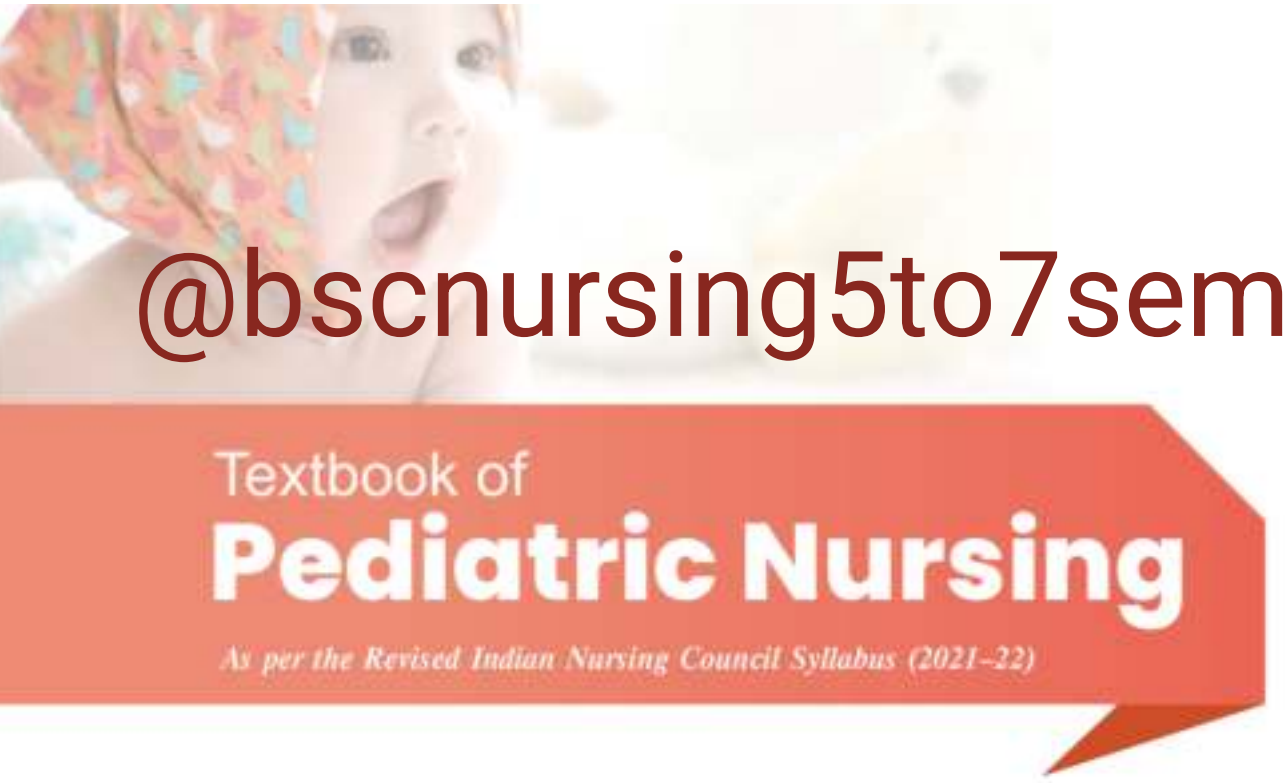

Textbook of Pediatric Nursing

As per the Revised Indian Nursing Council Syllabus (2021–22)

Chapter 17

Gastrointestinal Disorders

Chapter Outline

- Introduction
- Cleft Lip and Palate
- Gastroesophageal Reflux Disease
- Hypertrophic Pyloric Stenosis
- Choledochal Cyst
- Extrahepatic Biliary Atresia (EHBA)
- Intestinal Disorders
- Abdominal Wall Defects
- Anorectal Malformation
- Hepatitis
- Lactose Intolerance
- Malabsorption Syndrome
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- Appendicitis
- Worm Infestation

INTRODUCTION

The common surgical gastrointestinal (GI) problems found in children include cleft lip and palate, pyloric stenosis, megacolon, gastroesophageal reflux disease (GERD), omphalocele, etc. and the medical conditions include diarrhea, hepatitis, and malnutrition. The common symptoms visible in GI disorders include vomiting (bilious/non bilious), constipation, diarrhea, abdominal distension, different colored stools (pale, red, currant jelly) etc. The usual tests done to identify GI problems include X-rays, Barium studies, Biopsy, Endoscopy, etc. the goal of management is to maintain appropriate nutrition in children and maintain fluid and electrolyte balance.

Let us discuss few important disorders in detail.

CLEFT LIP AND PALATE

Definition

Cleft lip and cleft palate are facial and oral malformations that occur very early in pregnancy, while the baby is developing inside its mother. Clefting results when there is not enough tissue in the mouth or lip area, and the tissue that is available does not join together properly.

A cleft lip is a physical split or separation of the two sides of the upper lip and appears as a narrow opening or gap in the skin of the upper lip. This separation often extends beyond the base of the nose and includes the bones of the upper jaw and/or upper gum.

A cleft palate is a split or opening in the roof of the mouth. A cleft palate can involve the hard palate, and/or the soft palate.

Cleft lip and cleft palate (Fig. 17.1) can occur on one or both sides of the mouth. Because the lip and the palate develop separately, it is possible to have a cleft lip without a cleft palate, a cleft palate without a cleft lip, or both a cleft lip and cleft palate together.

Etiology

Cleft lip and palate can be due to heredity, drugs, viruses, prenatal exposure to teratogens, environmental factors or other toxins. It may occur along with other syndromes or birth defects.

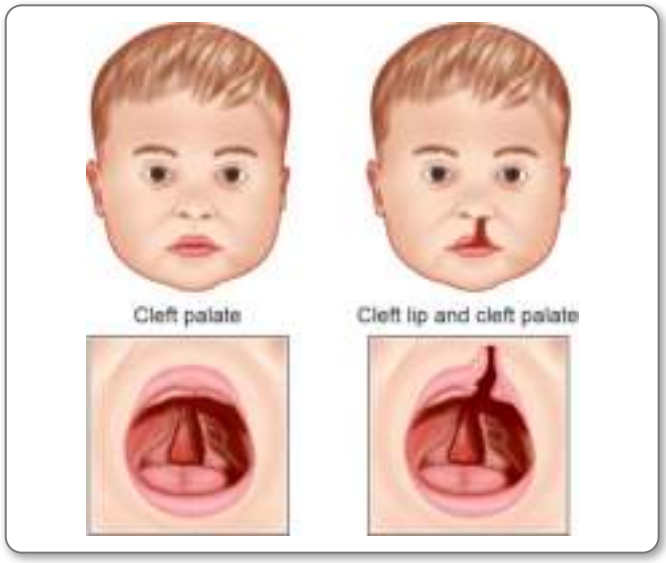

Figure 17.1: Cleft palate and cleft lip

Figure 17.2: Logan bar

Pathophysiology

Defects may be partial or complete, unilateral or bilateral, may involve just lip, palate or both. Defects originate in second month of pregnancy when the front and sides of the face and the palatine shelves fuse imperfectly.

Clinical Manifestations

A cleft lip may be just a small notch in the lip. It may also be a complete split in the lip that goes all the way to the base of the nose. A cleft palate can be on one or both sides of the roof of the mouth. It may go the full length of the palate. Other symptoms include misaligned teeth and change in nose shape (amount of distortion varies).

Diagnosis

Prenatal ultrasound can sometimes determine if a cleft exists in an unborn child. If the clefting has not been detected in an ultrasound prior to the baby's birth, a physical examination of the mouth, nose, and palate confirms the presence of cleft lip or cleft palate after a child's birth.

Management

Surgery is done for cleft lip (CL) and cleft plate (CP):

Cleft lip: It requires one or two surgeries depending on the extent of the repair needed. The initial surgery is usually performed by the time a baby is 3 months old.

Cheiloplasty: It is the correction of cleft lip, done between birth and 3 months of age in anticipation of tooth eruption. Steri strips or Logan bar (Fig. 17.2) are applied postoperatively to relieve tension off suture line.

Cleft palate: Repair of a cleft palate often requires multiple surgeries over the course of 18 years. The first surgery to repair the palate, i.e., staphylorrhaphy is done when the baby is between 6–12 months old, before speech development. The initial surgery creates a functional palate, reduces the chances that fluid will develop in the middle ears, and aids in the proper development of the teeth and facial bones.

Children with a cleft palate may also need a bone graft when they are about 8 years old to fill in the upper gum line so that it can support permanent teeth and stabilize the upper jaw. About 20% of children with a cleft palate require further surgeries to help improve their speech.

Once the permanent teeth grow in, braces are often needed to straighten the teeth.

Additional surgeries may be performed to improve the appearance of the lip and nose, close openings between the mouth and nose, help breathing, and stabilize and realign the jaw. Final repairs of the scars left by the initial surgery are performed during adolescence, when the facial structure is more fully developed.

Nursing Management

Preoperative

- Assess the ability to suck or swallow
- Feed in upright position, burp frequently
- Position on side after feeding
- Use large holed nipple or rubber tipped syringe for CL
- Administer gavage feeding
- Make sure infant can use cup before palate repair
- Feed with cleft palate nipple or Teflon implant
- Use ESSR technique, i.e., enlarged nipple, stimulate suck by rubbing nipple on lower lip and rest after each swallow to allow for complete swallowing
- Warn parents against pathogens

Postoperative

- Monitor for respiratory distress
- Maintain patent airway
- Cleft lip: Do not put the child in prone position
- Cleft palate: Place on abdomen or side
- No oral temperatures
- No straws, pacifiers, spoons or fingers
- Prevent suture line from trauma
- Use of elbow restraints. Prevent crying
- Start on clear liquids with cup, then soft diet
- No tooth brushing
- Rinse suture line with water after each feed
- Provide soft toys

Prognosis

Although treatment may continue for several years and require several surgeries, most children with a cleft lip and palate can achieve normal appearance, speech, and eating. However, some people may have continued speech problems.

Complications

- Speech defects
- Dental and orthodontic problems, dental cavities, displaced teeth
- Nasal defects
- Alterations in hearing, recurrent ear infections like Otitis media
- Increased risk of aspiration, upper respiratory infections
- Lip deformities, nasal deformities
- Shock, guilt and grief for the parents that may interfere with parent child bonding

GASTROESOPHAGEAL REFLUX DISEASE

Definition

Gastroesophageal Reflux Disease (GERD) is a digestive disorder that affects the lower esophageal sphincter (LES), leading to abnormal reflux of gastric contents into the esophagus (Fig. 17.3).

Etiology

GERD is commonly due to transient or permanent changes in the barrier between the esophagus and the stomach. This can be due to incompetence of the LES, transient LES relaxation, impaired expulsion of gastric reflux from the esophagus, or association with a hiatal hernia. The usual cause is babies' immature digestive system. Most infants stop having acid reflux by the time they reach their first birthday. Some children do not outgrow acid reflux, however, and continue to have it into their adolescence.

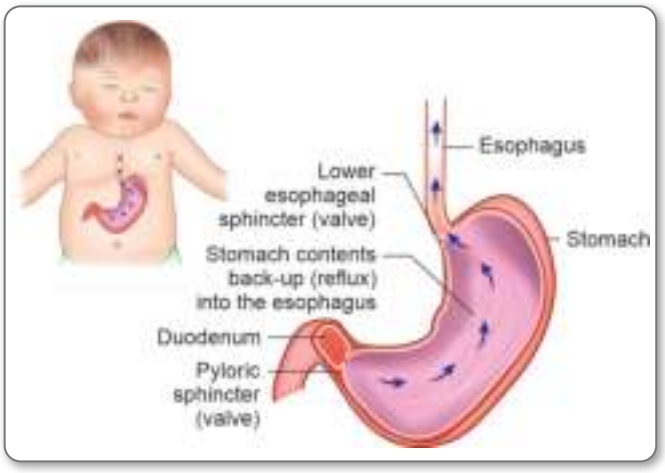

Figure 17.3: Gastroesophageal reflux disease

Risk Factors

- Hiatus hernia increases the likelihood of GERD due to mechanical and motility factors.
- Zollinger-Ellison syndrome, which can be present with increased gastric acidity due to gastrin production.
- Hypercalcemia, which can increase gastrin production, leading to increased acidity.
- Scleroderma and systemic sclerosis, which can feature esophageal dysmotility.

Pathophysiology

In normal digestion, the LES opens to allow food to pass into the stomach and closes to prevent food and acidic stomach juices from flowing back into the esophagus. Gastroesophageal reflux occurs when the LES is weak or relaxes inappropriately, allowing the stomach's contents to flow up into the esophagus. Obesity, tight-fitting clothes; and increased acidity or production of gastric acid can contribute to the problem.

Clinical Manifestations

GERD may be difficult to detect in infants and children. GERD in children may cause repeated vomiting, effortless spitting up, coughing, and other respiratory problems. Inconsolable crying, failure to gain adequate weight, refusing food and bad breath are also common. Children may have one symptom or many symptoms. No single symptom is universally present in all children with GERD.

Diagnosis

A detailed history taking is vital to the diagnosis.

Endoscopy: Image of peptic stricture or narrowing of the esophagus near the junction with the stomach can be seen. This is a complication of chronic gastroesophageal reflux disease, and can be a cause of dysphagia, or difficulty swallowing.

Other investigations which can be done are Barium swallow X-rays, esophageal manometry, 24-hour esophageal pH

monitoring and Esophagogastroduodenoscopy (EGD). EGD involves the insertion of a thin scope through the mouth and throat into the esophagus and stomach to assess the internal surface of the esophagus, stomach and duodenum. Biopsies can be performed during gastroscopy and these may show:

- Edema and basal hyperplasia (non-specific inflammatory changes)
- Inflammation
- Goblet cell intestinal metaplasia or Barrett's esophagus.
- Elongation of the papillae
- Thinning of the squamous cell layer
- Dysplasia or pre-cancer.
- Carcinoma.

Treatment

It involves lifestyle modifications i.e. maintaining appropriate weight and elevating the head of the bed. Foods like coffee, alcohol, and excessive amounts of Vitamin C supplements are stimulants of gastric acid secretion. Taking these before bedtime especially can promote evening reflux.

Some tips to prevent GERD are:

- Take small frequent meals.
- Avoid eating for 2 hours before bedtime
- Avoid smoking, soft drinks, chocolate, peppermint and spicy foods
- Avoid acidic foods like oranges and tomatoes
- Avoid cruciferous vegetables: onions, cabbage, cauliflower, broccoli, Brussel sprouts
- Milk and milk-based products contain calcium and fat, so should be avoided before bedtime.
- Avoid lying down after a meal.

Positional therapy: It refers to elevation of the head of the bed. Food avoidance before bedtime and elevation of the head of the bed helps in relief from GERD. Elevating the head of the bed can be accomplished by using blocks or with other items: plastic or wooden bed risers which support bed posts or legs, a bed wedge pillow, or an inflatable mattress lifter that fits in between mattress and box spring. The height of the elevation is critical and must be at a minimum of 6–8 inches (15–20 cm) in order to be at least minimally effective in hindering the backflow of gastric fluids.

Medical Management

- Antacids, e.g., aluminum hydroxide, magnesium hydroxide. Antacids before meals can reduce gastric acidity (increase the pH). Alginic acid may coat the mucosa as well as increase the pH and decrease reflux.
- Histamine H_2 Antagonists, such as ranitidine, cimetidine and famotidine can reduce gastric secretion of acid.
- Proton pump inhibitors such as omeprazole are the most effective in reducing gastric acid secretion, as they stop the secretion of acid at the source of acid production, i.e., the proton pump. To maximize effectiveness of this medication, the drug should be taken half an hour before meals.
- Prokinetics (Cisapride, domperidone) strengthen the LES and speed up gastric emptying.

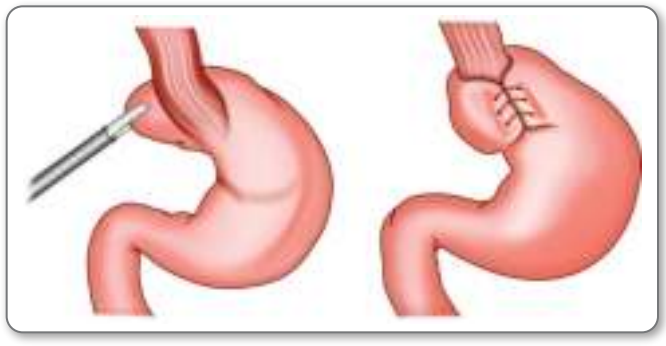

Figure 17.4: Nissen fundoplication

Surgical Management

The standard surgical treatment, sometimes preferred over long time use of medication, is the *Nissen fundoplication* (Fig. 17.4). The upper part of the stomach is wrapped around the LES to strengthen the sphincter and prevent acid reflux and to repair a hiatal hernia. The procedure is often done laparoscopically.

Complications

Barrett's esophagus: It is a type of dysplasia, which can lead to carcinoma. Due to the risk of chronic heartburn progressing to Barrett's, EGD is recommended every 5 years. GERD has been linked to laryngitis, chronic cough, pulmonary fibrosis, earache, and asthma, as well as to laryngopharyngeal reflux and ulcers of the vocal cords.

HYPERTROPHIC PYLORIC STENOSIS

Definition

Pyloric stenosis (Fig. 17.5) also known as infantile hypertrophic pyloric stenosis (IHPS), is a narrowing of the pylorus, the opening from the stomach into the small intestine. The pylorus of the stomach is a small, narrow muscular sphincter through which food passes into the duodenum after it has been partially digested in the stomach. This narrowing is believed to be caused by a combination of muscular hypertrophy, spasm and edema of the mucous membrane.

Etiology

No definitive cause for hypertrophic pyloric stenosis has been found. However, various environmental and hereditary factors have been implicated. Suspected environmental factors include infantile hypergastrinemia, abnormalities in the myenteric plexus innervation, cow's milk protein allergy, and

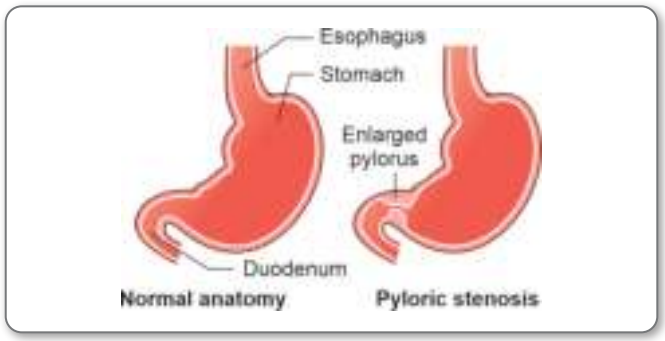

Figure 17.5: Pyloric Stenosis

exposure to macrolide antibiotics. Hereditary factors may also play a role; hypertrophic pyloric stenosis occurs in as many as 7% of infants of affected parents.

Pathophysiology

Normally, a muscular valve (pylorus) between the stomach and small intestine holds food in the stomach until it is ready for the next stage in the digestive process. In pyloric stenosis, there is diffuse hypertrophy and hyperplasia of the smooth muscle of antrum of the stomach and pylorus blocking food from reaching the small intestine.

The antral region is elongated and thickened to as much as twice its normal size. In response to outflow obstruction and vigorous peristalsis, stomach musculature becomes uniformly hypertrophied and dilated. Pyloric stenosis can lead to forceful vomiting, dehydration and weight loss. Babies with pyloric stenosis may seem to be hungry all the time.

Gastritis may occur after prolonged stasis. Hematemesis is occasionally noted. The patient may become dehydrated as a result of vomiting and develop marked hypochloremic alkalosis.

Clinical Manifestations

Infant will present with **nonbilious projectile vomiting** at 4–8 weeks. An enlarged pylorus, classically described as an "olive," can be palpated in the right upper quadrant or epigastrium of the abdomen. Peristaltic waves can be frequently noted passing from left to right during or immediately following a feeding. The baby in the early stage of the disease remains hungry and sucks vigorously after episodes of vomiting. Prolonged delay in diagnosis can lead to dehydration, poor weight gain, malnutrition, metabolic alterations, and lethargy. Thus the typical features include projectile vomiting, visible peristalsis, and a palpable pyloric tumor.

Diagnosis

- Infants with severe vomiting can develop profound hypochloremia and hypokalemia. The classic biochemical abnormality in hypertrophic pyloric stenosis is hypochloremic, hypokalemic metabolic alkalosis.
- **Ultrasonography:** Pyloric muscle thickness and pyloric channel length is measured. Muscle wall thickness 3 mm or greater and pyloric channel length 14 mm or greater are considered abnormal in infants younger than 30 days.
- **Barium upper GI (UGI) study:** It demonstrates an elongated pylorus with antral indentation from the hypertrophied muscle. The UGI study may demonstrate the "double track" sign when thin tracks of barium are compressed between thickened pyloric mucosa or the "shoulder" sign when barium collects in the dilated prepyloric antrum. After UGI barium study, irrigating and removing any residual barium from the stomach is advisable to avoid aspiration.

Management

Surgical Management

Surgery: Ramstedt pyloromyotomy is the standard procedure of choice for hypertrophic pyloric stenosis because it is easily performed and is associated with minimal complications. The usual approach is via a right upper quadrant transverse incision that splits the rectus muscle and fascia.

Laparoscopic pyloromyotomy has a significantly shorter recovery time compared with open pyloromyotomy. However, open pyloromyotomy has a higher efficacy and fewer complications. Endoscopic pyloromyotomy is a simple procedure and can be performed as an outpatient procedure.

Nursing Management

Preoperative Care

Repeated episodes of vomiting can cause progressive dehydration and loss of hydrogen chloride from the gastric juices. Preoperative management is directed at correcting the fluid deficiency and electrolyte imbalance.

- Administer an initial fluid bolus of 10 mL/kg with lactated Ringer solution or 0.45 isotonic sodium chloride solution. Continue intravenous (IV) therapy at an initial rate of 1.25–2 times the normal maintenance rate until adequate fluid status is achieved.
- Adequate amounts of both chloride and potassium are necessary to correct metabolic acidosis. Unless renal insufficiency is a concern, initially add 2–4 mEq of KCL per 100 mL of IV fluid. Adequate chloride for resuscitation can usually be provided by 5% dextrose in 0.4% sodium chloride solution.
- Monitor urine output and electrolyte values. (Serum chloride level of 90 mEq/L or greater is usually adequate to proceed with surgical intervention).
- Insert NG tube for gastric decompression. Ensure tube is in stomach.
- Before induction of anesthesia, aspirate the infant's stomach with a large-caliber suction tube to remove any residual gastric fluid or barium. Saline irrigation is occasionally necessary to remove a large quantity of barium.

Postoperative Care

- Maintain patent airway.
- Observe for shock.
- Continue IV maintenance fluid until the infant is able to tolerate enteral feedings. Feedings can begin within 8 hours following surgery. Gradually increasing the volume and strength of feedings is recommended.
- Although schedules that advance the volume of feeds more quickly or those that begin with ad lib feeds are associated with more frequent episodes of vomiting, they do not increase morbidity and actually may decrease the time to hospital discharge.
- Administer histamine receptor blockers, e.g., ranitidine.
- Administer analgesics, e.g., acetaminophen.
- Infants can be discharged from hospital care once they can remain hydrated and have adequate enteral intake.
- Monitor for complications like:
  - **Undetected mucosal perforation:** The infant develops fever, tenderness in the abdomen, and abdominal distention.
  - **Bleeding:** Observe for venous oozing from the myotomy site.
  - Persistent vomiting: Incomplete pyloromyotomy can lead to persistent vomiting until after the second week postsurgery.

CHOLEDOCHAL CYST

Definition

Choledochal cyst is a congenital cystic dilatation of all or some portion of intrahepatic and extrahepatic biliary tree (Fig. 17.6). Different types of choledochal cysts can be found depending on the location and type of enlargement.

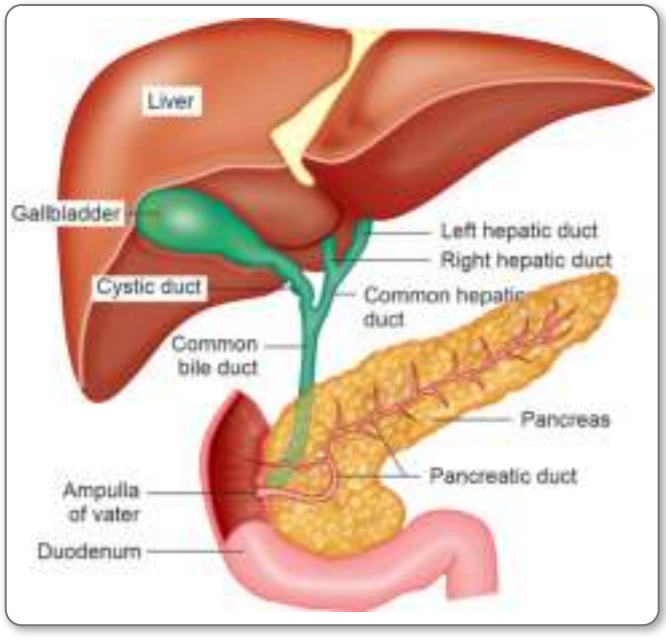

Figure 17.6: Biliary system

Pathophysiology

Normally the bile produced by liver is drained by right and left hepatic ducts into common hepatic duct. Common hepatic duct and cystic duct from gall bladder joins to form common bile duct. Choledochal cyst causes decrease in drainage of bile from the abnormal bile duct. The majority of choledochal cysts are present in childhood. The patient presents with jaundice. The poor drainage causes infections in the bile duct in many patients. Some patients develop repeated attacks of pancreatitis since the pancreatic duct may enter into the abnormal bile duct. The cyst wall consists of fibrous tissue without muscle. It may contain up to 2 liters of fluid. It is often associated with distal common bile duct stenosis which leads to cholestasis and cholangitis. If not diagnosed, they can progress to biliary fibrosis, cirrhosis, portal hypertension and liver failure.

Majority of patients with choledochal cysts have an anomalous junction of the common bile duct with the pancreatic duct (anomalous pancreatobiliary junction [APBJ]). An APBJ is characterized when the pancreatic duct enters the common bile duct 1 cm or more proximal to where the common bile duct reaches the ampulla of Vater.

The APBJ allows pancreatic secretions and enzymes to reflux into the common bile duct. In the relatively alkaline conditions found in the common bile duct, pancreatic proenzymes can become activated. This results in inflammation and weakening of the bile duct wall. Severe damage may result in complete denuding of the common bile duct mucosa.

Anomalies associated with cysts:

- Anomalous junction of the pancreatic and common bile duct
- Distal bile duct stenosis
- Intrahepatic ductal dilation
- Abnormal histologic findings of CBD
- Hepatic histological findings from normal to cirrhotic

Types of Choledochal Cyst

According to Todani and coworkers, five major classes of choledochal cysts exist, (i.e., types I-V), with subclassifications for types I and IV, (i.e., types IA, IB, IC; types IVA, IVB) (Fig. 17.7).

- Type I cysts are the most common and represent 80–90% of choledochal cysts. They consist of saccular or fusiform dilatations of the common bile duct, which involve either a segment of the duct or the entire duct. Type IA is saccular in configuration and involves either the entire extrahepatic bile duct or the majority of it. Type IB is saccular and involves a limited segment of the bile duct. Type IC is more fusiform in configuration and involves most or all of the extrahepatic bile duct.

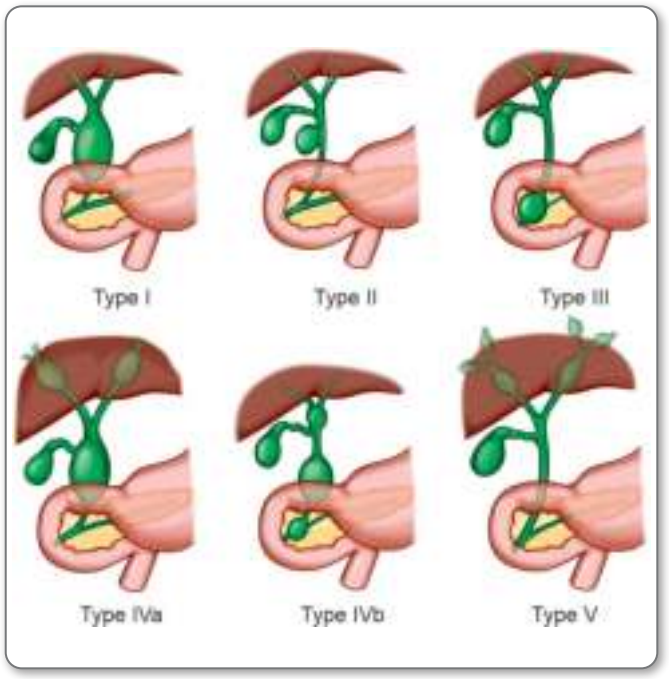

Figure 17.7: Types of choledochal cysts

- Type II choledochal cysts appear as an isolated diverticulum protruding from the wall of the common bile duct. The cyst may be joined to the common bile duct by a narrow stalk.
- Type III choledochal cysts arise from the intraduodenal portion of the common bile duct and are also known as choledochocele.
- Type IVA cysts consist of multiple dilatations of the intrahepatic and extrahepatic bile ducts. Type IVB choledochal cysts are multiple dilatations involving only the extrahepatic bile ducts.
- Type V (Caroli disease) consists of multiple dilatations limited to the intrahepatic bile ducts.

Clinical Manifestations

- 25% present as neonates with prolonged jaundice and cholestasis
- In early infancy, there are signs of complete extra hepatic biliary obstruction like jaundice, hepatomegaly, acholic stools
- 75% later in childhood present with triad of abdominal pain, abdominal mass and intermittent jaundice

Diagnosis

- **Lab findings:** Conjugated bilirubinemia, elevated alkaline phosphatase, transaminases, and gamma glutamyl transpeptidase.
- **CAT scan:** To define dimensions of cyst and extent of involvement
- **HIDA scan:** A hepatobiliary iminodiacetic acid (HIDA) scan to evaluate the function of the gallbladder and the bile ducts. It is also referred to as cholescintigraphy. In this procedure, a radioactive tracer is injected in vein. The tracer travels to liver and then into the bile ducts. A special scanner placed over abdomen tracks the movement of the tracer through biliary tract and makes images of the liver, gallbladder and bile ducts. A HIDA scan is used to diagnose obstruction of bile duct due to gall stones, tumors, diseases of GB and bile leaks.
- **MRCP:** Magnetic resonance cholangiopancreatography (MRCP) is a medical imaging technique which uses magnetic resonance imaging to visualize the biliary and pancreatic ducts in a non-invasive manner. This procedure can be used to determine if gallstones are lodged in any of the ducts surrounding the gallbladder. Stimulation with secretin may enable the acquisition of dynamic MRCP images. Compared to ERCP it is less invasive, but direct intervention is not possible.
- **ERCP:** Endoscopic retrograde cholangiopancreatography, or ERCP, is a specialized technique used to study the ducts of the gallbladder, pancreas and liver. During ERCP, an endoscope is passed through mouth, esophagus and stomach into the duodenum. After seeing the common opening to ducts from the liver and pancreas, a catheter is passed through the endoscope and into the ducts. A contrast material (dye) is injected into the pancreatic or biliary ducts and X-rays are taken.
- **Ultrasound:** Abdominal ultrasound usually reveals the cyst.

Complications

- Recurrent cholangitis
- Hepatic fibrosis
- Biliary cirrhosis and portal hypertension
- Rupture with biliary peritonitis
- Pancreatitis
- Hepatic abscess
- Gall stones
- Carcinoma of the biliary tree

Surgical Management

The treatment for choledochal cysts is surgical. The treatment of choice for a type I choledochal cyst is complete excision of the cyst with construction of a **Roux-en-Y biliary-enteric anastomosis** to restore biliary continuity with the gastrointestinal tract.

Type I and II cysts: Cyst should be resected and a **hepaticojejunostomy** performed. It prevents anastomotic stricture and malignancy in the cyst and postoperative cholangitis is uncommon.

Type III cysts: Therapy for type III choledochal cysts, depends on the size of the lesion. Choledochoceles with a diameter of 3 cm or smaller may be approached endoscopically and effectively treated by means of sphincterotomy.

Choledochoceles larger than 3 cm in diameter are often associated with some degree of duodenal obstruction. These cysts are excised surgically by using a transduodenal approach. If the pancreatic duct is found to be entering the choledochocele, it must be reimplanted into the duodenum after the cyst is excised. Cholecystectomy and choledochojejunostomy or choledochoduodenostomy is done.

Type IV choledochal cysts: The dilatated extrahepatic duct is completely excised, and a Roux-en-Y biliary-enteric anastomosis procedure is performed. No therapy is specifically directed at the intrahepatic ductal disease, except if intrahepatic ductal strictures, hepatolithiasis, or hepatic abscesses are present. In these patients, interventional radiologic techniques can be performed. If the disease is limited to specific hepatic segments or a lobe, these may be resected.

Type V choledochal cyst, or caroli disease: It is defined only by the dilatation of the intrahepatic ducts. If dilatation is limited to a single hepatic lobe, usually the left, the affected lobe is resected. Liver transplantation may be considered for patients with bilobar disease and signs of biliary cirrhosis, portal hypertension, or liver failure.

Prognosis

If diagnosed early, liver fibrosis regresses and normal hepatic function can be expected. Prognosis is poor if advanced disease with portal hypertension.

Nursing Management

Preoperative Management

Knowledge deficit r/t disorder, diagnostic modalities and treatment options

- Assess family's level of functioning
- Provide support to family
- Explain about disorder
- Encourage questioning. Explain to the child based on his developmental age
- Repeat and reinforce the information about diagnosis, prognosis, surgical procedure and perioperative course
- Listen patiently to the concerns of child and caregiver.

On the Day of Surgery

- Keep the child NPO from 12 midnight
- Give a savlon sponge bath in the morning
- Change to hospital garments with identification tags
- Keep the file ready with all investigation reports
- Ensure written consent is obtained
- Administer Vitamin K I/M (5 mg)
- Administer first dose of antibiotics (Inj cefotaxime 400 mg, Inj amikacin 75 mg, Inj metrogyl 125 mg)
- Administer IV fluids antibiotics IV fluids (N/3 in 5% Dextrose + MVI + KCl (1:100) @ 50 mL/hr from 12 MN)
- Administer preop medications
- Shift patient to OT safely

Postoperative Management

Alteration in comfort r/r incisional pain and surgery done

- Assist in activities of daily living
- Administer analgesics as per order (anamol suppository 160 mg TDS)
- Keep NG tube on continuous drainage to prevent gastric distension
- Provide comfortable position

Potential for alteration in fluid and electrolyte imbalance r/t IV infusion and NPO status of the child

- Prepare and administer IV fluids as per order. Add KCl 1:100. Maintain accurate drop rate
- 5Check daily weight
- Maintain strict I/O chart
- Monitor for signs of dehydration or overhydration
- Check blood pressure

Prone to infection r/t break in skin integrity like suction drain, IV line

- Encourage hand washing by caregivers
- Administer antibiotics as per order.
- Keep IV site clean, change IV cannula q 24 hours
- Practice aseptic technique while performing any procedure
- Follow standard precautions
- Monitor subhepatic suction drainage for color and amount
- Monitor temperature and WBC levels

Anxiety r/t unfamiliar surroundings and the surgery done

- Provide sensory and tactile stimulation
- Administer analgesics as per order to relieve pain
- Encourage parents to participate in child's care especially mother
- Explain according to his level of understanding before performing any procedure
- Allow child to keep his favorite toy with him
- Allow sibling to visit during visiting hours

Altered heath maintenance r/t lack of knowledge about diagnosis, surgery done and home care

- Teach mother about hygiene
- Teach how to take care of operative site and perform wound dressings
- Teach about the signs of complications, e.g., jaundice, abnormal color of stools, ascites, bleeding, respiratory distress so that the surgeon can be notified.

Discharge Teaching

- Inform about regular follow up
- Explain the medications to be taken
- Teach to monitor signs of complications like jaundice, pain, acholic stools, etc.

EXTRAHEPATIC BILIARY ATRESIA (EHBA)

Biliary atresia is characterized by obliteration or discontinuity of the extrahepatic biliary system, resulting in obstruction to bile flow. The disorder represents the most common surgically treatable cause of cholestasis encountered during the newborn period. If not surgically corrected, secondary biliary cirrhosis invariably results.

Etiology

Age: Biliary atresia is a disorder unique to the neonatal period.
Sex: Extrahepatic biliary atresia is more common in females than in males.

Pathology

The pathology of the extrahepatic biliary system widely varies in these patients, and the following classification is based on the predominant site of atresia (Fig. 17.8):

- Type I involves obliteration of the common duct; the proximal ducts are patent
- Type II is characterized by atresia of the hepatic duct, with cystic structures found in the porta hepatis
- Type III (>90% of patients) involves atresia of the right and left hepatic ducts to the level of the porta hepatis.

Clinical Presentation

The clinical presentation of neonatal cholestasis is remarkably similar in most infants. Typical symptoms include variable degrees of jaundice, dark urine, and light stools. In most cases, acholic stools are not noted at birth but develop over the first few weeks of life. Appetite, growth, and weight gain may be normal. Hepatomegaly may be present early, and the liver is often firm or hard to palpation. Splenomegaly is common, and an enlarging spleen suggests progressive cirrhosis with portal hypertension.

Diagnosis

- Consider biliary atresia in all neonates with direct hyperbilirubinemia.
- Ultrasonography may demonstrate absence of the gallbladder and no dilatation of the biliary tree.

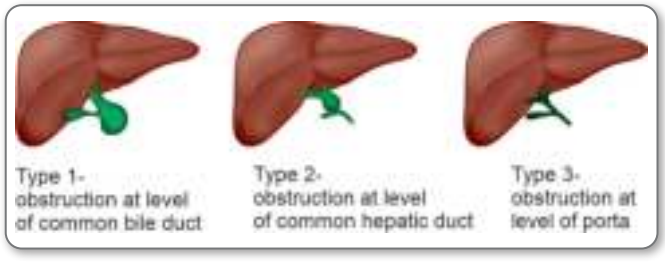

Figure 17.8: Extrahepatic biliary atresia (EHBA)

- **Intraoperative cholangiography:** To demonstrate anatomy and patency of the extrahepatic biliary tract.

Medical Management

In patients with chronic cholestatic conditions and bile duct patency, ursodeoxycholic acid, (i.e., ursodiol, UDCA) has shown to enhance bile flow. To prevent cholangitis postoperatively, prophylaxis with trimethoprim-sulfamethoxazole is used on a long-term basis.

Surgical Management

Once biliary atresia is suspected, surgical intervention is the only mechanism available for a definitive diagnosis (intraoperative cholangiogram) and therapy (Kasai portoenterostomy). During surgery, fibrotic biliary tract remnant is identified, and the patency of the biliary system is assessed.

INTESTINAL DISORDERS

The common surgical intestinal disorders seen are Megacolon, Intususception, Malrotation and Volvulus.

Hirschsprung's Disease (Congenital Aganglionic Megacolon)

Definition

Hirschsprung's disease is an abnormal dilation of the colon caused by absence of parasympathetic nerve ganglion cells in the distal bowel. The dilatation is often accompanied by a paralysis of the peristaltic movements of the bowel. In more extreme cases, the feces consolidate into hard masses inside the colon, called fecalomas (literally, fecal tumor), which can require surgery to be removed.

Etiology

Genetic causes: The disease is generally sporadic, although incidence of familial disease has been increasing. The genes associated with Hirschsprung's disease include the glial cell-derived neurotrophic factor gene, the endothelin-B receptor gene, and the endothelin-3 gene.

Hirschsprung's disease is associated with Down syndrome, congenital deafness, malrotation, gastric diverticulum, and intestinal atresia.

Pathophysiology

In Hirschsprung's disease, there is absence of enteric neurons within the myenteric and submucosal plexus of the rectum and/or colon. Enteric neurons are derived from the neural crest and migrate caudally with the vagal nerve fibers along the intestine. These ganglion cells arrive in the proximal colon by 8 weeks of gestation and in the rectum by 12 weeks of

gestation. Arrest in migration leads to an aganglionic segment. This results in clinical Hirschsprung disease.

The distal portion of bowel is unable to transmit regular peristaltic waves, which are coordinated with the proximal portion of the bowel. When a stool reaches the diseased area, it is not transmitted down the colon, but accumulates in the segment just proximal to this area, forming a functional obstruction. The bowel above the obstructed portion eventually becomes hypertrophied in its attempts to transmit the stool. The pathophysiology is depicted in Figure 17.9.

Clinical Manifestations

Neonate with megacolon presents as abdominal distention, failure of passage of meconium within the first 48 hours of life, and repeated vomiting which is bile stained. A family history of a similar condition is present in about 30% of cases.

Children may be malnourished. Poor nutrition results from the early satiety, abdominal discomfort, and distention associated with chronic constipation.

Older infants and children typically present with chronic constipation, offensive odor and ribbon like stools. This constipation often is refractory to usual treatment protocols and may require daily enema therapy.

Hirschsprung enterocolitis is a fetal complication of Hirschsprung disease. Enterocolitis typically presents with abdominal pain, fever, foul-smelling bloody diarrhea, and vomiting. If not recognized early, enterocolitis may progress to sepsis, transmural intestinal necrosis, and perforation.

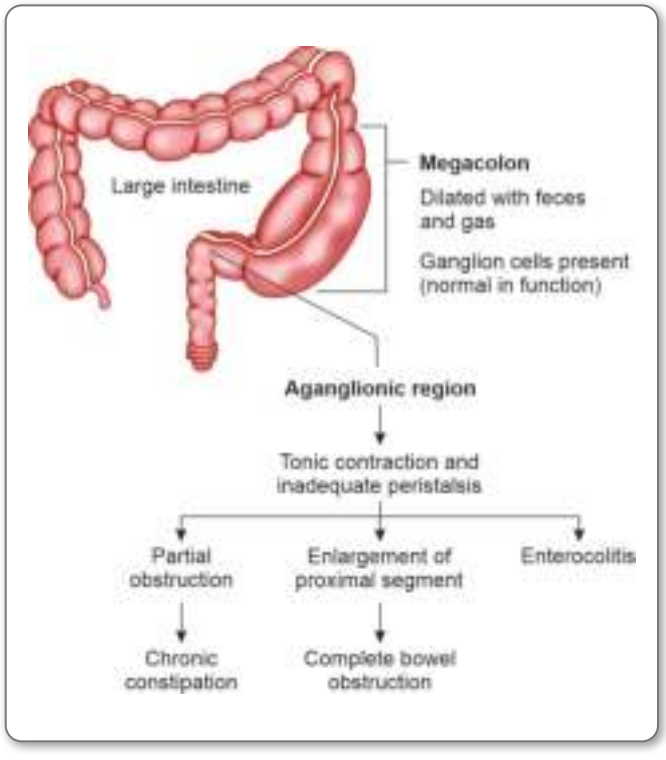

Figure 17.9: Pathophysiology of megacolon

Diagnosis

- **CBC count:** Elevation of WBC count may suggest enterocolitis.
- Plain abdominal radiography to be done if any signs or symptoms of abdominal obstruction are present.
- **Barium enema:** It may help establish the diagnosis by identifying a transition zone between a narrowed aganglionic segment and a dilated and normally innervated segment.
- **Rectal manometry:** In older children who present with chronic constipation and an atypical history for either Hirschsprung disease or functional constipation, anorectal manometry can be helpful in making or excluding the diagnosis. Children with Hirschsprung disease fail to demonstrate reflex relaxation of the internal anal sphincter in response to inflation of a rectal balloon.
- **Rectal biopsy:** The definitive diagnosis of Hirschsprung disease depends on histological review of rectal tissue. Biopsy specimens are examined for the presence or absence of ganglion cells in the submucous plexus or myenteric plexuses.

Preoperative Management

- Assist in diagnosis (rectal biopsy) for confirmation and treatment plan.
- Initial therapy includes intravenous hydration, withholding of enteral intake, and intestinal and gastric decompression.
- Decompression can be accomplished through placement of a nasogastric tube and either digital rectal examination or normal saline rectal irrigations 3–4 times daily.
- Administer broad-spectrum antibiotics to patients with enterocolitis.
- Counsel the family regarding available surgical options. If the child is to undergo a staged procedure or have a permanent ostomy, provide preliminary instructions about ostomy care to the family.

Surgery

The surgical options vary according to the patient's age, mental status, ability to perform activities of daily living, length of the aganglionic segment, degree of colonic dilation, and presence of enterocolitis.

- The first stage of treatment includes a temporary, transverse or sigmoid **colostomy** at the level of normal bowel.
- The bowel caliber is restored to normal, child is brought back to optimum health and nutritional status.
- The final procedure consists of dissection and removal of the nonfunctional bowel and anastomosis.
- Finally the temporary colostomy is closed.
- The single-stage pull-through procedure may be performed with laparoscopic, open, or transanal techniques. This procedure is generally performed after the newborn has had rectal irrigations at home and has passed the physiologic nadir.

- The common surgeries done are:
  - **Swensons:** In this there is combined abdomino-perineal approach. It leaves smallest amount of aganglionic bowel. Careful dissection of rectal wall is done to avoid injury to pelvic nerves.
  - The Swenson procedure was the original pull-through procedure used to treat Hirschsprung disease. The aganglionic segment is resected down to the sigmoid colon and rectum, and an oblique anastomosis is performed between the normal colon and the low rectum.
  - **Duhamel:** In this abdominal approach is followed. Retro-rectal pull through is done to avoid potential nerve injury. Ganglionic bowel is brought posteriorly and anastomosed end to end with aganglionic segment. Neorectum with anterior aganglionic and posterior ganglionic walls is formed.
  - **Soave:** The ganglionic bowel is pulled through the aganglionic bowel. Aganglionic bowel surrounds normal bowel. Avoids injury to pelvic nerves.

Postoperative Management

- Observe child for normal bowel function
- Maintain skin care of colostomy and anal areas
- Follow-up closely to assess healing and complications. Observe for postoperative complications like intermittent fecal soiling and incontinence, anastomotic leak, stricture formation, intestinal obstruction, and enterocolitis. Outpatient dilations may be necessary to alleviate strictures and should be expected in patients who undergo a single-stage pull-through procedure in the newborn period.
- Postoperatively, routine colonic irrigation and prophylactic antibiotic therapy (ampicillin, gentamicin, and metronidazole) is done to decrease the risk of developing enterocolitis. For patients who develop enterocolitis, nasogastric decompression, intravenous fluids, antibiotics, and colonic lavage may be necessary. Sodium cromoglycate, a mast cell stabilizer, has also been reported to benefit these patients.
- **Diet and activity:**
  - The patient should have nothing by mouth for 6–8 hours prior to operation.
  - Postoperatively, the patient will receive intravenous fluids and antibiotics; however, nothing may be administered by mouth until the child passes flatus or stool which signifies return of bowel function.
  - Upon resumption of bowel function, tube feeding or formula/breast milk may be resumed. Clear liquids are delivered by mouth, and the diet may be advanced until the feeding goals are met. Feedings are usually initiated 24–48 hours after the creation of a colostomy. The patient may be discharged from the hospital upon attaining full feedings.

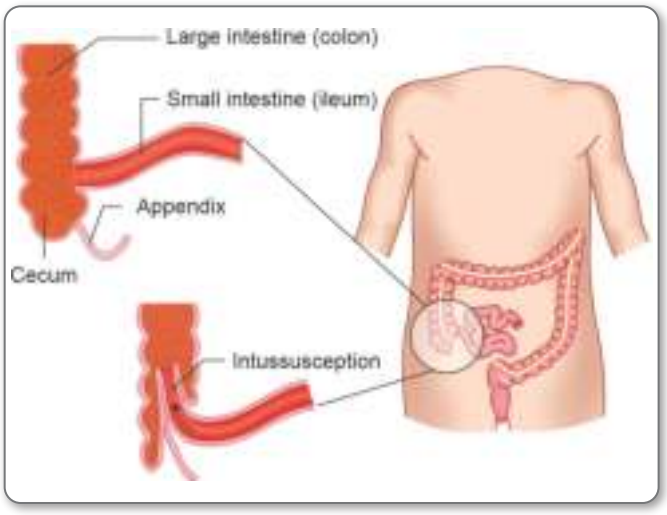

Figure 17.10: Intussusception

  - Diets consisting of fresh fruits, vegetables, and high-fiber may improve postoperative bowel function.
  - With regard to activity, limit physical activity for about 6 weeks to allow the incisions to heal properly especially in older children.

Intussusception

Definition

Intussusception (Fig. 17.10) is telescoping or invagination of a bowel segment into self, common site is ileocecal valve.

Causes

Cause is unknown, it may be associated with viral infections and may also result from polyps, hyperactive peristalsis.

Pathophysiology

- Bowel segment invaginates into itself, ileum invaginates into cecum and colon. Peristalsis propels it along the bowel pulling more bowel with it. There is inflammation and swelling at affected side. Obstruction and necrosis occur due to occlusion of blood supply to bowel.

Complications

- Bowel obstruction, strangulation of intestine, gangrene
- Bowel perforation, peritonitis
- Shock, death

Assessment

- The **onset is usually abrupt**. A previously healthy infant or child suddenly experiences acute abdominal pain with vomiting and passage of brown stool. There may be periods of comfort between acute attacks.
- As condition worsens, painful episodes increase.
- The stools become **red and resemble currant jelly** because of the mix of blood and mucus.

- Palpable **sausage-shaped abdominal mass in upper right quadrant.**
- Distended, tender abdomen

Diagnosis

- Abdominal X-ray
- Increased WBC

Management

- **NG tube:** For decompressing stomach.
- **Hydrostatic reduction:** Air pressure or solution of **barium** or water soluble contrast is introduced to the rectum. Force from fluid or air moves invaginated bowel back into its original position.

Surgery

Indications of surgery are as follows:

- Failure of hydrostatic reduction
- Recurrent intussusception
- Signs of shock or peritonitis
- **Direct manual reduction:** Intussusception is pulled back through bowel
- **Resection of affected bowel** if gangrenous or strangulated

Preoperative Management

- Monitor vitals
- Maintain fluid and electrolyte balance
- Monitor stool pattern

Postoperative Management

- Monitor signs of infection
- Managing the child's pain and maintain N/G tube patency
- Assess vital signs.
- Check for abdominal distension and listen for bowel sounds every 4 hours.
- After normal bowel function returns, clear liquid feedings are begun.
- Feedings are advanced to half strength milk and other foods as the infant or child tolerates it.

Malrotation

During embryonic life, rotation of duodenojejunal loop and rotation of ceco-colic loop has to take place simultaneously for normal intestinal development. Failure to complete normal rotation and fixation of the bowel leads to malrotation (Fig. 17.11). Superior mesenteric artery (SMA) may get strangulated. When the normal 270 degree rotation is interrupted duodenum lies behind SMA. Cecum does not reach right iliac fossa, adhesions form, running from cecum across the duodenum to the right lateral wall of abdomen - Ladd's Bands. Intestinal obstruction due to malrotation is volvulus.

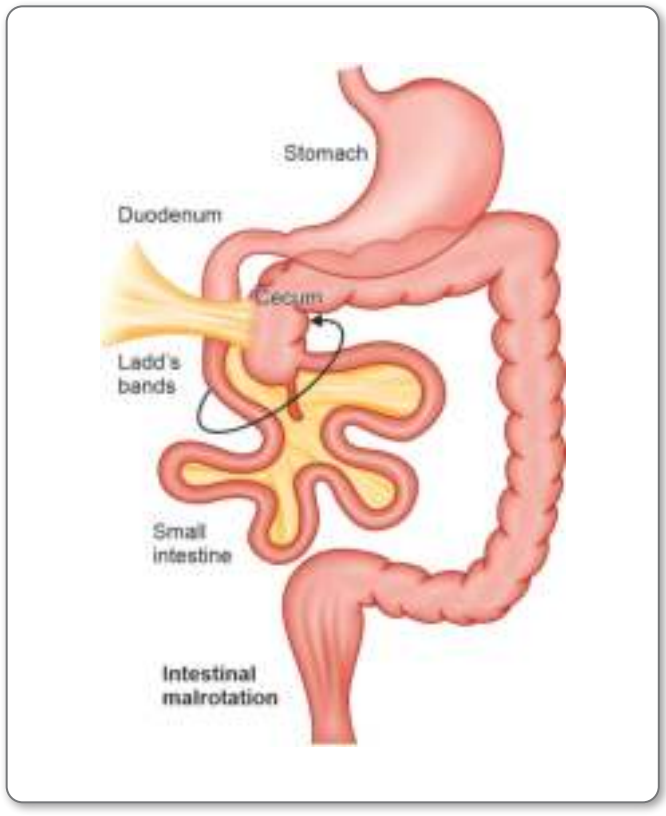

Figure 17.11: Malrotation

Causes

Sex: Male predominance is observed in neonatal presentations at a male-to-female ratio of 2:1. No sexual predilection is observed in patients older than 1 year.

Age: Traditional teaching suggests that as many as 40% of patients with malrotation present within the first week of life, 50% in the first month, and 75% in the first year.

Complications

Midgut volvulus, intestinal ischemia, bowel necrosis, perforation, shock, respiratory failure, death.

Signs and Symptoms

- The most common symptoms are recurrent abdominal pain and malabsorption syndrome
- Abdominal distension, tender abdomen
- Bilious vomiting
- Melena, currant jelly stools

Diagnosis

- An elevated or decreased WBC count may indicate systemic inflammation or sepsis. Metabolic acidosis provides evidence for ongoing ischemia as observed with necrotizing enterocolitis or strangulated bowel.
- Upper GI series, ultrasound, CT scan

Management

Surgery

A classic Ladd procedure is described as reduction of volvulus (if present), division of mesenteric bands, resection of necrotic bowel, placement of small bowel on the right and large bowel on the left of the abdomen, and appendectomy. Ladd's procedure can be done laparoscopically also. Appendicectomy is performed if the normal anatomical placement of the appendix is disrupted or dissection of the peritoneal bands causes damage to the appendiceal vessels.

Preoperative Management

- Correct hypovolemia, metabolic imbalances
- Administer antibiotics
- Insert an NG tube in all patients with bilious emesis and suspected malrotation. Adjust the NG tube to low intermittent suction in order to decompress the bowel proximal to any obstruction that may be present.
- Foley's catheterization
- Prevention of hypothermia

Postoperative Management

- IV fluids and electrolytes and blood transfusion if needed
- NG tube decompression should be continued until bowel function returns to normal
- Broad-spectrum antibiotics
- Pain may be managed with morphine, acetaminophen, distraction techniques
- Nutrition is TPN through central vein, gastrostomy feedings
- Monitor child for wound infection, sepsis, short bowel syndrome as bowel is resected, developmental delays, failure to thrive and vitamin deficiencies

Volvulus

The bowel twists around itself along with the tissue that surrounds it (mesentery) (Fig. 17.12). The twisting causes a blockage so that the contents of the bowel can no longer pass through. It can also cut off the blood supply to that part of the bowel, causing that section to die if not treated quickly.

Symptoms

Pain, knees drawn to chest, absence of stool, green emesis

Surgical Management

Ladd's procedure: The gut is untwisted and positioned so that it is unlikely to twist again. Any tight bands formed around the guts are cut. Often the appendix is removed too.

ABDOMINAL WALL DEFECTS

Abdominal wall defects include omphalocele, gastroschisis, umbilical hernia, inguinal hernia and hydrocele.

Omphalocele

Omphalocele (Fig. 17.13) is a congenital malformation in which intra-abdominal contents herniate through the umbilical cord. It results from failure of the abdominal contents to return to the abdomen when the abdominal wall begins to close by the 10th week of gestation. The protrusion is covered by a translucent sac into which the umbilical cord inserts. Rupture of the sac results in evisceration of the abdominal contents. Intestines within an intact omphalocele are protected from the amniotic fluid; hence, these babies tolerate feedings promptly after the abdominal wall defect is closed. Giant omphalocele can cause pulmonary hypoplasia. Omphalocele may also contain liver along with intestines.

Omphalocele is often associated anomalies such as cardiac defects, genitourinary anomalies, chromosomal defects, craniofacial abnormalities and diaphragmatic abnormalities. They are also associated with increased maternal age, twins, trisomy 13, 18 and 21 and elevation of maternal serum alpha-fetoprotein (MSAFP).

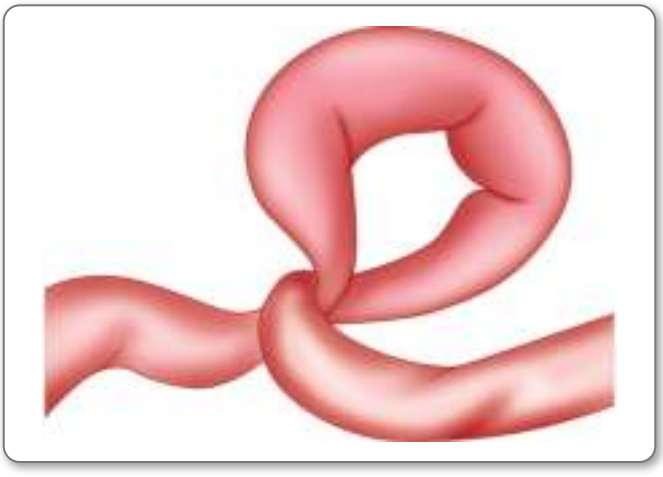

Figure 17.12: Volvulus

Figure 17.13: Omphalocele

Management

- Immediately after birth, the sac is covered with sterile gauze soaked in normal saline to prevent drying of abdominal contents; a layer of plastic wrap is placed over the gauze to provide additional protection against moisture loss.
- Primary repair, for small omphaloceles, the herniated organs are placed back into the abdominal cavity and the defect is completely closed in one operation.
- Staged repair also called Schuster procedure, when the omphalocele contains liver and other organs, a mesh fabric is sewn to the fascia (connective tissue) and muscle on each side of the omphalocele defect (Fig. 17.14). The two pieces of fabric are then sewn together over the defect, and the omphalocele sac remains intact. The organs are gradually returned to the abdominal cavity and the mesh is continuously tightened over the course of days or weeks. Once all of their organs are back in the abdomen, the mesh can be removed and the final closure is performed.
- For **giant omphaloceles**, 'paint and wait' is the safe approach. Topical antimicrobials are applied to the omphalocele membrane, and the infant's torso is wrapped with an elastic bandage, (e.g., ACE bandage). Healing occurs by epithelialization and wound contracture. These babies may require ventilatory assistance and tracheostomies.
- Monitor vital signs every 2–4 hours, particularly temperature because the infant can lose heat through the sac. Key preop and post op interventions are listed in Table 17.1.

Figure 17.14: Staged repair of omphalocele with mesh

Table 17.1: Preoperative and postoperative care in omphalocele

Preoperative management	Postoperative management
• Maintain nothing by mouth (NPO) NPO status. • Administer IV fluids as prescribed to maintain hydration and electrolyte balance. • Monitor for signs of infection. • Handle the infant carefully to prevent rupture of the sac. • Maintain normothermia	• Control pain • Prevent infection, administer broad spectrum antibiotics • Maintain fluid and electrolyte balance • Ensure adequate nutrition • Maintain normothermia

Figure 17.15: Gastroschisis

Gastroschisis

Both gastroschisis and omphalocele allow herniation of intra-abdominal contents through the abdominal wall. In gastroschisis, there is herniation of intestines lateral to umbilical ring usually right of umbilicus (Fig. 17.15). Peritoneal sac is not present.

Diagnosis

There will be increased MSAFP levels (normal 40 mg/L). Diagnosis can also be established with prenatal ultrasound and amniocentesis. Amniotic fluid acetyl cholinesterase has been reported as positive in 80–100% of cases with gastroschisis.

Surgery

Surgery is preferred within first 24 hours. The intestines are covered with cling wrap to prevent hypothermia and infection. First stage is an operative placement bowel in silo. Intermittent tie down of silo is done. It pushes bowel in abdominal cavity. When bowel reaches skin level, defect is closed (Fig. 17.16).

Complication

Fluid loss, infection, hypothermia.

Management

Key preop and postop care are listed in Table 17.2.

Umbilical Hernia

An **umbilical hernia** (Fig. 17.17) is a protrusion of the bowel through an abnormal opening in the abdominal wall that is usually reducible with a finger. In children, hernias most commonly occur at the umbilicus and also through the inguinal canal. Most umbilical hernias do not cause any symptoms and do not require surgical repair until approximately age 5 years.

Inguinal Hernia

Inguinal hernia refers to a painless inguinal when the infant cries or coughs. For inguinal hernia, elective herniorrhaphy is indicated swelling that is reducible (Fig. 17.18). Swelling may disappear during periods of rest and is most noticeable to prevent incarceration and subsequent strangulation.

Figure 17.16: Cling wrap and silo

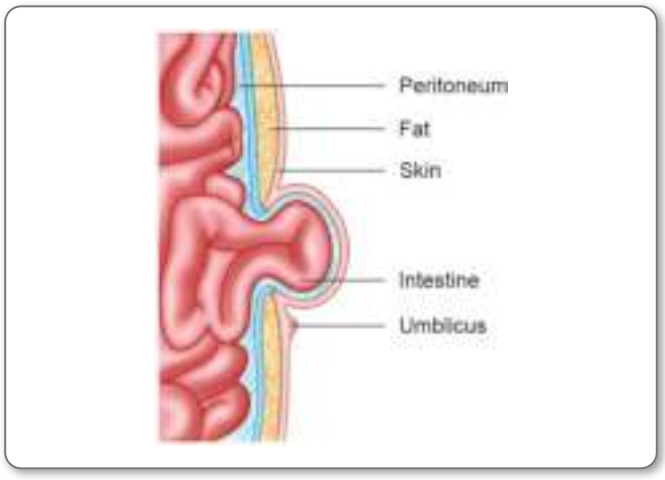

Figure 17.17: Umbilical hernia

Table 17.2: Preoperative and postoperative care in gastroschisis

Preoperative care	Postoperative management
• Prevent hypothermia: Cover the intestines with a nonadherent, semi-permeable membrane (e.g., plastic cling wrap). Wrap dry Kerlex dressing around the intestines, including the infant's torso, so that the intestines are situated just above the abdominal wall defect. Then, place the infant in a bowel bag. This technique minimizes the loss of heat and moisture from the exposed intestines, as well as protects the mesentery from twisting or stretching. • Hydration: Administer IV fluids, monitor weight of infant, intake output, lab tests (CBC, ABG, electrolytes), BP, urine output • Orogastric tube: To prevent gastric distention from swallowed air. • Prevent infection: Sterile barriers, antibiotics, bath silo with warm antibiotic solution, monitor for redness, drainage. • Respiratory support: SaO_2 >95%, avoid bag and mask ventilation (distends bowel), auscultate breath sounds bilaterally.	• Respiratory support • Infection prevention • Fluid therapy • Pain management • Skin care • GI decompression • Nutrition – gastrostomy feedings, TPN

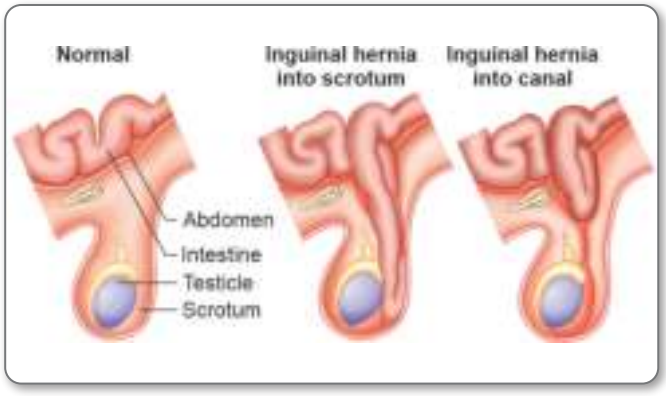

Figure 17.18: Inguinal hernia

Incarcerated Hernia

Incarcerated hernia occurs when the descended portion of the bowel becomes tightly caught in the hernial sac, compromising blood supply. It is a medical emergency requiring surgical repair. Child may present with irritability, tenderness at site, anorexia, abdominal distention, and difficulty defecating. The protrusion cannot be reduced, and complete intestinal obstruction and gangrene may occur.

Hydrocele

Hydrocele refers to presence of abdominal fluid in the scrotal sac (Fig. 17.19). It is of two types: non-communicating and communicating (Table 17.3). A hydrocele usually goes away on its own within six to twelve months of age. If the hydrocele does not resolve on its own, then surgical repair is needed to prevent further complications. The surgery to remove a hydrocele (hydrocelectomy) is done under general or regional anesthesia. An incision is made in the scrotum or lower abdomen to remove the hydrocele. Postoperative Interventions for hernia and hydrocele is described in Table 17.4.

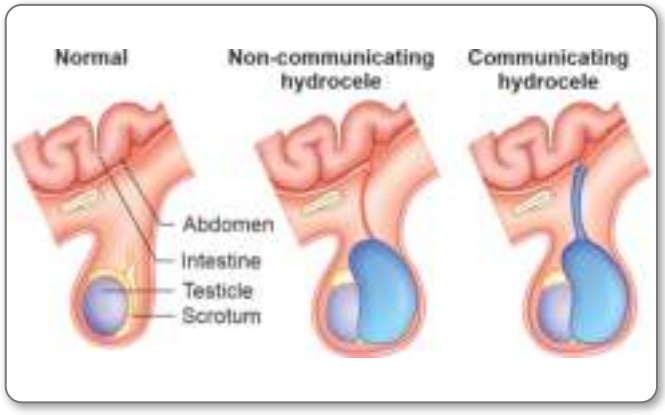

Figure 17.19: Hydrocele

Table 17.3: Types of hydrocele

Non-communicating hydrocele	Communicating hydrocele
• It occurs when residual peritoneal fluid is trapped in the scrotum with no communication to the peritoneal cavity. • It usually disappears by age 1, as the fluid is reabsorbed.	• It is associated with a hernia that remains open from the scrotum to the abdominal cavity. • Assessment: A bulge in the inguinal area or the scrotum that increases with crying or straining and decreases when the infant is at rest. Parents may also report the bulge is smaller in the morning but increases in size throughout the day.

Table 17.4: Postoperative interventions for hernia and hydrocele

Hernia	Hydrocele
• Monitor vital signs • Assess for wound infection • Monitor for redness or drainage • Monitor input and output and hydration status • Advance the diet as tolerated • Administer analgesics as prescribed	• Provide ice bags a scrotal support to relieve pain and swelling • Instruct the parents that tub bathing needs to be avoided until the incision heals • Instruct the parents that strenuous physical activities need to be avoided • Advise parent that the scrotum may not immediately return to normal size

ANORECTAL MALFORMATION

Definition

Malformations that occur congenitally due to abnormal development of genitourinary and pelvic organs are called anorectal malformations. With this defect, the anus and rectum do not develop properly. ARM encompasses multiple congenital anomalies of the rectum, urinary and reproductive structures with varying degree of complexity. Most children with ARM have abnormal communication with rectum, GU tract and perineum called fistulas.

Etiology

Any kind of congenital malformation can lead to anorectal malformation. Anorectal malformation may be seen with some of these genetic syndromes or congenital problems, i.e., VACTERL association, digestive system abnormalities, urinary tract abnormalities or abnormalities of the spine.

Pathophysiology

The anus and rectum originate from the embryological structure cloaca. Lateral growth of cloaca forms the urorectal septum that separates the rectum dorsally from the urinary tract ventrally. The rectum and urinary tract separate completely by the 7th week of gestation. Anomalies occur when separation fail to occur. With an anorectal malformation, several abnormalities can occur, including the following:

- The anal passage may be narrow
- A membrane may be present over the anal opening
- The rectum may not connect to the anus
- The rectum may connect to part of the urinary tract or the reproductive system through a passage called a fistula

The treatment for the malformation depends on the type of abnormality present.

Signs and Symptoms

- Abdominal distension, vomiting
- Anal opening very near the vagina
- Baby does not pass first stool within 24–48 hours after birth
- Missing or moved opening to the anus
- Meconium passes out of the vagina, base of penis, scrotum, or urethra

Classification of Anorectal Malformations (ARM)

Wingspread classification (Table 17.5) was established in 1984 according to the level of the arrest of rectal descent and patient's sex. Peña proposed a classification in 1995 (Table 17.6) based on the presence and position of the fistula. The position of the fistula was used to determine operative management. Male and female anomalies are shown in Figures 17.20 and 17.21 respectively.

Description of Anomalies

- **Perineal fistula:** When rectum opens into perineum, Low malformation

Table 17.5: The wingspread conference classification, 1984

Level of anomaly	Male	Female
High	**Anorectal agenesis** • Rectovaginal fistula • Without fistula • Rectal atresia	**Anorectal agenesis** • Rectovaginal fistula • Without fistula • Rectal atresia
Intermediate	Rectourethral fistula Anal agenesis without fistula	• Rectovestibular fistula • Rectovaginal fistula • Anal agenesis without fistula
Low	• Anocutaneous (perineal) fistula • Anal stenosis (perineal) fistula	• Anovestibular (perineal) fistula • Anocutaneous • Anal stenosis
Miscellaneous	• Rare malformations	• Persistent cloacal anomaly • Rare malformations

Table 17.6: Pena classification, 1995

Males	Female
Perineal fistula	Perineal fistula
Rectourethral fistula Bulbar Prostatic	Vestibular fistula
Rectovesical fistula	Persistent cloaca <3 cm common channel >3 cm common channel
Imperforate anus without fistula Rectal atresia	Imperforate anus without fistula Rectal atresia

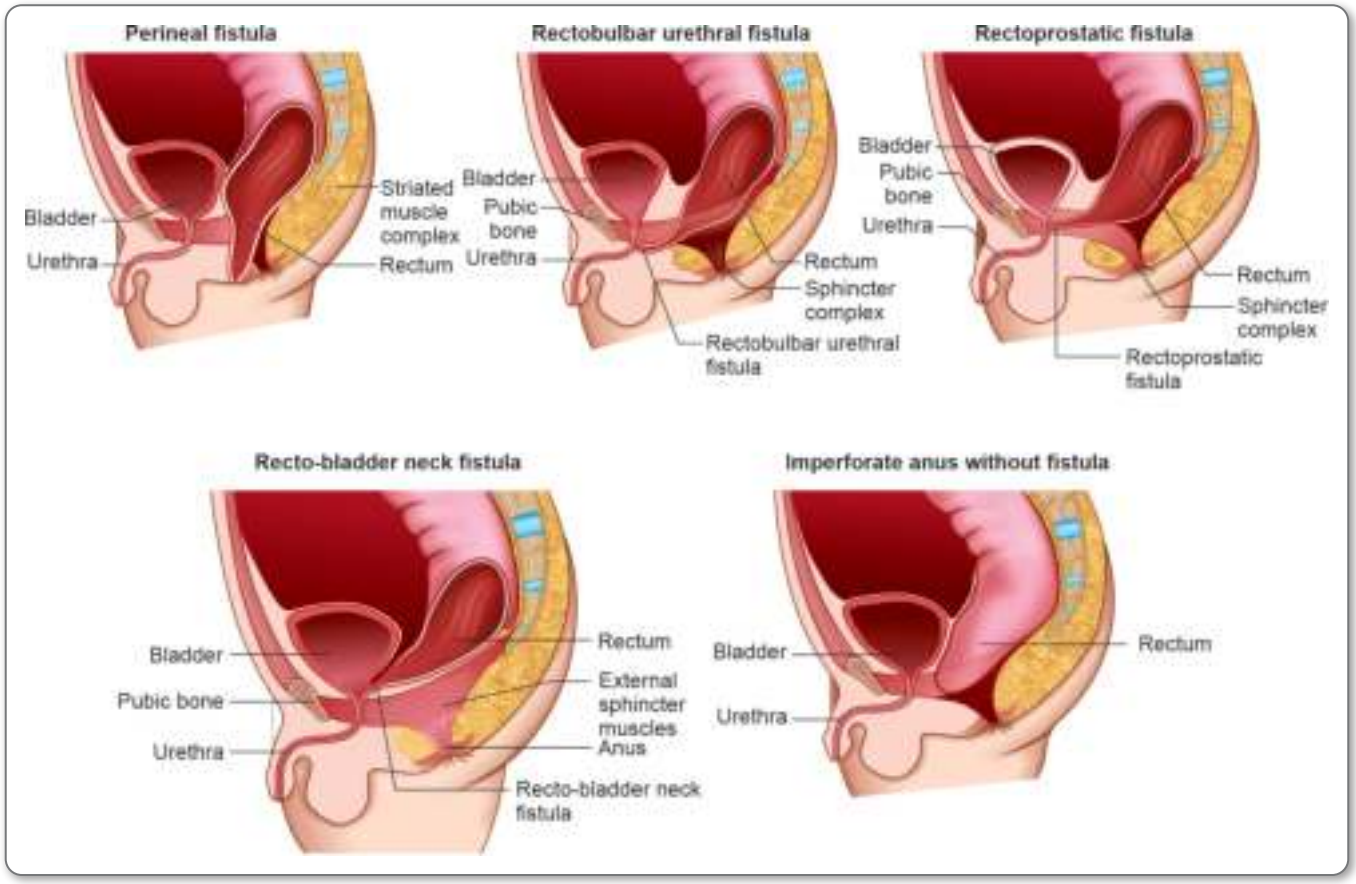

Figure 17.20: Anomalies in males

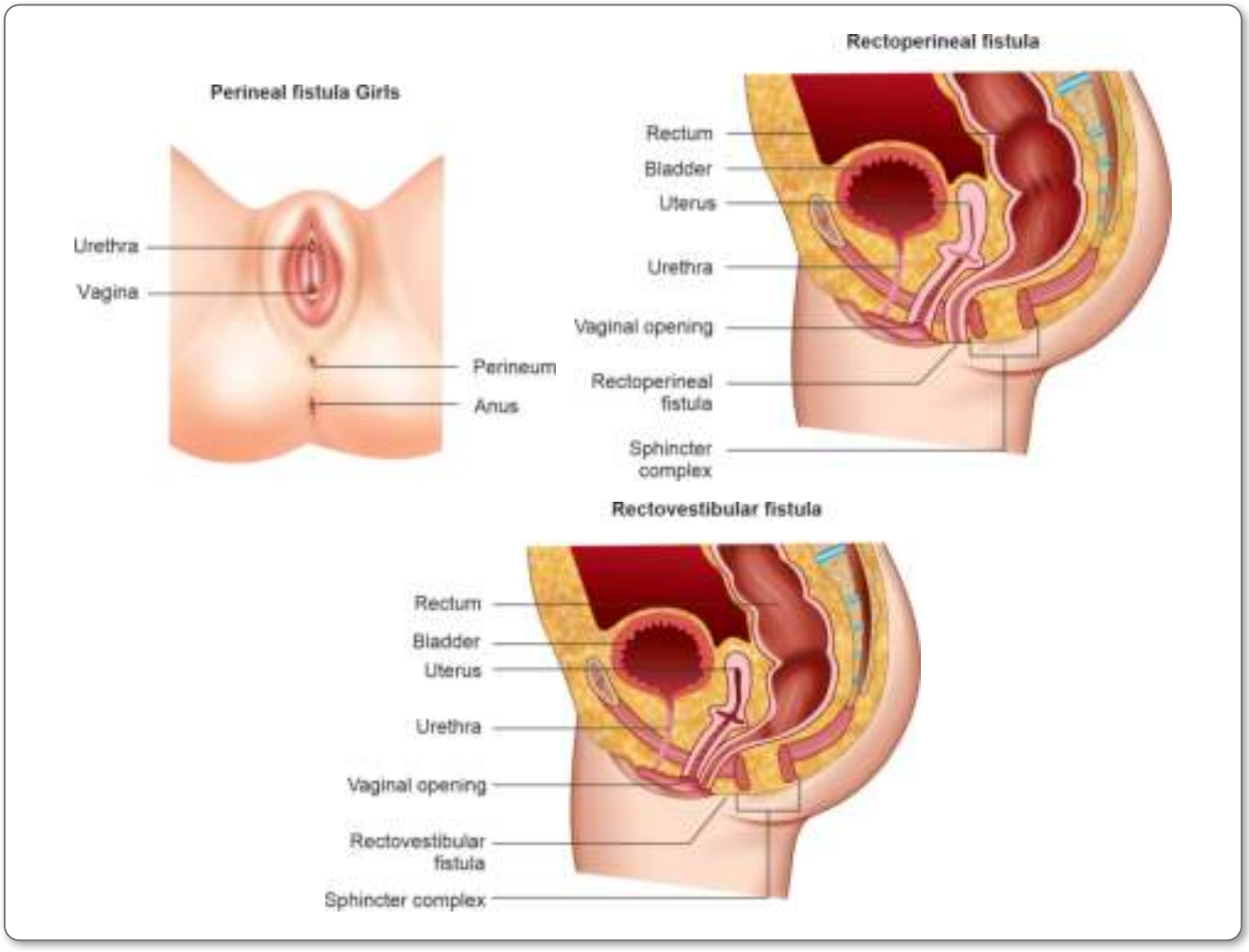

Figure 17.21: Anomalies in females

- **Rectourethral bulbar fistula:** Rectum communicates with the lower posterior portion of the urethra called bulbar urethra.
- **Rectourethral prostatic fistula:** Rectum communicates with the upper part of the posterior portion of the urethra passing through prostatic tissue
- **Rectobladder neck fistula:** Highest defect in male. Rectum communicates with bladder neck. There is abnormal sacrum.
- **Imperforate anus without fistula:** Rectum is completely blind, ends 2 cm from perineum. No meconium in urine. It is associated with Down syndrome.
- **Vestibular fistula:** Rectum opens through an abnormally narrow orifice located in the vestibule of the genitalia, outside the hymen
- **Vaginal fistula:** Meconium comes out through the vagina from inside the hymen orifice
- **Persistent cloaca:** Rectum, vagina and urinary tract are fused together into a single common channel that communicates exteriorly through a single perineal orifice located at normal urethral site
- **Cloacal exstrophy:** It is a rare malformation and includes omphalocele, 2 exstrophied hemi bladders with cecum between them, imperforate anus and abnormal sexual structures

Diagnosis

- **Physical assessment:** Check the patency of anus and rectum. Rectourinary fistula is suspected if there is meconium in the urine
- **Abdominal ultrasound:** To determine the existence of other malformation
- **Intravenous pyelogram and cystourethrogram:** For patients with high malformations to identify anomalies of urinary tract.
- **Invertogram:** To assess height of defect

Management

The treatment of an anorectal malformation may depend on the extent of the problem and the overall health of the child

and medical history. The type and number of operations necessary depend on the type of abnormality the infant has, including the following:

- **Low defects or perineal fistulas or imperforate anus: Anoplasty:** Infant is kept NPO for 4 hours. Fistula is moved to anatomically correct position, at the center of anal sphincter. Larger anal opening is created. Nothing is inserted in rectum. Antibiotic ointment is applied on perineum TDS for 2 weeks. Administer IV antibiotics for 2–3 days. Continue breastfeeding. Anal dilatations at 2 weeks.
- **Lack of rectal/anal connection, with or without a fistula:** These infants need a series of operations in order to have the malformation repaired.

First, a surgery is done to create a **colostomy**. With a colostomy, the large intestine is divided into two sections and the ends of intestine are brought through openings in the abdomen. The upper section allows stool to pass through a stoma and then into a collection bag. The lower section allows mucus that is produced by the intestine to pass into a collection bag. Colostomy does not impair the digestion of child and he/she can grow before time for the next surgery. Also, when the next surgery is done on the lower section of intestine, there will not be any stool present to infect the area. Care of the colostomy is important during this time.

Posterior Sagittal Anorectoplasty (PSARP): This attaches the rectum to the anus and is usually done within the first few months of life. The colostomies remain in place for a few months after this operation so the area can heal without being infected by stool. Even though the rectum and anus are now connected, stool will leave the body through the colostomies until they are closed with surgery. A few weeks after surgery, parents may be performing anal dilatations to help the child get ready for the next phase.

Two to three months later, an operation is done to **close the colostomies**. The child is not allowed anything to eat for a few days after surgery while the intestine is healing. Several days after surgery, the child will start passing stools through the rectum. At first, stools will be frequent and loose. Diaper rash and skin irritation can be a problem at this stage. Within a few weeks after surgery, the stools become less frequent and more solid, often causing constipation. A high-fiber diet can be provided to manage constipation.

Toilet training should be started at the usual age, which is generally when the child is between two and three years old. However, children who have had anorectal malformations repaired may be slower than others to gain bowel control. Some children may not be able to gain good control over their bowel movements, while others may be chronically constipated, depending on the type of malformation and its repair.

Nursing Management

- Assessment of the child immediately after birth.
- Check if the child has passed stools after 24 hours of birth.
- For children who have undergone anoplasty, provide frequent dressing change.
- Provide side-lying position with hips extended or supine position with legs extended at 90 degrees.
- Administer perineal care to the child.
- For children with colostomy, perform frequent dressing change and meticulous skin care. Assess stoma for infection or any other complications.

HEPATITIS

Hepatitis is an acute or chronic inflammation of the liver that may be caused by a virus, alcohol, a medication reaction, or another disease process.

Types

Hepatitis A, B, C, D, E, G

Causative organism of hepatitis is Hepatitis virus (A, B, C, D, E, and G), CMV, EB Virus, Herpes Simplex virus, Varicella zoster, enterovirus, adenovirus, and parvovirus.

Mode of Transmission (Table 17.7)

- **Feco-oral route:** No hand washing, eating contaminated food or drinks
- **Parenteral:** Razors, needle stick injury, blood transfusions
- Sexual
- **Perinatal:** Mother to baby

Symptoms

A child with hepatitis will exhibit flu like symptoms to pallor, nausea, vomiting, dark colored urine and clay colored stools.

Table 17.7: Types and mode of transmission of hepatitis

	Type A	Type B	Type C	Type D	Type E	Type G
Mode of Transmission	Feco-oral route	Parenteral route—Needle stick injury Perinatal Sexual—blood, body fluids	Parenteral Perinatal Sexual	It only happens with hepatitis B	Feco-oral route	Parenteral

Diagnosis

Diagnosis can be established in case there is elevated liver enzymes, prolonged PT, hyperbilirubinemia and lower serum albumin level. Other tests which can be done are LFT, U/S, MRI, CT scan and Liver Biopsy.

Complications

Liver failure and hepatocellular carcinoma

Management

Supportive, Antivirals, Liver transplant

Prevention

Vaccination is available for Hepatitis B.

Hepatitis A

Hepatitis is a self-limited disease. It has only acute phase.

- Mode of Transmission is a feco-oral route. Viral replication occurs in the liver, leading to hepatic injury. Liver injury is represented in three ways:
  1. Direct cellular injury that elevates serum liver enzyme levels
  2. Cholestasis that causes jaundice and hyperbilirubinemia
  3. Inadequate liver function
- **S/S:** The patient's initial symptoms during the prodromal period include low-grade fever, nausea, vomiting, decreased appetite, and abdominal pain. Older children are more likely to report pain in the right upper quadrant.
- **Diagnosis:** Elevated alanine aminotransferase (ALT), aspartate aminotransferase (AST), increased bilirubin, lower serum albumin levels and prolong PT

Nursing Interventions

- Enforce strict hand washing.
- Hospitalization is required if there is evidence of coagulopathy or fulminant hepatitis.
- Follow standard precautions and enteric precautions during hospitalization.
- Provide enteric precautions for at least 1 week after the onset of jaundice with HAV.
- The hospitalized child usually is not isolated in a separate room unless he or she is fecally incontinent and items are likely to become contaminated with feces.
- Children are discouraged from sharing toys.
- Instruct the parents to disinfect diaper-changing surfaces thoroughly with bleach solution.
- Maintain comfort and provide adequate rest and sleep.
- Provide a low-fat, well-balanced diet.
- Inform the parents that because HAV is not infectious 1 week after the onset of jaundice, the child may return to school at that time if he or she feels well enough.
- Inform the parents that jaundice may appear worse before it resolves.
- Instruct the parents about the signs of worsening child's condition, changes in neurological status, bleeding, and fluid retention.

Hepatitis B

Most HBV infection in children is acquired **perinatally**. Newborns are at risk if the mother is infected with HBV or was a carrier of HBV during pregnancy. Possible routes of maternal-fetal (newborn) transmission is leakage of the virus across the placenta late in pregnancy or during labor, ingestion of amniotic fluid or maternal blood, and breastfeeding, especially if the mother has cracked nipples. The severity in the infant varies from no liver disease to fulminant (severe acute course) or chronic active disease. HBV infection occurs in specific high-risk groups, i.e.,:

- Children with hemophilia or other disorders requiring multiple blood transfusions
- Children or adolescents involved in IV drug abuse, institutionalized children, preschool children in endemic areas
- Children who are involved in sexual activity

Infection with HBV can cause a carrier state and lead to eventual cirrhosis or hepatocellular carcinoma in adulthood. Clinicopathologic syndromes include the following:

- **Acute asymptomatic infection with recovery:** Child is asymptomatic and there is serologic evidence of infection
- **Acute hepatitis with resolution:** It consists of anicteric or icteric phase (Table 17.8)
- Chronic hepatitis, with or without progression to cirrhosis
- Fulminant hepatitis with massive liver necrosis
- Coinfection with hepatitis D virus (hepatitis delta virus)

Diagnosis

- Detection of immunoglobulin M (IgM) which denotes active infection and IgG which denotes past infection.

Table 17.8: Phases of hepatitis

Prodromal or anicteric phase	Icteric phase
• Lasts 5–7 days • Absence of jaundice • Anorexia, malaise, lethargy, easy fatigability • Fever (especially in adolescents) • Nausea and vomiting • Epigastric or right upper quadrant abdominal pain • Arthralgia and rashes (more likely with hepatitis B virus) • Hepatomegaly	• Jaundice, which is best assessed in the sclera, nail beds, and mucous membranes • Dark urine and pale stools • Pruritus

- Hepatitis B – HBsAg and antibody + , Hepatitis B DNA-DNA PCR
- Liver biopsy

Medical Management

Antiviral agents: Tenofovir, Lamivudine

Prevention for Hepatitis B

- Follow standard precautions.
- Wear gloves while touching bandages, tampons, and linens.
- Cover all open cuts or wounds.
- Don't share razors, toothbrushes, nail care tools, or pierced earrings with anyone.
- Make certain that any needles for drugs, ear piercing, or tattoos – or tools for manicures and pedicures – are properly sterilized.
- Clean up blood with one part household bleach and 10 parts water.
- Educate for safe sex practices
- Screening for blood donation

Vaccination

- Hepatitis A vaccine and hepatitis B vaccine
- Hepatitis B vaccine is given at 0, 1, 6 months, I/M, 0.5 mL. Vaccine uses hepatitis B surface antigen (HbsAg) to stimulate the production of antibodies in noninfected individuals (active immunity).
- PEP- Hepatitis B Ig provides passive immunity and may be effective in preventing infection after a 1-time exposure (should be given immediately after exposure), such as an accidental needle puncture or other contact of contaminated material with mucous membranes; immunoglobulin should also be given to newborns whose mothers are positive for hepatitis B surface antigen.

LACTOSE INTOLERANCE

Definition

Lactose intolerance is defined as inability to tolerate lactose due to absence or deficiency of lactase, an enzyme found in the secretions of the small intestine that is required for the digestion of lactose.

Assessment

- Symptoms occur after the ingestion of milk or other dairy products.
- Abdominal distention
- Crampy, abdominal pain; colic
- Diarrhea and excessive flatus

Interventions

- Eliminate the dairy product which is causing symptoms, or administer an enzyme tablet replacement.
- Provide information to the parents about enzyme tablets that predigest the lactose in dairy products or supplement the body's own lactase.
- Substitute soy-based formulas for cow's milk formula or human milk.
- Allow milk consumption as tolerated.
- Instruct the child and family that the child should drink milk with other foods
- Encourage consumption of hard cheese, cottage cheese, and yogurt, which contain the inactive lactase enzyme.
- Encourage consumption of small amounts of dairy foods daily to help colonic bacteria adapt to ingested lactose.
- Instruct the parents about the foods that contain lactose, including hidden sources.
- A child with lactose intolerance can develop calcium and vitamin D deficiency. Instruct the parents about the importance of providing these supplements.

MALABSORPTION SYNDROME

Malabsorption syndrome refers to a number of disorders in which the small intestine cannot absorb enough of certain nutrients and fluids. These may include proteins, carbohydrates, and fats, micronutrients (vitamins and minerals), or both.

Causes

Factors that may cause malabsorption syndrome include:

- Damage to the intestine from infection, inflammation, trauma, or surgery
- Prolonged use of antibiotics
- Other conditions such as celiac disease, Crohn's disease, chronic pancreatitis, or cystic fibrosis
- Lactase deficiency, or lactose intolerance
- Congenital defects such as biliary atresia
- Diseases of the gallbladder, liver, or pancreas
- Parasitic diseases
- Radiation therapy, which may injure the lining of the intestine
- Certain drugs that may injure the lining of the intestine, such as tetracycline, colchicine, or cholestyramine

Symptoms

Symptoms are related to deficiency of absorption of following nutrients:

- **Fats:** Light-colored, foul-smelling stools that are soft and bulky. Stools are difficult to flush and may float or stick to the sides of the toilet bowl.
- **Protein:** Dry hair, hair loss, or fluid retention.
- Bloating, gas, or explosive diarrhea.
- **Certain vitamins:** Anemia, malnutrition, low blood pressure, weight loss, or muscle wasting.

- Women may stop menstruating, and children may not grow properly. Their weight or rate of weight gain may be significantly below that of other children of a similar age and gender.

Diagnosis

- Stool tests to measure fat.
- Blood tests to measure the level of specific nutrients such as vitamin B-12, vitamin D, folate, iron, calcium, carotene, phosphorus, albumin, and protein.
- Biopsy of small intestine.
- Imaging tests to look for structural problems of small intestine

Treatment

- Treatment is supportive based on the findings.
- Loperamide may be administered for diarrhea
- Monitor for signs of dehydration, like increased thirst, low urine output, and dry mouth, skin, or tongue.
- In case of lactose intolerance, advise to avoid milk and other dairy products or take a lactase enzyme tablet.
- Enzyme supplements to help body absorb the nutrients
- Vitamin supplements

CELIAC DISEASE

Celiac disease is also known as gluten enteropathy or celiac sprue. There is intolerance to gluten, the protein component of wheat, barley, rye, and oats. There is accumulation of the amino acid glutamine, which is toxic to intestinal mucosal cells. Intestinal villous atrophy occurs, which affects absorption of ingested nutrients.

Symptoms of the disorder occur most often between the ages of 1 and 5 years. There is usually an interval of 3 to 6 months between the introduction of gluten in the diet and the onset of symptoms. Strict dietary avoidance of gluten minimizes the risk of developing malignant lymphoma of the small intestine and other GI malignancies.

Assessment

- Acute or insidious diarrhea
- Steatorrhea
- Anorexia
- Abdominal pain and distention
- Muscle wasting, particularly in the buttocks and extremities
- Vomiting
- Anemia
- Irritability
- Celiac crisis is precipitated by fasting, infection, or ingestion of gluten. There is profuse watery diarrhea and vomiting. It can lead to rapid dehydration, electrolyte imbalance, and severe acidosis.

Interventions

- Maintain a gluten-free diet, substituting corn, rice, and millet as grain sources.
- Instruct the parents and child about lifelong elimination of gluten sources such as wheat, rye, oats, and barley.
- Administer mineral and vitamin supplements, including iron, folic acid, and fat-soluble vitamins A, D, E, and K.
- Teach the child and parents about a gluten-free diet and about reading food labels carefully for hidden sources of gluten.

APPENDICITIS

Appendicitis is inflammation of the appendix. When the appendix becomes inflamed or infected, perforation may occur within hours, leading to peritonitis, sepsis, septic shock, and potentially death. Treatment is surgical removal of the appendix before perforation occurs.

Assessment

- Pain in periumbilical area that descends to the right lower quadrant
- Abdominal pain that is most intense at McBurney's point (Fig. 17.22).
- Referred pain indicating the presence of peritoneal irritation
- Rebound tenderness and abdominal rigidity
- Elevated white blood cell count
- Side-lying position with abdominal guarding (legs flexed) to relieve pain
- Difficulty in walking and pain in the right hip

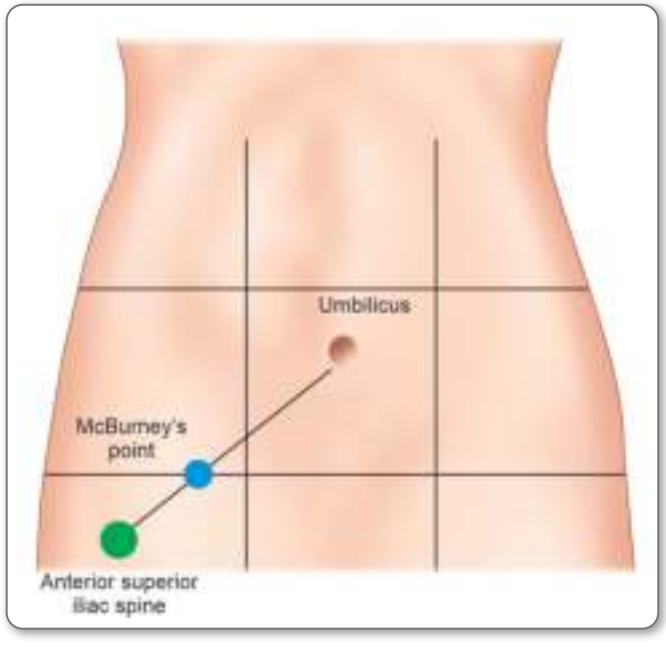

Figure 17.22: McBurney's point

- Low-grade fever
- Anorexia, nausea, and vomiting after pain develops
- Diarrhea

Complication

Peritonitis: It can result from a perforated appendix. There is increased fever, progressive abdominal distention, tachycardia and tachypnea, pallor, chills, restlessness and irritability. An indication of a perforated appendix is the sudden relief of pain and then a subsequent increase in pain accompanied by right guarding of the abdomen.

Surgical Management

Appendectomy: It refers to removal of appendix

Preoperative Care

- Maintain NPO status.
- Administer IV fluids and electrolytes as prescribed to prevent dehydration and maintain electrolyte balance.
- Monitor for changes in the level of pain.
- Monitor for signs of a ruptured appendix and peritonitis.
- Avoid the use of pain medications so as not to mask pain changes associated with perforation.
- Administer antibiotics as prescribed.
- Monitor bowel sounds.
- Position in a right side-lying or low to semi-Fowler's position to promote comfort
- Apply ice packs to the abdomen for 20 to 30 minutes every hour if prescribed.
- Avoid the application of heat to the abdomen.
- Avoid laxatives or enemas

Postoperative Interventions

- Monitor vital signs, especially temperature.
- Maintain NPO status until bowel function returns, advance the diet gradually as tolerated and as prescribed when bowel sounds return.
- Assess the incision for signs of infection, redness, swelling, drainage, and pain.
- Monitor drainage from the drain, which may be inserted if perforation occurred.
- Position the child in a right side lying or low to semi-Fowler's position with the legs slightly flexed to facilitate drainage.
- Change the dressing as prescribed and record the type and amount of drainage.
- Perform wound irrigations if prescribed.
- Maintain NG tube suction and patency of the tube if present.
- Administer antibiotics and analgesics as prescribed.

WORM INFESTATION

Common infections in children are giardiasis and pinworm infestation. Giardiasis is caused by protozoa and is prevalent among children in crowded environments, such as classrooms or day care centers. Mode of transmission is feco-oral route (soil, water). It is caused by Giardia intestinalis, Giardia lamblia, or Giardia duodenalis), found on surfaces or in soil, food, or water that has been contaminated with feces) from infected humans or animals. Stool specimens from three or more collections are used for diagnosis. Giardiasis and Pinworm infestation are explained in Table 17.9.

Table 17.9: Overview of worm infestations

	Giardiasis	Pinworms
Causative organism	Giardia intestinalis, Giardia lamblia, or Giardia duodenalis	A small, thin, white roundworm (nematode) called Enterobius vermicularis
Symptoms	• Diarrhea and vomiting • Anorexia • Failure to thrive • Abdominal cramps with intermittent loose stools and constipation • Steatorrhea	• Intense perianal itching • Irritability, restlessness • Poor sleeping • Bed wetting
Management	• Medications are not usually prescribed for children younger than 2 years. • Drugs, metronidazole, tinidazole, and nitazoxanide • Caregivers should wash hands meticulously. • Provide education to family and caregivers regarding sanitary practices.	• Perform a visual inspection of the anus with a flashlight 2 to 3 hours after sleep. • The tape test is the most common diagnostic test. • Educate the family and caregivers regarding the tape test. A loop of transparent tape is placed firmly against the child's perianal area; it is removed in the morning and placed in a glass jar or plastic bag and transported to the laboratory for analysis. • All members of the family are treated for the infection. • Teach the family and caregivers about the importance of meticulous hand washing and about washing all clothes and bed linens in hot water. • Deworming, either mebendazole (500 mg), pyrantel pamoate, or albendazole (400 mg). Any of these drugs are given in one dose initially, and then another single dose of the same drug two weeks later.

Summary

The red flags for children with GI system disorders include abdominal pain, vomiting, weight gain or weight loss, bloody stools, constipation, diarrhea, fever, fistulas, oral ulcers, etc. Treating GI disorders involve plenty of rest, medicines, maintaining hydration, BRAT (bananas, rice, applesauce and toast) diet, and avoiding spices. Sometimes surgery may be needed.

Assess Yourself

1. Name the surgeries done for:
   - Cleft lip .....
   - Cleft palate .....
   - Pyloric stenosis .....
   - GERD.....
   - Megacolon .....
   - Choledochal cyst .....
   - High ARM .....
2. Full form of VACTERL syndrome is .....
3. List the preventive strategies for transmission of Hepatitis.
4. Pain in McBurney's point is seen in .....