to convert after HFOV
4. Metabolic acidosis (pH <7.15) and lactate >5 mmol/L
5. Systemic hypotension resistant to fluid and inotropes with a urine output <0.5 mL/kg/h over 12 to 24 hours

Contraindications of ECMO therapy are as follows:

1. Gestational age less than approximately 34 weeks
2. Weight less than approximately 2 kg
3. Ongoing bleeding disorder or uncorrectable coagulopathy
4. Intraventricular hemorrhage.

The outcome benefit of the ECMO procedure in patients with CDH has not been well established. A two-center study in two cities (Boston and Toronto) concluded that the overall survival rate with HFOV as rescue was equivalent to the overall survival rate in patients with ECMO as rescue. In both groups, permissive hypercapnia significantly increased survival. The leading cause of mortality was pulmonary hypoplasia and associated anomalies.[54,55]

There appears to be no difference in survival benefit between the two modes of ECMO (VV [venovenous] and VA [venoarterial]) in CDH patients. The optimal timing of repair while on ECMO has not been identified yet, but early repair may improve survival as it decreases complications.

In recent years, a fetal therapy for CDH, named fetoscopic endoluminal tracheal occlusion (FETO), has been practiced. The procedure prevents egress of fluid from the lung and thus increases pulmonary pressure. The increased pressure promotes proliferation of pulmonary tissue, increases the total alveolar surface area, and helps to mature the pulmonary vasculature.

Surgical Management

Surgical repair of CDH is deferred until the pulmonary status is stabilized, since mortality rate in these infants is primarily related to pulmonary hypoplasia and persistent pulmonary hypertension. Despite generalized agreement in the benefits of delayed surgery, specific parameters of cardiorespiratory stability and timing of surgery remain unconfirmed. The usual practice is to control the pulmonary hypertension, thus requiring lower ventilator settings with low PIP, minimal shunting, and low FiO_2.

The CDH-EURO consortium states that surgical repair should be performed when the following criteria are met:[56]

- Mean arterial blood pressure normal for gestational age
- Preductal saturation levels between 85% and 95% with a fractional inspired oxygen <50%
- Serum lactate <3 mmol/L
- Urine production >2 mL/kg/h

Other set of recently reported parameters include the following:[57]

- Oxygen saturation >92% (FiO_2 <0.5)
- Mean arterial pressure >45 mm Hg
- Pulmonary artery pressure less than two-thirds of the systemic pressure

- Weaning iNO (<10 ppm)
- Hemoglobin >10 g/dL

Surgical approaches to repair CDH include minimally invasive approach (thoracoscopy) and open repair. Thoracoscopic approach has the advantages of better visualization of the defect and mobilization of herniated contents to the abdomen, which happens due to thoracic insufflation with CO_2.

Disadvantages of thoracoscopic repair are a higher 1-year recurrence rate, longer operative time, and need to adjust ventilatory parameters due to absorbed CO_2. The defect is repaired with prosthetic material (Gore-Tex) or muscle flap (abdominal wall muscles).

In many centers, surgical repair of CDH is performed in the neonatal ICU to avoid accidental disconnection of different lines and significant changes in ventilatory support.

Anesthetic Management

Two important goals of anesthetic management in CDH patients: avoiding volutrauma and preventing worsening pulmonary hypertension

Standard ASA monitors

Pulse oximetry in right upper extremity (preductal)

Arterial line (right radial artery; preductal)

Position of ETT tube to be checked

NG tube placement

Central venous line access or an extra-large gauge intravenous line placement for volume replacement.

Maintenance fluid with glucose-containing solution continuation

ICU ventilator use for continuation of ventilation during surgery; pressure control ventilation with PIP <25 cm H_2O

Maintenance of anesthesia with fentanyl (10–30 mcg/kg) and inhalational agent as tolerated; in case of ventilation with NICU ventilator, propofol infusion (100–150 mcg/kg/min) and fentanyl infusion (2 mcg/kg/h)

Neuromuscular blocking agent (rocuronium) use to keep the patient paralyzed

Nitrous oxide avoidance

Avoidance of hypothermia, acidosis, and hypoxia (triggers of pulmonary hypertension)

At the end of surgery, patient to be kept intubated and sedated, and transferred to the neonatal intensive care unit

APPENDICITIS

Acute appendicitis is the most common surgical urgency/emergency in children, with approximately 70,000 pediatric cases each year in the United States.[58] It occurs in approximately 1 to 4 per 1000 children per year.[59] The estimated life risk is reported to be 7% to 8%.[60] Appendicitis is more common in children aged 12 to 18 years, and less common in children less than 5 years of age.

The pathogenesis of appendicitis is thought to be due to direct luminal obstruction by a fecalith, lymphoid hyperplasia, or tumor, which leads to a series of pathological changes including distension of lumen, venous congestion, edema, bacterial infections, reduced blood supply, ischemia, necrosis, and gangrene. Recent studies suggest genetic factors, and environmental factors also might play a role.[60] Bacterial infections with agents such as *Escherichia coli*, *Salmonella*, *Shigella*, and Bacteroides might play an important role in initiating the development of appendicitis in some

cases.

The classic presentation of appendicitis includes abdominal pain with anorexia, nausea, and vomiting. Abdominal pain begins as a dull periumbilical pain, shifting to the right lower quadrant of the abdomen. The classic presentation is noted in less than 50% of patients. In other children, it may start with some flu-like symptoms, eg, anorexia and malaise, and then rapidly progressing to abdominal pain and fever. Continuation of these symptoms may lead to perforation of appendix within 36 to 48 hours after initial presentation of the illness. Due to initial absence of typical symptoms along with difficulty in elicitation of definitive signs in pediatric patients, the incidence of complications such as perforation, abscess, and gangrene are high. The signs of appendicitis include localized tenderness, rebound tenderness, and guarding in the right lower quadrant. The point of tenderness in patients with appendicitis is classically described as McBurney's point, at the junction of the lateral one-third and medial two-thirds of the line joining the umbilicus and right anterior superior iliac spine. A scoring system (Pediatric Appendicitis Scores) has been developed to assist in the diagnosis of appendicitis in pediatric patients; components of the system include eight relevant clinical and laboratory data such as fever >38°C, anorexia, nausea/vomiting, migration of pain, right lower quadrant tenderness, cough/percussion/hopping tenderness, leukocytosis >10,000 cells/µL, and polymorphonuclear neutrophilia >7500 cells/µL. Transabdominal ultrasound is the most common diagnostic modality for appendicitis, because it does not expose the patient to radiation and is less expensive than computed tomography (CT). In some centers, there is increasing reliance on CT scan, and it has become the most accepted diagnostic strategy.

In the majority of cases, appendectomy is the standard management. Preoperative antibiotics with broad coverage of gram-negative bacteria, IV fluids, analgesics, and antipyretics are usually started after diagnosis. Laparoscopic appendectomy over open appendectomy is the preferred surgical approach.

Anesthetic Management

1. Preoperative evaluation should include assessment of vital signs, BMI, associated comorbidities (obesity, obstructive sleep apnea, symptoms of gastroesophageal reflux), current medications, and NPO status.
2. Standard American Society of Anesthesiologists (ASA) monitors: Arterial line may be considered in patients with abdominal sepsis and peritonitis.
3. Rapid sequence induction with propofol (2–3 mg/kg), and succinylcholine (1.5–2 mg/kg) or rocuronium (1 mg/kg)
4. Endotracheal intubation: Video laryngoscopy and other airway management equipment should be accessible in patients with anticipated difficult airway.
5. Nasogastric tube is used to decompress the stomach.
6. Inhalational agents (sevoflurane or desflurane) with oxygen/air are used for maintenance of anesthesia. Nitrous oxide should be avoided.
7. Intraoperative narcotics (fentanyl 1–2 mcg/kg, morphine 0.05–0.1 mg/kg) and antiemetics (dexamethasone 0.2–0.5 mg/kg and ondansetron 0.1–0.15 mg/kg) should be considered.
8. Postoperative analgesia:
    - Infiltration of surgical wound with local anesthetics at the completion of surgery
    - Acetaminophen (15 mg/kg IV)
    - Dexmedetomidine (0.5–1 mcg/kg IV) at the completion of surgery
    - Ketorolac (0.5 mg/kg IV)
    - Morphine (0.05–0.1 mg/IV)
    - Regional anesthesia: TAP block; rectus sheath block may be considered at selected cases
    - Patient-controlled analgesia (PCA) with opioids for ruptured cases

AGANGLIONIC MEGACOLON (HIRSCHSPRUNG DISEASE)

Hirschsprung disease or congenital intestinal agangliosis, named after Dr. Harold Hirschsprung, is the most common cause of intestinal obstruction in neonates. The disease occurs in 1 in 5000 live births and is more common in males. It may be associated with other congenital anomalies including Down syndrome, tumors of neural crest origin (neurofibromatosis), microcephaly, mental retardation, multiple endocrine neoplasia type 2 (MEN 2) syndrome, DiGeorge syndrome, and congenital hypoventilation syndrome.[61] The malformation primarily occurs to the structures which are embryologically derived from the neural crest.

Hirschsprung disease is characterized by the absence of ganglionic cells in the myenteric and submucosal plexuses of the bowel wall. The myenteric plexus is formed from the vagal neural crest cells which migrate in a craniocaudal direction at 5th to 12th weeks of gestation. Failure of migration of the precursor cells to the distal gut is thought to be the primary etiology of Hirschsprung disease. The short segment variety of the disease, where the aganglionic segment primarily involves the rectosigmoid region, accounts for 80% of the cases. Peristalsis cannot propagate through this aganglionic segment, causing distension of the bowel proximal to the segment. The other 10% to 15% of patients have long-segment disease which involves the area proximal to the sigmoid colon.

Healthy full-term infants pass meconium within 48 hours after birth. Any baby that fails to pass meconium within this timeframe should be investigated for Hirschsprung disease. Progressive intestinal obstruction causes abdominal distension, failure to feed, and vomiting. The intestinal obstruction may lead to stasis and bacterial overgrowth with *Clostridium difficile*, *Staphylococcus aureus*, and coliforms, leading to enterocolitis and diarrhea with foul smelling and bloody stools, fever, obstruction, and sepsis. Enterocolitis is the major cause of death in Hirschsprung disease and the mortality rate may be as high as 30% in patients with enterocolitis. Failure to thrive, constipation, episodes of obstruction, and blood-flecked diarrhea (suggestive of enterocolitis) are common symptoms.

Diagnosis of Hirschsprung disease is confirmed by histological and histochemical tests of the specimen obtained from a rectal biopsy. Initial treatment is decompression of the gastrointestinal tract, fluid resuscitation, and administration of broad-spectrum antibiotics. Decompression of the colon may be carried out by rectal catheter (with irrigation) and temporary ostomy.

Definitive surgery is a single-stage pull-through procedure which is usually performed after a few months. The aganglionic bowel with the transitional zone is resected, and the normal functioning bowel is moved down and connected to the dentate line of the anal canal. Functions of the anal sphincter are preserved. Laparoscopy is used for definitive repair in pull-through procedures.

Anesthetic Management

1. Preoperative evaluation: Presence of any other coexisting disease (Down syndrome, neurofibromatosis, etc.) should be ruled out. Vital signs, airway, IV access, hydration (fontanelles, capillary refill), and oxygenation should be assessed. Premedication with midazolam if appropriate.
2. Anesthetic management depends on the clinical status of the patient (hypovolemic, in shock state) or nature of surgery (initial ostomy for decompression or primary pull through).
3. A single-shot spinal anesthesia may be considered if it is a short procedure for biopsy and colostomy in selected patients.
4. General anesthesia is the most common modality for anesthesia management.
5. Standard ASA monitors: Arterial line may be considered in patients for fluid resuscitation, support vasopressor infusion, and continuous monitoring of electrolytes or other laboratory values.
6. A single-shot caudal block may be considered for postoperative analgesia. Caudal catheters are sometimes difficult to maintain because of the area of surgery involved.
7. Glycopyrrolate (10 mcg/kg to prevent reflex bradycardia during intubation) and fentanyl (1 mcg/kg)
8. Induction: Rapid sequence induction should be considered with propofol or ketamine (in volume-deficient patient) with succinylcholine (1.5–2 mg/kg) or rocuronium (1 mg/kg) in patients with obstruction, or perforation of intestines.

9. Oral endotracheal intubation with appropriate-sized cuffed endotracheal tube.
10. Additional intravenous access for hypovolemic patients.
11. Maintenance of anesthesia with sevoflurane/desflurane with oxygen and air. Nitrous oxide should be avoided.
12. Fluid resuscitation with normal saline, lactated ringer's solution, 5% albumin if warranted.
13. Vasopressor (phenylephrine, dopamine, norepinephrine, dobutamine) consideration to maintain hemodynamic stability.
14. Extubation at the completion of procedure.
15. Morphine (0.05–0.1 mg/kg), dexmedetomidine (0.5 mcg/kg IV), acetaminophen (15 mg/IV) for postoperative analgesia.

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