

Neonatal Alimentary System

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GASTROINTESTINAL DEVELOPMENT

The gastrointestinal (GI) tract has multiple functions that include the digestion, absorption, processing, and transport of dietary nutrients. Digestion depends in part on exocrine secretions from the salivary glands, pancreas, and liver. These solid organs also secrete hormones into the bloodstream that serve endocrine roles throughout the body. The GI tract provides a mucosal barrier, preventing systemic access to the normal enteric flora and potential pathogens. Many gastrointestinal functions are not fully developed in the term human newborn, and this immaturity is even more pronounced in the premature infant.

Development of the GI tract can be divided into six stages, beginning with conception and progressing

to full digestive capability, which occurs well into the 1st year of life. These stages summarize the development of the GI system in regard to organ morphology, digestion, absorption and secretion, hormone regulation, and immunologic capabilities.¹

- Stage I. Organogenesis: embryogenic development
- Stage II. Chemoarchitectural: basic structure and epithelial formation
- Stage III. Differentiation: differentiation of epithelial and mesothelial cells
- Stage IV. Maturation: preparation of the gut for extrauterine life
- Stage V. Neonatal phase of adaptation: introduction of feeding
- Stage VI. Transitional diet: transition from milk to solids

The first step in the embryologic development of the GI system is the formation of a hollow internal cavity that is created by the infolding of the embryonic disk before the end of the 1st month of gestation. This early, primitive tract, divided into three regions based on the source of the blood supply, ultimately gives rise to the neonate's GI tract (Table 1).

Table 1. Structure of the Embryonic GI Tract From Primitive Gut¹⁻³

Foregut	Midgut	Hindgut
Celiac axis blood supply	Superior mesenteric artery blood supply	Inferior mesenteric artery blood supply
Esophagus	Distal 2/3 of duodenum	Distal portion of transverse colon
Stomach	Small intestine	Descending colon
Proximal 1/3 of duodenum	Cecum	Sigmoid colon
Liver	Ascending colon	Rectum
Gallbladder	2/3 of transverse colon	
Pancreas		

The GI tract of the healthy neonate is thought to be functionally ready for extrauterine life by 36 weeks' gestation. GI motility, while not fully mature, is adequate for successful propulsion of food through the esophagus, stomach, and intestine. Enzyme, gastric acid, and bile acid secretion are likewise adequate, but undergo further maturation after birth. Enterocyte function, as well as gut perfusion, can handle successful absorption and transport nutrients. Postnatal development of the GI tract is influenced by genetic, neuroendocrine, hormonal, and luminal factors, including enteral nutrients.

The high-risk premature neonate, however, faces the consequences of an immature GI tract. Depending on the degree of prematurity, these consequences include altered motility and digestion, immature immunologic response, and decreased rate of mucosal cellular proliferation and migration.³

ENTERAL FEEDINGS

The two major factors affecting postnatal GI adaptation and maturation are gestational maturity and the type of feedings provided. The "gold standard" of nutrition for the newborn is breast milk. However, when feeding breast milk is not feasible, specialized infant formulas and supplements are available to provide nutrition. The perinatal development of GI diseases, defects, and anomalies will also affect the delivery of adequate nutrition to the neonate.

NURSING ISSUES

Nursing care using nursing process tools, care maps, standards of care, and quality assessment/assurance/improvement programs should focus on the issues specifically identified with each condition. Pain management and collaborative care measures also are an essential component of nursing care for all infants. One of the primary goals in the management of these diseases and anomalies is successful enteral feedings.^{2,4-7}

COMMON GASTROINTESTINAL DISEASES, DEFECTS, AND ANOMALIES

Tracheoesophageal Fistula and Esophageal Atresia

Normal Embryology

Normal development of the trachea at about 4 weeks of gestation includes the complete division of the cranial section of the foregut into the respiratory and digestive tracts. The formation of the tracheo-esophageal septum results in two separate and distinct hollow formations: the trachea and the esophagus.

Etiology and Pathogenesis

Tracheoesophageal fistula (TEF) results from the abnormal division of the foregut into trachea and esophagus due to incomplete fusion of tracheo-esophageal folds during embryogenesis. Such abnormalities in the separation of these two structures also may lead to esophageal atresia (EA). However, the embryonic origins of EA with or without TEF are still unclear.⁸

Epidemiology

TEF and its variants affect approximately 1 in 2,000 to 5,000 live births.^{2,8,9} The most common defect is EA with a distal TEF (Table 2 and Figure). Forty percent to 60% of affected infants have associated anomalies, which include vertebral, anal, limb, genitourinary, and cardiac abnormalities.² Great care should be taken to identify and evaluate additional anomalies in this population.

EA with distal TEF	85%
EA without TEF	8%
Isolated TEF	4%
EA with proximal TEF	2%
EA with distal and proximal TEF	<1%

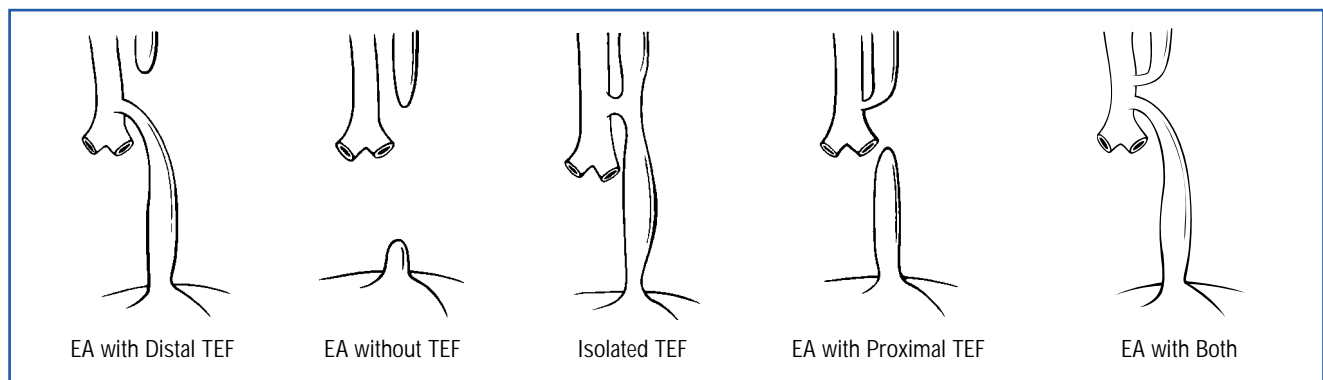


Figure. Types of TEF.

Morbidity/Mortality

Infants with the highest survival rate (95%) are full-term infants who are diagnosed early in their hospital course and whose defect(s) is aggressively treated to prevent morbidity.^{2,9} Premature neonates carry the greatest risk for death, particularly those with additional anomalies.¹⁰ Premature infants with other associated anomalies have a 50% to 60% mortality rate; those with cardiac defects have up to a 78% risk of death.^{2,9,10}

Assessment and Diagnosis

History

Polyhydramnios occurs in 50% to 75% of infants with TEF. Prenatal ultrasonography may be helpful for diagnosing EA with or without TEF, although <50% of these defects are identified by this means. Absence of a fluid-filled abdomen in the fetus coupled with the presence of polyhydramnios should arouse suspicion of EA. GI structures can be accurately identified only during the 2nd and 3rd trimesters.

Signs and Symptoms

Physical assessment. The infant may appear normal in the delivery room. However, if an orogastric tube is attempted and cannot be passed, the clinician should be alerted to the likelihood of EA. The hallmark signs and symptoms of TEF are excessive accumulation of secretions, regurgitation, and respiratory distress with accompanying history of polyhydramnios. A proximal fistula often produces respiratory distress, such as coughing, choking, and cyanosis, during attempts to feed. A distal fistula can produce progressive abdominal distention secondary to the accumulation of air in the stomach. Total atresia may present with a scaphoid abdomen. Findings on auscultation vary with each type of defect and the presence and location of a fistula.

Diagnostic Tests

Roentgenographic findings. EA with or without TEF is diagnosed clinically with roentgenologic assistance. Contrast studies, as a rule, are contraindicated because of potential problems with aspiration. Tracheoesophageal abnormalities can often be roentgenographically identified by the placement of a roentgenopaque catheter. Isolated TEFs usually require further study.

Treatment/Management

Prevention and Clinical Management

Preventive measures to minimize complications or exacerbation of symptoms include maintenance of a patent airway, appropriate positioning specific for the defect, and continuous suctioning to prevent aspiration. Early detection and aggressive preventive management prior to surgical correction are the mainstays of medical management.

Surgical Management

Surgical intervention consists of ligation of the TEF and end-to-end anastomosis of the esophagus whenever possible. When primary anastomosis of the esophagus cannot be performed, several options are available. The initial surgical approach is to ligate the fistula and place a gastrostomy. Definitive repair can then be done at a later date in smaller neonates when growth permits the anastomosis. If anastomosis is impossible, esophageal lengthening and/or colonic interposition is a later option.

Common postsurgical complications include anastomotic breakdown and leakage, refistulization, esophageal stricture, and gastroesophageal reflux. Undetected gastroesophageal reflux has its own morbidity and must be identified and managed appropriately. Esophageal replacement with colonic interposition carries a significant risk of complications.¹¹

Nursing Management

Nursing management includes routine neonatal care, along with the specific issues listed in Table 3.

Table 3. Nursing Issues and Focal Points of Care: TEF

Early recognition
NPO/secretion management: Decompression with continuous suctioning
Appropriate positioning for the type of defect
Assessment/management of nutritional status
Postoperative care:
• Gastrostomy tube
• Fistula care
Ostomy care
Gastroesophageal reflux surveillance

Abdominal Wall Defects

OMPHALOCELE AND GASTROSCHISIS

Description

Two abdominal wall defects—omphalocele and gastroschisis—are similar in appearance and presentation in the neonatal period, yet are two distinct anatomical defects. The omphalocele is a ventral wall defect with herniation of abdominal contents through the umbilical ring, covered by an amnioperitoneal membrane. It is variable in size and can be large enough to allow evisceration of the liver and other organs. Gastroschisis is a full-thickness defect of the anterior abdominal wall that permits extrusion of uncovered abdominal organs into the amniotic cavity without involvement of the umbilical cord.

Normal Embryology

At approximately 4 weeks of gestation, there is cranio-caudal and lateral infolding of the embryonic disk, forming structures of the abdominal wall. By the end of 5 weeks, the umbilical ring is completed via the migration and fusion of the cranial, caudal, and lateral folds. By the end of 10 weeks, the intestines return to the abdomen (reduction of the midgut hernia), with the small intestine returning first and the large intestine returning while undergoing further counterclockwise rotation (180°). The cecum is the last structure to return.

Etiology and Pathogenesis

Omphaloceles are midgut malformations that are obvious at birth and result from the failure of the intestine to return to the abdomen during embryologic formation.

Several theories about the pathogenesis of gastroschisis have been proposed^{2,12} and can be summarized as follows:

- A primary insult causing incomplete lateral infolding of the embryonic disk results in an incompletely formed abdominal wall. This is the most widely accepted theory for gastroschisis.
- Abdominal contents rupture through the embryonic wall during the accelerated growth phase, and an intrauterine vascular accident occurs with occlusion of the omphalomesenteric artery and subsequent loss of circulation to the cord. This vascular accident then leads to a sequence of events that includes cord necrosis, abdominal wall opening, and evisceration of abdominal contents.
- Gastroschisis may be a variation of the omphalocele, with early intrauterine membranous rupture, reabsorption of the remaining membrane, and with re-formation of the umbilicus around the repositioned vessels.
- Finally, it is possible that gastroschisis is within a spectrum of intrauterine rupture of an umbilical hernia, as has been viewed ultrasonographically in a few cases.

Epidemiology

Omphalocele occurs in 1 of every 5,000 to 6,000 live births, compared to 1 of 10,000 to 50,000 occurrences for gastroschisis. Omphalocele carries a much greater risk than gastroschisis of accompanying anomalies and syndromes (Table 4). Comparative findings are summarized in Table 5.^{2,12-15}

Table 4. Anomalies and Syndromes Associated With Omphalocele and Gastroschisis^{2,12-15}

Omphalocele	Gastroschisis
Congenital heart defects	Malrotation
Genitourinary anomalies	Intestinal atresia
Vertebral malformations	Gastrointestinal obstruction
Intestinal atresia	Vertebral malformations
Midgut volvulus	Limb-body wall complex (lethal)
Meckel's diverticulum	
Intestinal duplication	
Malrotation	
Gastrointestinal obstruction	
Diaphragmatic and upper midline defects	
Trisomy 13 and trisomy 18	
Beckwith-Wiedemann syndrome	
Pentalogy syndrome	

Table 5. Comparative Incidence of Omphalocele and Gastroschisis^{2,12-15}

Omphalocele	Gastroschisis
Occurrence	Occurrence
1 in every 5,000 to 6,000 live births	1 in 10,000 to 50,000 live births
Male > Female	No gender preference
Associated anomalies/syndromes in 50%-77% of cases	Right-sided defect > left-sided
Intrauterine rupture of amnioperitoneal membrane = 11%-23%	Associated anomalies—up to 26%
Risk Factors	Risk Factors
Prematurity = 30%-33%	Prematurity = 58%
SGA = 19%	Low birth weight = 92%
	Malrotation = 100%

The overall mortality rate for omphalocele is 30% and is related to the size of the defect, associated anomalies, and degree of prematurity and/or low birth weight.^{2,15} In contrast, mortality from gastroschisis (13% to 28%) is directly related to the defect itself, the absence of peritoneal protection, and subsequent sequelae. Early death in gastroschisis, as a rule, is due to shock, sepsis, or hypothermia, whereas

late deaths are attributed to sepsis, respiratory failure, and nutritional issues related to GI dysfunction.

Assessment and Diagnosis

History

Omphalocele and gastroschisis can be identified in the perinatal period by abdominal ultrasound and are usually discovered incidentally.¹³ Characteristic ultrasound findings in omphalocele include the presence of a midline abdominal wall defect that extends into the base of the umbilical cord and surrounding content sac. With gastroschisis, findings include free-floating herniated bowel in the amniotic fluid without an intact peritoneal membrane.

Because of the association of omphalocele with other anomalies and syndromes, a detailed genetic history should be pursued.

Signs and Symptoms

Physical assessment. Initial assessment should focus on differentiating omphalocele from gastroschisis. Both defects are readily apparent at birth (Table 6). Differentiation is based on the presence or absence of the umbilical ring, identification of an intact sac or sac remnants if ruptured, and site of umbilical insertion. The umbilical ring is absent with an omphalocele and present in gastroschisis. A membranous sac or remnants of a sac can be found in omphalocele but not with gastroschisis. In a majority of patients with gastroschisis, the defect is located to the right of the umbilicus, and there is a normal umbilical insertion.

Treatment/Management

Clinical Management

Clinical management of omphalocele and gastroschisis share many elements, reflecting the similarity of the defects. These types of defects readily allow

thermal, fluid, and protein loss and each of these losses should be addressed in a timely and effective manner. This is especially important when surgical intervention is delayed. Care includes prevention of infection with surveillance and antibiotics; decompression of the gut; and prevention of hypothermia. This is particularly important in an infant with gastroschisis; hypothermia occurs in up to 67% of cases.² The use of topical agents to promote eschar formation and epithelialization is controversial and varies from institution to institution.

Surgical Management

Surgical correction of both defects depends on three factors: the size of the defect, accompanying anomalies or syndromes, and the infant's clinical tolerance of the procedure. Obviously, primary closure is preferred, but if this is not possible, a staged repair must be performed. Before a staged repair, placement of a prosthetic silicone silo is necessary for the gradual reduction of abdominal contents. In complicated cases in which associated congenital anomalies interfere with final ventral wall closure, a skin-flap closure over the remaining herniated viscera can be performed. Final ventral hernia repair is then delayed for 6 to 12 months.

Omphalocele. Primary closure in omphalocele is possible when the defect is ≤ 5 cm.² Definitive surgery is accomplished by returning the viscera to the abdominal cavity and closing the defect. Staged repair is required when the defect is too large for primary closure or when the neonate cannot tolerate the primary procedure. The infant may develop respiratory distress if the repair compromises air exchange and lung expansion.

Table 6. Signs and Symptoms of Omphalocele and Gastroschisis^{2,12}

Omphalocele	Gastroschisis
Variable herniation of contents into umbilicus Viscera covered by a thin, transparent membrane (peritoneum + amnion) Umbilical ring absent	Protrusion of intestine with or without colon As a rule, major organs remain in the abdomen; however, evisceration can occur Defect generally located to right of the umbilicus
Defect size ranging from 2 to 15 cm	Defect size ranging from 2 to 5 cm
Abdominal cavity generally underdeveloped and small, depending on the size of the defect	Abdominal cavity underdeveloped and small
Matted edematous bowel if sac rupture occurred in utero	Eviscerated bowel is edematous and matted as a result of exposure to amniotic fluid No identifiable bowel loops

Gastroschisis. A gastroschisis is usually closed with a staged repair as a result of the intrauterine amniotic fluid exposure and its effect on the viscera and bowel. Surgical enlargement of the defect is necessary to inspect and explore the entire gastrointestinal tract and allow adequate space for return of the abdominal contents. Virtually all such defects are accompanied by malrotation of varying degree, and it is sometimes necessary to perform resection and anastomosis due to intestinal atresia or infarction. If peritonitis or inflammation is present, primary anastomosis is contraindicated, necessitating an enterostomy with delayed anastomosis pending final closure.

Nursing Management

Nursing management includes routine neonatal care, along with the specific issues listed in Table 7.

Table 7. Nursing Issues and Focal Points of Care: Omphalocele and Gastroschisis
<p>Early recognition of associated anomalies</p> <p>Focal points of preoperative care:</p> <ul style="list-style-type: none"> • Decompression of GI tract • Defect/sac care to protect eviscerated organs • Hydration/fluid and electrolytes • Thermoregulation • Infection surveillance • Nutritional surveillance • Comfort measures <p>Focal points of postoperative care:</p> <ul style="list-style-type: none"> • Silo care • Respiratory assessment and support • Fluid and electrolyte shift—third spacing • Circulatory assessment for compromise of venous return secondary to vena cava impingement and hypovolemia • Ileus and cholestasis evaluation

Gastrointestinal Obstruction

ATRESIA AND STENOSIS

Normal Embryology

At approximately 4 weeks’ gestation, portions of the primitive gut—the caudal part of the foregut, the cranial portion of the midgut, and the splanchnic mesenchyme—give rise to the duodenum. The foregut is responsible for the development of the upper portion of the duodenum and celiac artery.

The midgut gives rise to the small intestine past the opening of the bile duct and superior mesenteric artery. During the 5th week of development, the duodenal lumen is obliterated as a result of lining-cell proliferation and normally recanalizes by the end of the embryonic period.

Etiology and Pathogenesis

Atresia is the complete disruption and obstruction of the lumen of the intestine or alimentary tract secondary to a congenital loss of continuity in the bowel lumen. In contrast, stenosis is a narrowing of the lumen that may result in an incomplete obstruction. Types of intestinal atresia are described in Table 8.^{12,16}

Table 8. Types of Intestinal Atresia ^{12,16}
<p>Type I</p> <p>Intact-membrane (mucosa and submucosa) obstruction Muscularis and serosa intact No external indication of obstruction Change in diameter of bowel: distal to actual lesion</p> <p>Type II</p> <p>More significant gaps in continuity of bowel or greater atretic area Short fibrous bands connecting proximal and distal segments</p> <p>Type IIIa</p> <p>No connecting tissue between segments Usually associated V-shaped gap in mesentery</p> <p>Type IIIb</p> <p>Proximal small-bowel atresia Absence of distal superior mesenteric artery Absence of dorsal mesentery “Apple peel” or “Christmas tree” defect and foreshortening of small bowel distal to atresia: familial association High jejunal or distal duodenal Distal ileum wrapped around a thin vascular stalk, usually consisting of the ileocolic artery Large gap in mesentery Intestinal malrotation Microcolon</p> <p>Type IV</p> <p>Multiple atresias Bowel may be foreshortened Familial association</p>

Duodenal atresia is the failure of recanalization of the lumen, resulting in segmental occlusion. In contrast, duodenal stenosis is the result of incomplete recanalization of the lumen during the 8th week of embryogenesis.

Jejunioileal atresia is the result of ischemic necrosis of the fetal intestine, thought to be secondary to mesenteric vascular compromise from volvulus, intussusception, or an internal hernia. The area of necrosis is reabsorbed, resulting in intestinal segment loss and blind proximal and distal ends.

Epidemiology

Duodenal Atresia

Duodenal atresia occurs in 1 in 10,000 to 1 in 40,000 births and accounts for 50% or more of all duodenal obstructions. Approximately 50% of cases occur in premature and/or low-birth-weight infants; 30% to 70% of affected infants present with associated anomalies (see Table 9), and up to 15% of patients have two or more anomalies. Thirty percent occur in infants with Down syndrome and 20% have annular pancreas. Twenty-five percent to 50% of these cases have a prenatal presentation of polyhydramnios.^{2,12,16-18}

Table 9. Duodenal Atresia and Associated Anomalies^{2,12,16-18}

Esophageal atresia
Malrotation of the midgut
Annular pancreas
Biliary atresia
Preduodenal portal vein
Imperforate anus
Renal anomalies
Congenital heart disease
Intrauterine growth retardation
VACTERAL/VATER (vertebrae, anorectal, cardiovascular, tracheoesophageal, renal, and limb abnormalities)

The mortality rate for this intestinal anomaly ranges from 7% to 35% and is generally related to the associated anomalies, complications of prematurity, and infection.^{2,18}

Jejunioileal Atresia

Jejunioileal atresia is far more common than duodenal atresia, with rates from 1 in 332 to 1 in 5,000 births; 25% to 38% of affected infants are premature. Associated anomalies are less common in this type of intestinal atresia and are generally restricted to the gastrointestinal tract. Malrotation with volvulus,

gastroschisis, and meconium ileus are the most frequent associations. As a rule, jejunioileal lesions occur equally in the jejunum and ileum. Although predominantly single lesions, multiple lesions are present 6% to 20% of the time.^{2,12,16}

Mortality is 10% with this lesion and increases with complicated types IIIb and IV. Infants presenting with the “apple peel” or “Christmas tree” (see Table 8) deformity have a 54% death rate. Survival rates depend on associated anomalies and prematurity, as well as the type of lesion, length of small bowel remaining, nutritional status, complications related to anastomotic dysfunction or obstruction, stricture and/or adhesions, and total parenteral nutrition (TPN)-related liver disease.

Colonic Atresia

Colonic atresia is the rarest defect, occurring in approximately 1 in 40,000 live births, and accounting for only 10% of all atresias. This lesion can be overlooked in surgery, as inspection of the surface of the bowel often reveals no abnormality. Although the association is rare, colonic atresia can occur with Hirschsprung’s disease.¹⁶

Assessment and Diagnosis

History

Pertinent prenatal history is similar for all atresias. Polyhydramnios occurs in approximately 15% to 50% of all GI obstructions. It is most common (>50%) with duodenal involvement and rarest in ileal atresia.¹⁶ There is a positive familial history with type IIIb “apple peel” and type IV multiple atresias, and an increased risk of small intestine obstruction in twins, particularly monozygotic twins.¹⁹ A review of antenatal ultrasounds is advisable, since they can often assist with postnatal diagnosis. The diagnosis needs to be confirmed postnatally with standard diagnostic studies.

Signs and Symptoms

Physical assessment. A gastric aspirate > 10 to 15 mL obtained in the delivery room is suggestive of obstruction. Postnatal clinical signs are listed in Table 10. The degree of abdominal distention depends largely on the level and type of obstruction. Timing of the onset of vomiting and passage of stool is important. If the site of obstruction is proximal, the vomiting usually occurs earlier, with the potential of normal meconium passage. With distal obstructions, vomiting often occurs within a few hours of birth, and there is failure to pass stool. A majority of duodenal obstructions occur below the ampulla of Vater, and bilious vomiting can be expected as a presenting symptom. With distal obstruction, visible and palpable bowel loops may be present.

Table 10. Signs and Symptoms of Intestinal Obstruction ^{2,12,16,18}

General Findings	
Large gastric aspirate in delivery room and ongoing Bilious vomiting Failure to pass meconium if lesion is distal Abdominal distention Significant gastric aspirates Emesis Prolonged jaundice	
Duodenal Obstruction	Jejunioileal Obstruction
Bilious vomiting in 1st day of life if lesion proximal but below ampulla of Vater Limited abdominal distention in distal lesions, scaphoid appearance of lower abdominal cavity	Presentation in 1st few days of life Bilious vomiting Generalized abdominal distention

Diagnostic Tests

Roentgenographic findings. If intestinal obstruction is being considered as a diagnosis, a plain abdominal roentgenograph should be the first diagnostic test ordered. In duodenal obstruction, the appearance of the classic “double bubble” reveals air in the stomach and first duodenal segment proximal to the obstruction, suggesting atresia. The presence of fluid levels may interfere with these findings, and removal of gastric contents via a nasogastric tube and injection of 30 to 60 mL of air will assist in the diagnosis. The absence of air distal to the duodenum suggests atresia. Small isolated pockets of air past the duodenum indicate stenosis. Minimal gas distally in the jejunum or ileum does not rule out atresia, and may be the result of gas moving through pancreatic ducts. Contrast studies are indicated to rule out malrotation with volvulus and to further define the pathology.

In jejunioileal obstruction, the plain abdominal roentgenograph routinely demonstrates dilated small-bowel loops with air-fluid levels. Generally, the more distal the obstruction, the greater the distention. The presence of intraperitoneal calcification is suggestive of prenatal perforation and meconium peritonitis.

Differential diagnosis. The differential diagnosis for intestinal atresia includes colonic atresia, Hirschsprung’s disease, malrotation and volvulus, and meconium ileus.¹⁶ To rule these out and to identify the presence of incomplete or partial

obstructions (duodenal stenosis or web, and annular pancreas), a lower gastrointestinal series followed by an upper GI series should be performed. With colonic atresia, a rectal biopsy is advised at the time of surgery to rule out Hirschsprung’s disease.

Treatment/Management

Clinical Management

Preoperatively, traditional stabilization measures should be instituted with particular attention to intravenous access, fluids and electrolytes, bowel/gut decompression, and close surveillance for infection/sepsis. Ruling out malrotation with volvulus is a priority. Associated anomalies and prematurity may require considerable stabilization before surgery; however, surgical emergencies take precedence (see “Malrotation with volvulus,” page 10).

Surgical Management

Surgical intervention with duodenal atresia/stenosis is accomplished by removing the atretic bowel and performing either an end-to-end or side-to-side duodenoduodenostomy or duodenojejunostomy, depending on the level of obstruction. Additional stenosis or atresia should be ruled out, and a gastrostomy is often performed for decompression. With most jejunioileal atresia/stenosis, surgical correction is accomplished with a simple end-to-end anastomosis after resection of all dilated and atretic portion(s) of bowel, and tapering if necessary (see Table 11 for surgical options).^{7,12,16} If a large amount of bowel is resected, short-bowel syndrome may result. Placement of a feeding jejunostomy tube via the gastrostomy has been found to be effective for feeding infants with high atretic lesions.¹²

Table 11. Surgical Strategies for Intestinal Obstruction ^{7,12,16}

Duodenal Obstruction	Jejunioileal Obstruction
Rule out additional stenosis or atresia	Multiple atresias repaired at primary surgery
Resection of atretic bowel	Resection of dilated proximal and atretic distal bowel
End-to-end or side-to-side duodenoduodenostomy	Surgical tapering (enteroplasty)
Gastrostomy/nasogastric tube placement	Simple end-to-end anastomosis
	End-to-oblique side anastomosis
	Ostomy placement when > 2:1 ratio between size of dilated proximal intestine to distal nondilated segment
	Placement of feeding jejunostomy at surgery if necessary

Postoperative Care

Postoperative care usually consists of traditional stabilization, gut decompression, and nutritional support. TPN is usually instituted early (24 to 48 hours after surgery) and continued until enteral feedings are successfully established and appropriate weight gain is documented.

Postoperative complications generally are related to surgery, sepsis, TPN, or short-bowel syndrome. Surgical complications include anastomotic dysfunction with obstruction, stricture, and adhesions. Prolonged use of TPN can result in liver compromise and failure. Short-term and long-term nutritional implications in infants with short bowel necessitate considerable collaborative management and support.

Nursing Management

Nursing management includes routine neonatal care, along with the specific issues listed in Table 12.

Table 12. Nursing Issues and Focal Points of Care: Intestinal Obstruction

Early recognition of defect and any associated anomalies

Focal points of preoperative care:

- Gastric decompression
- Hydration/fluids and electrolytes
- Infection prevention/surveillance

Focal points of postsurgical care:

- Gastric decompression and ostomy care
- Nutritional status and support
- Infection prevention/surveillance
- Accurate documentation of fluid losses

MALROTATION WITH AND WITHOUT VOLVULUS

Normal Embryology

Normally, the intestines return to the abdominal cavity at about 10 weeks of gestation. In doing so, they undergo a 270° counterclockwise rotation around the superior mesenteric artery axis, followed by attachment to the posterior abdominal wall.

Etiology and Pathogenesis

When normal embryology is interrupted or arrested during intestinal reentry, rotation, and fixation, malrotation or nonrotation can result. Because arrest of the normal embryologic sequence can occur at any time, a variety of anomalies occur:

- **Nonrotation:** The duodenal-jejunal loop ends up on the right side of the abdomen and the cecal-colic loop on the left, with a broad mesentery base. The duodenum is separate from the cecum.

- **Malrotation:** The cecum rests in the right upper quadrant near the duodenum and the duodenal-jejunal loop remains to the right of the midline. The mesenteric base is a narrow, thin pedicle centered on the superior mesenteric artery and vein. The vein may run to the left of the artery, rather than to the right. This defect carries the highest risk for volvulus.
- **Reverse, or clockwise, rotation:** This can result in the duodenum overlying and obstructing the transverse colon.
- **Intestinal volvulus:** This is the most serious possible complication of malrotation. The intestine becomes obstructed by kinking, knotting, or twisting. Secondary ischemia, bowel necrosis, perforation, and peritonitis may result. Intestinal and lymphatic obstruction can occur and can result in necrosis of the midgut, with grave consequences. Lymphatic obstruction presents as chylous ascites.
- **Duodenal obstruction:** This can occur secondary to peritoneal bands (Ladd's bands), from the cecum to the right upper quadrant.

Epidemiology

Malrotation is a relatively rare abnormality, and the exact statistical incidence is not known. Two thirds of the cases of malrotation are accompanied by volvulus. Malrotation with volvulus is found predominantly in males, and 24% of affected infants have other GI anomalies that include, but are not limited to, intestinal atresia, annular pancreas, and Meckel's diverticulum, as well as urinary tract malformations. Congenital heart disease is found in 8% to 24% of patients. These patients are also prone to heterotaxia, an abnormal arrangement of organs or other parts of the body. The prognosis is excellent in uncomplicated cases, with a mortality rate of 2%. With intestinal necrosis, mortality increases to as high as 65%. Up to 85% of cases of malrotation in infants present in the neonatal period, and approximately 50% of affected infants with volvulus present with symptoms during the 1st week of life. For a summary of anomalies associated with nonrotation and malrotation, see Table 13.^{2,12,16}

Assessment and Diagnosis

History

The onset and pattern of symptoms vary with the degree and nature of the obstruction. Typically, the neonate is initially well with no indications of problems. Recurrent, intermittent signs and symptoms suggest a partial obstruction or a total but reversible obstruction.

Nonrotation	Malrotation
Gastroschisis	Congenital diaphragmatic hernia
Congenital diaphragmatic hernia	Hirschsprung's disease
Omphalocele	Omphalocele
	Gastroschisis
	Duodenal atresia
	Intussusception (occasional)
	Mesenteric cysts
	Annular pancreas
	Meckel's diverticulum
	Urinary tract malformations
	Heterotaxia

Signs and Symptoms

Malrotation. Most infants with malrotation are likely to feed initially and to pass meconium. They may develop jaundice, anorexia, and symptoms consistent with upper intestinal obstruction (see Table 14).^{2,12,16}

Malrotation with volvulus. Midgut volvulus usually occurs during infancy, but can occur at any age and often presents in a sudden fashion. A presentation consistent with duodenal obstruction often accompanied by jaundice is common.

When clinical manifestations (see Table 14) such as sudden onset of bilious vomiting occur in a previously normal newborn, volvulus should be suspected and assessed immediately. Other signs and symptoms include rectal bleeding and abdominal distention, which indicate vascular compromise. In the case of lymphatic obstruction, chylous ascites becomes evident. Shock and cardiovascular collapse indicate a catastrophic event related to complete volvulus and bowel necrosis.^{2,12,20}

Diagnostic Tests

Malrotation with or without volvulus is predominantly diagnosed roentgenographically. The most frequent roentgenographic presentation is that of intestinal obstruction. Sonography may demonstrate mesenteric inversion (superior mesenteric vein left of the superior mesenteric artery) and the “whirlpool” sign (superior mesenteric vein encircling the artery in a clockwise direction), suggesting malrotation.²¹

Roentgenographic findings. Plain films of the abdomen can suggest the diagnosis but are not definitive by themselves. Duodenal obstructions generally occur early in fetal development, giving rise to greater bowel distention and a classic “double bubble” x-ray appearance representing gas-fluid levels. Jejunal obstruction may result with x-rays showing several gas-filled loops of bowel in the upper left quadrant and calcifications in the various areas of involvement. Contrast studies with barium or absorbable contrast agents, particularly the upper GI series, are the most accurate tools of diagnosis. Volvulus often affects the distal third of the duodenum; the findings are often similar to those with duodenal and jejunal obstruction.

Malrotation with midgut volvulus is a surgical emergency. It must be suspected and verified or ruled out in all cases of upper intestinal obstruction in infants. The two studies most consistently used to confirm this diagnosis are the barium enema and upper GI barium contrast study.

Barium enema demonstrates obstruction at the mid-transverse colon and location of the cecum in the right upper quadrant. An upper GI barium contrast study shows abnormal positioning of the duodenojejunal junction, a distal duodenal obstruction or “corkscrew” appearance of the distal duodenum or proximal jejunum, absence of the normal ligament of Treitz to the left of the midline, and a barium column ending with a “beaking” effect. Isolated

Malrotation	Malrotation With Intermittent or Partial Obstruction	Volvulus
Clinically silent At risk for: • Intermittent or partial obstruction • Volvulus	Protracted bilious vomiting Abdominal pain Anorexia/failure to thrive Signs of partial or total obstruction/duodenal Jaundice	Progressive or sudden onset of bilious vomiting in previously healthy neonate Rectal bleeding/blood-tinged mucousy diarrhea ± Abdominal distention Chylous ascites In older children—anorexia and right upper quadrant fullness Shock

asymptomatic malrotation without volvulus is often revealed with barium contrast studies; typical findings are a dilated duodenum, abnormal position of the duodenojejunal junction, jejunal loops in the right side of abdominal cavity, and malpositioned cecum.

Treatment/Management

Clinical Management

The goals of immediate preoperative care are twofold: stabilization that does not preempt surgery, and preservation of as much bowel as possible. Early stabilization includes provision of intravenous access, bowel decompression, fluid and electrolyte maintenance and/or correction, resuscitation from shock (respiratory, cardiac, and metabolic), and administration of antibiotic therapy.

Surgical Management

Surgical management depends entirely on operative findings. The goals are correction of the obstruction and recovery of as much bowel as possible. A summary of surgical procedures is included in Table 15.^{2,12,16}

Table 15. Surgical Management Highlights: Malrotation and Volvulus^{2,12,16}

<p>Untwisting/detorsion of bowel (counterclockwise rotation around axis of superior mesenteric artery and vein)</p> <p>Division of cecal bands (Ladd's bands) between cecum and paraduodenal region</p> <p>Ladd's procedure: dissection between cecum and right colon on the left and duodenum on the right, with mesenteric broadening around the superior mesenteric artery and vein (can be delayed if second definitive surgery planned within 12-18 hours after initial surgery)</p> <p>Straightening of duodenal loop with positioning on the right side of the abdomen and placement of the colon on the left side</p> <p>Removal of appendix</p> <p>With volvulus, staging may be necessary to determine viability of ischemic bowel; nonviable bowel is removed in initial surgery; a second surgery follows, usually 24 hours later, to reassess bowel viability that was initially questionable</p>
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Nursing Management

Nursing management includes routine neonatal care, along with the specific issues listed in Table 16.

MECONIUM ILEUS

Meconium ileus is a mechanical obstruction of the distal small bowel and colon by abnormally thick and viscous meconium. This abnormal meconium is the result of pancreatic exocrine-ecrine dysfunction or insufficiency and abnormal intestinal secretion, predominantly related to cystic fibrosis.

Table 16. Nursing Issues and Focal Points of Care: Malrotation and Volvulus

<p>Early suspicion of possible volvulus:</p> <ul style="list-style-type: none"> • Prevention of short-bowel syndrome <p>Focal points of preoperative care:</p> <ul style="list-style-type: none"> • Gastric decompression • Fluid and electrolyte stabilization <p>Focal points of postoperative care:</p> <ul style="list-style-type: none"> • Gastric decompression • Fluid and electrolyte stabilization if necessary • Nutrition
--

Etiology and Pathogenesis

Cystic fibrosis is the most common underlying cause of meconium ileus. Cystic fibrosis is an autosomal recessive genetic disorder that is characterized by pancreatic dysfunction and enzyme deficiency, and production of highly viscous and tenacious meconium. It is more often associated with chronic progressive pulmonary disease seen in the pediatric population, but it affects all exocrine glands. The intraluminal accumulation of this abnormal meconium antenatally is responsible for meconium ileus.^{22,23} Meconium ileus not associated with cystic fibrosis is generally related to other pancreatic disorders (eg, pancreatic ductal stenosis, partial pancreatic aplasia).

Epidemiology

The incidence of meconium ileus varies between 1 in 1,150 and 1 in 5,000 live births. Ninety-five percent of infants who present with meconium ileus have cystic fibrosis; conversely, up to 25% of patients with cystic fibrosis demonstrate meconium ileus. As many as 33% of cases are associated with small-bowel obstruction. Cystic fibrosis is predominantly seen in Caucasians, is rare in African Americans, and is essentially absent in Asians.

The short-term prognosis is dependent on surgery and the development of complications. Infant mortality and morbidity are related to malabsorption and malnutrition. The long-term prognosis is related to the progression of the pulmonary disease.

Assessment and Diagnosis

History

A family history of cystic fibrosis or meconium ileus can be obtained in up to 33% of cases and should be a primary question when obtaining a history. There is a 10% to 33% association with prematurity. An antenatal history of polyhydramnios and intrauterine growth retardation would be suggestive as well. The

even distribution of feces in the colon observed roentgenographically in adults and older children is rarely seen in neonates before 2 weeks of age. Such an appearance before this age suggests bowel pathology, most likely related to Hirschsprung's disease, meconium ileus, or necrotizing enterocolitis (NEC). Clinically, meconium ileus may be uncomplicated or simple, or complicated, as seen in Table 17.^{12,16,23}

Uncomplicated	Complicated
67% occurrence	33% occurrence
Symptoms 24 to 48 hours after birth	Appearance of symptoms dependent on lesion
Involves distal small bowel:	Features of uncomplicated form plus any of the following:
<ul style="list-style-type: none"> • Dilated proximal small bowel • Thickened bowel walls • Microcolon (narrow, empty distal colon) 	<ul style="list-style-type: none"> • Intestinal volvulus • Atresia • Necrosis • Perforation • Meconium peritonitis • Meconium pseudocyst • Scrotal and inguinal calcifications

Signs and Symptoms

Common clinical features associated with uncomplicated and complicated forms of meconium ileus are those associated with obstruction.

Diagnostic Tests

Roentgenographic findings. Roentgenographic findings for uncomplicated meconium ileus are consistent with those for ileal atresia, Hirschsprung's disease, and small left-colon syndrome. Characteristics include low intestinal obstruction, multiple distended proximal bowel loops, inspissated "soap bubble" and "ground glass" appearance in the right lower quadrant, and little or no air-fluid level on upright films. Up to one third of cases show no differentiating roentgenographic features. Complicated meconium ileus may be suggested by free peritoneal fluid and reveal calcifications on plain films. Bowel loops usually vary significantly in size and are unevenly distributed, based on their location.

Roentgenographic confirmation depends largely on the contrast enema (see "Clinical Management"). The usual findings with this study are normally positioned microcolon, reflux into the terminal ileum, and outlined inspissated meconium.

Differential diagnosis. The differential diagnosis includes ileal atresia, Hirschsprung's disease, neonatal small left colon, and meconium plug syndrome. The diagnosis is confirmed by the medical or surgical discovery of the classic sticky meconium and confirmation of the diagnosis of cystic fibrosis. Distention of intestinal mucosal goblet cells is a hallmark in the histologic evaluation of meconium ileus. Once meconium ileus is definitively diagnosed, cystic fibrosis becomes a clinical diagnosis as well. The two diagnostic tests for this disease are the sweat chloride test and genotyping of the CFTR gene.

Treatment/Management

Clinical Management

Immediate management of meconium ileus consists of gastric decompression and fluid and electrolyte correction and maintenance. Additional considerations include ongoing nutritional support, respiratory care, and genetic and supportive counseling for the family, most of which are comprehensively addressed when the infant is referred to a regional center for the management of cystic fibrosis.

Nonsurgical treatment of uncomplicated cases of meconium ileus can be accomplished at the time of the contrast study, provided the criteria for this procedure (Table 18) are met. Historically, hypertonic solutions have been used for the procedure, but they are not without complications because of the induced fluid shift and risk of hypovolemic shock. Although the success of this therapy is variable, the benefits are thought to outweigh the risks. A safer, but more expensive, method is the use of low-osmolarity or iso-osmolar contrast media mixed with *N*-acetylcysteine.²⁴

Exclusion of other causes of distal intestinal obstruction
Exclusion of complications of meconium ileus
Availability of fluoroscopic surveillance at the time of procedure
Antibiotic coverage
Stable-patient status following successful fluid resuscitation
Surgical team prepared for complications and possible surgical intervention

Surgical Management

Surgical treatment of uncomplicated meconium ileus is necessary if the contrast enema is not therapeutic, if obstruction and distention increase, or if a complication or perforation occurs during the procedure. Surgical intervention entails opening the bowel and removing the meconium by irrigation, placing an ileostomy at the proximal end of the obstructed segment, and, in some cases, inserting a T tube for postoperative irrigation with acetylcysteine.^{8,23}

Most complicated cases of meconium ileus require surgery. The type of surgery depends on the complications found at the time of surgery.

Nursing Management

Nursing management includes routine neonatal care, along with the specific issues listed in Table 19.

Table 19. Nursing Issues and Focal Points of Care: Meconium Ileus

Screening for cystic fibrosis
Early initiation of appropriate referrals
Stabilization:
• Gastric decompression
• Establish fluid and electrolyte balance
• Monitor respiratory status, cardiovascular status
Nutrition intervention
Ostomy care
Cystic fibrosis support:
• Genetic referral and counseling
• Respiratory care
• Parental support and education about ostomy care, respiratory care, and enzyme supplementation; assess need for counseling and nutrition education

HIRSCHSPRUNG'S DISEASE/ NEONATAL AGANGLIONOSIS

Normal Embryology

During the 5th to 7th week of gestation, craniocaudal migration of neurocrest cells to the colon is accomplished. Complete migration can occur as late as 12 weeks. This results in the normal innervation of the colon and normal peristalsis.

Etiology and Pathogenesis

Hirschsprung's disease, also known as neonatal aganglionosis, congenital megacolon, or aganglionic megacolon, is a neuroganglionic defect in the innervation of the colon or portion(s) of the colon

that renders it dysfunctional or ineffective in the evacuation process. This defect results from the absence of myenteric and submucosal ganglionic cells (aganglionosis) and subsequent loss of parasympathetic innervation.

The cause of the neuroganglionic defect is not clear, but is thought to be related to²⁵:

- Failure of neuronal crest cell migration in the usual craniocaudal fashion resulting in the absence of ganglionic cells in the distal colon
- Failure of neuroblasts to develop into normal, mature ganglia
- Degradation of normally developed cells, possibly as a result of an ischemic insult

Aganglionosis results in the loss of inhibitory parasympathetic nerves in the myenteric plexus. This results in a chemical inhibition of segment relaxation of the normally contracted enteric smooth muscle of the colon, which leads to a lack of peristalsis and functional obstruction. This disease always involves the anus and has variable distal colonic involvement. Rectosigmoid lesions are present in 75% to 80% of cases, and 8% to 10% have total colonic involvement.

Patients with long-segment involvement, which is rare, are at risk for zonal aganglionosis or segmental aganglionosis or "skip areas."

Hirschsprung's disease is considered a heterogeneous genetic condition with autosomal-dominant, autosomal-recessive, and polygenic subtypes. Eighty percent of cases are attributed to the latter two subtypes.^{2,25}

Epidemiology

The incidence of Hirschsprung's disease varies between 1 in 5,000 and 1 in 10,000 live births. Sixteen percent to 25% of children with chronic constipation (defined by individual stooling patterns) are diagnosed with Hirschsprung's disease. It has no racial association. Males are at increased risk, with a male-to-female ratio of approximately 4:1. There is a familial occurrence of 7% to 30%, especially with long segmental lesions, and a 2% to 7% risk among family members. Mortality rates range from 4% to 33%, depending on the presence or absence of secondary enterocolitis and associated syndromes.^{2,12,25-27}

Hirschsprung's disease is specifically associated with a number of syndromes, particularly Down syndrome (see Table 20). Associated cardiac defects

have been reported in as many as 50% of patients with Hirschsprung's disease.^{2,12,14,25-27}

Congenital Anomalies	Syndromes
Gastrointestinal: <ul style="list-style-type: none"> • Imperforate anus • Malrotation Genitourinary: <ul style="list-style-type: none"> • Inguinal hernia • Asymptomatic disorders <ul style="list-style-type: none"> - Cryptorchidism - Urinary bladder diverticulum Cardiac: <ul style="list-style-type: none"> • Ventricular septal defect • Tetralogy of Fallot • Patent ductus arteriosus Central nervous system: <ul style="list-style-type: none"> • Mental retardation 	Frequently seen in: <ul style="list-style-type: none"> • Down syndrome • Smith-Lemli-Opitz syndrome • Waardenburg syndrome

Assessment and Diagnosis

History

A familial history of Hirschsprung's disease is the most common finding in the neonate with this disorder. In older children, delayed toilet training, chronic constipation, fecal impaction, and soiling are consistently found in the history, along with poor weight gain and abdominal distention. Retrospectively, the vast majority of older children diagnosed with this disease have developed symptoms within the first few months of life.

Signs and Symptoms

Presentation may occur in the immediate newborn period after birth or several weeks later. Presenting symptoms are consistent with those of low intestinal obstruction: abdominal distention, bilious emesis, and a history of delayed passage of meconium > 24 to 48 hours. However, some neonates present with enterocolitis, secondary electrolyte imbalance, dehydration, and shock. Infants who also have Down syndrome and/or a delayed diagnosis of Hirschsprung's disease beyond the 1st week of life have the greatest risk for enterocolitis in the neonatal period. In older children, clinical characteristics reflect chronic obstruction and fecal retention secondary to

increased anal sphincter tone. Chronic abdominal distention, constipation, and failure to thrive are common findings in these children. Colitis is less common in older children. A comprehensive list of signs and symptoms is presented in Table 21.^{12,25,26}

Neonates	Infants and Children
Intestinal obstruction: <ul style="list-style-type: none"> • Failure to pass meconium within first 48 hours • Abdominal distention • Bilious vomiting Enterocolitis: <ul style="list-style-type: none"> • Abdominal distention • Diarrhea/foul-smelling bloody stools • Explosive diarrhea with rectal examination • ± Fever • Lethargy • ± Shock Protein-losing enteropathy Dehydration Electrolyte imbalance Septicemia	Chronic constipation, fecal impaction, and soiling Vomiting Abdominal distention Failure to thrive or poor weight gain Diarrhea Perforation of appendix or colon Anal fissures with bleeding Delayed toilet training Secondary ureteral compression—retention, hydronephrosis, and hydronephrosis Empty rectum on digital examination Enterocolitis: <ul style="list-style-type: none"> • Abdominal distention • Explosive watery stools • Fever • Hypovolemic shock

Timing. The timing of diagnosis and the presence or absence of enterocolitis generally determine prognosis. In the past, delayed diagnosis was common. Early diagnosis reduces complications and morbidity. Short-segment involvement poses the greatest risk for delayed diagnosis. Up to 41% of cases are diagnosed within the 1st month of life, but 8% are not diagnosed until 3 to 7 years of age (see Table 22).^{25,26}

8% to 41% within 1st month of life
64% by 3rd month
35% to 80% in 1st year
8% delayed diagnosis at 3 to 7 years of age

Diagnostic Tests

Roentgenographic findings. Plain film findings are consistent with intestinal obstruction, typically with dilated bowel loops to the level of the obstruction. Absent pelvic gas in the prone position is another common finding. Hirschsprung's disease in the neonate is accurately diagnosed by contrast enema in about 80% of cases. Barium retention, although not diagnostic, is frequently an additional finding, and delayed films should be considered. It is recommended that no bowel preparation or digital rectal examinations be performed prior to the barium enema to preserve the appearance of the transitional zone. A transitional zone, the area between the normal and abnormal segments of the bowel, may not be present in the newborn. As a rule, the rectal diameter is smaller than that of the sigmoid colon. In the ultrashort-segment disease where the length of the affected segment is <5 cm, colonic narrowing may not be demonstrated by the barium enema; manometry and rectal biopsy are required for definitive diagnosis.^{24,25,28}

Anal manometry. Anorectal manometry is used as a diagnostic tool when contrast studies fail to establish a diagnosis, particularly with ultrashort-segment disease. Anorectal manometry assesses basal anal sphincter tone and relaxation in response to increased rectal ampullary pressure (balloon distension). In Hirschsprung's disease there is typically increased resting tone and contraction upon balloon distension. This tool may be unreliable in the neonatal population, especially in premature neonates, and in cases where previous rectal manipulation has occurred. Outside of the neonatal population, anal manometry has a diagnostic accuracy of $\geq 90\%$.^{12,25,26}

Rectal biopsy. The definitive diagnostic tool for Hirschsprung's disease is the rectal biopsy, which establishes the absence of ganglion cells in submucosal and myenteric plexus. Rectal biopsies can be obtained with a special suction biopsy tube or surgically when a full-thickness muscle sample is desired. The ability to make a definitive diagnosis is close to 100%, providing adequate tissue is sampled, appropriate staining techniques are used, and expert interpretation is available. Infants with meconium plugs should undergo rectal biopsy to rule out Hirschsprung's disease, as these two disorders can coexist. In the critically ill neonate, the biopsy can often be accomplished at the time of surgical decompression.

Differential Diagnosis

The differential diagnosis of Hirschsprung's disease in the immediate newborn period includes conditions leading to intestinal obstruction, such as meconium plug syndrome, meconium ileus, microcolon, jejunoileal atresia, and malrotation. Consideration should also be given to anorectal malformations, necrotizing enterocolitis, and sepsis with ileus. In older infants and children, possibilities include cystic fibrosis, hypothyroidism, and various rare forms of neuronal dysplasias.^{2,12,25,26}

Treatment/Management

Clinical Management

Neonatal clinical care is geared toward the early detection of Hirschsprung's disease and prevention of enterocolitis, perforation, and peritonitis. Supportive care that includes gut decompression, the prevention and treatment of any existing fluid and electrolyte imbalance, and antibiotic treatment of sepsis is important until surgical intervention can be accomplished.

Surgical Management

Surgical treatment is aimed at decompression, fecal diversion, assessment of the level(s) of involvement, and removal or bypass of the aganglionic bowel with anastomosis of normal intestine. A temporary colostomy is usually placed during the initial surgery, especially in neonates. Definitive surgery is accomplished when the infant reaches a weight of at least 10 kg and an age of 6 to 12 months. The colostomy is placed proximal to the aganglionic segment of bowel for decompression and diversion of fecal contents. Biopsies from along the colon are obtained to determine the level of involvement. The definitive repair includes resection of affected areas and anastomosis of the normal bowel to the anus in a pull-through procedure. Primary repair is an option but is determined by the age of the patient and the extent of aganglionosis.^{25,29}

Postsurgical complications vary, depending on the clinical course and the type of surgical procedure used. The most common complications include perioperative infections, anal stricture, and enterocolitis. Satisfactory outcome is reported in up to 90% of cases. Long-term problems with continence may develop and approximately 1% of patients will require a permanent colostomy. Patients with total colonic involvement are expected to have less successful outcomes, and those with the rare cases of both small and large intestinal aganglionosis have the poorest outcomes and highest mortality rates.²⁵

Nursing Management

Nursing management includes routine neonatal care, along with the specific issues listed in Table 23.

Table 23. Nursing Issues and Focal Points of Care: Hirschsprung's Disease

Establish diagnosis:

- Family history
- Failure to pass stool in first 48 hours of life
- Signs and symptoms of obstruction

Focal points of preoperative care:

- Gastric decompression
- Fluid and electrolyte, respiratory, and cardiovascular surveillance
- Monitoring for signs and symptoms of fluid shift
- Biopsy surveillance/bleeding
- Daily enemas **not** recommended; contraindicated during the neonatal period due to the risk of enterocolitis, perforation, peritonitis, and septicemia
- Preoperative bowel care

Focal points of postoperative care:

- Assessment and surveillance for:
 - Respiratory compromise
 - Abdominal distention
 - Hemorrhage
 - Wound site
 - Infection/enterocolitis/sepsis
- Ostomy care
- Nutrition support

of associated internal defects. A vast majority of these defects include some type of fistula from the distal rectum to the perineum or genitourinary system. They are a result of abnormal development of the urorectal septum. Imperforate anus is the result of the failure of rectal descent.

Epidemiology

The incidence of anorectal malformations is approximately 1 in 5,000 live births. Twenty percent to 75% of patients with this disorder have associated anomalies. Anorectal malformations are found frequently as part of several syndromes and sequence malformations, most notably the VACTERAL/VATER association. Genitourinary defects are present in 25% to 50% of children with anorectal malformations; of these, 4% have lethal renal agenesis and/or dysplasia. Approximately half of anorectal malformations have spinal dysraphism. Thirty-three percent of anorectal malformations are associated with bone and sacral anomalies. Cardiac and central nervous system abnormalities can also occur. The mortality rate is 20% and varies with the type of defect and associated anomalies.^{2,8,12,14,31,32}

Assessment and Diagnosis

Signs and Symptoms

Presenting signs and symptoms depend on the type of defect and associated anomalies, although a pattern of low intestinal obstruction is typical. In the neonate, visceral pain may not be identified. Vomiting may or may not occur, but if it is present, it characteristically changes over time. Initially, stomach contents are a yellow-green and bilious, then change to a greenish-brown, and eventually turn feculent. Abdominal distention and failure to pass stool in the neonate are also features of anorectal defects. Imperforate anus is obvious on physical exam. However, anal stenosis and imperforate anal membranes may be present with what appears to be a normal perineum. Anal stenosis is often noted upon closer inspection. The anus is microscopic in appearance, and significant narrowing is noted on digital examination. Passage of meconium may also occur with the digital exam. A membranous lesion may not be apparent until the infant presents with signs and symptoms of obstruction. The presence of a fistula is often documented by finding meconium in the urine or around the vaginal outlet.

Clinically, lesions can be separated into two categories: high and low. High lesions lie above the puborectal component of the levator ani complex, with the rectum terminating above the supralelevator. Low lesions occur below the puborectal component of the levator ani complex with the rectum terminating below the translevator (infralevator). Determination

ANORECTAL MALFORMATIONS

Normal Embryology

The superior two thirds of the anal canal comes from the hindgut, the blood supply of which is derived from superior rectal arteries. The inferior third develops from the proctodeum, the blood supply for which is derived from the inferior rectal arteries. During the 4th to 6th week of embryogenesis, the anorectum develops as a result of the separation of the primitive cloaca into anterior urogenital sinus and posterior intestinal canal by the urorectal septum and lateral folding to create separate urinary and rectal segments. An interruption of this development leads to incomplete migration of the rectum to its normal perineal position.³⁰

Etiology and Pathogenesis

Anorectal malformations or imperforate anus encompasses a spectrum of malformations resulting in an anomalous or absent anal opening, as well as a variety

of the type of lesion starts with recognition of the absence of a normal anus and is completed by determining the presence of associated anomalies and the level of the obstruction. Most high lesions end with a fistula from bowel to bladder, urethra, or vagina. Clinical presentations for the two kinds of lesions are presented in Table 24.^{2,12,16,31,32}

Table 24. Clinical Lesion Classification: Anorectal Malformation ^{2,12,16,31,32}	
Low Lesions (Infralevator)	High Lesions (Supralevator)
Males: appearance of meconium at the perineal level Females: occurrence of anocutaneous or anovestibular fistula at the posterior fourchette or vagina	More common in males Meconium in the urine without evidence of any other passage of meconium Absent anal opening/internal anal sphincter Absent perineal fistula Associated genitourinary tract, spinal, cardiac anomalies

Diagnostic Tests

Roentgenographic findings. Lateral x-rays taken with the infant in a head-down position historically have been used for diagnosis and staging of anorectal malformations. However, they are no longer considered diagnostic because they do not definitively identify the level of the lesion with certainty. Roentgenographic evaluation of this defect should include evaluation for those anomalies associated with intermediate and high lesions as well as evaluation of the anorectal lesion itself. This includes specific screening for VACTERAL/VATER-associated anomalies and genitourinary defects. Computed tomography and ultrasonography are used to assist with the diagnosis, but they do not provide a definitive diagnostic evaluation. Magnetic resonance imaging has gained considerable favor as a single test for determining lesions of this type. Roentgenographs and/or ultrasound of the spinal cord should be considered for all infants with imperforate anus to rule out spinal cord involvement. Ultrasound and MRI of the urinary tract system permit visualization of anomalies; the retrograde urethrogram can determine the level of fistula in males, and voiding cystourethrograms can confirm the presence of a suspected fistula.

Treatment/Management

Clinical Management

The treatment of this type of defect is based on the level of the lesion and the presence of associated

anomalies. Presurgical care should focus on intestinal decompression and monitoring or correcting fluid and electrolyte imbalance. Neonates with a urorectal fistula are at risk for hyperchloremic acidosis due to colonic absorption of chloride in urine.

Surgical Management

The surgical treatment of anorectal defects is dictated by type of the defect. Low defects are often corrected by dilation procedures that may or may not be accompanied by minor perineal surgery. Intermediate and high defects may require serial surgeries that include a temporary colostomy and a pull-through procedure. The ultimate goal in all cases is to provide bowel function and continence that are as close to normal as possible.²⁹

The prognosis subsequent to surgery is dependent on the type and level of the lesion and the occurrence of postsurgical complications. Common complications include anastomotic stricture, recurrent rectourinary fistula, rectal mucosal prolapse, constipation, and incontinence. The level of the original lesion often determines the degree of incontinence; patients with low lesions generally achieve continence early in life, whereas those with high lesions rarely do so before late adolescence. Incontinence has psychosocial implications that cannot be ignored during childhood, and the issue needs to be discussed with parents in the newborn period.

Nursing Management

Nursing management includes routine neonatal care, along with the specific issues listed in Table 25.

Table 25. Nursing Issues and Focal Points of Care: Anorectal Malformations

Early detection: <ul style="list-style-type: none"> • Identification of associated anomalies, syndromes, and sequences
Focal points of preoperative care: <ul style="list-style-type: none"> • Prevention of distention, gastric decompression • Electrolyte surveillance (hyperchloremic acidosis) • Prevention of urinary tract infection
Focal points of postoperative care: <ul style="list-style-type: none"> • Wound care/assessment • Surveillance for complication • Colostomy care (higher defects) • Urine inspection for recurrent fistula • Electrolyte balance • Parental education

INTUSSUSCEPTION

Etiology and Pathogenesis

Intussusception is a telescoping, prolapse, or invagination of a portion of the intestine into itself resulting in an acquired intestinal obstruction.

It can occur in the fetus and presents as intestinal atresia in the neonatal period.³³ The etiology of intussusception is uncertain, although some cases (5%-13%) have a predisposing defect or lesion known as a lead point. Lead points are more common in older infants and children and include, but are not limited to: ileal stenosis and atresia in premature infants, Meckel's diverticulum, neoplasm(s), polyps, and hypertrophied ileal Peyer's patches.^{2,20,33} The ileocecal valve is the most common site for an intussusception, occurring in 90% of all cases, with a segment of the ileum prolapsing into the colon. Other types, named after the portions of intestine involved, include ileoileocolic, jejunojejunal, jejunoileal, and colocolic. A history of intercurrent infection is common, implying that the pathogenesis of this disorder is related to altered motility or hyperplasia of lymph nodes that can then serve as a "lead point."

Epidemiology

It is unknown whether there are any embryological influences on the development of intussusception. The condition is rare in the neonatal period. Intussusception is the second most common cause of intestinal obstruction in infants and children. It is more common in males, and two thirds of cases occur in the 1st year of life.

Mortality rates have been reported to be as high as 40% in term newborns and 23% in premature neonates, which is higher than in older infants and children.^{2,33} This difference is probably because it is not uncommon for the diagnosis and treatment to be delayed or overlooked in this age group, which has a direct impact on prognosis. Table 26 contains a list of the epidemiologic characteristics of intussusception.^{2,20,33}

Table 26. Epidemiology of Intussusception^{2,20,33}

Second most common cause of intestinal obstruction
4:1 male to female
2/3 of cases occur in 1st year of life
Rare in neonatal period; incidence unknown
23% mortality rate in premature infants
Mortality rate as high as 40% in term newborns
2%-20% recurrence rate:
• 10% subsequent to an earlier nonoperative reduction
• 1% subsequent to operative reduction
In neonates, majority are ileocecal (terminal ileum)
Prognosis dependent on timing of diagnosis and treatment

Assessment and Diagnosis

Signs and Symptoms

The clinical presentation differs in premature neonates and full-term neonates or infants. In the premature neonate, the typical picture of presentation is similar to that of NEC. Predominant signs include bilious emesis (or nasogastric drainage), marked abdominal distention, but without pneumatosis intestinalis, and passage of rectal blood without evidence of systemic inflammation. The full-term neonate or infant is usually in good health, or perhaps just recovering from a gastrointestinal or respiratory infection. The presenting signs and symptoms are a direct result of mechanical obstruction and compression of mucosal blood vessels. These include severe episodes of colic or abdominal pain with the following characteristics:

- Occurring at 15- to 20-minute intervals
- Uncontrollable crying and drawing up of the legs
- Pallor

Colicky episodes are followed by periods of listlessness, vomiting, and characteristic "currant jelly" stools. However, it should be noted that one third of infants do not pass blood or mucus. Often, a right upper quadrant or midepigastic abdominal mass (sausage-shaped) can be palpated.

Diagnostic Tests

Roentgenographs may assist in the diagnosis. However, findings are not always present. Positive findings include the roentgenographic appearance of small-bowel obstruction, an intraluminal mass, a small amount of feces in the colon, and a paucity of gas, especially in the right lower quadrant. Ultrasound screening is somewhat successful in diagnosing intussusception. Typical findings include a "doughnut" appearance on cross-sectional view, and a "pseudokidney" appearance on the longitudinal section.²¹ The contrast enema under fluoroscopy is diagnostic and can be therapeutic. It is generally recommended that this procedure not be used for reduction in premature infants because of a greater risk of perforation. The types of contrast media used include barium, water-soluble contrast, and air. Most recently, the use of air as a contrast agent has gained renewed popularity.^{20,33,34}

Treatment/Management

Clinical Management

Stabilization is the cornerstone of care if sepsis or shock is present. Fluid loss and blood sequestration in the affected intestine must be managed with fluid and volume resuscitation as indicated. Routine management includes fluid surveillance, bowel decompression, and evaluation for sepsis.

Hydrostatic pressure reduction of an intussusception during contrast enema is attempted in children and adults. This particular therapy is contraindicated in infants younger than 3 months of age because of the risk of perforation, and in other cases where signs and symptoms indicate peritonitis or pneumoperitoneum.

Surgical Management

Surgical reduction is the recommended therapy for the neonatal population, particularly high-risk neonates. Open manual reduction is attempted initially, with resection and anastomosis if unsuccessful. The bowel is inspected, necrotic tissue is removed, and lead points, if identified, are resected. Due to the chance of recurrence, postsurgical surveillance is paramount.

Nursing Management

Nursing management includes routine neonatal care, along with the specific issues listed in Table 27.

Table 27. Nursing Issues and Focal Points of Care: Intussusception

Infection/sepsis
Fluid and electrolytes
Shock/hypovolemia
Recurrence of intussusception
Postsurgical care dependent on resection and loss of bowel

CONGENITAL HYPERTROPHIC PYLORIC STENOSIS

Normal Embryology

The foregut gives rise to the stomach between the 4th and 5th weeks of gestation, initially as a dilatation of the caudal portion. Subsequently, the dilatation enlarges and the stomach rotates and assumes its normal position.³⁰

Etiology and Pathogenesis

Congenital hypertrophic pyloric stenosis (HPS) is a condition in which the circular and longitudinal muscle layers of the distal sphincter of the stomach are hypertrophied. This results in significant narrowing of the pyloric canal and subsequent obstruction.

The etiology of congenital HPS is unknown; however, evidence suggests there is a genetic predisposition that is polygenic in nature and gender-linked.^{2,12,30}

Many cases are thought to be acquired postnatally, with no evidence of hypertrophy at birth. Recent data suggest neonatal exposure to erythromycin to be a significant risk in the development of HPS.³⁵

Epidemiology

The average incidence of HPS is 2 per 1,000 live births (see Table 28).

Table 28. Incidence of HPS^{2,12,36,37}

2 per 1,000 live births
Males at 5 times greater risk than females
Greater occurrence in firstborn infants
20% incidence in firstborn males of mothers with a history of HPS
5% incidence in infants of mothers with a history of HPS
Most common in Caucasians
5% to 15% of affected infants have concurrent hiatal hernia and reflux
Up to 7% have other major malformations

Additional risk factors and associated findings that occur with this lesion are listed in Table 29.

Table 29. Associated Risk Factors With HPS^{2,12,35-38}

Prematurity rare, but incidence is increasing
Higher frequency of intestinal/gut anomalies: <ul style="list-style-type: none"> • Malrotation • Esophageal and duodenal atresia • Short, small intestine • Urinary tract defects • Anorectal anomalies
Blood groups O and B
Maternal stress in last trimester
Prolonged transpyloric jejunal tube feedings in premature infants with reflux
Long-term prostaglandin exposure
Antenatal exposure to doxylamine succinate-pyridoxine hydrochloride
Neonatal exposure to erythromycin

Mortality is extremely low with uncomplicated pyloric stenosis as the result of early diagnosis and prevention of malnutrition, dehydration, and related electrolyte disturbances.

Assessment and Diagnosis

History

An affected infant generally has a relatively normal course during the first 2 weeks of life. Due to the increased familial risk of HPS, family history should receive close attention. A few children present earlier or follow a less typical course.

Signs and Symptoms

Course. Vomiting typically begins around the 2nd or 3rd week of life. The vomiting is nonbilious and frequently projectile, with increasing frequency over the course of approximately 1 week. Left untreated, there is progression to dehydration, constipation, electrolyte imbalance (hypochloremic hypokalemic alkalosis), and failure to thrive. Infants with cardiac disease receiving long-term prostaglandin therapy also may develop gastric outlet obstruction.

The characteristic vomiting of HPS is never bile-stained and often contains milk. Early in the course of HPS, the infant may be irritable, anxious, and hungry. If electrolyte depletion or malnutrition sets in, lethargy and malaise ensue. The feeding behavior of infants with this disease is characterized by ravenous feeding in the first few minutes and then, as the stomach volume increases, anxiety, irritability, and vomiting. Gastric peristalsis before vomiting may or may not be apparent on examination.

Physical assessment. Signs and symptoms of dehydration may be present. This is particularly common in the later course of the disease due to weight loss and emaciation. Behaviors vary, depending on the stage at which the infant is evaluated. Physical findings often include a palpable tumorlike mass below the liver, most frequently felt immediately following a bout of emesis.

Diagnostic Tests

The diagnosis of HPS can be made on the basis of characteristic physical findings of epigastric peristaltic waves and the palpable “olive-like” tumor. Questionable cases are confirmed by roentgenologic and/or ultrasonic studies.

Roentgenographic findings. Plain films demonstrate pyloric holdup, a dilated stomach, and absence of gas distal to the pylorus. This method may be helpful in a differential diagnosis, although it is no longer considered a standard tool in diagnosing HPS. In cases where the clinical and roentgenographic findings are suggestive, ultrasound is usually the initial diagnostic procedure of choice.

Differential diagnosis. The differential diagnosis of nonbilious vomiting in the absence of typical physical findings includes other forms of anatomic gastric outlet obstruction (eg, cystic pyloric duplication and ectopic pancreatic tissue), pylorospasm secondary to sepsis, gastroesophageal reflux (GER), metabolic disorders (eg, adrenogenital syndrome, organic acidemias, hyperammonemia, and galactosemias), and intracranial lesions.

Ultrasound findings. Typical findings include an elongated, narrow pyloric channel (string sign) surrounded by thickened muscle.^{28,34,39}

Treatment/Management

Clinical Management

Clinical management is centered on supportive care before surgery. Priorities include the prevention or treatment of dehydration, metabolic disturbance(s), and hyperbilirubinemia, and the prevention of aspiration.

Surgical Management

A pyloromyotomy extending the length of the hypertrophic muscle along the anterior pyloric wall is the definitive correction for HPS. The procedure is often done laparoscopically. Complications related to this surgery are wound dehiscence secondary to malnutrition, incomplete myotomy, and associated GER.

Nursing Management

Nursing management includes routine neonatal care, along with the specific issues listed in Table 30.

Table 30. Nursing Issues and Focal Points of Care: HPS

Focal points of preoperative care:

- Early recognition of dehydration, fluid and electrolyte disturbances, and malnutrition
- Prevention of distention, vomiting, and aspiration

Focal points of postsurgical care:

- Hydration, fluid, and electrolytes
- Nutrition
- Suture line assessment
- Temporary NG for decompression
- GER surveillance

Necrotizing Enterocolitis

Etiology and Pathogenesis

NEC is an acquired idiopathic disease of the gastrointestinal tract characterized primarily by ischemia and necrosis. Necrosis of the mucosal and submucosal layers can occur anywhere in the bowel, but predominantly in the ileum and proximal colon.

The causes and pathogenesis of NEC are not well understood, but are likely to be multifactorial. These factors include prematurity, GI ischemia, enteral alimentation, and microorganisms.

Prematurity is the most compelling factor implicated in this disease process. Approximately 90% of cases

occur in premature neonates.^{2,40,41} This is thought to be due to several factors, most notably immature GI structure, immature secretory function, limited regenerative capacity, immature host defense mechanisms, and impaired circulatory adaptation.

Once considered the primary insult in NEC, GI ischemia is now thought to have a secondary role. Bacterial exposure, endotoxin release, and release of inflammatory mediators are now considered to play a greater role in the pathogenesis of this disorder.^{41,42}

Enteral alimentation introduces a substrate for the growth of bacteria implicated in the pathogenesis of NEC. Aggressive advancement of early enteral feedings (> 20 kcal/kg per day) has been strongly associated with this disease.⁴¹ On the other hand, low-volume non-nutritive feedings have not been associated with NEC and may reduce morbidities linked with fasting.⁴¹⁻⁴³

Microorganisms may play a significant role in NEC via infectious processes as well as by the release of endotoxin and provocation of an inflammatory response on the part of the infant's immune system. The resultant cytokine, prostaglandin, and leukotriene release contribute to intestinal injury.^{41,42}

Microbial Agents

Infectious agents associated with NEC fall into four general categories: those attributed to sepsis syndromes, toxin-producing bacteria, other miscellaneous bacteria, and enteropathogenic viruses. These agents are listed in Table 31.^{2,41,42,44} It should be emphasized that occasional cases of NEC are not associated with the isolation of any pathogens.

Sepsis Syndromes	Enteropathogenic Toxin-Producing Bacteria	Other Bacteria/Yeast	Enteropathogenic Viruses
Enterovirus <i>Klebsiella</i> <i>Staphylococcus aureus</i> <i>Torulopsis</i>	<i>Clostridium difficile</i> <i>Escherichia coli</i> enterotoxigenic <i>Salmonella</i> <i>Staphylococcus coagulase-negative</i>	<i>C butyricum</i> <i>C perfringens</i> <i>Enterobacter</i> <i>E coli</i> <i>Klebsiella</i> <i>Pseudomonas aeruginosa</i> Yeast	Coronavirus Rotavirus

Epidemiology

The incidence of NEC varies between geographic regions and institutions. It occurs in approximately 10% of neonates weighing < 1500 g at birth.^{2,42}

Gastrointestinal Morbidity/Mortality

NEC is the most common surgical emergency in neonates and has significant short- and long-term morbidities. Short-term morbidity includes intestinal perforation, sepsis, shock, and disseminated intravascular coagulation. Long-term morbidity in this population includes stricture with or without surgery, short-bowel syndrome, enterocyst formation, and malabsorption. Although rare, inflammatory polyps and enteric fistulas can also occur.^{2,40,42}

Mortality rates as a rule are inversely related to birth weight, typically ranging from 9% to 28%. However, some reports of rates are as high as 60%.^{2,40-42} Recurrence of NEC is thought to range from 4% to 6%.²

Assessment and Diagnosis

History

Prenatal, perinatal, and neonatal risk factors all can predispose neonates to NEC. Drug exposure, particularly to cocaine, adds to the risk. Perinatal factors include any condition that might increase the risk of asphyxia, hypoxia, or infection. Neonatal risk factors, in addition to prematurity and exposure to intestinal ischemia, include hematologic abnormalities, indomethacin exposure, and a history of feedings (see Table 32).⁴⁵ No association between NEC and sex, race, geography, or season has been found.² The risk factors previously mentioned should serve as red flags during history taking and should be thoroughly explored.

Table 32. Neonatal Risk Factors and Clinical Identifiers of NEC^{2,40-42,46}

Timing, type, amount, and additional characteristics of feedings
Presence and patterns of apnea
Hypoxic episodes
Cyanosis
Respiratory distress syndrome
Polycythemia
Umbilical venous or arterial catheter
Hyperosmolar enteral medications

Signs and Symptoms

NEC is classified on the basis of clinical presentation described by the modified Bell's criteria. These criteria describe the six stages of NEC, as shown in Table 33.⁴¹

Patterns of Presentation and Classification

NEC can present in either a sudden or an insidious manner (see Table 34). Sudden-onset NEC is a catastrophic event in term or preterm infants, whereas

the insidious presentation evolves over days in stable, growing premature infants. As a rule, age of onset is inversely related to birth weight and gestational age; early onset occurs more often in more mature infants and late onset occurs more often in more preterm infants. The latter may be a result of the delayed introduction of enteral feedings.^{2,44}

Stage IA: Suspected NEC	Stage IB: Suspected NEC
Temperature instability Apnea Bradycardia Lethargy Roentgenographic findings varying from normal to intestinal dilation and mild ileus Increasing residuals Abdominal distention Emesis Guaiac-positive stool	All clinical signs in Stage IA Plus Frank rectal blood
Stage IIA: Proven NEC/Mildly III	Stage IIB: Proven NEC/Moderately III
All clinical signs in Stage IB Plus Absent bowel sounds ± Abdominal tenderness Intestinal dilation Ileus <i>Pneumatosis intestinalis</i>	All clinical signs in Stage IIA Plus Mild metabolic acidosis Mild thrombocytopenia Definite abdominal tenderness ± Abdominal cellulitis ± Right lower quadrant mass Definite ascites on roentgenography
Stage IIIA: Advanced NEC/ Severely III/Intact Bowel	Stage IIIB: Advanced NEC/ Severely III/Perforated Bowel
All clinical signs in Stage IIB Plus Hypotension Bradycardia Severe apnea Combined acidosis (respiratory and metabolic) DIC (disseminated intravascular coagulation) Neutropenia Signs of generalized peritonitis Marked abdominal tenderness and distention	All clinical signs in Stage IIIA Plus Pneumoperitoneum

Insidious (Preterm)	Sudden (Term/Preterm)
Stable growth pattern	Catastrophic event
1- to 2-day evolution	Acute illness
Feeding difficulties	Respiratory decompensation
Change in stool pattern	Shock
Abdominal distention	Acidosis
Guaiac-positive stools	Abdominal distention
	Positive blood cultures

NEC often is classified as either endemic or epidemic. Endemic cases occur in a sporadic fashion, whereas epidemics occur in clusters in a nursery and appear to be associated with an infectious etiology. Epidemics have generally been associated with lower rates of case fatalities, higher birth weights and Apgar scores, and fewer perinatal complications.

Diagnostic Tests

Roentgenographic findings. Roentgenographically, NEC is diagnosed by the presence of pneumatosis intestinalis (gas-filled intramural cysts). Until this pathognomonic sign is present, the x-ray may display signs consistent with developing NEC. Early findings in the development of NEC include fixed and dilated bowel loops with thickened walls (indicating edema) and generalized bowel distention. With progressive NEC, pneumatosis intestinalis develops. Pneumatosis intestinalis is found in approximately 85% of affected infants. Additionally, submucosal or subserosal cystic air collections may be evident as the disease progresses. In the later stages, extensive pneumatosis intestinalis, air in the portal venous system, and free air in the peritoneum are often evident. The presence of portal venous air in an extremely-low-birth-weight infant is associated with full-thickness bowel necrosis, extensive bowel involvement, and, consequently, a high mortality rate.⁴⁴

Treatment/Management

Prevention

Prenatally, prevention of prematurity is the foremost goal. If that fails, antenatal steroids for the prevention of respiratory distress syndrome should be considered. Antenatal steroids provide the neonate with a boost of glucocorticoids, which appear to induce intestinal maturation, increase enzyme activity for digestion and absorption, and decrease mucosal inflammation.⁴⁰⁻⁴²

Prevention of NEC is a multifaceted endeavor determined by the infant's degree of prematurity and the degree of stabilization required to establish gut homeostasis.

Trophic induction. Once the neonate's cardiorespiratory state is stable, the clinician needs to consider the introduction of substrate to the gut. Evidence supports the use of "trophic feeding," a low-volume feeding to prevent villus atrophy and promote GI function. This type of induction is more for its local GI effect than for its nutritional value, as calories are negligible. The substrate used for trophic feedings varies according to individual and institutional/unit practices and may range from sterile water to breast milk or dilute infant formula.

Immunoglobulins. Breast milk is thought to protect the gut in several ways. The protective factors include provision of antibodies, macrophages, lymphocytes, complement components, growth and anti-inflammatory factors, as well as beneficial effects on bacterial colonization.

When the neonate's condition permits the introduction of enteral feedings, three factors should be considered: type of substrate, advancement regimen, and limitation of hyperosmolar preparations. Type and concentration of formula should be carefully determined. However, it should be kept in mind that even breast milk conveys the aforementioned advantages. A cautious approach to advancement is recommended, limiting advancement to less than 20 kcal/kg per day, as aggressive enteral feeding of greater than 20 kcal/kg per day has been associated with NEC.⁴¹ The contribution of drugs and additives to the osmolarity of feedings should always be assessed, and their administration should be adjusted accordingly.

Epidemic precaution. From a preventive perspective, handwashing is the first line of defense. Infection-control measures should be in place. If an outbreak is suspected, consultation with the institution's infection control team will determine the rules and procedures for managing the situation.

Clinical Management

Clinical management depends largely on the severity and stage of the disease and the clinical presentation. Once NEC is suspected, the initial aim of therapy is twofold: to stabilize the compromised neonate and to prevent further injury to the gut, as summarized in Table 35. Laboratory and roentgenologic tests to be considered in the initial phase of diagnosis are listed in Table 36. Once the neonate is stabilized, long-term issues such as nutrition can be addressed. Initiation and advancement of parenteral nutrition should take priority in this phase of recovery.

Table 35. Initial Clinical Intervention for NEC

Prevention of Injury to the Gut	Early Stabilization
Bowel rest/NPO Bowel decompression	Respiratory stabilization Hemodynamic support: <ul style="list-style-type: none"> • Establish fluid and electrolyte balance • Maintain adequate hematocrit Antibiotic therapy

Table 36. Initial Laboratory and Roentgenologic Evaluation

Baseline Laboratory Studies	Baseline Roentgenologic Studies
Stool guaiac Complete/differential blood count, platelets Coagulation profile Blood gases Electrolytes, glucose Bacterial cultures of blood, CSF, and urine if appropriate	Abdominal x-rays: <ul style="list-style-type: none"> • Anterior-posterior and left lateral decubitus • Serial films for comparison

Surgical Management

Once the diagnosis of NEC is established, early surgical consultation is advisable. Perforation with or without pneumoperitoneum is an absolute indication for surgery. Relative indicators are profound and unremitting clinical deterioration, portal venous gas, abdominal wall erythema, fixed abdominal mass, and persistently dilated bowel loops. Surgery generally entails evaluation of bowel integrity and removal of necrotic bowel. Sections of bowel with questionable viability may be left for a "second look" surgery. The extent and findings of surgery often dictate ostomy placement.

Postsurgical complications are not uncommon and include breakdown of the stoma or wound, strictures, altered metabolic status, and short bowel syndrome (SBS) (Table 37). Advancements in management of patients with SBS, including parenteral nutrition and the advent of bowel transplantation, have markedly changed the prognosis for this population. Factors that influence ultimate outcome include percentage of remaining bowel and colon, presence or absence of the ileocecal valve, frequency of episodes of recurrent sepsis, number of surgical interventions, gastrointestinal bacterial overgrowth and associated enterocolitis, success in advancement of enteral feedings, and quality of nutritional care and surveillance.^{2,40-42} Total parenteral nutrition (TPN)-related complications also can

Stoma		Wound		Short Bowel	Metabolic, TPN-Related	Stricture
Retraction	7%	Infection	8%	23% of hospital survivors	27%	31%
Prolapse	5%	Dehiscence	5%			
Hernia	2%	Fistula	2%			
Total	14%	Total	15%			

introduce significant morbidity and mortality when TPN is received for extensive lengths of time. These complications include sepsis, cholestasis, cholelithiasis, cirrhosis, liver failure, osteopenia, and central line embolization.

Nursing Management

Nursing management includes routine neonatal care, along with the specific issues listed in Table 38.

Prevention of NEC
Early recognition
Prevention of further gut injury: <ul style="list-style-type: none"> • Bowel rest • Decompression
Stabilization
Nutritional support
Postsurgical surveillance and intervention for complications

Inguinal Hernia

Normal Embryology

The time frame for development of the inguinal canal extends from 7 weeks' gestation to the descent of the testes and ovaries at 28 weeks. Development of the urinary and genital systems are closely related and parallel, with both systems derived from intermediate mesoderm, mesothelium of the peritoneal cavity, and the endoderm of the urogenital sinus. At approximately 28 weeks the testes descend from the posterior abdominal wall to the deep inguinal rings. The ovaries descend from the posterior abdominal wall to just inferior to the pelvic brim.³⁰

Etiology and Pathogenesis

Inguinal hernia is the most common defect requiring surgery in children and one of the most common in the neonatal population. Recognition and intervention for this defect is important to prevent the development of incarceration and strangulation of the hernia. Incarceration occurs when herniated bowel and/or gonads are trapped in the inguinal canal, scrotum, or labium majus. When this happens,

strangulation, impingement, or compression may lead to a compromise of blood flow to the bowel, testicle, or ovary.

Inguinal hernias are due to the failure of closure of the communication between the tunica vaginalis and the peritoneal cavity, resulting in the patency of the canals and, depending on the diameter of the defect, subsequent movement of abdominal contents through the defect. Bowel, omentum, and testis are generally the hernia contents found in boys. Bowel, omentum, fallopian tube, and ovary can be found in girls. Inguinal hernia can be classified as either direct or indirect. The indirect type is much more common in infants and children, with abdominal contents following the processus vaginalis pathway through the internal ring lateral to the inferior epigastric vessels. In contrast, the direct inguinal hernia is extremely rare in infancy and early childhood. It allows abdominal contents to enter the canal through its posterior wall, medial to the inferior epigastric vessels. With complete hernias, contents extend all the way to the tunica vaginalis, testis, and scrotum or soft tissue of the labium majus. Incomplete hernias can partially extend down the pathway previously described (tunica vaginalis, spermatic cord).

Epidemiology

The incidence of inguinal hernia varies depending on gestational age, ranging in frequency of no more than 5% in full-term infants and 30% in premature neonates. The risk of inguinal hernia in males is six times greater than in females; the frequency in males is 1 in 50 live births. Involvement is right-sided in 60% compared to 25% on the left; 8% to 10% are bilateral. This defect demonstrates a familial tendency and may be associated with genitourinary (GU) defects (see Table 39).^{4,47,48}

Assessment and Diagnosis

History

Family history and physical appearance of the groin area are key in the history and physical examination. Intermittent swelling of the groin area, particularly if exacerbated by crying or straining, should be noted. Strangulation of hernia contents occurs when circulation becomes compromised.

Table 39. Inguinal Hernia Epidemiology^{4,47,48}

6:1 male/female ratio
1 in 50 live male births
Full-term 3.5% to 5%
Higher frequency in:
<ul style="list-style-type: none"> • Premature infants (9% to 30%) <ul style="list-style-type: none"> - Bilateral involvement (up to 64%) - Connective-tissue disorders - Incarceration (31%) - Postsurgical respiratory complications • Bladder extrophy • Ascites • Ventriculoperitoneal shunts • Peritoneal dialysis • Cryptorchidism
Familial tendency
60% right side/25% left side/10% bilateral

Signs and Symptoms

Inguinal hernia typically presents as a groin mass that may or may not be reducible and may occur unilaterally or bilaterally. A simple inguinal hernia usually does not have accompanying pain or discomfort. In contrast, an incarcerated hernia is painful and will also present with the additional findings of obstruction and inflammation listed in Table 40. These signs and symptoms are directly related to the compression and compromise of testicular, ovarian, or intestinal perfusion. This condition represents a surgical emergency because it can rapidly lead to infarction of the involved testis/ovary and visceral contents. In females, ovarian involvement is more difficult to ascertain, although ovaries can sometimes be palpated in the canal and/or groin.

Table 40. Signs and Symptoms of Inguinal Hernia

Simple Hernia	Incarcerated Hernia
Mass in groin area: <ul style="list-style-type: none"> • ± Reducibility • Rare discomfort 	Vomiting Irritability Abdominal pain Loss of appetite Tender inguinal mass With or without abdominal distention Hernia, thick and nonreducible Ipsilateral edema of testis and spermatic cord

It is not unusual for a hydrocele to accompany an inguinal hernia, and in some instances these two entities are difficult to differentiate. Hydroceles do not appear and disappear, do not have significant associated swelling or differentiated mass, and, as a rule, cannot be easily reduced and may transilluminate positive for fluid. In addition to hydrocele, the differential diagnosis includes femoral hernia and an undescended testis in the inguinal canal. It should be noted that in a premature infant, depending on gestational age, it is not unusual to find an undescended testis in various stages of development.

Diagnostic Tests

Physical examination confirms the diagnosis of simple inguinal hernia in the majority of cases by palpation of the herniated contents or fluid with gentle reduction or manual palpation. Exploration of the contralateral inguinal area of the groin when correcting a unilateral hernia remains controversial. This is because the risk of contralateral development of hernia as a result of surgical exploration is approximately 10%, in addition to a risk for injury and testicular atrophy. Laparoscopic evaluation of contralateral incarcerated hernia also has been utilized by some surgeons.⁴⁹

Treatment/Management

Clinical Management

Manual reduction should be done whenever possible. If incarcerated, management prior to surgery is determined by the infant's presenting symptoms and age, ranging from routine care and comfort measures to treatment for shock in the case of advanced incarceration (see Table 41). If vomiting is present, fluid and electrolyte surveillance and intervention may be required. Pain management must be addressed on a case-by-case basis.

Table 41. Nursing Issues and Focal Points of Care: Inguinal Hernia

Focal points of preoperative care: <ul style="list-style-type: none"> • Patient comfort measures • Fluid and electrolyte assessment if vomiting present • Pain assessment • Observation for intestinal obstruction or incarceration Focal points of postoperative care: <ul style="list-style-type: none"> • Suture line care/assessment • NG decompression if necessary • Positioning for suture line support: on side or supine with head turned

Surgical Management

In healthy neonates and infants, outpatient surgery for simple hernia is common. In cases of strangulation, emergency repair is necessary. Surgery may be delayed for infants for whom anesthesia poses a risk and the hernia poses little risk of incarceration. Categories in which anesthetic risk is increased include extreme prematurity, cardiac disease, and respiratory disease. It is not uncommon for surgical correction to be performed before discharge from the newborn intensive care unit in neonates who have recovered from critical illness. Prognosis is excellent, even in cases of incarcerated hernia, when the diagnosis of incarceration is made early. The previous practice in North America of delaying surgery until the infant is older than 10 weeks of age, weighs more than 10 lb, and has a hemoglobin greater than 10 g is now rare, according to a survey conducted by the American Academy of Pediatrics.⁴⁹ In addition, surgical intervention in girls has been found to be inconsistent, relative to the unknown risk of ovarian ischemia in cases of irreducible ovaries. Less than 50% of pediatric surgeons treated ovarian torsion and/or incarcerated ovaries as a surgical emergency.⁴⁹

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Neonatal Alimentary System



Malrotation With Ladd's Bands and Volvulus



Esophageal Atresia With Tracheoesophageal Fistula



Hypertrophic Pyloric Stenosis



Intussusception



Hirschsprung's Disease/ Neonatal Aganglionosis



Strangulated Inguinal Hernia



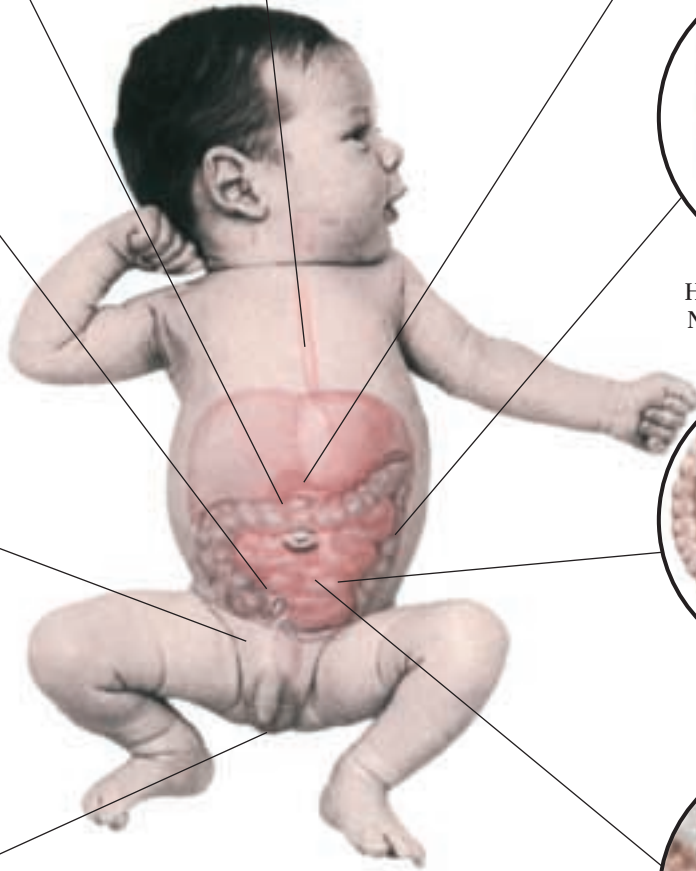
Meconium Ileus



Imperforate Anus



Intestinal Atresia



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Similac® With Iron
Infant Formula