“ ЗАТВЕРДЖЕНО”

на методичній нараді кафедри

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професор\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_А.Ф. Левицький

**THEME No 2**

**Congenital gastrointestinal abnormalities: Hypertrophic pyloric stenosis, Duodenal atresia, Jejuno-ileal atresia and stenosis, Hirschsprung's disease, Anorectal abnormalities.**

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**Overview**

Most congenital GI anomalies result in some type of intestinal obstruction, frequently manifesting with feeding difficulties, distention, and emesis at birth or within 1 or 2 days. Some congenital GI malformations, such as malrotation, have a very good outcome, whereas others, such as congenital diaphragmatic hernia, have a poor outcome, with a relatively high mortality rate of 10 to 30%.

A common type of anomaly is atresia, in which a segment of the GI tract fails to form or develop normally. The most common type is esophageal atresia, followed by atresia in the jejunoileal region and in the duodenum.

Immediate management includes bowel decompression (by continuous nasogastric suction to prevent emesis, which can lead to aspiration pneumonia or further abdominal distention with respiratory embarrassment) and referral to a center for neonatal surgery. Also vital are maintenance of body temperature, prevention of hypoglycemia with IV 10% dextrose and electrolytes, and prevention or treatment of acidosis and infections so that the infant is in optimal condition for surgery.

**Educational aims:**

The aim of this part of module is to provide help in performing early diagnosis and management of children with congenital gastrointestinal abnormalities.

**A student must know:**

1. Relevant anatomy of the intestine and abdominal cavity in children.
2. The classic clinical manifestation of the congenital gastrointestinal abnormalities.
3. Features of clinical presentations of congenital gastrointestinal abnormalities in newborns.
4. Lab studies and imaging studies in children with congenital gastrointestinal abnormalities.
5. Differential diagnosis of different congenital gastrointestinal abnormalities.
6. Complications of congenital gastrointestinal abnormalities.
7. Different surgical technique under congenital gastrointestinal abnormalities.
8. Preoperative management in children with congenital gastrointestinal abnormalities.
9. Advantages and disadvantages of laparoscopic techniques under congenital gastrointestinal abnormalities.
10. Postoperative complications after surgery of congenital gastrointestinal abnormalities.
11. Postoperative management after congenital gastrointestinal abnormalities.

**A student must be able to:**

1. Look for:
* location of anus in neonates
* evidence of pyloric stenosis on the abdominal wall
* signs of abdominal distension
1. Define the dehydratation stage in neonate and calculate the liquid and electrolyte volumes for rehydratation
2. Make the physical exam of children
3. Evaluate the quantity and quality of meconium in neonates
4. Put nasogastric tube
5. Interpret the results of abdominal x-ray and ultrasound diagnosis and
6. Perform the enema before the irrigography

**Terminology**

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| --- | --- |
| ***Term*** | ***Definition*** |
| Hypertrophic pyloric stenosis | is narrowing of the opening from the stomach to the first part of the small intestine known as the duodenum, due to enlargement of the muscle surrounding this opening (the pylorus, meaning "gate"), which spasms when the stomach empties. |
| “Bull’s eye” sign | characteristic appearance of pyloric stenosis on ultrasound on cross section of the pyloric channel |
| Double-bubble sign | An x-ray gas shadows essentially pathognomonic for duodenal obstruction |
| Intestinal atresia | malformation where there is a narrowing or absence of a portion of the intestine |
| Hirshprung’s disease | is a form of megacolon that occurs when part or all of the large intestine or antecedent parts of the gastrointestinal tract have no ganglion cells and therefore cannot function |
| Acetylcholinesterase (AChE) staining techniques | investigation of choice to evaluate suction biopsies to show an increased activity in the parasympathetic nerves of the affected zone as well as neurofibrils within the lamina propria and muscularis mucosa |
| VACTERL association  | Association of vertebral and spinal cord, Anorectal, Cardiac, TracheoEsophageal, Renal and other urinary tract, Limb anomalies |

CONTENT

***Hypertrophic pyloric stenosis***

**Introduction**

Infantile hypertrophic pyloric stenosis (IHPS) is a common surgical cause of vomiting in infancy. Historically, it was described as a disease entity in 1888 by Harald Hirschsprung. Gastrojejunostomy was used to treat this disease until 1912, when extramucosal muscle-splitting pyloromyotomy was described by Ramstedt. This procedure has dramatically changed the outcome of infants with IHPS.

**Demographics**

The reported incidence of IHPS is 1–4 per 1,000 live births.5 There is a male-to-female ratio of 4:1, with reported ratios ranging from 2.5:1 to 5.5:1.

**Aetiology**

Despite the frequency of pyloric stenosis, the aetiology remains unclear. Genetic predisposition acting in conjunction with environmental factors is the most widely accepted explanation; however, debate still continues as to whether it is a congenital or acquired disease. Breast-feeding has been suggested as offering some immunity to the disease. First-born children have been noted to be more likely affected, and a familial link is seen with a greater than fivefold increase in the risk in first-degree relatives. The genetics explaining this are likely to be polygenic, as no single locus has been identified. Male and female children of affected mothers carry a 20% and 7% risk, respectively, of developing the condition, whereas male and female children of affected fathers carry a 5% and 2.5% risk, respectively. Furthermore, an association is seen in twins, with concordance among monozygotic twins of 0.25–0.44, and in dizygotic twins of 0.05–0.10.

**Pathophysiology**

Pyloric stenosis is characterized by hypertrophy of the pyloric musculature, leading to a mechanical obstruction of the gastric outlet in the affected infant. Thus, hypertrophied pyloric antral muscle fibres protrude distally into the duodenal lumen, producing a reflection of duodenal mucosa. Infants with a diagnosis of pyloric stenosis will show characteristically low chloride and hydrogen ions as measured in the serum. The loss of gastric secretions secondary to protracted vomiting will result in dehydration. As a result, through aldosterone-stimulated absorption, potassium is excreted in the urine in an attempt to conserve sodium. As potassium depletion worsens, sodium resorption across the renal tubule is then achieved in exchange for a hydrogen ion, thereby creating paradoxical aciduria. Classically, this results in the occurrence of a hypochloraemic hypokalaemic metabolic alkalosis. In severe cases with diagnostic delay, hypoglycaemia and hypoalbuminaemia can be observed. It is known that the pyloric hypertrophy will eventually resolve, but this takes a long period of time; the infant would usually succumb to the electrolyte derangement and dehydration before this happened.

**Clinical Presentation**

Infants with pyloric stenosis usually present with a gradual onset of worsening nonbilious vomiting, beginning between 3 to 6 weeks of age. The pattern of vomiting can vary, but often it progresses to the characteristic “projectile” vomiting. Infants may present in the early stages of the disease and be treated for reflux disease or undergone numerous formula changes before the diagnosis is made. Delay in diagnosis can result in significant electrolyte imbalance, weight loss, and failure to thrive.

The typical clinical features include the following:

• Nonbilious vomiting is usually forceful and postprandrial.

• The infant is hungry after vomiting and eager to feed, only to vomit again.

• Weight loss occurs in severe cases.

• Signs of dehydration present in cases of repeated vomiting.

• Scaphoid abdomen especially noted after recent vomiting.

• Visible peristalsis may be observed in the upper abdomen, usually moving from the left hypochondrium towards the right side.

• A palpable mass is present in the right upper quadrant (90% in experienced hands); this is best appreciated while the infant is being fed with clear fluid.

**Differential Diagnosis**

The differential diagnosis of pyloric stenosis includes:

• gastro-oesophageal reflux;

• viral enteritis;

• pylorospasm;

• duodenal stenosis/duodenal web;

• raised intracranial pressure.

**Clinical Diagnosis**

Depending of the time to presentation, the clinical picture can vary enormously from a well-hydrated baby to an emaciated infant. Weight loss and dehydration coupled with an insatiable appetite lead to a characteristic facies, with a furrowed brow, wrinkled appearance, and prominent sucking pads. In some infants, the distended stomach may be identifiable in the hypochondrium, with active peristaltic activity visible through thin abdominal wall. On examination, a mobile, ovoid mass, commonly referred to as an “olive”, is palpable in the epigastrium or the right upper quadrant.

If the pylorus is palpated by an experienced clinician, no further imaging is necessary. In some cases, however, other structures may be confused with hypertrophied pylorus, including the caudate lobe of the liver, the right kidney, the vertebrae, or an orogastric tube in the distal stomach. If there is any doubt, or in the absence of a palpable “olive”, diagnostic imaging can be helpful.

**Ultrasonography**

In situations where doubt exists, examination by ultrasound (US) should be performed. This would normally confirm the presence of a pyloric “tumour”. The characteristic appearance of pyloric stenosis on ultrasound is that of a “doughnut” or “bull’s eye” on cross section of the pyloric channel. Pyloric dimensions with positive predictive value greater than 90% are muscle thickness greater than 4 mm and a pyloric channel length greater than 17 mm. These limits may be lower in infants younger than 30 days of age.

An experienced sonographer will recognize periods of relaxation in infants with pylorospasm, commonly confused with pyloric stenosis at examination. Pylorospasm has been hypothesized to be an early stage of IHPS, but this has not been proven.

**Upper Gastrointestinal Contrast Study**

In an occasional case where doubt still persists after US examination, an upper gastrointestinal (UGI) series may be done. The UGI series would show a narrow pyloric channel, the so-called “string sign” and the “shoulder sign”, caused by the impression of the pylorus into the stomach.

**Serum Electrolytes**

Serum electrolytes should be measured immediately when the patient arrives in hospital. If vomiting has been ongoing for several days, serum electrolytes are frequently deranged. The nature of derangement is a spectrum, ranging from mild to severe hyponatraemia, hypochloraemia, hypokalaemia, and metabolic alkalosis. The degree of elevation of serum urea is directly related to the severity of dehydration. Infants presenting late are often malnourished and may have somedegree of anaemia, which may require correction. Therefore, a haemogram and full blood count are warranted.

**Treatment**

**Correction of Electrolyte and Fluid Depletion**

Patients with pyloric stenosis may have severe electrolyte disturbances, so the serum electrolytes should always be estimated. Mild electrolyte disturbances can be corrected preoperatively with 0.45% normal saline with 5% dextrose solution. Severe disturbances require correction with 0.9% normal saline bolus of 10 to 20 ml/kg, followed by administration of 0.9% saline in 5% dextrose solution. Potassium can be added if necessary when adequate urine output (1.5–2 ml/kg per hour) is established and under electrocardiogram (ECG) monitor. Fluid should be administered at a rate of 25–50% above maintenance.

Following resuscitation and correction of electrolyte imbalance, maintenance IV with 0.45% saline in 5% dextrose with 20-mmol potassium chloride should be given at 25-50% above the standard rate.

Meticulous care and time should always be taken to correct fluid and electrolyte depletion before any surgical correction. It is important to emphasize that mortalities from pyloric stenosis are attributable to fluid and electrolyte problems.

**Nasogastric Decompression**

Once diagnosis is made, all feeds are stopped. It is helpful to aspirate all gastric content by nasogastric tube (NGT). Frequently, this content comprises milk curds, which may require lavage with saline to adequately evacuate the stomach. Keeping the stomach empty would help prevent aspiration from vomiting. Once the stomach is emptied, the NGT is either closed off or removed to avoid worsening electrolyte depletion by aspirating gastric content. In the West, gastric lavage is not routinely performed. An NGT is passed, size 8 Fr or above. Gastric losses are monitored and replaced milliliter for milliliter with 0.9% saline. To avoid iatrogenic hyperkalaemia, no potassium is added to the replacement fluid.

**Surgical Correction**

Surgical correction of pyloric stenosis is not an emergency, and therefore the electrolyte disturbances can and should be meticulously corrected before operation. Occasionally, children with pyloric stenosis will have jaundice due to a transient impairment of glucuronyl transferase activity. This is self-limited once postoperative feeding is initiated.

Infants undergoing pyloromyotomy are assumed to have a full stomach and the anaesthesiologist should keep this in mind. Both the anaesthesiologist and surgeon should be vigilant during the operation to prevent aspiration of gastric juice. The stomach must be evacuated in the operating room, particularly if NGT had not been inserted earlier.

Preoperative antibiotics are controversial; data supporting their use with the standard right upper quadrant incision are scant. They may be of benefit when performing the operation through the umbilical skinfold.

**Operative Details**

The standard operation is the Ramstedt pyloromyotomy. Classically, the operation has been approached through a right upper quadrant muscle-splitting approach17. Alternatively, the approach may be via a supra-umbilical transverse skinfold incision.

1. Once the peritoneum is entered, the omentum is retrieved into the wound and elevated to lift the transverse colon. This manoeuvre enables the surgeon to identify the antrum of the stomach. The lower third of the stomach is then gently elevated using moist gauze to deliver the pyloric mass into the wound (Figure 3).

2. A vertical incision is then made into the mid anterior surface through the serosa and superficial muscularis, beginning about 1–2 mm from the pyloroduodenal junction to a point 0.5 cm into the lower antrum.

3. The underlying firm fibres are then divided using blunt dissection with a clamp, rounded end of a scalpel blade, or special Benson’s pyloromyotomy spreader. Special care is taken to prevent mucosal perforation, especially at the lower end of the incision. Upward protrusion of the gastric mucosa indicates relief of the obstruction.

Mucosal perforation usually occurs at the duodenal end and is indicated by the appearance of bilious fluid. When this occurs, repair is done by using interrupted fine monofilament long-term absorbable sutures placed transversely and covered with omentum. If the closure of the mucosal perforation compromises the pyloromyotomy, which rarely happens, a fresh pyloromyotomy is done at about 45°–90° of the first incision. Air is then instilled through the NGT to check the integrity of the duodenal mucosa.

**Laparoscopic pyloromyotomy** can be performed with similar efficiency and surgical outcome as traditional open pyloromyotomy. Improved cosmesis and avoidance of wound complications are major benefits of this procedure, and a tendency towards less postoperative emesis is a potential benefit that deserves further investigation.

**Postoperative Management**

Postoperative nasogastric decompression is not necessary unless the mucosa has been entered and repaired. Several feeding schedules have been advocated after surgery. Traditional structured feeding regimens as opposed to more rapid initiation and advancing feeding schedules are probably unnecessary. Feedings are begun 4 to 6 hours after operation, normally with low-volume balanced electrolyte or dextrose solution initially, rapidly advanced to full feeds of formula over the next 12- to 24-hour period. If the patient vomits, which is common after this procedure, the same volume feed that caused the emesis can be repeated. The patient is usually discharged the day after operation.

**Surgical Complications**

Intraoperative risks include bleeding, infection, and mucosal perforation. Postoperative complications include wound infection and dehiscence in about 1%. Persistent vomiting beyond 48 hours, thought to be due to gastricatony, occurs in about 3%. Unrecognized perforation during pyloromyotomy is a serious but rare problem demanding immediate reoperation.

**Outcome**

The majority of infants go on to make a full recovery postoperatively and need no further medical input. After a surgical pyloromyotomy, the pyloric muscle subsides to a normal size and, when viewed during subsequent operations, is usually visible only as a fine line over the pylorus at the site of the myotomy. Incomplete pyloromyotomy may occur, but it is difficult to diagnose in the early postoperative phase. Imaging studies done postoperatively are difficult to interpret and usually not helpful. If complete gastric-outlet obstruction is present on a contrast study, repeated pyloromyotomy is necessary. Mortality is rare, but when it occurs, it is usually from fluid and electrolyte depletion in infants presenting late, and inadequately corrected electrolyte problems before surgery.

***Duodenal atresia***

**Introduction**

Congenital duodenal obstruction may be due to intrinsic or extrinsiclesions. Intrinsic duodenal obstruction may be caused by duodenal atresia,stenosis, diaphragm with or without perforation, or by a wind-sockweb or membrane that balloons distally. Extrinsic duodenal obstructionmay be caused by malrotation with Ladd’s bands or a preduodenal portalvein or annular pancreas. The annular pancreas itself is not believedto be the cause of obstruction, as there is usually an associated atresiaor stenosis in these patients.

Duodenal obstructions usually occur in the second part of theduodenum. They are believed to result from a developmental errorduring early foetal life within the area of intense embryological activityinvolved in the creation of the biliary and pancreatic structures. Thus,the obstruction usually occurs at or below the ampulla of Vater.

Duodenal obstruction is associated with prematurity (46%) andmaternal polyhydramnios (33%). In addition, there is a high incidenceof specific associated anomalies, including Down syndrome (>30%),malrotation (>20%), congenital heart diseases (20%), and othergastrointestinal tract (GIT) and renal anomalies. Along with prematurityand low birth weight, these associated anomalies are known to besignificant risk factors contributing to mortality in patients withduodenal atresia. Of note, the presence of Down syndrome itself doesnot influence the outcome of these babies.

**Demographics**

Theincidence of duodenal obstruction is reported to be 1 in 5,000–10,000births in most reports. Duodenal obstruction and jejunoileal atresiarank among the two most common causes of intestinal obstruction inlarge series in the population.

**Aetiology**

It has been demonstrated that from gestational weeks 5 to 10, the duodenumis a solid chord. Intrinsic obstructions result from failure of vacuolizationand recanalization. An annular pancreas results from fusion of the anteriorand posterior anlage, forming a ring of pancreatic tissue that surrounds thesecond part of the duodenum. Extrinsic obstructions result from a variety ofdisorders of embryologic development specific to the pathology.

**Clinical Presentation**

**Prenatal**

Duodenal obstruction is readily diagnosed by prenatal ultrasound.Antenatal care with prenatal ultrasonography should therefore be offeredto pregnant women in all circumstances. Duodenal obstruction presentsup to gestational week 20 with a double-bubble phenomenon due to thesimultaneous distention of the stomach and the first part of the duodenum.In more than 30% of cases, maternal polyhydramnios is present,and in some cases, serial amniotic aspiration has been reported as necessary.In facilities where ultrasound is not available, a high index of suspicionmust be maintained in cases of maternal polyhydramnios. Pregnancycan last near to maturity, and spontaneous delivery is usually the case.

**Postnatal Symptoms and Signs**

The most common presenting features arebilious vomiting and feeding intolerance. Dehydration and electrolytedepletion rapidly ensue if the condition is not recognized and intravenoustherapy is not begun. Aspiration and respiratory failure may follow.Repeated nonbilious vomiting is seen in cases of supra-ampullaryobstruction (20%). Patients with a web or partial stenosis can surviveto present in a much delayed fashion.Physical signs are nonspecific but can include upper-abdominaldistention with scaphoid lower abdomen. Additionally, in theappropriate clinical context, observation of typical Down syndromefeatures should raise suspicion towards duodenal obstruction as thecause for neonatal intestinal obstruction. Finally, a careful physicalexam should concentrate on recognizing signs of significant congenitalheart disease (e.g., cyanosis, murmurs), which could complicateperioperative management.

**Investigation**

In tertiary perinatal centres where a prenatal diagnosis has already beenestablished, no further diagnostic work-up is typically necessary.In doubtful cases or in other settings, a plain abdominal x-rayis the key method for diagnosis. An x-ray showing double-bubblegas shadows is essentially pathognomonic for duodenal obstruction(Figure 6). If no double bubble is seen, instillation of 10–15 mlof air immediately prior to a plain abdominal radiograph may helpto demonstrate these findings. In cases of stenosis or perforatedmembranes, air may be seen in the distal GIT. Water-soluble contrastradiography is confirmatory, but it is generally needed only in cases ofincomplete obstruction. Radiographic findings of annular pancreas areusually indistinguishable from other forms of duodenal obstruction.

The most important differential diagnosis is duodenal obstructiondue to malrotation, resulting in volvulus of the midgut loop or extrinsiccompression related to Ladd´s bands across the duodenum. When noprenatal diagnosis is available, contrast radiography may be helpful todifferentiate between these entities and can demonstrate the absence ofthe normal C-shaped curve of the duodenum or a classic “bird’s-beak”shape secondary to a volvulus. When the diagnosis still remains indoubt, prompt laparotomy is warranted because undiagnosed volvuluscan result in gangrene of the entire midgut within hours.

If available, in cases of incomplete obstruction, oesophagogastroduodenoscopy (EGD) can be done to prove the existence of an intrinsicobstructing membrane. An endoscopic approach to membrane resectioncan be utilized.

**Management**

**Preoperative Care**

The intensity of preoperative care is typically proportionate to thetime from birth until hospital presentation. Initial therapy consists ofnasogastric decompression and appropriate replacement of fluid andelectrolytes. Most of these newborn patients are premature and smallfor their gestational age, so special care must be taken to preserve bodyheat and to avoid hypoglycaemia, especially in cases of very low birthweight, congenital heart disease, and respiratory distress syndrome.When incubators are unavailable, the “kangaroo” method of nursingthese children offers the best hope for survival.

**General Intraoperative Considerations**

General anaesthesia with endotracheal intubation is required. The mostcommonly utilized incision is a muscle-cutting, transverse, right upperquadrant incision. However, some centres are now employing minimalaccess laparoscopic methods for repair of duodenal obstruction.

A side-to-side duodenoduodenostomy is the standard repair forduodenal stenosis, atresia, or obstruction due to a preduodenal portalvein. In 1977, Kimura and colleagues described a modification of thisprocedure, known as the diamond-shaped duodenoduodenostomy.Inthis technique, a horizontal incision is made across the distal aspect ofthe proximal, dilated bowel, and a lengthwise incision is made alongthe proximal aspect of the distal, small-calibre bowel. This can achievea greater diameter of the anastomosis for better emptying of the upperduodenum. In some cases, duodenojejunostomy can be an alternativeand may afford an easier repair with minimal dissection. The choice ofsurgical procedure is largely based on the preference of the surgeon.

When an annular pancreas associated with duodenal obstruction isencountered, the treatment of choice is performance ofa duodenoduodenostomy between the segments of duodenum aboveand below the area of the ring of pancreas. One should never considerdivision of the pancreatic ring because that could result in a pancreaticfistula while the underlying stenosis or atresia of the duodenum wouldremain unchanged.

In the case of an endoluminal membrane, duodenotomy andresection of the membrane can be done after localisation of the ampullaof Vater. Alternatively, bypass of the membrane can be performed via aduodenoduodenostomy, if desired.

**Postoperative Considerations and Complications**

Intravenous infusions are continued for the postoperative period. Usinga transanastomotic tube laying deep in the jejunum, feeding can bestarted as early as 48 hours postoperatively. Where available, parenteralnutrition via a central or peripherally inserted catheter can be veryeffective for longer-term nutritional support if transanastomotic enteralfeeding is inadequate, not feasible, or not tolerated by the patient. Allpatients have a prolonged period of bile-stained gastric aspirate. This ismainly due to the ineffective peristalsis of the distended upper duodenum.The commencement of oral feeding is dependent upon a decreasein the volume of gastric aspirate and is often delayed for up to severalweeks. Patients who have a severely prolonged return of duodenalfunction and have exceptionally marked dilatation of the proximalduodenum may benefit from reoperation and tapering of the proximalsegment, although this is rare.Anastomotic leak, intraabdominal sepsis, and wound complicationsalso are rare.

**Prognosis**

Although prognosis of intestinal atresia in general is good, an overall mortality of 7% for duodenal obstruction is shown in large series. Associated congenital anomalies are identified as an independent risk factor for an impaired clinical course. Low birth weight and the problems of prematurity further increase mortality risk.

***Intestinal atresia and stenosis***

**Introduction**

Atresias of the jejunum and ileum are common causes of bowel obstruction in the neonate, with a third of infants born prematurely or small for their gestational age. Stenoses are much less common and seldom present in the newborn period due to delay in diagnosis.

**Demographics**

Jejuno-ileal atresia has a prevalence rate of approximately 1:330–1:1,500 live births, with a third of infants either born prematurely or small for date.

**Aetiology/Pathophysiology**

Our present understanding of the aetiology of intestinal atresias is based upon the classic experimental work of Louw and Barnard reported in 1955. These investigators observed that ligating mesenteric vessels and causing strangulated obstruction in foetal dogs resulted in atretic lesions of the small intestine that were similar to those observed clinically in human neonates. Thus, atresias and stenoses of the small intestine are believed to be due to an ischaemic insult. This aetiologic mechanism explains the frequent association of atresias with mesenteric defects and with other conditions that may cause strangulated obstruction of the intestinal tract (e.g., volvulus, intussusception, internal hernias, and gastroschisis). An ischaemic aetiology may also explain why intestinal atresia is associated with maternal smoking and vasoconstrictor drug exposure during pregnancy.

The morphological classification into four types has both prognostic and therapeutic implications (Figure 10):

• Stenoses occurs in 11%.

• Type I atresia (23%) is a transluminal septum with proximal dilated bowel in continuity with collapsed distal bowel. The bowel is usually of normal length.

• Type II atresia (10%) involves two blind-ending atretic ends separated by a fibrous cord along the edge of the mesentery with mesentery intact.

Type IIIa atresia (15%) is similar to type II, but there is a mesenteric defect and the bowel length may be foreshortened.

• Type IIIb atresia (19%) (“apple peel” or “Christmas tree” deformity) consists of a proximal jejunal atresia, often with malrotation with absence of most of the mesentery and a varying length of ileum surviving on perfusion from retrograde flow along a single artery of supply.

• Type IV atresia is a multiple atresia of types I, II, and III, like a string of sausages. Bowel length is always reduced. The terminal ileum, as in type III, is usually spared.

The immediate consequence of an atresia is dilatation of the bowel for a variable distance proximal to the first occlusion encountered. This dilated bowel, even when the obstruction is relieved by resection and anastomosis or stoma formation, remains dilated, having inefficient prograde peristalsis. Surgical strategies to overcome this include back resection of this bowel to a normal-calibre intestine or reduction in diameter by various tapering manoeuvres.

**Clinical Presentation**

However, atresias of the proximal jejunum are frequently associated with polyhydramnios. Therefore, many of these patients are born prematurely and often are small for their gestational age, the latter due to inability to absorb nutrients from the amniotic fluid in patients with proximal intestinal obstructions.

Intestinal atresia should be suspected in any newborn showing evidence of bowel obstruction (bilious vomiting, abdominal distention, and failure to pass meconium). Aspiration of >25 ml of fluid from the stomach via a nasogastric tube (NGT) is very suggestive of obstruction. Antenatal ultrasound scanning may show dilated loops of bowel with vigorous peristalsis, which is diagnostic of obstruction. Polyhydramnios may develop but it is more commonly seen in duodenal and oesophageal obstructions. The more distal the atresia, the more generalized the abdominal distention. After aspiration of gastric contents, the abdomen will be less distended and visible peristalsis may be observed. There is usually a failure to pass meconium, and typically small-volume gray mucoid stools are passed. Abdominal tenderness or peritonitis develops only with complications of ischaemia or perforation. This commonly occurs with a delay in diagnosis and is due to increased intraluminal pressure from swallowed air and secondary volvulus of the bulbous blind-ending bowel at the level of the first obstruction.

**Physical Examination**

Findings on physical examination are frequently not very revealing. Most patients will have some degree of abdominal distention. The amount of distention will vary, depending upon the level of obstruction. Patients generally do not have abdominal tenderness or an abdominal mass. Therefore, the presence of these findings suggests a complicated obstruction associated with ischaemia or prenatal perforation, or that the cause of obstruction may be malrotation with midgut volvulus.

**Investigations**

In most patients, a simple abdominal x-ray with anteroposterior (AP) and either cross-table or left lateral decubitus projection are adequate to make the diagnosis based upon the presence of dilated, air-filled intestinal loops and air-fluid levels. In addition, plain abdominal x-rays will suggest the level of obstruction based upon the number of dilated bowel loops. The presence of multiple dilated bowel loops without air-fluid levels suggests the possibility of meconium ileus, particularly if the intestinal content has a “ground glass” appearance. A single very dilated loop with a large fluid level is often indicative of atresia.

The differential diagnosis includes other causes of intestinal obstruction in the neonate. In patients with evidence of a proximal complete obstruction, the differential diagnosis is limited and no additional diagnostic studies are required. In patients with multiple dilated bowel loops, suggesting a distal obstruction, the differential diagnosis includes several conditions for which surgical intervention may not be required. Therefore, in these patients a contrast enema may be helpful to look for evidence of a meconium plug or meconium ileus, which may respond to nonoperative management. In addition, a contrast enema may demonstrate findings suggestive of Hirschsprung’s disease, which would direct initial management towards obtaining confirmatory tests for this disease. A contrast enema showing a patent colon is helpful in that demonstration of colonic patency by injection of saline at operation—a sometimes tedious procedure—is not required.

In patients with intestinal stenoses, plain abdominal x-rays may demonstrate proximal bowel dilatation; however, in most patients a gastrointestinal contrast meal or enema is required to confirm and locate the site of partial obstruction.

**Management**

All patients should receive judicious fluid hydration prior to operative intervention. In addition, a nasogastric or orogastric tube should be passed to empty the stomach and decrease the risk of vomiting with aspiration. In general, patients with intestinal atresias have a low risk of associated cardiac anomalies, so that preoperative special investigation is not required unless the patient has clinical evidence of a serious cardiac defect.

At exploration, the site of the most proximal atresia is readily identified as the site of marked change in intestinal calibre. The outer wall of the intestine at the site of obstruction may appear intact or there may be an associated defect in continuity of the intestine and the mesentery. Generally, surgical treatment requires excision of the ends of the intestine involved in the atresia. It is also important to look for distal sites of obstruction, which can occur in up to 20% of patients and may not be immediately obvious due to lack of caliber change beyond the proximal atresia. These distal points of obstruction can be identified by flushing the distal intestinal lumen with saline to confirm intestinal continuity to the level of the rectum.

After resection of the atretic segment, the surgeon is faced with the difficult task of re-establishing continuity between intestinal segments with marked size discrepancies. Another consideration is the potential dysmotility of the proximal markedly dilated segment, which may result in delayed intestinal function and problems with bacterial overgrowth. Therefore, in patients with a relatively short segment of severely dilated proximal intestine, resection of the dilated segment with re-establishment of continuity by end-to-end anastomosis is a good option. However, in patients with long segments of proximal intestine that are significantly dilated, resection of the whole involved segment may result in inadequate remaining intestinal length to allow absorption of enteric nutrients (i.e., short bowel syndrome). Therefore, these patients frequently are treated by either imbrication or tapering enteroplasty of the proximal dilated segment. To date, no randomized studies have compared the outcomes for patients with intestinal atresias with or without the addition of an enteroplasty or plication. In patients for whom the atresia is just distal to the duodenojejunal flexure, it may be advantageous to resect the dilated bowel, derotate, and taper the duodenum with primary anastomosis. This facilitates passage of a transanastomotic feeding tube and early restoration of foregut function. The total residual length of bowel should be measured with a tape and recorded, as this gives some guidance as to prognosis.

Patients who have multiple atresias (type IV) or an apple-peel deformity (type IIIb) present particularly challenging management problems. These patients may require multiple anastomoses and frequently will experience long-term delays in return of intestinal function. In addition, many of these patients will have short bowel syndrome due to inadequate residual intestinal length. In general, the formation of stomas is unnecessary and should be avoided because dilated bowel does not reduce in caliber, and fluid and electrolyte losses may be severe.

**Postoperative Complications**

The most common postoperative complication is a functional obstruction at the site of anastomosis. Unfortunately, this complication may be due to the underlying intestinal dysmotility associated with this anomaly and may not be preventable by changes in surgical technique. Other less commonly observed complications include anastomotic leak and adhesive obstructions. Obstructions due to missed distal unrecognized atresias should not occur and can be prevented by proper evaluation at the time of the initial operation.

**Prognosis and Outcomes**

Most patients with intestinal atresia do not have associated life-threatening anomalies. Therefore, the primary factor that impacts mortality is the ability to support the nutritional needs of the patient during the postoperative period while awaiting adequate bowel function to allow enteral alimentation. In centres where parenteral nutritional support is feasible, these patients can be supported for prolonged periods of time while awaiting gastrointestinal function. However, in centres without these resources, patient mortality will be higher and primarily attributable to malnutrition. The judicious use of nasojejunal or gastrostomy transanastomotic feeding tubes for enteral feeding may be life saving.

***Hirshprung’s disease***

**Introduction**

Hirschsprung’s disease (HSCR or HD) may be defined as a functional intestinal obstruction resulting from the congenital absence of parasympathetic ganglion cells in the myenteric plexus of the distal bowel. The initial description of congenital aganglionosis (Hirschsprung’s disease) by Harald Hirschsprung, heralded as the Father of Danish Paediatrics, in 1886 could not have anticipated the worldwide interest it would evoke. Successful treatment of the condition had to wait for 50 years until the pivotal role of the distal aganglionic segment and its vital role in the pathophysiology of Hirschsprung’s disease was identified. The development of successful surgical management in the same year has made HSCR one of the success stories of paediatric surgery in the modern era.

**Demographics**

Hirschsprung’s disease is a congenital cause of functional intestinal obstruction mostly diagnosable at birth but often presenting late in resource-poor environments.

The HSCR incidence worldwide is approximately 1 in 5,000 live births. The male-to-female ratio is approximately 4:1 overall worldwide, but appears to vary between the various ethnic groups represented in South Africa. The male-to-female ratio is also affected by the length of the aganglionic segment, which approaches 1:1 in total colonic aganglionosis (TCA). An overall male predominance is apparent, with incomplete gene penetrance and a variable phenotype.

Most babies with HSCR are born to mothers with a normal antenatal history (92%) with the majority having good Apgar scores. HSCR is mostly associated with big babies (average birth weight of 3,129 gm), and has been generally thought to be rare or absent in premature babies. This widely held belief has not been substantiated in recent studies, although our series had 12.2% who weighed less than 2.5 kg at birth, but in only 3 (1.9%) could a gestational age <37 weeks (28, 32, 33 weeks) be documented.

**Aetiology/Pathophysiology**

The normal migration of neuroblasts in a cephalo-caudal direction within the bowel reaches the rectum by 12 weeks. Should the normal development be disturbed, a part of the colon will lack the normal ganglion cells and result in a functional loss of coordinated peristalsis, as occurs in HSCR.

At a molecular level, HSCR essentially appears to result from disruption of normal signalling during development. As a result, the cues controlling the migration of the neural crest cells go awry, resulting in aganglionosis of the distal bowel.

Current evidence would appear to support a large genetic component in HSCR aetiology. The patterns of conditions associated with HSCR have already been of great value in revealing many of the genetic components of the condition. Known genetic variations have been identified in at least 12% of HSCR cases which is higher than the expected in the normal population. Although much progress has been made in assessing possible mechanisms by which this gene malfunction may be involved in the pathogenesis of HSCR, the aetiology of HSCR is complex probably involving both genetic and microenvironmental influences in the development of the clinical phenotype. The extent of the complexity is shown by the number of genes implicated in its pathogenesis (at least nine). This is hardly surprising, as the signals governing cell migration and development in the embryo are extraordinarily complicated, and signalling molecules are notorious for crosstalk and redundancy.

Although the reason for the incomplete migration and development of ganglion cells is as yet not completely clear, the identification of major susceptibility genes (namely, the REarranged during Transfection (RET) gene and the Endothelin B receptor gene (EDNRB) and other genetic variations) have helped in understanding the aetiology of the condition. Ongoing research has identified a number additional HSCR susceptability genes (the EDNRB ligand EDN 3, the glial cell line derived neurotrophic factor (GDNF) situated at chromosome 5p12-13, and its related GFRα). In addition, the recently described association with PHOX2B and the SOX-10 gene on chromosome 22q13 and appears to synergise with the endothelin system in very long aganglionic segments.

**Pathology**

Hirschsprung’s disease results in a functional obstruction of the bowel with dilatation of the proximal colon and hypertrophy of the muscles (i.e., megacolon).

Macroscopically, HSCR features a narrow aganglionic segment and a transitional zone, and then the dilated proximal portion with a thickened bowel wall as a result of hypertrophy of the muscular wall of the intestine.

HD classically affects the rectum and sigmoid (70%), but can also involve a long portion of colon and can affect the entire large bowel. In 20% of cases, a long segment of colon is affected, with the total colon being aganglionic in 8–10%.

Extensive ultra-long aganglionosis with extensive small bowel involvement is uncommon, occurring in <2%. The functional abnormality always includes the internal anal sphincter and extends proximally for short or long distances, depending on the type.

HSCR, although regarded at one stage as a relatively easy diagnosis, has on occasion become “one of the most difficult diagnoses in Paediatric Surgery”,10 mainly due to difficulties in interpreting the histological, clinical, and radiological findings.

**Histologic Diagnosis**

The classic histological picture of Hirschsprung’s disease is the absence of ganglion cells in intramuscular myenteric (Auerbach’s) plexus and the submucosal Meissner’s plexus. In addition, proliferation of peripheral nerves may also be seen in the affected bowel. The method of suction biopsy initially described by Helen Noblett11 allows specimens of the submucosal and mucosal layers to be obtained with minimal discomfort and without anaesthesia. A number of innovations to the biopsy forceps over the years have culminated in the availability of a superior tool with disposable capsules for specimen taking. This method, although reliable, is underutilized due to the expense of the disposable capsules. A more durable tool of the oldfashioned type is still manufactured, but can be difficult to locate.

Biopsy specimens are taken at 2 and 4 cm (3 and 5 cm in older children). Failure to obtain adequate diagnostic yield on rectal suction biopsy will necessitate a full thickness biopsy, which causes problems during surgical procedures that require mucosal stripping (e.g., endorectal pull-through procedures). The biopsy specimens are then snap frozen and sectioned into slices ±15 μm thick.

**Acetylcholinesterase Increase**

Acetylcholinesterase (AChE) staining techniques remain the investigation of choice to evaluate suction biopsies to show an increased activity in the parasympathetic nerves of the affected zone as well as neurofibrils within the lamina propria and muscularis mucosa. This technique remains the gold standard on rectal suction biopsies in Europe and many parts of the world. Interpretation of AChE staining may be influenced by the different patterns of AChE seen (particularly in neonates). The classic type A staining shows prominent AChE positive nerve fibrils throughout the lamina propria. The type B pattern shows similar AChE neurofibrils in the muscularis mucosa and neighbouring lamina propria.

The morphologic diagnosis of HSCR therefore rests on the following:

• Absence of ganglion cells in the submucosal layer (and/or intermyenteric (Auerbach’s) plexus)

• Presence of the enlarged peripheral nerve trunks in the submucosa

• Increased AChE staining—proliferation of neurofibrils in the lamina propria and the muscularis mucosa (absent in normally innervated intestine).

**Clinical Features**

The clinical evaluation of the patient remains the most important diagnostic step in the diagnosis of HSCR.

**History**

A delay in passage of meconium is the most important neonatal observation. Normal babies pass meconium within 24 hours, and a small percentage will also pass meconium by 48 hours. A significant delay in the passage of meconium after birth indicates a congenital cause. Any baby who has a delay in the passage of meconium of more than 24 hours or who passes little meconium should be investigated for HSCR. Other signs in the neonate are a functional intestinal obstruction and bile-stained vomiting, often from day 2. A family history of HSCR or severe constipation are othersignificant factors. A family history is present in 4%, with a 61% possibility of a long segment. Other associated anomalies account for 16% of the cases of HSCR.

**Physical Presentation**

The clinical presentation depends not only on the length of the aganglionosis

but also the age of the patient. Neonate (50–80%)

• Most cases (>90%) present with signs in the neonatal period but are sometimes overlooked in poorly resourced health situations.

• Intestinal obstruction presents with bile-stained vomiting andabdominal distention (often by day 2).

• Delayed passage of meconium is a presenting feature in more than 80% of patients with Hirschsprung’s disease.

• Hirschsprung’s-associated enterocolitis (HAEC) (16%) presents with bloody diarrhoea and mucus associated with abdominal distention and vomiting. The majority of cases of enterocolitis present during 2–4 weeks after birth. This is an important diagnosis because it accounts for 53% of the mortality arising from Hirschsprung’s disease.

Older child (+10–50%)

• Some patients have early onset of mild constipation followed by acute low intestinal obstruction. The early onset of chronic constipation (often since birth) is an indication to exclude HSCR. Stools when passed are irregular and passed with great difficulty.

• Abdominal distention occurs in almost 100% (may be marked).

• Megacolon of the proximal colon is a classic sign of HSCR.

• The child does not develop normally and is often thin and malnourished. In contrast to the rectal findings in chronic constipation, the rectum is often empty on examination in HSCR and should the examining finger push beyond the aganglionic zone, there is an explosive evacuation of soft stool.

• Hirschsprung’s-associated enterocolitis can also occur in the older patient or after surgery. This may lead to toxic megacolon.

• Secondary urogenital problems may occur in patients as a result of the chronic obstruction (e.g., vesicoureteric reflux, hydronephrosis).

**Diagnostic Investigations**

Abdominal X-rays

The diagnostic accuracy of abdominal x-rays is 52%.

• Look for signs of low intestinal obstruction and distended bowel loops of different calibres.

• Erect plates can demonstrate in fluid levels.

• A lateral view may demonstrate the narrow rectum.

**Contrast Enema**

• A contrast enema is diagnostic in two-thirds of patients if low pressures are used. Irrigation of the colon before the contrast must not be performed as it may result in possible decompression of the megacolon or the distended bowel.

• The narrow aganglionic segment is shown with a dilated proximal bowel segment, and a transitional zone is diagnostic.

• Care must be taken not to apply pressure in neonates, as the bowel can easily be distended

The aganglionic segment may be irregular, demonstrating a sawtooth appearance, probably as a result of muscular fasciculations.

• A delay in the clearing of contrast (barium sulphate) within 24 hours is also a reliable sign, and a follow-up x-ray should be performed the following day.

**Manometry**

In normal people, distention of the rectum results in the reflex relaxation of the internal sphincter (rectosphincteric reflex; see Figure 19).

This is absent on manometry in patients with Hirschsprung’s disease.

***Differential Diagnosis***

Differential diagnosis includes the following:

• Small left colon syndrome, particularly in diabetic mothers

• Meconium plug syndrome in neonates.

• Chronic idiopathic intestinal pseudo-obstruction (CIIP, or CHIPS)

• Other dysganglionosis of the gastrointestinal tract

• Acquired megacolon (generally at >1 year of age), resulting from anal fissure, anal or rectal stricture, anorectal malformations, tumour, or psychogenic reasons.

**Management**

The standard surgical management of Hirschsprung’s disease varies among centres. The initial aim of management is to confirm the diagnosis. The patient is resuscitated and antibiotics are given (some children with enterocolitis have clostridium difficile infection). The second aim is to correct the problem (i.e., relieve the intestinal obstruction) by means of definitive surgery. The aim of definitive surgery is to resect the abnormal aganglionic bowel and to anastomose normally ganglionated bowel to the rectum without affecting continence. Fluid and electrolyte losses are corrected and antibiotics commenced, if required.

**Principles of Surgical Management and Results**

A defunctioning colostomy (colostomy located at site of normal ganglionated bowel as determined by the presence of ganglion cells on frozen section) is the traditional way of relieving obstruction. This is then followed by a definitive pull-through procedure 3 to 9 months later.

Although a covering colostomy appears to decrease the incidence of infection, complications from the colostomy are not uncommon, including skin excoriation and bleeding from the stoma and prolapse.

In modern practice, temporary decompression is first attempted by means of washouts with warm saline. Should this be successful, it can be continued until definitive surgery can be performed.

The recent swing to management by bowel irrigation techniques allows for early one-age surgery to be performed at a much earlier stage (mostly still within the neonatal period). Colostomy may still be necessary in resistant cases or if nursing care is unreliable.

Definitive pull-through surgical procedures have undergone numerous modifications since the original description by Swenson. Most of these modifications are based on the original concept described in 1948 by Swenson and Bill4 and adhere to the principle of removing the functionally obstructive segment of aganglionic bowel and reanastomosis. In recent years, the endorectal pull-through (ERP) described by Soave14 has gained popularity.

**Neonatal Pull-Through**

The current trend is for primary anal ERP either as a primary procedure or as a laparoscopically assisted pull-through procedure once the patient has been stabilized. Many argue for the latter because the laparoscope allows for histological mapping to identify the level of the transitional zone by biopsy It appears to make the procedure considerably easier in the neonate. It would appear that neonatal pull-through is a safe and feasible method of treatment of Hirschsprung’s disease and is suitable for those patients diagnosed in the neonatal period. Unfortunately, this applies to fewer HSCR cases in much of Africa. The remainder, who present with grossly distended megacolon, are probably best managed by surgical diversion of the faecal stream, allowing the chronically distended bowel to return to a normal calibre lumen before definitive surgical correction. The major disadvantage of the one-stage neonatal pull-through procedure is that the determination of normal bowel is entirely dependant on frozen section histopathological evaluation. An expert pathological service is essential for this procedure to be carried out safely; the time taken for this procedure may vary but may add considerable delay. In addition, longer aganglionic segments present certain technical difficulties, resulting in a significant number of conversions to standard operative procedures.

**Definitive Surgical Procedures**

Five surgical procedures have received reasonably wide acceptance.

*Swenson Procedure*

This was the original operation described by Swenson in 1948. It involves resection of the aganglionic segment deep into the pelvis and direct endto- end anastomosis of the proximal colon to the anorectal canal.

*Duhamel Procedure*

In the Duhamel procedure (retrorectal pull-through), the lower but aganglionic rectum is retained and the ganglionated bowel brought posteriorly and anastomosed to the aganglionic remnant in a side-toside anastomosis.

*Soave Procedure*

The Soave procedure (extramucosal endorectal pull-through), along with its variations, is the most frequently performed procedure in the world for short-segment Hirschsprung’s disease. It has more recently been popularized as a laparoscopic-assisted or anal approach.

The procedure involves an extramucosal resection of a retained aganglionic rectal segment. The rectal mucosa is removed and a muscular cuff retained. The ganglionated colon is brought through this cuff and anastomosed to the dentate line in the rectum, thus forming an endorectal pull-through.

Transanal Pull-Through

The transanal pull-through approach is through the anus, thus avoiding abdominal scars. It is currently in vogue in many parts of the world, being mostly suited to a short aganglionic segment. It is similar to the Soave procedure, but is performed in reverse through the anus.

The technique involves the patient being placed in lithotomy and the rectum irrigated until clean. Retraction sutures are placed to expose the rectal mucosa and open the anus. Submucosal dissection is commenced 3–5 mm from the dentate line and the cut line controlled by multiple fine traction sutures. Following the completion of the submucosal dissection, the rectum is transected. The dissection is continued proximally until the peritoneal reflection where the sigmoid colon is mobilized and delivered. Following histological confirmation of ganglion cells in the proximal bowel, the aganglionic segment is resected, and a sutured anastomosis is performed (Fig.11.8).

Many recommend a laparoscopic first stage to mobilize the bowel and perform biopsies to facilitate the resection margins.

**Postoperative Complications of Hirschsprung’s Surgery**

Early postoperative complications appear to occur at a similar rate regardless of the age at surgery. The majority of reports of neonatal correction of Hirschsprung’s disease describe initial experiences, and a paucity of long term-data exists. The Toronto group15 have shown that the complication rate of single-stage repair was unaffected by whether the repair was performed prior to or later than the neonatal period, and the short-term outcome was similar for those weighing less than 4 kg. There is a paucity of long-term evaluations, although one large study16 reported a 6% incidence of postoperative enterocolitis and a 4% stricture rate. The overall long-term development of function (continence, sexual, and psychological) remains largely unevaluated.

*Early Complications*

Early postoperative complications include anastomotic insufficiency, stenosis, prolonged ileus, adhesive obstruction, intestinal obstruction, and retraction of the neorectum. Wound sepsis may be present to varying degrees, and other complications of sepsis or pelvic or presacral abscesses may be evident. Acidosis may be associated with excessive fluid and electrolyte losses in long-segment disease, and enterocolitis associated with Hirschsprung’s disease may be present. Early complications following the Soave procedure are a reported higher rate of anastomotic leakage (4–7.7%) and stenosis (9.4–23.7%) compared with other procedures.

*Late Complications*

It is essential that the operative details are available in the assessment of long-term complications, so that like can be compared with like. The evolving nature of the surgery of Hirschsprung’s disease should be borne in mind, as it has evolved from the original three-stage procedure to a two-stage and now one-stage (mostly neonatal) pull-through (often laparoscopic assisted). These technical advances have largely altered the nature and frequency of early complications, and it would appear from available evidence that the newer modifications give a lower incidence of enterocolitis and stricture formation. Reliable data and evaluation of late complications are largely unavailable regarding constipation incidence, continence, social integration, and sexual function, among others.

It is generally recognised that the major long-term complications in the postoperative period following Hirschsprung’s surgery are constipation and Hirschsprung’s-associated enterocolitis. The long-term incidence of constipation approaches 9% following almost all surgical procedures, but this figure may be considerably influenced by certain procedures with a well-established higher incidence of constipation (e.g., the Rehbein procedure). The incidence of postoperative constipation is one of the most practical methods of measuring successful therapy, although its assessment is often highly subjective. The true incidence may , in fact, be hidden, due the fact that many patients receive some form of treatment (e.g., stool softeners, etc.). Postsurgical obstructive symptoms must be separated from HAEC because diarrhoea and enterocolitis may persist into the postoperative period (especially following extensive gastrointestinal (GI) involvement such as TCA).

The incidence of HAEC may also be influenced by obstruction due to the presence of stenosis or cuff strictures. Constipation and obstructive symptoms may be related to a number of possible causes, which include a residual aganglionic segment, sphincter achalasia, associated dysganglionosis of the enteric nervous system, strictures, restrictive cuff following ERP, retained spur following Duhamel procedures, “acquired” aganglionosis, and other functional causes. Most large series include a small number of patients with incomplete resection of the aganglionic segment (namely, 2.2% for the Soave procedure; 3.6% and 3.8% for the Swenson and Rehbein techniques, respectively; and 1.2% following the Duhamel procedure), and repeat biopsies may be required to ascertain the status of the pullthrough segment.

In many cases of postsurgical obstructive symptoms, failure to identify a cause on routine clinical and pathologic investigations suggests some degree of sphincter achalasia due to the failure of the internal sphincter to relax. Sphincter achalasia may be difficult to treat, and previous attempts at repeated anal dilatation have had mixed success. We have found the topical application of a glyceryltrinitrate paste to be fairly effective, and it is a cheap alternative to the injection of botulin toxin advocated by some.

Although the majority of patients do well following the modified surgical techniques currently employed for treating HSCR, some patients experience some degree of lack of control on follow-up. The incidence of incontinence following the ERP technique appears to be low, although sufficient long-term follow-up is as yet not available to assess the long-term outcome of neonatal or the transanal ERP techniques. Intermittent soiling may be associated with constipation, diarrhea, and a faeculoma, thus appearing to be related to overflow rather than inadequate sphincters. Some may be attributed to ill-advised sphincterotomy (largely historical).

**Management of Special Problems**

Enterocolitis management

Avoid surgery in the acute stage of HAEC because it may result in an increased incidence of complications. Management of HAEC includes resuscitation, antibiotics, decompression (6-hourly), and colostomy when stabilized.

Total colonic aganglionosis

Total colonic aganglionosis poses special problems due to the length of the bowel involved. It is usually treated by means of an ileostomy, a subsequent anal pull-through procedure with preservation of some aganglionic bowel, and anastomosis by means of a long side-to-side anastomosis.

Other

Obstructive symptoms may occur in the postoperative phase with recurrent episodes of abdominal distention associated with watery diarrhea.

**Prognosis and Outcome**

Ethical Issues

• Hirschsprung’s disease is surgically correctable, and the majority of patients with the disease can live productive, satisfying lives.

• Physical growth and development generally approximate normal.

• Intellectual function is mostly good.

• Most HSCR patients (93%) achieve acceptable anorectal function, given sufficient time to adjust.

Long-term functional results are excellent in some, good in the majority, and poor in approximately 15–30%.

• Functional results depend on the length of aganglionosis, procedure performed, surgical complications, social circumstances, family support, and associated anomalies, among other factors.

• Psychological problems may be magnified in those with poor support systems.

• Ethical issues do arise in cases of very long aganglionic segments, leading to intestinal failure.

• Ethical issues may be pertinent in certain disabling associated anomalies.

• Genetic counselling (if not handled correctly) also has the potential for giving rise to ethical issues, but should not be handled in isolation.

***Anorectal malformations***

**Introduction**

Anorectal malformations (ARMs) occur commonly throughout the world. Even under the best of circumstances, children undergoing operative treatment for ARMs may have lifelong bowel management problems of constipation, incontinence, and encopresis.

**Pathophysiology**

A description of the embryological events resulting in ARMs is beyond the scope of this chapter. ARMs occur in an estimated 1 in 4,000 births worldwide, but this estimate is based on data from more developed countries where accurate birth records are available. To the paediatric surgeon sitting in a grossly overcrowded surgery clinic in a major African teaching hospital, the incidence of ARMs appears much higher. This, however, may be an artifactual observation because most children with ARMs are referred to these tertiary referral centres, whereas children with other less complicated operative problems are managed locally without referral.

The term “imperforate anus” has traditionally been used to describe all anorectal abnormalities in females and males. Although imperforateanus implies that the anus never opened anywhere, a purely blind anal pouch is actually rare. Usually the rectum has opened either onto the perineum or into the genitourinary tract. The spectrum of abnormalities is quite broad, and therefore this chapter uses the term “anorectal malformations” for all of these abnormalities with further clarification to describe the specific malformation. “Imperforate anus” then refers to the specific portion of any anomaly where the rectum does not open properly through the anal musculature.

**Diagnosis**

The diagnosis of ARM should be made during the newborn physical examination. Some children are not born in health care facilities, however, and the absence of an anus (Fig. 11.9) may not be appreciated by the family until hours or days after birth, when it is noted that the child’s abdomen is distending and the infant has not passed meconium.

When consulted for a newborn with ARM, surgeon should initially note the child’s gender and examine the perineum carefully to see whether there is any evidence of meconium from a perineal fistula or along a midline raphe or whether the child has a true bucket-handle deformity. If the child is male and there is no initial evidence of visible meconium, the child should be observed for 12 hours or more to see whether meconium appears in the perineum. During this time, gauze is placed over the penis so that the urine can be examined for evidence of meconium. If there is meconium in the urine, the diagnosis of a rectourinary tract fistula is made, and no further diagnostic procedures are indicated.

If, however, there is no meconium identifiable in the urine or on the perineum, an invertogram is performed because a very small percentage of children (more common in children with Down syndrome) will have a blind pouch without a fistula. The child is placed in a prone jackknife position with the buttocks higher than the rest of the body for at least 30 minutes. After that time, a radio-opaque object (drop of barium or coin) is placed on the anal dimple, a cross-table lateral x-ray is performed, and the distance between the rectal air bubble and the perineal skin is measured. If the distance is less than 1 cm, the lesion can be treated as a low lesion, but any distance greater than 1 cm should be managed as a high lesion. In females, the labia should be grasped for traction and the posterior portion of the vestibule (just external to the hymen) examined for a rectovestibular fistula. In persistent cloaca, there is only one perineal opening, and therefore the separate openings for urethra, vagina, and rectum are not visible.

When the diagnosis of ARM is made, the child should be examined for other components of the VACTERL (Vertebral and spinal cord, Anorectal, Cardiac, TracheoEsophageal, Renal and other urinary tract, Limb) complex of anomalies. Diagnostic modalities must be appropriate for the particular locale. An orogastric tube is used to test for patency of the esophagus. An ultrasound, if available, is the best way to initially assess the urinary tract for abnormalities.

**Treatment**

Initial determination of the particular type of ARM will determine the proper initial treatment for the newborn. For many years, defects were classified as high, mid, or low. For the purpose of simplifying the treatment protocol for African paediatric surgeons, lesions in this chapter will be classified only as high or low. A colostomy is recommended as the initial treatment for high lesions, whereas low lesions can be treated primarily with an anoplasty. Low lesions that can be treated without a colostomy include those with evidence of meconium in the perineum or a bucket-handle lesion and those with a blind pouch less than 1 cm from the anal dimple, as demonstrated on invertogram. The most common lesions, including rectourinary tract fistulas in males and rectovestibular fistulas in females, should be treated as high lesions with an initial colostomy.

Alberto Peña has repeatedly emphasized the importance of a double-barrel colostomy to achieve total diversion of faeces. This is particularly important in males because there is usually a fistula between the distal colon and the urinary tract, and undiverted stool in the distal colon may cause repeated urinary tract infections (UTIs). It is best to place the colostomy in the distal descending or proximal sigmoid colon to make the distal limb shorter than it would be in a transverse colostomy. Many male children with ARMs have urine flow from the urethra into the distal colon and then out the mucous fistula (distal stoma), and if urine stays in the colon for long periods of time, it can cause a significant metabolic acidosis. However, if the distal sigmoid is mistakenly selected for the colostomy, the distal colon may be too tethered to properly come down to the perineum and may require taking down the colostomy at the time of anorectoplasty.

A recommended method for creation of a colostomy is to make a transverse muscle transecting incision in the left abdomen just below the level of the umbilicus. The sigmoid colon is identified and traced proximally and distally to be sure that it is truly the sigmoid and not the transverse colon. A point is selected in the proximal sigmoid area, and the colon is transected at this point. The proximal sigmoid is less likely to prolapse because the distal descending colon is fixed to the left lateral peritoneal reflection. After transecting the colon, it is quite important to irrigate the distal colon with warm normal saline to remove all meconium, taking care that the effluent does not get into the peritoneal cavity. If this meconium is not properly removed at the time of colostomy, the meconium will desiccate and form large impacted faecal rocks, which complicate subsequent anorectoplasty. In order to prevent prolapse, the two stomas are secured to the peritoneum and fascia at opposite ends of the wound by using a minimum of six small absorbable sutures (3/0-5/0 vicryl) for each stoma. Approximately 2 cm of each stoma should protrude past the skin level. There is no need to mature these small stomas because they will spontaneously mature, and attempts at operative maturation may occlude the lumen. One or two sutures (3-0 vicryl, polydiaxanone, nylon, silk) are used to approximate the peritoneum, fascia, and muscle between the stomas to minimize parastomal herniation.

A skin bridge is created between the stomas by using interrupted, rapidly absorbable small sutures. The bridge must be wide enough to totally separate the stomas. When the child reaches approximately 2–3 months of age, and if the child appears in very good nutritional status as evidenced by a weight of 8–10 kg, a definitive posterior sagittal anorectoplasty (PSARP) can be considered. In males, a distal colostogram should be performed prior to operation to determine the site of the fistula. This is performed by inserting a Foley catheter into the mucous fistula and inflating the balloon enough to occlude the colon lumen. The child is placed in a lateral position on the x-ray table with the hips flexed. Under fluoroscopy, if available, a water-soluble contrast medium is injected to fill the distal rectum and adequately identify the place of entry into the urinary tract. If fluoroscopy is not available, proper timing of injection with a plain film x-ray can usually adequately define the fistula.

**Posterior Sagittal Anorectoplasty Technique**

The technique of PSARP, popularized by Alberto Peña in the 1980s, is currently the most commonly utilized procedure for repairing ARMs. A urinary catheter is inserted before definitively positioning the patient. Sometimes the catheter goes through the fistula and into the rectum instead of into the urinary tract. If no urine is obtained from the catheter, the balloon should not be inflated in case it has curled up in the urethra. If the catheter is in the rectum, it can later be manipulated into the bladder. PSARP is performed with the child in a prone, jackknife position with all pressure points properly padded.

The insulation is removed for a distance of 1 cm from both ends of both pieces of the wire. The wire itself can be sterilized by soaking in an appropriate sterilizing solution. One end of both wires is connected to the stimulator. The other ends of the wire are left close to, but not touching, each other. This then serves as the handpiece used to touch the patient. Nonsterile personnel push the “continuous tetany” button on the stimulator whenever the surgeon wants to stimulate the patient with the handpiece to assess muscle contraction. It is important during PSARP operations that anaesthesia personnel not administer a muscle relaxant because this impairs the use of any muscle or nerve stimulator. If a muscle relaxant is absolutely necessary for intubation, it must be a very short-acting one because the muscle/nerve stimulator should be used early in the operation to define the usculature before any incision is made.

After mapping out the perineal musculature with a stimulator, temporary stay sutures are placed on either side of the midline at the anterior and posterior limits of the anal muscle complex (sphincter muscle) to identify the limits of the complex later in the operation if needed. A posterior sagittal incision is performed from the coccyx to the perineal body area, preferably with a needle-tip electrocautery placed on “cutting” current. After incising the skin, the deeper tissues can be incised by using the “coagulation” setting. The stimulator is used frequently to ensure that a true midline incision is being performed with equal musculature on either side. The incision is carried in the midline through the parasagittal fibres, the muscle complex, and the levator muscle.

Deep to the levator, the white appearance of the rectum should be visualized. Attempts at dissecting around the rectum at this time should be avoided. Distally, the rectum is opened longitudinally to view the inside of the rectal lumen. Anteriorly, the small fistula into the urinary tract is seen. Traction sutures are placed, and the anterior rectal wall is very carefully dissected from the posterior urethral wall. This is the most difficult part of the operation. After the initial dissection from the urethral wall itself, the dissection becomes easier above the prostate at the level of the bladder itself. The urinary fistula site should be carefully closed with an absorbable, fine suture. The rectum is then dissected circumferentially, staying close to the rectum itself so as to avoid damage to surrounding nerves, muscle, and urinary tract structures. Fibrotic neurovascular bands are taken down to further free the rectum. When the rectum is freed enough to come to the perianal skin without significant tension, the closure is performed. Significant tension will cause the neoanus to retract, resulting in lack of epithelial continuity with subsequent scarring and the need for another operation. The muscle anterior to the neoanus, including the perineal body, is closed with fine (4/0, 5/0 vicryl), absorbable suture. The colon is placed deep to the levator muscle, and the levator approximated in the midline, taking a bite of the posterior colon wall also.

**Bowel Management Programs**

Even under the best of circumstances, some children—particularly those with high ARMs—suffer from constipation or faecal incontinence caused by encopresis (escape of liquid stool around a large, hard faecal impaction). Bowel management programs have been initiated by hospitals in the United States to help children with these problems. The aim of these programs is to return children to normal lives at school in unsoiled underwear. The overriding principle behind bowel management is that the child learns to evacuate in a socially acceptable place (home) at a socially acceptable time (before or after school). The primary technique used to achieve continence is an enema regimen.

The child is given enemas each night or early in the morning at home to completely evacuate the colon. The child can then go to school with confidence, knowing that there will be no stool accidents. After a successful enema regimen has been established, an antegrade continent enema (ACE) operation (known also as an ACE procedure) can be considered. This procedure uses an appendicostomy as a conduit for administering antegrade enemas for evacuating the colon. This technique has been utilized with great success in achieving continence in children who were previously social recluses due to their bowel management problems.

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**Tests for initial level of knowledge**

1. In the newborn soon after birth secretion of foamed saliva from nose and mouth was observed, stomach increased in epigastric region, cyanosis and increase of dyspnea. When tried to catheterization naso-gastric tube didn’t go further. What’s the congenital pathology you are dealing with?
A. \*Atresia of the esophagus with fistula to the respiratory tract
B. Atresia of the duodenum
C. Atresia of the small intestine
D. Atresia of the esophagus is without fistula
E. Atresia of the colon

2. In a child of 2 months of age there was vomiting with fermented milk from 3-4 weeks, delayed stool, oliguria, visible peristalsis of the stomach. What additional diagnostic procedure is necessary to confirm the diagnosis of "pylorostenosis"?
A. Excretory urography
B. \*Ultrasound

C. Irrigography
D. Review X-ray
E. Thermography

3. The most informative method for diagnosing the Ladd syndrome is:
A. Review X-ray of the abdominal cavity organs
B. Ultrasonography
C. \*Irrigography

D. Chest X-ray
E. Bronchoscopy

4. At examination of the child in the delivery room, found that a child weighing 2800 grams, the stomach was bloated in the epigastric region, while a 30 ml yellowish color secretion was recieved from gastric tube. There was no meconium. On the review X-ray - absence of pneumatization of the intestine was observed, two levels of gas and fluid were found in the epigastrium. What pathology you are dealing with?
A. Atresia of the colon
B. Atresia of the esophagus
C. Atresia of the ileum
D. \*Atresia of the duodenum
E. Atresia of the rectum

5. A newborn girl with a body weight of 2800 g secreted 25 ml of green colored gastric juice through naso-gastral tube. There was no meconium, after enema; we received clean enemic water with a lump of grey couloured mucous; the abdomen is enlarged in the epigastric region and sunk down. After 2 hours there was vomiting with bile. The stomach has become sunken, with palpation soft is not painful. Peristalsis of intestines is not heard. On the review X-ray, the presence of two levels of fluid with gas, respectively, in the stomach and in the 12-intestine and lack of pneumatization in the lower intestine. In the irrigogram - location of colon was anatomically correctly located. What is the next tactic?
A. Ultrasonography

B. Dynamic observation
C. Conservative treatment
D. Consultation of the gastroenterologist
E. \*Transfer to a specialized children's surgery department

6. At the irrigogramm in a 3-year-old child with Hirschsprung's disease, there is a narrowing in the area of ​​the rectum and the sigmoid colon. What is the anatomical form of Hirschsprung's disease in this patient?
A. \*Rectosigmoidal
B. Rectal
C. Subtotal
D. Total
E. Segmental

7. In the newborn baby during the examination, immediately after birth, an absence of anus was discovered, the urine is sometimes clear, sometimes with meconium mixtures. What kind of anorectal defect?
A. Atresia of the anus, without fistula
B. \*Atresia of the anus with a fistula in the bladder
C. Atresia of the anus with a fistula in urethra
D. Atresia of the anus with a fistula at the entrance of vagina
E. Hirschsprung's disease

8. A child aged 1.5 years is having constipation from birth, abdominal distension, hypotrophy, stuffiness. The irrigograph shows narrowing in the region of the rectum with suprastenotic enlargement of the colon above this place. With anomanometry anorectal reflex is negative. Your diagnosis.
A. Dolikhosigma
B. Diaphragmal hernia
C. \*Hirschsprung's disease
D. Colonoptosis
E. Atresia of the colon

9. What are the tactics of the first stage of surgical treatment in the atresia of the anus with a fistula at the entrances of vagina?
A. Bougieing
B. Proctoplasty
C. Conservative treatment
D. \*Colostomy
E. Observation

10. In a newborn child with a severe form of Hirschsprung’s disease after enema, it was observed that abdomen was bloated, muscles of the anterior abdominal wall were tenseand noperistalsis of the intestineand onpercussion,and positiveperitoneal symptoms were seen. What is the cause of these symptoms?
A. Hepatic coma
B. Sepsis
C. Pneumothorax
D. Pneumomediastinum
E. \*Perforation of the intestine

***Keys for tests:***

**No 1.**

**A. \*Atresia of the esophagus with fistula to the respiratory tract.**The abundant secretion of the foamed saliva from the mouth and nose and cyanosis and the increasing dyspneais a characteristic symptom for esophageal atresia. Impossibility of catheterization of stomach confirms the presence of esophageal atresia. Bloatting of the abdomen in the epigastric region during the first minutes indicates the presence of fistula in the respiratory tract.

**B. Atresia of the duodenum.**Abundant secretion of foamed saliva from the mouth and nose and cyanosis with increasing dyspnea is not a characteristic symptom of atresia of the duodenum. In atresia of the duodenum, frequent vomiting is characteristic. The naso-gastral catheter freely passes into the stomach.

**C. Atresia of the small intestine**.
Abundant secretion of foamed saliva from the mouth and nose and cyanosis with increasing dyspnea is not a characteristic symptom of atresia of the small intestine. In atresia of the small intestine, vomiting is characteristic. The naso-gastral catheter freely passes into the stomach.

**D. Atresia of the esophagus is without fistula.**Abundant secretion of foamed saliva from the mouth and nose and cyanosis with increasing dyspnea is a characteristic symptom for esophageal atresia. Impossibility of catheterization of stomach confirms the presence of esophageal atresia. However, bloating in the epigastric region during the first minutes will never occur in non fistular form of esophageal atresia.

**E. Atresia of the colon**.
Abundant secretion of foamed saliva from the mouth and nose and cyanosis with increasing dyspnea is not a characteristic symptom of atresia of the colon. In atresia of the colon, vomiting is characteristic. The naso-gastral catheter freely passes into the stomach.

**No 2.**

**A. Excretory urography.**Excretory urography is informative only for the pathology of the urinary system, and does not support any symptoms characteristic of pylorostenosis

**B. \*Ultrasound.**To confirm the diagnosis of "pylorostenosis", the most informative and safe is ultrasonography

**C. Irrigography**.
Irrigography are informative only for the diagnosing the pathology of the colon, and does not confirm any symptoms characteristic of pylorostenosis

**D. Review X-ray.**With review X-ray of the abdominal cavity, it is possible to detect only a gas bubble of the stomach, which is not sufficient for the diagnosis of pylorostenosis. After that you need to conduct additional diagnostic procedures for final diagnosis.

**E. Thermography**.
Thermography does not confirm any symptoms characteristic of pylorostenosis

**No 3.**

**A. Review X-ray of the abdominal cavity organs**.
A review X-ray of the abdominal cavity organs provides information on the presence of two gas bladders suspected of one of the causes of high intestinal obstruction, but this is not sufficient for the diagnosis of the Ladd syndrome. After that, you need to conduct additional diagnostic procedures for final diagnosis.

**B. Ultrasonography**.
Ultrasonography, one can only suspect volvulus, which is not enough to diagnose the syndrome of the Ladd. After that you need to conduct additional diagnostic procedures for final diagnosis.

**C. \*Irrigography.**

Irrigography, which determines the high position of the intestine in the left hypochondrium, is the most informative method for diagnosing the syndrome of the Ladd syndrome.

**D. Chest X-ray**.
A review X-ray of the chest organs is not an informative method for the diagnosis of intestinal obstruction.

**E. Bronchoscopy.**

Bronchoscopy is not an informative method for diagnosing intestinal obstruction.

**No 4.
A. Atresia of the colon.**

In the atresia of the colon on the revive X-ray, multiple levels of gas and fluid are characteristic in all sections of the abdominal cavity. Catheterization of the stomach, you can get 10 ml of clear content. The abdomen is evenly bloated all over.

**B. Atresia of the esophagus**.
For the atresia of the esophagus, in the review X-ray it is not characteristic the presence of two levels of gas and fluid in the epigastrium, despite the absence of pneumatization of the intestine. Catheterization of the stomach is also impossible.

**C. Atresia of the ileum**.
At atresia of the ileum, in the review X-ray it is not characteristic the presence of more than two levels of gas and fluid in the upper abdominal cavity, and the absence of pneumatization of the intestine. Catheterization of the stomach, you can get 10-15 ml of contents with bile mixture. The stomach is asymmetrically bloated.

**D. \*Atresia of the duodenum.**

The presence of two levels of gas and liquid in the epigastrium, with the absence of pneumatization of the intestine, on the review X-ray is a characteristic feature of atresia of the duodenum. In addition, in the atresia of the duodenum, the stomach is bloated in the epigastric region, and catheterization of the stomach, you can get up to 30 ml secretion of yellowish color.

**E. Atresia of the rectum.**

In the atresia of the rectum on the revive X-ray, multiple levels of gas and fluid are characteristic in all sections of the abdominal cavity. Catheterization of the stomach, you can get 10 ml of clear content. Atresia of the rectum is confirmed by the impossibility of installing the catheter through the anus. The abdomen is evenly bloated all over.

**No 5.**

**A. Ultrasonography.**

All of the above symptoms indicate the presence of atresia of the duodenum. Ultrasonography will detect a possible accompanying pathology of the abdominal cavity and retroperitoneal space that does not affect tactics of further treatment, for transfer to a specialized children's surgical institution for further treatment

**B. Dynamic observation**.
All of the above symptoms indicate the presence of atresia of the duodenum. Dynamic observation of this pathology will lead to the appearance of terrible complications (hypotrophy, aspiration pneumonia, etc.), which will worsen the condition, and worsen the conditions for further surgical treatment

**C. Conservative treatment**.
All of the above symptoms indicate the presence of a duodenal atresia in a child that can be only treated surgically.

**D. Consultation of the gastroenterologist.**All of the above symptoms indicate the presence of atresia of the duodenum. Consultation of the gastroenterologist is inappropriate for the decision of the further treatment tactics.

**E. \*Transfer to a specialized children's surgery department.**

All of the above symptoms indicate the presence of atresia of the duodenum, requiring a transfer to a specialized children's surgical department for further treatment

**No 6.
A. \*Rectosigmoidal**.
The presence of narrowing in the region of the rectum and sigmoid gut in the irrigograph in a child with a Hirschprung's disease indicates its rectosigmoidal form.

**B. Rectal.**In the rectal form of the Hirschsprung's disease, there is a narrowing of colon only in the area of the rectum.

**C. Subtotal**.
In the subtotal form of the Hirschsprung’s disease, there is a narrowing of colon in the area above the rectum and sigmoid colon

**D. Total**.
In the total form of the Hirschsprung's disease, there is a narrowing of colon throughout the colon

**E. Segmental.**

In the segmental form of the Hirschprung's disease, there is a narrowing only in the area of a particular segment.

**No 7.**

**A. Atresia of the anus, without fistula**.
In the non-fistular form of the atresia of the anus, urine is always clean

**B. \*Atresia of the anus with a fistula in the bladder**.
Absence of an anal aperture, sometimes excretion of clean urine, sometimes with the mixture of meconium indicates atresia of the anus with fistula in the bladder.

**C. Atresia of the anus with a fistula in urethra.**At the atresia of the anus with fistula in the urinary tract, urine excretion will always be with mixture of meconium.

**D. Atresia of the anus with a fistula at the entrance of vagina**.
At the atresia of the anus with a fistula at the entrance of vagina, urine is always clean. It is also possible to detect fistula at the entrance of vagina.

**E. Hirschsprung's disease.**

In the Hirschsprung's disease, the anus is always in a natural place. Urine is always clean

**No 8.
A. Dolikhosigma.**

In the irrigography with dolikhosigma, there is no characteristic narrowing in the region of the rectum with suprastenotic enlargement of the colon above this place. With anomanometry anorectal reflex is positive. There will not be typical presence of hypotrophy and dyspnea.

**B. Diaphragmal hernia**.
At the irrigography with diaphragmatic hernia, there is no characteristic narrowing in the region of the rectum with suprastenotic enlargement of the colon above this place. With anomanometry anorectal reflex is positive.

**C. \*Hirschsprung's disease.**

The above-mentioned symptom is characteristic only for the Hirschsprung's disease

**D. Colonoptosis**.
Colonoptosis is manifested in adolescence. At the irrigography in colonoptosis, there is no characteristic narrowing in the region of the rectum with suprastenotic enlargement of the colon above this place. With anomanometry anorectal reflex is positive. There won’t be typical presence of hypotrophy and dyspnea.

**E. Atresia of the colon.**

Atresia of the colon is manifested during the first days of life. At the irrigogram there will be a "microcolon" till the level of atresia, above this place the contrast will not go.

**No 9.
A. Bougieing**

Bougieingof the atresia of the anus with a fistula at the entrance of vagina can be only a preparatory stage for the colostomy.

**B. Proctoplasty**.
Proctoplasty is the second stage of surgical treatment for atresia of the anus with a fistula at the entrance of vagina

**C. Conservative treatment**.
Treatment of the atresia of the anus with a fistula at the entrance of vagina is only surgical

**D. \*Colostomy.**

The first stage of surgical treatment in the atresia of the anus with a fistula at the entrance of vagina is the colostomy.

**E. Observation**.
Observation tactics in the treatment of atresia of the anus with a fistula at the entrance of vagina is inappropriate and not recommended.

**No 10.**

**A. Hepatic coma.**

These symptoms are not characteristic for the hepatic coma

**B. Sepsis**.
Sepsis in this situation may already be a consequence of septic complications of the Hirschsprung's disease (enterocolitis), or because of an undiagnosed perforation of the intestine.

**C. Pneumothorax**.
These symptoms are not characteristic for pneumothorax

**D. Pneumomediastinum.**These symptoms are not characteristic for pneumomediastinum

**E. \*Perforation of the intestine.**

The cause of these symptoms in this child is perforation of the intestine

**Tests for final level of knowledge**

1. The examination of newborn in obstretic room showed gastric catheter occlusion. Vertical review x-ray with contrast catheter shows the catheter turns back in “blind” end of esophagus. There is no gas inside the bowels and gastrum. What is a reason of this symptoms?

A. \*Аtresia of esophagal without fistula

B. Atresia of oesophagus with lower tracheo-esophagal fistula

C. Congenital stenosis of esophagus

D. Atresia of esophagus with upper and lower tracheo-esophagal fistula

E. Isolated tracheo-esophagus fistula

2. Two-month baby was approched to pediatric department with vomiting with fermented milk after each feeding and low body weight. Baby’s condition is serious with hypotrophy 2 stage. Abdomen palpitation was performed. “Sand-watch” was seen, gastric peristaltic is visible. Whats the possible diagnosis?

A. Atresia of esophagus

B. \*Pylorostenosis

C. Ring-shape pancreas

D. Gastric volvulus

E. Diaphragmatic hernia

3. Newborn girl with 2800.0 g body was examined. 25.0 ml of greenish fluid was received from gastric catheter. There was no meconium excretion. After enema clean water with gray coloured mucous lumps was excreted. The abdomen is enlarged in epigastrium and sinked in lower part. After 2 hours vomiting with bile occurred. The abdomen became sinked ,soft and painless during palpating. Bowels movement are not heard. Which diagnostic procedure should be performed for verification of diagnosis ?

A. Ultrasound examination

B. Irrigography

C. \*Review X-ray and irrigography

D. Repeat gastric catheterization

E. Neurosonography

4. Newborn was hospitalized on 3rd day with often vomiting with bile and general weakness. Examination showed enlarged asymmetric abdomen in epigastria. Grey meconium was excreted from the rectum. Chest and abdomen vertical X-ray shows two gas bubbles. Which the possible diagnosis you think?

A.Paralytic bowel obstruction

B.Pylorostenosis

C.Lower bowel obstruction

D. \*Duodenal obstruction

E.Pyleorospasm

5. The most effective treatment of membraneous atresia of duodenum is ?

A. Colostomy

B. Conservative treatment

C. Observation

D.Fibrogastroduodenoscopy

E. \*Duodenotomy and membrane excision

6. Two-weeks baby-boy was hospitalized with vomiting with green contents and with stains of blood, restless, swelling abdomen, defecation was absence, a little blood from the rectum. Newborn has tachycardia, tachypnoe and acrocianosis. Abdomen is asymmetric and “boat” form. Vertical abdominal X-ray shows two levels of fluid in upper parts of abdomen. Irrigography shows high position of appendix in a left epigastrium. Probable type of obstruction?

A. \*Ladd syndrome

B. Acute form of Hirschprung'sdisease

C. Atresia of ileum

D. Atresia of duodenum

E.Ring-shape pancreas

7. Newborn baby has vomiting with fecal contants. There was no excretion of gases or stool from anus. The abdomen is bloated and abdomen muscles are tensed. There is no bowel movements. Peritonial symptoms are positive. “Hepatic dullness” is not observed during percussion. Which dignostic precedure should be performed?

A. Abdominal ultrasound

B. \*Review X-ray of thoracic and abdominal cavityin vertical position

C. General blood test

D. ECG

E. Rectoromanoscopy

8. Newborn has acute form of Hirschprung's disease. Conservative treatment is not effective. Which operation should be performed?

A. Duamel operation

B. Swenson operation

C. \*Colostomy

D. Soave operation

E. Enterostomy

9. Newborn examination was performed in delivery room. Body weight is 2600.0 g. Normal abdomen. 10.0 ml of clean fluid was obtained during gastric catheterization. Meconium didn’t excrete. Catheter which was installed through anus went into 2 cm. X-ray shows pneumatization of bowel,levels of gas and fluid. What is a congenital disease?

A. Atresia of anus and rectum

B. Atresia of duodenum

C. Atresia of ileum

D. \*Atresia of rectum

E. Atresia of esophagus

10. In a newborn boy of 3 days old, with a weight of 2950 g, with Apgar score of 8-9 points, after 2 hours of birth, an abdominal enlargement with bloating was observed. Then the loops of the intestines with visible peristalsis are seen. There was no meconium till 48 hours. Anus was in its anatomical position. After the enema the meconium was hardly excreted. In irrigography,rectum and sigmoid colon are narrowed with suprastenotic expansion of all sections of the colon, colon is anatomically correctly placed. Your diagnosis.

A. Atresia of the anus and rectum

B. Atresia of the deodenum

C. Ring shaped pancreas

D. Malrotation of intestine

E. \*Hirschsprung's disease

***Keys for tests***

**No 1**

**A. \*Аtresia of esophagal without fistula.**

In the review X-ray , the contrast catheter turns back in the "blind" end of the esophagus, there is no gas in the stomach and intestine, which suggests the non fistular form of the esophageal atresia.

**B. Atresia of oesophagus with lower tracheo-esophagal fistula.**

Absence of gas in the stomach and intestine testifies to the non fistular form of atresia of the esophagus.

**C. Congenital stenosis of esophagus.**

In the congenital stenosis of the esophagus, the contrast catheter enters the stomach. There will be gas inside the bowels and gastrum.

**D. Atresia of esophagus with upper and lower tracheo-esophagal fistula.**

At the atresia of the esophagus with the upper and lower trachea, the esophageal fistula will be gas filling of the stomach and intestine. Contract catheter may get in trachea if the fistula is wide enough.

**E. Isolated tracheo-esophagus fistula.**

E.If the isolated trachea-esophageal fistula, the contrast catheter may get into the trachea (with a wide fistula).

**No 2.**

**A. Atresia of esophagus.**

The esophageal atresia manifests itself from the first hours after birth, when the foamed saliva discharges from the mouth and nose.

**B. \*Pylorostenosis.**

For pylorostenosis is characterized by the time of onset of the disease, vomiting with fermented milk, weight loss and apparent peristalsis of the stomach in the form of "sand watch"

**C.Ring-shape pancreas.**

A ring shaped pancreas causes upper intestinal obstruction, clinical manifestations of which appear from the first days of life. Apparent peristalsis of the stomach in the form of "sand watch" is not characteristic for this pathology.

**D. Gastric volvulus.**

The gastric volvulus is also manifested by upper intestinal obstruction, clinical manifestations of which appear from the first days of life. Apparent peristalsis of the stomach in the form of "sand watch" is not characteristic for this pathology.

**E. Diaphragmatic hernia.**

Diaphragmatic hernia is manifested by severe respiratory failure, the clinical manifestations of which appear from the first hours of life. At inspection typical boat-shaped squeezed stomach.

**No 3.**

**A. Ultrasound examination.**

Ultrasound as an independent method will not give a complete picture of obstruction but will help to exclude accompanying pathology

**B. Irrigography.**

Irrigography as an independent method will not give a complete picture of obstruction, so an recieved X-ray in the upright position is required.

**C. \*Review X-ray and irrigography.**

A review X-ray and irrigography will show two levels of fluid and gas (double bubble), and irrigography will show the wrong placement of the large intestine, indicating the syndrome of the Ladd.

**D. Repeating gastric catheterization.**

Repeating the catheterization of stomach does not help to exclude all possible causes of obstruction.

**E. Neurosonography.**

Neurosonography will determine the pathology of the brain. It’s not the diagnostic procedure for diagnosing intestinal obstruction

**No 4.**

**A.Paralytic bowel obstruction.**

Paralytic bowel obstruction occurs as a complication of peritonitis, or other generalised infections. There are no indications for the specified diseases. Bloating of the epigastric region of the abdomen is typical for high intestinal obstruction.

**B.Pylorostenosis.**

For pylorostenosis typical later on the onset of clinical manifestations (3rd week of life) A review X-ray in vertical postion will never have two levels of fluid and gas (double bubble).

**C.Lower bowel obstruction.**

For a low intestinal obstruction in the review X-ray, has multiple levels of liquid and gas (cloudbergs archs) will be characteristic.

**D. \*Duodenal obstruction.**

A review X-ray with two levels of fluid and gas (double bubble) indicates higher intestinal obstruction, the highest probability that is the obstruction will be at the level of duodenum.

**E. Pylorospasm.**

For pylorus spasm characteristic of vomiting gastric contents after each feeding, passage in the intestine is not disturbed.

**No 5.**

**A. Colostomy.**

Colostomy for duodenal membrane is not performed.

**B. Conservative treatment.**

Conservative treatment for deodenal membrane will not be successful as it requires surgery.

**C. Observation.**

Observation will only lead to loss of time may lead to lethal consiquences.

**D. Fibrogastroduodenoscopy.**

Fibrogastroduodenoscopy is a diagnostic procedure it’s not the treatment for duodenal obstruction.

**E. \*Duodenotomy and membrane excision.**

The operation of choice for membranous atresia of deudenum is a duodenotomy, excision of the membrane.

**No 6.**

**A. \*Ladd syndrome**

For the Ladd syndrome, vomiting is characterized by the content of green color with the stains of blood, significant anxiety, bloated stomach in the epigastric region, lack of independent defecation, small stains of blood from the rectum. Asymmetric stomach, boat-shaped. On the review X-ray of the abdominal cavity in an upright position with the Ledd syndrome, two levels of fluid in the upper abdominal cavity are determined. The irrigography determines the wrong placement of colon in the left hypochondrium.

**B. Acute form of Hirschprung's disease**

In the irrigography for the acute form of Hirshsprung's disease, the high location of the intestine in the left hypochondrium is not determined; it is characterized by narrowing of the site agangliosis with the transition to suprastenotic enlargement of the colon. Levels of fluid are not characteristic for acute form of Hirshprung's disease.

**C. Atresia of ileum**

For the atresia of ileum characterized by signs of lower intestinal obstruction, which starts later than upper intestinal obstruction, in rewiev X-ray we can see multiple international loops filled with liquid and gases .

**D.Atresia of duodenum**

For the atresia of the duodenum, a typical beginning of clinical manifestations in the first days of life. In the irrigography, the wrong placement of colon in the left hypochondrium is not seen.

**E.Ring-shape pancreas**

E. For the ring-shaped pancreas, a typical beginning of clinical manifestations in the early days of life. In the irrigography, the wrong placement of colon in the left hypochondrium is not seen.

**No 7.**

**A.Abdominal ultrasound**

Ultrasonic examination of the abdominal cavity is not informative diagnostic test for perforation if intestine.

**B. \*Review X-ray of thoracic and abdominal cavityin vertical position**

Review X-ray of thoracic and abdominal cavityin vertical position will reveal the presence of free gas under the diaphragmal domes it’s the most informative diagnostic test for perforation if intestine

**C. General blood test**

In the general analysis of blood, leukocytosis with a shift of the formula to the left is not informative diagnostic test for perforation of intestine.

**D. ECG**

ECG is not informative diagnostic test for perforation of intestine.

**E. Rectoromanoscopy**

Rectoromanoscopy is contraindicated if intestinal perforation is suspected.

**No 8.**

**A. Duamel operation**

Duhamel's operation is one of the variants for treatment of Hirschprung'sdisease is carried out routinely in the patients with typical manifestations of the Hirschsprung's disease.

**B. Swenson operation**

Svenson's operation is one of the variants for treatment of Hirschprung'sdisease is carried out routinely in the patients with typical manifestations of the Hirschsprung's disease.

**C. \*Colostomy**

Colostomy is an operation of choice in the acute form of Hirschsprung's disease.

**D. Soave operation**

Soave's operation is one of the variants for treatment of hirshprung disease is carried out routinely in the patients with typical manifestations of the Hirschsprung's disease.

**E. Enterostomy**

Enterostomy is shown only for the patients with total forms of Hirshprung disease.

**No 9.**

**A. Atresia of anus and rectum**

Atrasia of the anus and rectum appears at the primary examination in the delivery room, which is confirmed by the absence of an anal aperture.

**B. Atresia of duodenum**

For the atresia of the deodenum, an intrinsic pneumatization of the intestine throughout, but the rectum is passable

**C. Atresia of ileum**

Atresia of the ileum is typically pneumatisation of the intestines throughout, levels of gas and fluid, but the rectum is passable.

**D. \*Atresia of rectum**

Atresia of the rectum is confirmed by the impossibility of catheterization of the anus.

**E. Atresia of esophagus**

Atresia of the esophagus is manifested by signs of respiratory failure in the first place, and not by the phenomena of anorectal defect.

**No 10.**

**A. Atresia of the anus and rectum**

Atresia of the anus and rectum appears at the initial examination in the delivery room, which is confirmed by the absence of an anal aperture.

**B. Atresia of the deodenum**

For atresia of deodenum, a typical bloating in the epigastric region, boat shaped stomach. In irrigography - a microcolon.

**C. Ring shaped pancreas**

A ring shaped pancreas is manifested by high intestinal obstruction. Narrowing of the rectum and sigmoid gut with suprastenotic extension on irrigography is characteristic only in the case of Hirschsprung's disease.

**D. Malrotation of intestine**

In the irrigation with malrotation of the intestine, charecteristic is wrong wrong placement of colon in the upper parts of abdominal cavity in the area of duodenum.

**E. \*Hirschsprung's disease**

Symptoms of abdominal enlargement with bloating, contouring of loops of intestines with visible peristalsis, absence of meconium for more than 24 hours, difficulty of excretion of meconium after an enema, and narrowing of the rectum and sigmoid colon with suprastenotic enlargement in irrigography is pathognomonic symptom in the Hirschsprung's disease.