

Surgery in Africa - Monthly Review

Neonatal Surgical Emergencies

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A. Introduction

In countries where neonatal facilities and intensive care units are not available and limited numbers of pediatrics surgeons are available the burden of pediatric surgery in general and the challenging neonatal surgical problems as well falls on the hands of general surgeons who are practicing surgery in most difficult conditions.

In sub Saharan Africa the over all fertility rate is as high as 4 - 6%. This higher fertility rate is directly related to a higher prevalence of congenital anomaly. For surgeons engaged in primary pediatric surgical management particularly, knowledge of neonatal surgical emergencies helps to reduce morbidity and mortality as well reduces frustration. Full-text references and further readings can be found at the end of this Review and the interested reader is encouraged to access these.

B. Neonatal Physiologic Characteristics

Basic knowledge of neonatal metabolic changes and fluid and electrolyte management is a cornerstone in neonatal surgery, so it is worth discussing:

1. Water Metabolism

Water represents 70 and 80% of the body weight of the normal neonate and premature baby respectively. Total body water (TBW) varies inversely with fat content, and prematures have less fat deposits. TBW is distributed into extracellular fluid (ECF) and intracellular fluid (ICF) compartments. The ECF compartment is one-third the TBW with sodium as principal cation, and chloride and bicarbonate as anions. The ICF compartment is two-thirds the TBW with potassium the principal cation. The Newborn's metabolic rate is high and extra energy is needed for maintenance of body temperature and growth. A change in body water occurs upon entrance of the fetus to his new extrauterine environment.. There is a gradual decrease in body water and the extracellular fluid compartment with a concomitant increase

in the intracellular fluid compartment. This shift is interrupted with a premature birth. The newborn's body surface area is relatively much greater than the adults and heat loss is a major factor. Insensible water losses are from the lung (1/3) and skin (2/3). Transepithelial (skin) water loss is the major component and decreases as the newborn ages. Insensible water loss is affected by gestational age, body temperature (radiant warmers), and phototherapy. Neonatal renal function is generally adequate to meet the needs of the normal full-term infant but may be limited during periods of stress. Renal characteristics of newborns are a low glomerular filtration rate and concentration ability (limited urea in medullary interstitium) which makes them less tolerant to dehydration. The neonate is metabolically active and production of solute to excrete in the urine is high. The kidney in the newborn can only concentrate to about 400 mOsm/L initially (500 - 600 mOsm/L in the full-term compared to 1200 mOsm/L for an adult), and therefore requires 2 - 4 cc/kg/hr urine production to clear the renal solute load. The older child needs about 1 - 2 cc/kg/hr and the adult 0.5 - 1 cc/kg/hr.

2. Fluid and Electrolytes Concepts

Cellular energy mediated active transport of electrolytes along membranes is the most important mechanism for achieving and maintaining normal volume and composition of fluid compartments. Infants can retain sodium but cannot excrete excessive sodium. Electrolyte requirements of the full-term neonate are: Sodium 2 - 3 meq/kg/day, potassium 1 - 2 meq/kg/day, chloride 3 - 5 meq/kg/day at a rate of fluid of 100 cc/kg/24 hrs for the first 10 kg of weight. As a rule of thumb, the daily fluid requirements can be approximated too:

- **prematures** 120 - 150 cc/kg/24 hrs
- **neonates** (term) 100 cc/kg/24 hrs
- **Infants >10kg** 1000 cc+ 50cc/kg/24 hrs.

Special needs of preterm babies' fluid therapy are: conservative approach, consider body weight changes, sodium balance and ECF tonicity. They are susceptible to both sodium loss and sodium and volume overloading. High intravenous therapy can lead to patent PDA, bronchopulmonary dysplasia, enterocolitis and intraventricular hemorrhage. Impaired ability to excrete a sodium load can be amplified with surgical stress (progressive renal retention of sodium). Estimations of daily fluid requirements should take into consideration:

- (1) urinary water losses,
- (2) gastrointestinal losses,
- (3) insensible water losses, and
- (4) surgical losses (drains).

Blood Volumes estimates of help during surgical blood loss are:

premature 85 - 100 cc/kg,
term 85 cc/kg, and infant 70 - 80 cc/kg.

The degree of dehydration can be measured by clinical parameters such as: body weight, tissue turgor, state of peripheral circulation, depression of fontanelles, dryness of the mouth and urine output. Intravenous nutrition is one of the major advances in neonatal surgery and will be required when it is obvious that the period of starvation will go beyond five days. Oral feeding is the best method and breast is the best source. A newborn infant requires 100 - 200 calories/kg/day for normal growth. This is increased during stress, cold, infection, surgery and trauma. Minimum daily requirements are 2 - 3 gm/kg of protein, 10 - 15 gm/kg of carbohydrate and small amount of essential fatty acids.

Neonatal Surgical Emergencies present as respiratory distress, intestinal obstruction, or abdominal wall defects

C. Respiratory Distress

1. Congenital Diaphragmatic Hernia (CDH)

1. Definition: Diaphragmatic hernia is a failure of diaphragmatic development.

2. Embryology: Formation of the diaphragm occurs at 8 to 10 weeks in the fetus. During this time, the intestines returning into the abdomen will enter the chest if the diaphragm is not formed (ie, persistent pleuroperitoneal canal) and will prevent normal lung development. Intestines are also malrotated and subject to possible midgut volvulus postoperatively.

3. Nature of defects:

- a) There is a left-side to right side side predominance of 4:1
- b) Most defects are posterolateral (Bochdalek's hernia)
- c) Anteromedial defects (Morgagni's hernia) are rare.
- d) Pulmonary hypoplasia occurs on the ipsilateral side and also commonly occurs on the contralateral side.

4. Clinical Presentation

- a) The patient presents with respiratory distress. If extreme hypoplasia is present, respiratory distress is present early after delivery.
- b) Most patients present with respiratory acidosis and metabolic acidosis secondary to hypoxia, hypercapnia and hypothermia.

5. Diagnosis

- a) **Physical examination:** On examination, the patient's abdomen is scaphoid.
- b) **Radiography:** Radiographs reveal a bubbly bowel pattern in the chest and a lack of normal intestinal gas shadow

6. Treatment

The timing of the operation is still debated, but currently, most surgeons recommend waiting until the pulmonary hypertension has resolved and evidenced by resolution of preductal and postductal shunting and weaning to minimal support care.

a) Preoperative Management

- 1) Planned resuscitation
 - a) The patient should not be ventilated by mask.
 - b) The patient should be kept on oxygen and prepared for immediate orotracheal intubation.
- 2) The patient should be kept warm to prevent hypothermia. Warming lamps should be used.
- 3) An orogastric tube should be placed.
- 4) Determine pH and blood gas, type and crossmatch. Cutdown should be performed as needed.
- 5) Intravenous bicarbonate (1 to 2 mEq/kg) is almost always needed.
- 6) Extracorporeal membrane oxygenation may be necessary if blood gas values cannot be restored to near normal.

b) Operative Management

The CDH is repaired transabdominally through the subcostal incision in most major centres.

- 1) The bowel is reduced, a chest tube inserted, the defect is repaired, and a gastrostomy is performed.
- 2) The small hypoplastic lung is not expanded.

c) Postoperative Management

- 1) Ventilatory support at the lowest possible pressure is usually needed but should be discontinued as soon as possible.
- 2) The chest tube should be kept at underwater seal and at 2 cm H₂O suction.
- 3) The chest tube can usually be removed by the fifth post surgical day, if the patient is off ventilatory support.
- 4) pH and blood gas levels should be determined frequently.

7. Mortality

- a) When symptoms of diaphragmatic hernia present early (ie. less than 12 hours after birth), the mortality rate is 50%.
- b) When symptom onset is within 1 to 2 hours after birth, the mortality rate is 90%.

2. Esophageal Atresia

1. Definition: Esophageal atresia (EA) with distal tracheo-esophageal fistula (TEF) is the most common congenital anomaly of the esophagus, followed by EA without TEF also known as pure esophageal atresia and pure TEF. Incidence is 1 in every 2500 live births.

2. Embryology: The trachea and esophagus initially begin as a ventral diverticulum of the foregut during the third intrauterine week of life. A proliferation of endodermal cells appears on the lateral aspect of this growing diverticulum. These cell masses will divide the foregut into trachea and esophageal tubes. Whether interruption of this normal event leads to tracheo-esophageal anomalies, or whether during tracheal growth, atresia of the esophagus results because of the fistulous fixation of the esophagus to the trachea remains to be proven. Thirty percent of infants with this syndrome are premature.

3. Clinical presentation:

Respiratory distress due to:

- 1) Clinical signs include:
 - a) Excessive salivation
 - b) Choking during feeding
- 2) Patients with esophageal atresia may also present with:
 - a) Recurrent aspiration / reflux pneumonia
 - b) Right upper lobe pneumonia and atelectasis. Contrast studies are rarely needed and of potential disaster (aspiration). Correct dehydration, acid-base disturbances, respiratory distress and decompress proximal esophageal pouch (Replodge tube).

4. Associated Anomalies:

Evaluate for associated conditions which occurs in 30 % of patients. Such as **VACTERL** association (3 or more):

- Vertebral anomalies (ie. hemivertebrae, spina bifida)
- Anal malformations (ie. imperforate anus)
- Cardiac malformations (ie. VSD, ASD, Tetralogy Fallot)

- Tracheo-Esophageal fistula (must be one of the associated conditions)
- Renal Deformities (ie. absent kidney, hypospadia, etc)
- Limb dysplasia

5. Diagnosis

- 1) Diagnosis confirmed by observing a coiling nasogastric tube in a proximal pouch on radiograph.
- 2) Barium/water soluble gastrgraphin preferably / (0.5 ml) may be *carefully* instilled in the proximal pouch, only if there is diagnostic dilemma. During this procedure, we recommend the child to be accompanied by the doctor to the x-ray room.
- 3) Radiographs should include the neck and abdomen
 - a) If air is seen in the intestine, the diagnosis is esophageal atresia with distal tracheoesophageal fistula is the most likely diagnosis.

6. Treatment

1) Peri-operative Management

- a) If present, right upper lobe pneumonia and atelectasis should be corrected with antibiotics before surgery.
- b) The baby should be kept in a reverse Trendelenburg's position.

2) Operative Management

A gastrostomy should be performed as soon as possible

- a) Vigorous chest physical therapy and suctioning should be performed.
 1. The gastrostomy tube should be placed to gravity.
 2. Saliva should be suctioned from the blind proximal pouch either by continuous sump tube (Replegle) or by oral suctioning every 15 minutes.
- b) An extrapleural division and closure of the tracheoesophageal fistula with end-to-end anastomosis without undue tension should be performed. In certain types of tracheoesophageal fistula with esophageal atresia or in premature infants, a delayed anastomosis may be considered.

7. Outcome

Most important predictors of outcome if the patient presents immediately are: birth weight, severity of pulmonary dysfunction, and presence of major congenital cardiac disease

Complication after surgery: anastomotic leak, stricture, gastroesophageal reflux, tracheomalacia and recurrent TEF.

Increased survival: Is associated with improvements in perioperative care, meticulous surgical technique and aggressive treatment of associated anomalies.

D. Congenital Intestinal Obstruction

1. Logical Approach to Neonatal Intestinal Obstruction

1. Signs and Symptoms:

- a) Bilious vomiting is always abnormal.
- b) Abdominal distention (scaphoid abdomen possible).
- c) Delayed, scanty or no passage of meconium.
- d) Polyhydramnios in mother.
- e) Down's Syndrome
- f) Family history:
 - i) Hirschsprung's disease
 - ii) Diabetic mother
 - iii) Jejunal atresia

2. Plain roentgenograms of the abdomen

- a) Diagnostic in complete high intestinal obstruction - no gas in distal small bowel.
 - i) Double bubble in duodenal obstruction
 - ii) Few gas filled loops beyond duodenum indicates jejunal atresia.
- b) Many gas filled loops (requires 24 hours) indicates some form of low intestinal obstruction
 - i) Ileal atresia
 - ii) Meconium ileus (an unfortunate misnomer) - obstruction of the distal small intestine by thick undigested meconium.

- iii) Meconium plug syndrome - obstruction of colon by a plug of meconium.
- iv) Small left colon syndrome
- v) Hirschsprung's disease - congenital aganglionosis of colon starting from the rectum.
- vi) Colonic atresia.

- c) May be nonspecific in instances of malrotation of the intestines. This diagnosis must always be considered in neonates with unexplained bilious vomiting.
- d) Calcifications - at some time during fetal life meconium was (is) present in the abdomen.

3. Contrast enema will differentiate the various types of low intestinal obstruction.

- a) Microcolon - complete obstruction of the small bowel.
- b) Meconium plug syndrome - colon dilated proximal to an intraluminal mass .
- c) Hirschsprung's disease - although it may appear to be diagnostic, not reliable in the newborn. Proximal dilation with transitional zone and distally narrow segment.
- d) Small left colon syndrome - colon dilated to the splenic flexure, then becomes narrow.

4. Upper G.I. series - the procedure of choice in diagnosing malrotation of the intestines. In the past, a contrast enema was thought to be the disagnostic test of choice in instances of malrotation but the cecum and ascending colon can be in normal position in an infant or child with malrotation of the intestines.

Causes of Intestinal Obstruction in various age groups:

Newborn Causes: Intestinal atresias and stenoses (including imperforate anus and pyloric atresia), anomalies of rotation and fixation (including midgut volvulus), Hirschsprung's disease, meconium ileus, pseudocyst, meconium plug, abscess or adhesions from peritonitis, peritoneal bands, segmental volvulus, incarcerated hernia (inguinal, internal or diaphragmatic).

Infants to 24 months: Pyloric stenosis, incarcerated inguinal hernia, intussusception, Hirschsprung's disease, intestinal stenosis, congenital bands, duplications, cyst, omphalomesenteric duct remnant, internal hernia, midgut volvulus, trauma.

24 months or older: Incarcerated inguinal hernia, appendicitis, adhesions from prior surgery, duplications cysts, anomalies of rotation and fixation, trauma, granulomatous disease, tumors.

2. Intestinal Atresias

1. Duodenal atresia

- a) **Etiology:** The condition is probably the result of canalization failure.
- b) **Incidence:** Duodenal atresia is the most frequent type of intestinal obstruction, followed by jejunal atresia and then ileal atresia.
- c) **Associated Conditions:** High incidences of low birth weight (50%), Down's Syndrome (30%) and other major anomalies (30 - 50%) are associated with duodenal atresia. Most duodenal atresias are distal to the ampulla of Vater.
- d) **Clinical Presentation:** The patient usually presents with bilious vomiting shortly after birth.
- e) **Diagnosis:** Radiographs show a stomach and duodenum with a gasless abdomen ("double bubble"). If a delay in surgery is anticipated, it is imperative to differentiate from midgut volvulus, which requires immediate intervention.
- f) **Management:** Management is by duodenojejunostomy, duodenoduodenostomy, and occasionally, gastrostomy.

2. Ileal and Jejunal Atresia

- a) **Etiology:** Small bowel atresia is almost certainly the result of vascular occlusion (ie. intrauterine volvulus, intussusception) with aseptic necrosis and resorption of the gangrenous segment
- b) **Associated Conditions:** A high incidence of low birth weight (40%) and low incidence of other anomalies are noted. Small bowel atresia is associated with meconium ileus.
- c) **Clinical Presentation:** Patients present with bilious vomiting and abdominal distension.
- d) **Diagnosis:** Radiographs show many dilated loops with air-fluid levels. High jejunal atresia may present with a few air fluid level and no further gas beyond that.
- e) **Differential diagnosis** (*see above*)
- f) **Management:**
 - 1) Dehydration, along with acid-base and electrolyte imbalances, should be corrected.
 - 2) Laparotomy may be performed with resection of the proximal dilated end. End-to-end anastomosis is usually possible. The distal bowel should be evaluated for the additional atresia or stenosis by passage of a soft red rubber catheter or by injection of saline solution.

3. Colon Atresia

- a) **Incidence:** Rare

b) Management: Colostomy at the point of atresia

3. Meconium Ileus

1. Incidence: Meconium ileus accounts for almost one third of obstructions of the small intestine in neonates.

2. Epidemiology: The disorder occurs in approximately in 15% of infants with cystic fibrosis.

3. Clinical Presentation

- a) The diagnosis should be suspected in an infant who develops generalized abdominal distension, bilious vomiting, and failure to pass meconium in the first 24 to 48 hours after birth
- b) A family history of cystic fibrosis is not uncommon; a maternal history of polyhydramnios is present in 20 % of patients.
- c) The meconium may be palpable as a doughy substance in the dilated loops of distended bowel. The anus and rectum are typically narrow.

4. Imaging Studies:

- a) Plain abdominal radiograph demonstrates bowel loops of variable size.
 - i) Bowel contents have a soap-bubble appearance.
 - ii) Calcifications usually indicate meconium peritonitis resulting from an intrauterine intestinal perforation.
- b) A barium enema demonstrates a microcolon with inspissated meconium proximally.

5. Management:

- a) **Initial treatment:** Initial treatment is meglumine diatrizoate enemas and no surgery.
 - i) The patient should be intravenously hydrated.
 - ii) Under fluoroscopic control, a 50% solution of meglumine diatrizoate and water should be infused into the rectum and colon through a catheter.
 - iii) This procedure usually results in rapid passage of semi liquid meconium that continues during the next 24 to 48 hours.
 - iv) Multiple enemas may be required.
- b) **Surgical Treatment:** Surgery is indicated if:
 - i) the meglumine diatrizoate enemas do not relieve the obstruction.
 - ii) The infant appears too ill to delay operation.
 - iii) The diagnosis of meconium ileus is uncertain.
- c) **Post-surgical Management:**
 - i) All infants diagnosed with meconium ileus require an iontophoresis test to confirm a diagnosis of cystic fibrosis. This test is usually not practical before operation.
 - ii) All infants require vigorous postoperative pulmonary therapy.
 - iii) When oral feedings are begun, a pancreatic enzyme preparation is given with each feeding.

4. Malrotation

1. Etiology: The infant has compression of the second portion of the duodenum by Ladd's bands, which can potentially cause obstruction. For further information on the etiology, please see the: [Pediatric Surgery Handbook](#).

2. Incidence: Malrotation is a very common cause of intestinal obstruction in infants.

3. Clinical Presentation:

- a) Sudden onset of bilious emesis is the primary presenting sign; malrotation must be considered in every infant with bilious emesis.
- b) Abdominal distention is common but may be absent.
- c) Abdominal tenderness varies.
- d) On rectal examination, stool, if present, is guaiac positive.

4. Diagnosis

- a) Midgut volvulus is one of the most serious emergencies seen in these neonates or infants, and delay in diagnosis can result in loss of the entire midgut.
- b) Plain films of the abdomen are variable; a definitive diagnosis requires a contrast study.
 - 1. An upper gastrointestinal test is the preferred study and should be done in most cases.
 - 2. Occasionally, a barium enema is also helpful.
 - 3. These studies should be performed expeditiously because a few hours may be the difference between a totally reversible condition and loss of the entire midgut.

4. Contrast studies may be dispensed with in cases of shock or clear indication for exploration.

Surgical Management:

A right supraumbilical transverse incision is made so that the entire intestine can be delivered from the peritoneal cavity. Blood stained fluid indicates vascular compromise. Often chylous ascites is encountered, indicating lymphatic disruption from a volvulus. The colon is not immediately visible but is found at the base of the small bowel mesentery. The proximal duodenum is dilated and pass into the coils of the large bowel. The small intestine is often edematous and hemorrhagic and may appear frankly necrotic.

All volvuli are clockwise so that the small bowel must be rotated in *counterclockwise* fashion. One to three complete turns are necessary to reduce the volvulus and bring the transverse colon and the cecum into view. Peritoneal folds pass from the ascending colon to the duodenum and into the right lateral gutter. These folds, called **Ladd's band**, are incised along the medial aspect of the duodenum. Then the viability of the involved intestine and proper alignment of the intestine is checked.

Previously, it was recommended that the intestine be fixed by duodenal sutures to the right lower abdomen and colonic sutures to the left upper abdomen, but this has proved to be ineffective. The incidence of recurrent volvulus was 5% to 8%, whether or not fixation was accomplished.

Because of the abnormal location of the appendix in the left upper abdomen after widening of the mesentery, most surgeons perform an appendectomy.

Exploration and operative management can be carried out laparoscopically and by open approach but if there is any question about the completeness of the procedure done laparoscopically, conversion to an open procedure is in order.

The guiding principle when operating on patients with midgut volvulus with compromised bowel is rapid reduction of the volvulus and preservation of maximum length of intestine.

5. Hirschsprung's Disease (Congenital Aganglionic Megacolon)

a) Incidence: Hirschsprung's disease is a frequent cause of neonatal intestinal obstruction. It may also present during the first few years of life.

b) Etiology: The most common form is the absence of ganglion cells in the lower rectum.

- 1) This leads to ineffective conduction of peristalsis, resulting in a functional obstruction.
- 2) The aganglionic segment may extend more proximally and can involve the entire colon.

c) Clinical Presentation: Symptoms are nonspecific and may include episodic abdominal distension, diarrhea, and obstipation (which is not ordinarily seen in neonates) or constipation. Gash of air and/or stool comes out on withdrawal of rectal examining finger.

d) Diagnosis:

a) Radiological examination

- 1) A barium enema shows a narrow rectum with a dilated colon proximally. However, this finding is often absent in infants.
- 2) If the barium enema is normal and there is a high suspicion for Hirschsprung's disease, a plain radiograph of the abdomen should be obtained on the following day. Retained barium in the colon on this follow-up film is highly suggestive of Hirschsprung's disease.

b) Biopsy: The diagnosis is confirmed by rectal muscle biopsy (suction mucosal or full-thickness) showing an absence of ganglion cells in the submucosal plexus and hypertrophied nerve endings.

e) Management: Hirschsprung's disease may be initially managed with a temporary colostomy above the aganglionic segment, ie. leveling colostomy. If there is no facility for intra operative frozen section, it is good to do a right transverse colostomy. Recently, pediatric surgeons have performed primary pull-through procedures in the neonate.

6. Imperforate Anus

Classification: In both sexes, anorectal deformities are divided into *high, intermediate and low anomalies* as related to the level of the puborectalis portion of the levator ani muscle and whether there is a fistula to the urinary tract in male or the vagina in females. Cloacal anomalies, in which the urethra, vagina and the rectum all empty into a single conduit, are considered, as a separate category, because of the critical associated GU malformations.

The incidence of anorectal malformation is approximately 1 in 5000 live birth.

Initial management of the newborn with imperforate anus:

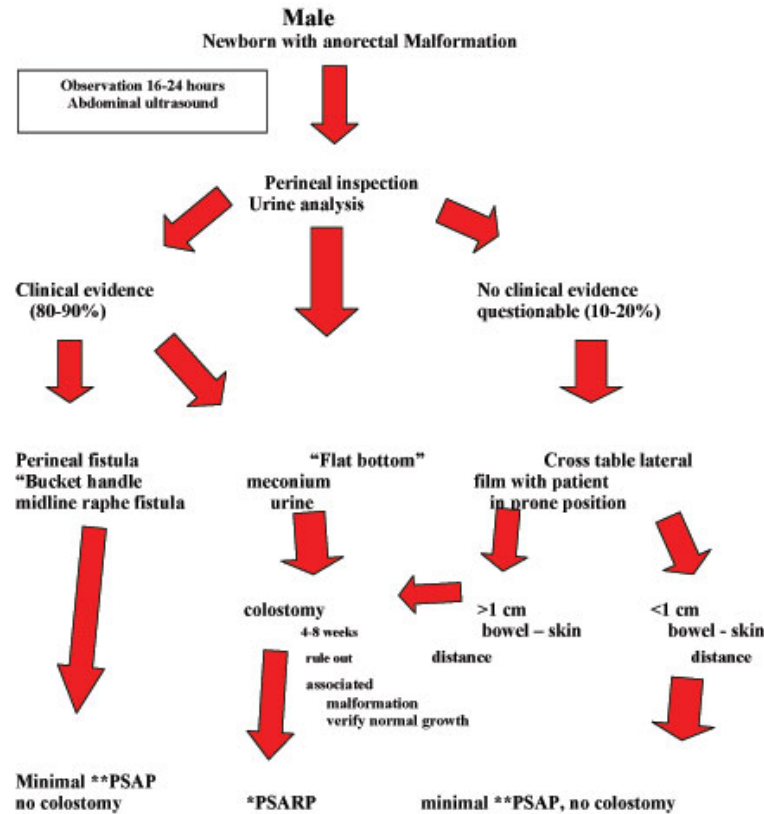
The first decision in the assessment of the newborn with an anorectal anomaly is whether a colostomy is required. Although it is always better to err on the side performing a colostomy, there are many anomalies in which one is unnecessary. The examination of the perineum is paramount because it may provide evidence of a low lying fistula or meconium beneath the membranous covering typical of a low lesion. In contrast , a flat or "rocker-bottom" perineum may be observed, which indicates poor sphincter or levator muscle development typical of a high anomaly. It is best to wait 24 hrs to allow progression of gas or meconium down close to or onto the perineum before finalizing the assessment. Of the female malformations, 95% are of the low variety, whereas most male anomalies are high.

Surgical management of imperforate anus:

1. Initial treatment in the neonate:

Male and female newborn who have low anomalies in the form of an anocutaneous fistula, ano vestibular fistula or anal stenosis usually can be treated initially with dilatation. In the past, anocutaneous fistulas regularly were dilated and repair performed several months later, but in more recent years, more pediatric centers have been performing anoplasty shortly after birth unless contraindicated by some other condition. Male and female infants suspected of intermediate or high-lying deformities and infants whose internal anatomy cannot be determined with certainty should have a colostomy performed. The most satisfactory approach is to perform colostomy at the junction between the descending colon and the sigmoid colon, leaving sufficient length to permit a subsequent pull-through procedure without having a take down the colostomy. Many surgeons believe that loop colostomy to be sufficient.

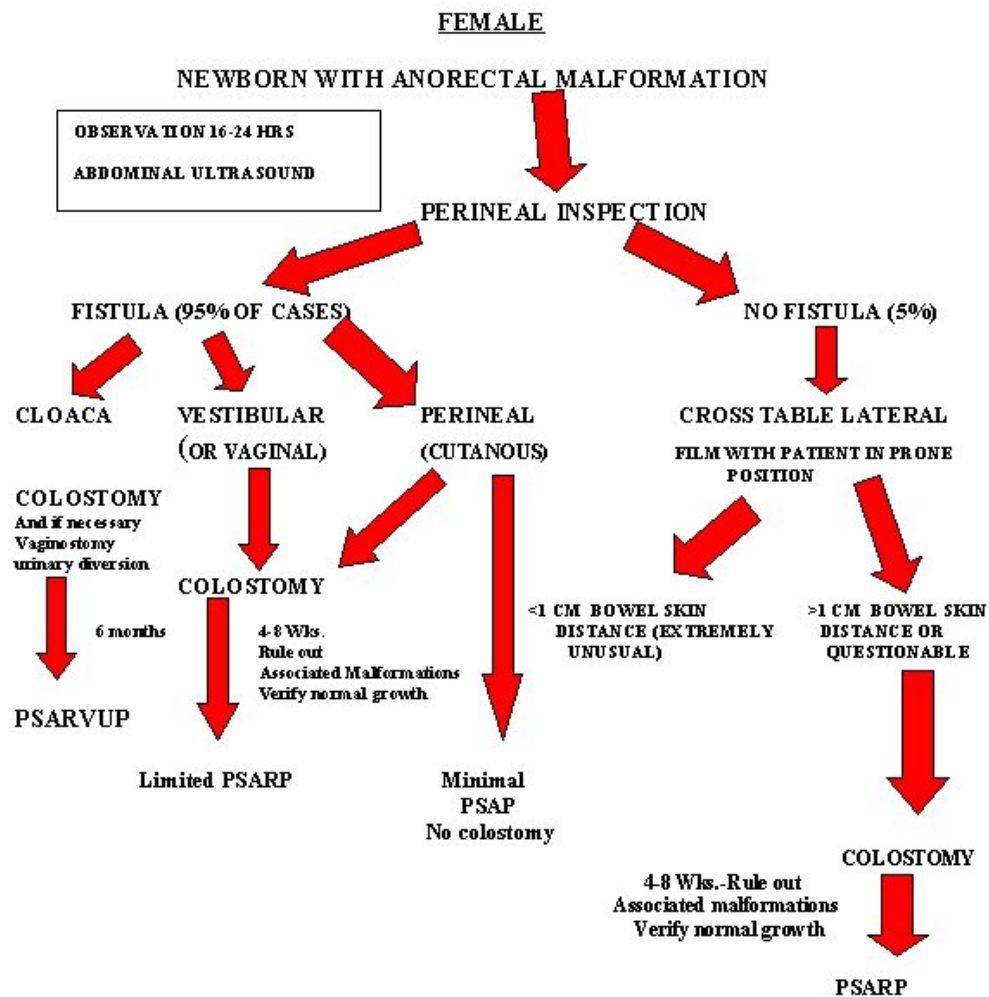
Algorithm for treatment of newborn boy with anorectal malformation



*PSARP :Posterior Saggital Ano Recto Plasty

**PSAP posterior Saggital Ano Plasty

Algorithm for treatment of newborn girl with anorectal malformation.



E. Abdominal Wall Defects

1. Omphalocele and Gastroschisis

a) Definition: Omphalocele is a covered defect of the umbilical ring into which abdominal contents have herniated. The defect is thought to occur in the third week of intrauterine life when the midgut elongates and resides in the yoke sac outside of the embryonic coelom.

Gastroschisis: Is a defect of the anterior abdominal wall just lateral to the umbilicus. The defect is almost always to the right of an intact umbilical cord and some cases are separated from the cord by an intact skin bridge. In contrast to omphalocele there is no peritoneal sac so that antenatal evisceration of the bowel occurs through a relatively small defect during intra uterine life. The irritating effect of the amniotic fluid (pH 7.0), which contains fetal urine and various growth factors, on the exposed bowel wall results in a chemical form of peritonitis characterized by a thick edematous membrane that is occasionally exudative.

b) Associated Anomalies: Associated anomalies should be ruled out, particularly in neonates with an omphalocele. The VACTERL constellation is often found in patients with omphalocele.

c) Management: Treatment begins immediately following delivery.

1. Medical Treatment:

- a) Hypothermia is usually the immediate life-threatening problem.
- b) Systemic intravenous antibiotics (ampicillin/gentamicin) are given to protect contaminated amnion and viscera. Infection can be devastating if a mesh closure is necessary.
- c) Intravenous hydration with balanced salt solution and colloid is essential.

2. Surgical Treatment - Omphalocele

- a) The sac or exposed intestines should be covered by a barrier-type dressing. A large circumferential dressing is applied last.
 - b) With gastroschisis in particular, it is essential that the bowel be supported, usually with the patient on his or her side and the bowel supported by towels, to prevent strangulation of the bowel and consequent bowel ischemia.
 - c) Gastrointestinal decompression by nasogastric tube is imperative to minimize further gastrointestinal distention and prevent aspiration of gastric contents.
 - d) Small defects (2 cm) can be managed by direct primary closure of the abdominal wall. Medium-sized defects are managed by careful removal of the sac at its base with suture ligation of the umbilical vein, the two umbilical arteries and the urachus. The liver and then the bowel are reduced into the abdomen. If the abdominal wall fascia cannot be approximated, skin closure with creation of a small ventral hernia may be used in some patients. Some may use prosthetic material to bridge the gap or staged-closure using a Dacron-reinforced silastic silo as a temporary locale for the bowel.
- Non-operative management using topical application of an escharotic (0.25% merbromin and 0.5% silver nitrate) agent is an alternative choice of treatment.

3. Surgical Management for Gastroschisis

The abdominal wall defect is enlarged 1 to 2 cm cephalad and caudad to improve the mechanical advantage for reducing the exposed viscera. Primary abdominal repair may not be possible in 40 - 50 % of cases. Too tight a closure may result in cardiorespiratory compromise from diaphragmatic elevation and ventilatory restriction, vena caval compression that reduces venous return, and diminished bowel perfusion leading to intestinal ischemia and necrosis. Some infants require a staged closure using a Dacron reinforced Silastic Silo as a temporary extra - abdominal housing.

As a result of prolonged adynamic ileus and exposure of the bowel to the amniotic fluid, it takes long for the bowel to commence normal peristalsis, necessitating, in almost all infants, TPN for adequate caloric support.

F. Hypertrophic Pyloric Stenosis

a) Definition: Pyloric stenosis is obstruction of the pyloric outlet of the stomach.

b) Epidemiology: Pyloric stenosis usually occurs in the first 3 to 6 weeks of life. It is extremely rare during the first week of life.

c) Clinical Presentation: Nonbilious vomiting (becoming projectile), cannot hold down water, leading to severe dehydration (metabolic alkalosis, decreased potassium and chloride ions). Serum pH is increased.

d) Diagnosis

1. Palpation of the pyloric olive: Contrary to the textbook description of its location in the right upper quadrant, the pyloric olive is more commonly found in the midline. If the pyloric olive can be felt, no further diagnostic tests are necessary.

- a) In an infant with a history that suggests pyloric stenosis, emptying the stomach with a nasogastric tube to make the olive easier to feel is recommended.
- b) Palpating the olive is impossible if the infant is crying.
 - i) Crying can be suppressed by giving the infant a pacifier or a small amount of oral electrolyte maintenance solution
 - ii) Patience on the part of the physician is important in this circumstance.

c) When the infant is not crying, the physician should stand at the infant's left side and hold up the baby's feet with his or her left hand to relax the infant's belly. The physician should then gently palpate the epigastrium with the extended middle finger of the right hand, being careful not to dig into the baby's abdomen.

2. **Barium Study** is used for the occasional infant in whom physical examination and ultrasound are not diagnostic.

Typical findings on barium study indicating pyloric stenosis are distended stomach with an elongated and narrowed pyloric canal - the "string" and the "double tract" sign.

3. **Ultrasonography** is the gold standard for diagnosis of pyloric stenosis.

a) If the history is strongly suggestive of pyloric stenosis but a mass is not palpable, an ultrasound is a good diagnostic test in experienced hands. The most commonly used criteria are a pyloric mass muscle thickness of 4 mm or more and pyloric channel length of 16 mm or more.

b) If pyloric stenosis is not the cause of vomiting, gastroesophageal reflux may also be diagnosed by ultrasound.

e) Treatment is surgical

1. Preoperative Management

Preoperative care is aimed at restoring fluid and electrolyte losses. A clinical assessment of the patient's hydration should be made, and serum electrolyte levels should be checked immediately on admission to rule out a serious hypokalemic, hypochloremic metabolic alkalosis. This should be corrected with appropriate potassium and chloride containing intravenous fluids before elective pyloromyotomy.

2. Pyloromyotomy

The Ramstedt-Fredet pyloromyotomy, performed with the infant under general anesthesia, is universally acceptable as the preferred operation. The stomach should be emptied again just before induction of anesthesia to minimize the risk of aspiration. A transverse skin incision followed by vertical splitting the right rectus muscle and fascia is used most frequently. The right upper abdominal transverse muscle splitting gridiron incision (Robertson) is used by many as well. Because it provides a better cosmetic appearance, a supraumbilical, curvilinear incision has been used by some surgeons.

After entering the abdomen, the omentum can be readily retrieved into the wound, which when elevated lifts the transverse colon and leads directly to the gastric antrum. The lower stomach may be elevated with gentle traction using moistened gauze to minimize slipping as the pyloric mass is delivered into the wound.

A vertical incision made on the mid anterior surface through the serosa and superficial muscularis extending from approximately 2 mm proximal to the pyloric vein (on an avascular area) to a point 0.5 cm onto the lower antrum. The circular muscle is opened bluntly using the Benson pyloromyotomy spreader or the back of the knife Handel. With completion of the myotomy, the gastric submucosa protrudes outwards, indicating relief of obstruction.

3. Postoperative Management

a) Feeding regimen:

i) The patient should be given nothing orally for 6 hours after surgery.

ii) Feeding can usually be initiated 6 to 8 hours postoperatively. Sugar water is generally given first, followed by formula or breast milk, using the following guidelines. This regimen can be advanced more rapidly or slowly depending on how the baby does;

a) Sugar Water, 30 mL every 2 hours, two times

b) Formula or breast milk

i) If sugar water is tolerated, the baby may be given half strength formula, 30 mL every 2 hours, two times. This is followed by full-strength formula every 4 hours at liberty.

ii) Breast milk may be substituted for formula but must be measured and fed by bottle.

iii) If the infant vomits, feedings should cease for 2 hours.

iv) All routine procedures (eg. taking vital signs, diaper changing, sponge bathing) should be completed before each feeding begins.

b) **Hospital Discharge:** Most infants may be discharged 24 hours after surgery.

c) **Surgical Complications:** Gastric and gastroesophageal reflux are the most common causes of persistent postoperative vomiting. Incomplete pyloromyotomy may be considered if vomiting continues for more than 7 - 10 days postoperatively, if it is forceful, and if it follows every feeding.

If the duodenum is inadvertently entered during the pyloromyotomy, and was noticed during the surgery, the perforated mucosa can be repaired or all layers should be closed and pyloromyotomy performed on other site, then the infant should remain on both nasogastric suction and intravenous antibiotics postoperatively for a minimum of 2 days.

G. Review Questions (Choose the single best answer for each question):

1. In Hirschsprung's disease, rectal biopsy typically shows which of the following patterns?

a) Absence of ganglion cells and nerve fibers

b) Absence of ganglion cells and presence of normal nerve fibers

- c) Absence of ganglion cells and presence of hypertrophied nerve fibers
- d) Presence of hypertrophied ganglion cells and normal nerve fibers

2. Which of the following statements concerning congenital esophageal atresia is FALSE?

- a) The lesion should be immediately suspected when a newborn infant aspirates the first feeding.
- b) In the most common form of the anomaly, air is absent from the gastrointestinal tract.
- c) More than 10% of affected infants have an imperforate anus.
- d) In affected infants who have pneumonia or cardiac difficulties, gastrostomy under local anesthesia is the initial surgery of choice.
- e) Stenosis at the site of anastomosis is the most common late complication.

3. A 19-day-old, full-term, previously healthy infant develops sudden onset of bilious vomiting at home. On examination in the emergency department, the infant appears ill. His abdomen is mildly tender but not distended, and he passes blood in his stool. The most likely diagnosis is:

- a) Pyloric stenosis
- b) Gastroenteritis
- c) Malrotation with midgut volvulus
- d) Necrotizing enterocolitis
- e) Jejunal atresia

4. A 26-day-old boy presents with a history of nonbilious vomiting. The child has lost 500 grams over the past week and appears clinically dehydrated. A palpable olive-sized mass is present in the mid-epigastrium. Laboratory data reveal serum sodium, 131 mEq/L; serum potassium, 2.8 mEq/L; serum chloride, 82 mEq/L; serum bicarbonate, 42 mEq/L; and a pH of 7.5. The most likely diagnosis is:

- a) Malrotation with a midgut volvulus
- b) Pyloric atresia
- c) Esophageal duplication
- d) Hypertrophic pyloric stenosis
- e) Appendicitis

5. The most appropriate maintenance fluid for a 9-kg infant is:

- a) 5% dextrose in 0.2 normal saline + 30 mEq/L of KCl at 36 mL per hour
- b) 5% dextrose in 0.5 normal saline + 30 mEq/L of KCl at 36 mL per hour
- c) Isolyte at 20 mL per hour
- d) 5% dextrose in 0.2 normal saline + 20 mEq/L of KCl at 45 mL per hour
- e) 5% dextrose in 0.5 normal saline + 20 mEq/L of KCl at 45 mL per hour

[Click here for correct answers](#)

Dr. Miliard Derbew
December 2005

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