PEDIATRIC SURGERY NOTES
LEVEL VI MBCHB
2019

COMPILED BY NAILA KAMADI

GLORY TO JESUS CHRIST

OUTLINE

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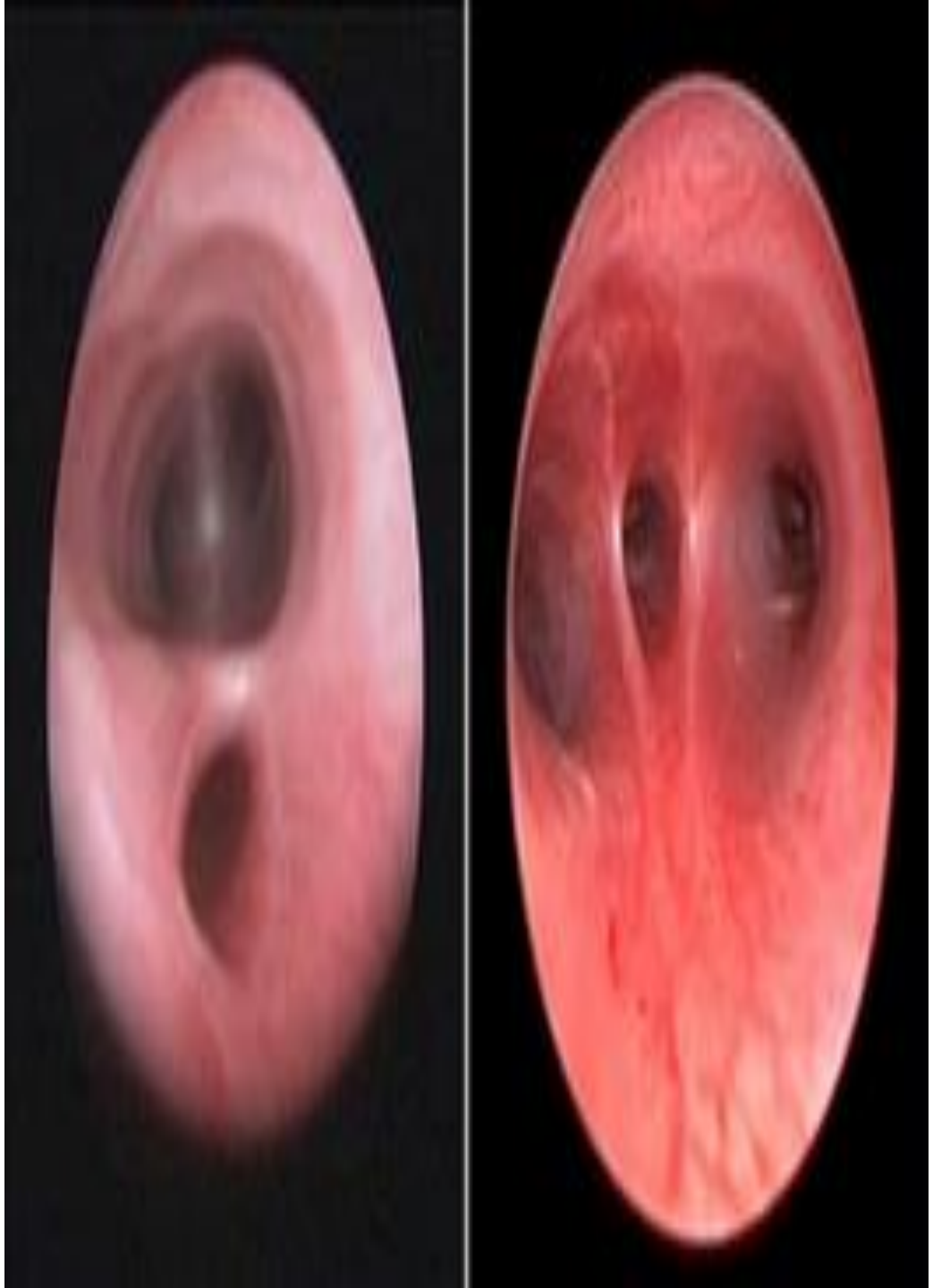


1. ESOPHAGEAL ATRESIA & TRACHEO – ESOPHAGEAL FISTULA (EA/ TEF)

BY: DR. MWIKA M. P.

TYPED BY NAILA KAMADI

Definition

- ✓ TEF is a ***congenital or acquired communication between the trachea & esophagus***.
- ✓ TEFs often lead to ***severe and fatal pulmonary complications***.
- ✓ Acquired TEFs occur secondary to:
  - ✓ Malignant disease, infection (esp. TB), ruptured diverticula, trauma, prolonged mechanical ventilation with an ETT or tracheostomy tube.

Embryology of congenital TEF

- ✓ Esophagus & trachea develop from the ***primitive foregut***.
- ✓ In a 4 – 6 week old embryo, the ***caudal part of the foregut forms a ventral diverticulum that evolves into the trachea***.
- ✓ Fusion of the longitudinal tracheoesophageal fold forms a ***tracheoesophageal septum*** that divides the foregut into a ventral laryngotracheal tube & a dorsal esophagus.
- ✓ ***Posterior deviation of the tracheoesophageal septum*** causes incomplete separation of the esophagus from the laryngotracheal tube resulting in a TEF.

Epidemiology

- ✓ Incidence: 1 in 2500 – 4,500 live births.
- ✓ 20 – 30% of neonates with TEF/EA are premature (BWT < 2000g) due to the following reasons:
  - ✓ Inability to swallow amniotic fluid → polyhydramnios that induces preterm labor.
  - ✓ Associated congenital anomalies may induce preterm labor
- ✓ M:F → 1.33 – 2.29:1; 60% higher prevalence in Caucasians; In Kenya, there is a high incidence in Kisii & Mombasa.

Cont.

- ✓ Although no definite cause exists for congenital TEF, an association with ***trisomies 18, 21 & 13*** has been reported.
- ✓ In addition, the use of ***decongestants that contain imidazoline derivatives*** by women during the 1st trimester of pregnancy has been linked to an ↑ risk of congenital TEFs.

Associations

- ✓ ***30 – 50%*** EA patients have associated congenital malformations.
- ✓ ~50% of all EA/TEF – associated anomalies are ***syndrome related, i.e.,***
  - ✓ Chromosomal
  - ✓ VACTERL
  - ✓ CHARGE
  - ✓ Fanconi anaemia
  - ✓ Opitz G/BBB syndrome
  - ✓ Goldenhar (Oculo – auriculo – vertebral, OAV) syndrome

VACTERL SYNDROME

- ✓ ***V***ertebral anomalies: ***Hemi – vertebra of the sacrum*** ***& accompanying spina bifida*** (MC)
- ✓ ***A***norectal malformations (ARMs)
- ✓ ***C***ardiac anomalies: ***PDA*** (MC; least serious & most subtle), also septal defects (VSD, ASD)
- ✓ ***T***racheo – ***E***sophageal Fistula
- ✓ ***R***enal malformations: ***VUR*** (MC); others are renal ectopia i.e., renal agenesis, horse shoe kidney
- ✓ ***L***imb anomalies: ***CTEV*** is the MC; Agenesis of the radial bone & fibula

Incidence of associated anomalies

Associated anomaly	Incidence (%)
Genitourinary	25 (MC)
Cardiovascular	~24
Gastrointestinal	~21
Musculoskeletal	~14
CNS	~7
VACTERL association	~20
Overall	50 – 70%

Gross – Vogt Anatomical classification of EA/TEF

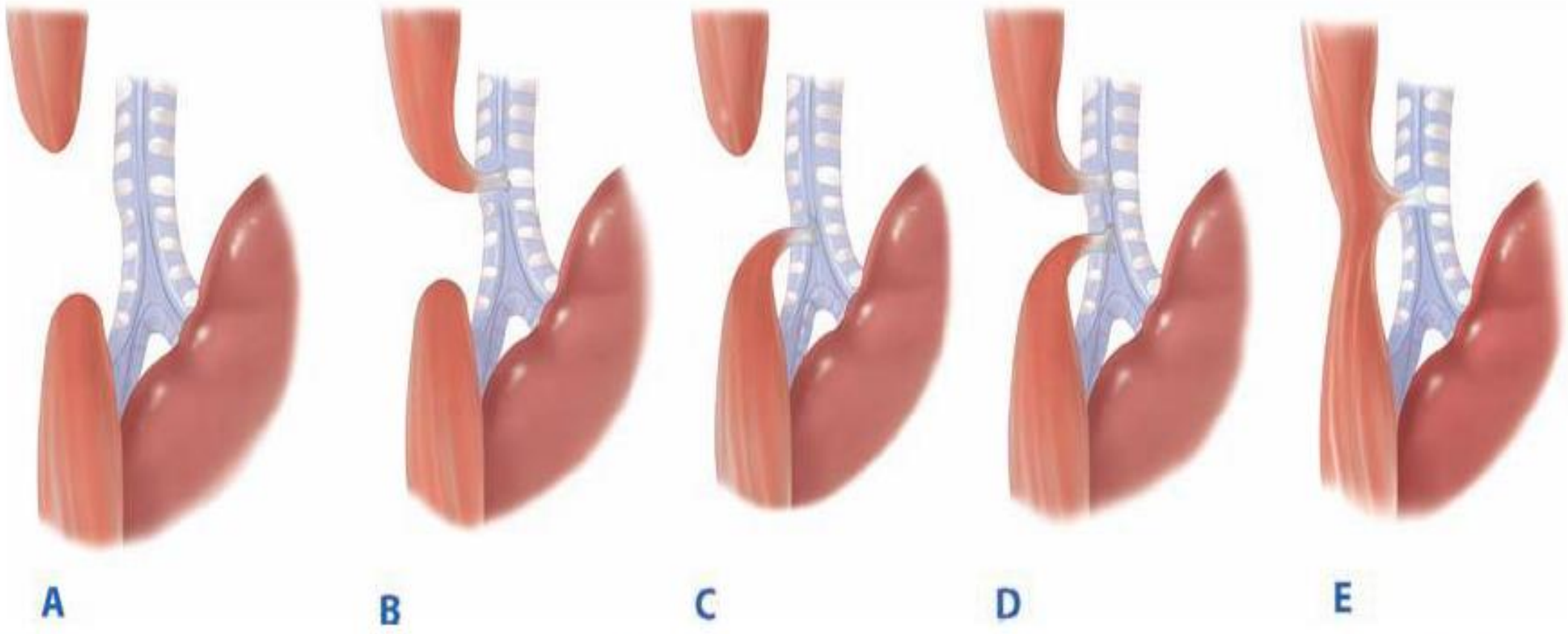
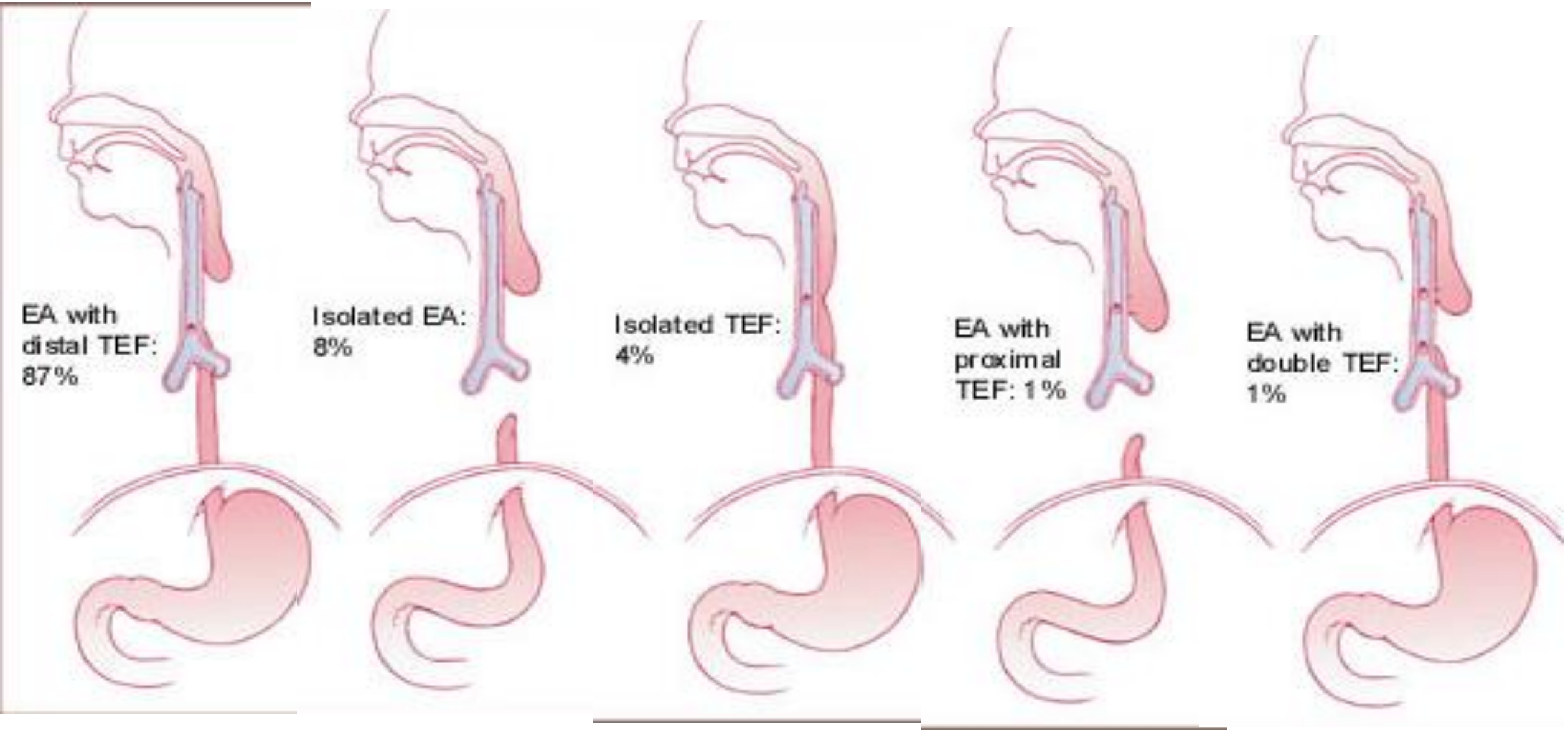


Figure 39-8. The five varieties of esophageal atresia and tracheoesophageal fistula. **A.** Isolated esophageal atresia. **B.** Esophageal atresia with tracheoesophageal fistula between proximal segment of esophagus and trachea. **C.** Esophageal atresia with tracheoesophageal fistula between distal esophagus and trachea. **D.** Esophageal atresia with fistula between both proximal and distal ends of esophagus and trachea. **E.** Tracheoesophageal fistula without esophageal atresia (H-type fistula).

Cont.

- ✓ The MC seen variety is ***EA with distal TEF (type C)***, which occurs in approximately ***85 – 87%*** of the cases.
- ✓ The next most frequent type is ***isolated EA without TEF (type A)***, occurring in ***8%*** of patients, followed by ***isolated/ H – type TEF without EA (type E)*** which occurs in ***4%*** of cases.
- ✓ ***EA with a double TEF between both proximal & distal ends of the esophagus & trachea (type D)*** is seen in approximately ***2%*** of cases & ***EA with a proximal TEF (type B)***, is seen in approximately ***1%*** of all cases.

Cont.


EA with distal TEF: 87%
Isolated EA: 8%
Isolated TEF: 4%
EA with proximal TEF: 1%
EA with double TEF: 1%

Clinical presentation

- ✓ Diagnosis is commonly made in the ***first 24h of life*** but it may be made either antenatally or later (delayed).
- ✓ Early diagnosis is key for better prognosis.
- ✓ By day 3 there are higher rates of liver injury due to starvation & increased activity of synthesis of repair proteins → this may progress to ***liver failure***.
- ✓ Delay in diagnosis may also allow for ***sepsis*** to develop.

Earliest signs:

- ✓ Copious, fine white frothy bubbles of mucus & saliva in the mouth & nose that recur despite suctioning.
- ✓ Rattling respiration
- ✓ Episodes of coughing/ choking in association with cyanosis. Symptoms worsen during feeding in the presence of a TEF.
- ✓ O/E: if a distal TEF is present: abdominal distention may occur secondary to collection of air in the stomach.

Cont.

- ✓ Other signs: cyanosis ± feeding, respiratory distress, inability to swallow, ***inability to pass a feeding/suction catheter through the mouth or nose into the stomach***.
- ✓ As the abdomen distends with air, the diaphragm is tamponaded worsening the pulmonary status.
- ✓ Aspiration of saliva from the upper pouch into the trachea further exacerbates the pulmonary compromise.
  - ✓ Atelectasis may occur → respiratory failure

Cont.

- ✓ In patients with EA/TEF, abnormal esophageal motility is always present because of abnormal development & innervation of the esophagus.
- ✓ Long term follow up studies have reported complications of: ***esophagitis***, ***Barrett esophagus*** & ***hiatal hernia***.

Differentials

- ✓ ***Pharyngeal pseudo diverticulum***: secondary to traumatic perforation of posterior pharynx from finger insertion into oropharynx during labor. Pneumo – mediastinum may develop.
- ✓ ***Tracheal agenesis***: present with respiratory distress; fatal in hrs. of birth; NGT can be inserted easily.
- ✓ ***Zenker diverticulum/ posterior hypo – pharyngeal diverticulum***

Complications

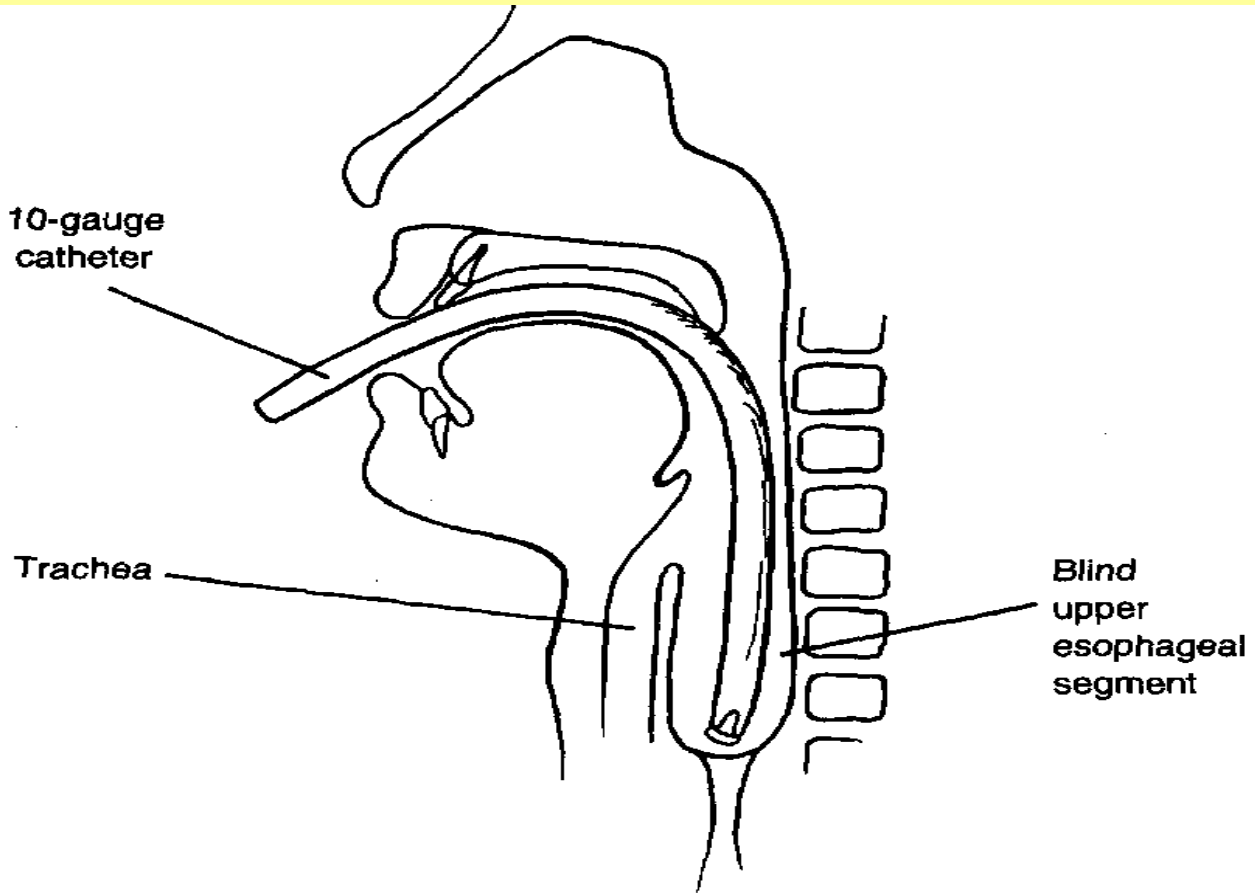
- ✓ Recurrent pneumonia
- ✓ Acute lung injury
- ✓ ARDS
- ✓ Lung abscess
- ✓ Poor nutrition
- ✓ Bronchiectasis from recurrent aspiration
- ✓ Respiratory failure
- ✓ Death

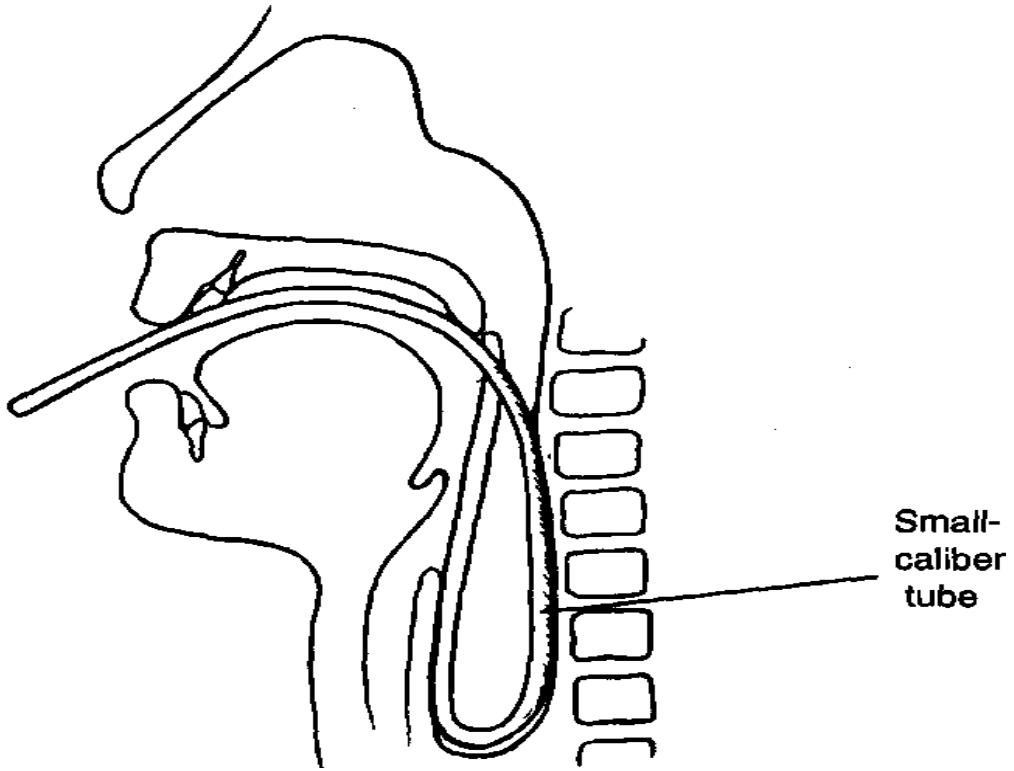
Prenatal diagnosis: Prenatal U/S

- ✓ U/S features of EA are identifiable on <10% of prenatal scans.
- ✓ Previous patient series reports show modest PPV of 20% and 50%.
- ✓ Non specific criteria:
  - ✓ Polyhydramnios
  - ✓ A distended esophageal pouch (mid – cervical)
  - ✓ Absent gastric bubble (in isolated esophageal atresia & esophageal atresia with a proximal pouch)
  - ✓ Absent fluid – filled stomach
  - ✓ Small abdomen
  - ✓ Lower than expected fetal weight

Post – natal diagnosis is clinical

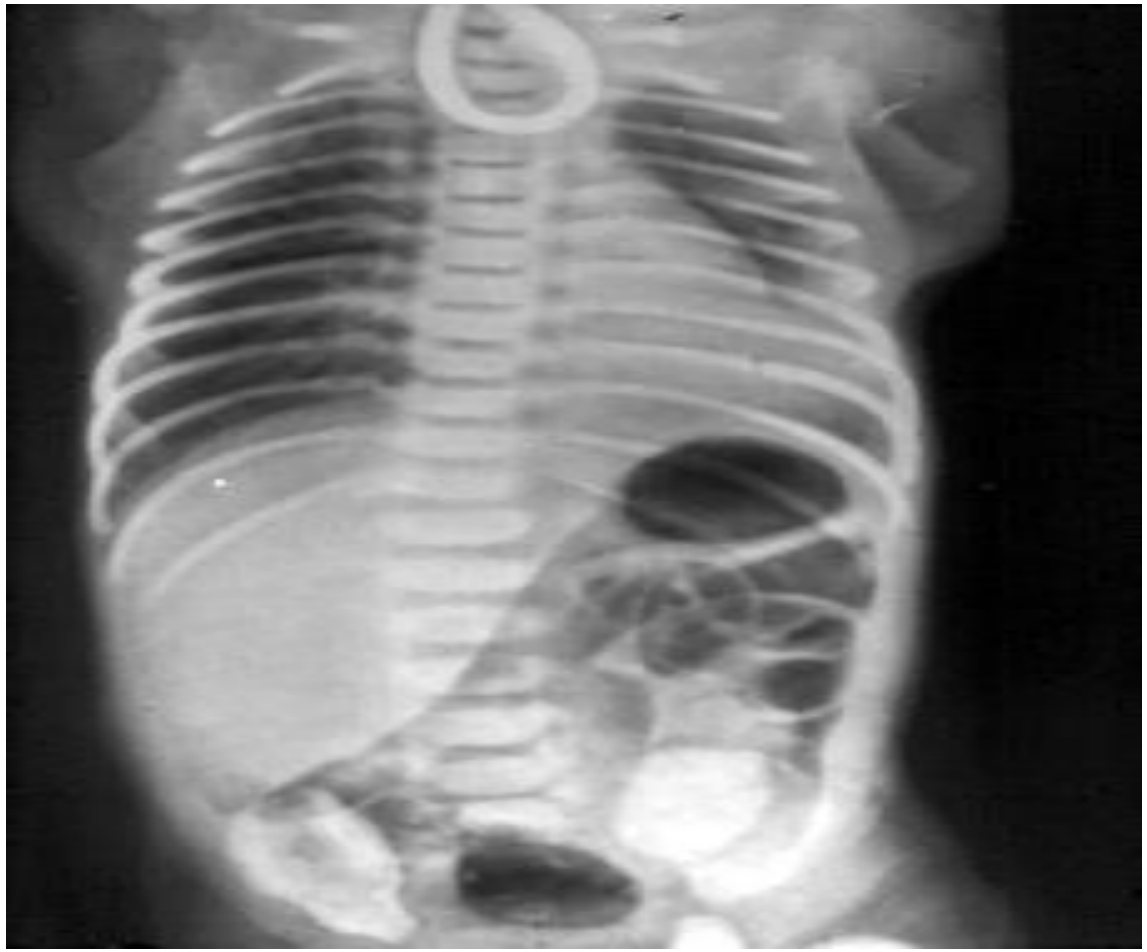
✓ Diagnosis of EA is confirmed when a ***10 gauge Fr catheter cannot be passed beyond 10cm from the gums***.





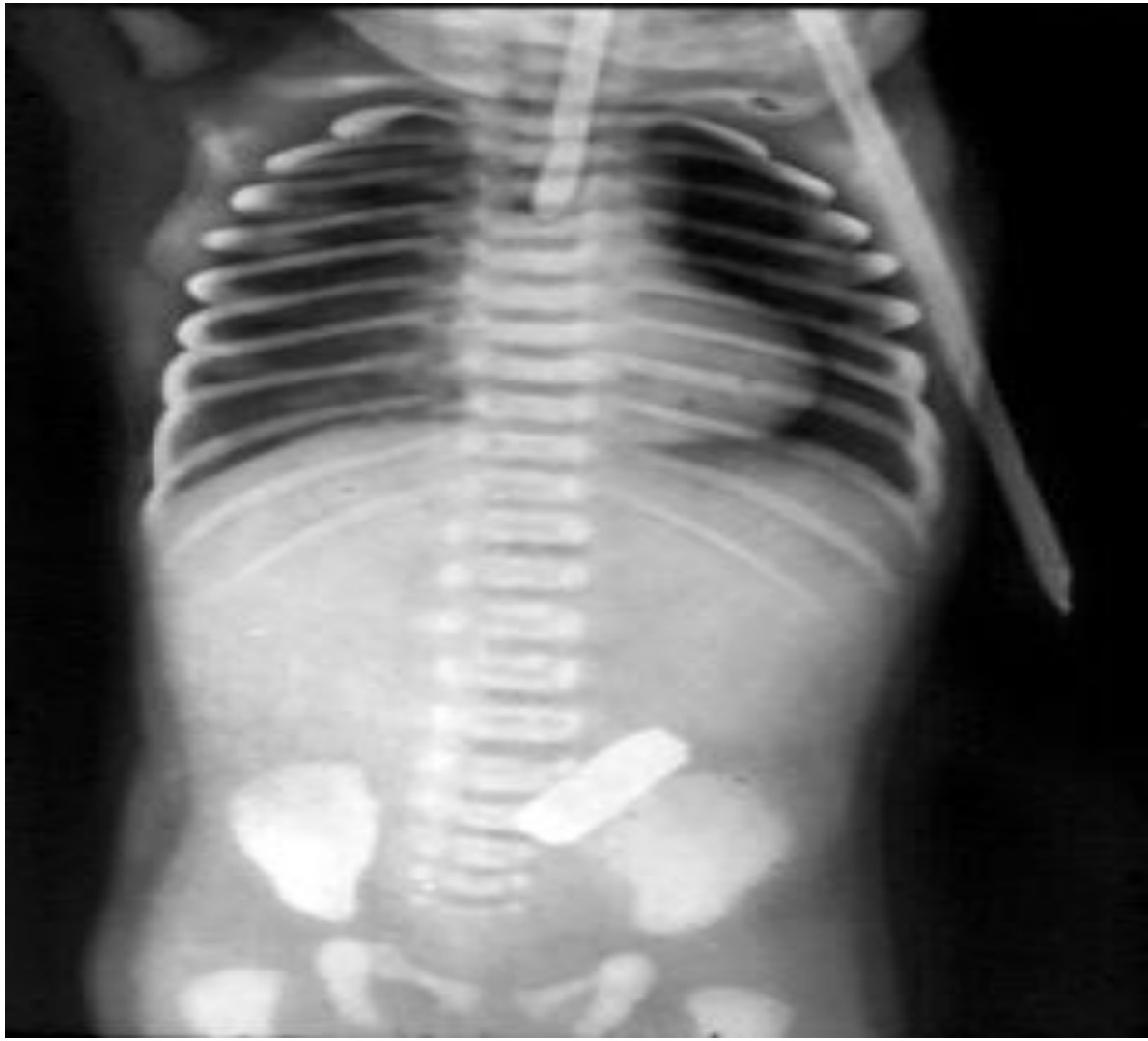
Post natal diagnosis: CXR

- ✓ A plain radiograph will confirm that the ***tube (should have a radio – opaque line) has not reached the stomach & has coiled in the mediastinum***. This indicates a concomitant EA.
- ✓ Gastric bubble is present indicated a distal TEF.



Gasless abdomen

- ✓ Seen in: ***pure atresia*** or a ***EA with a proximal TEF***.
- ✓ This is difficult to manage as the gap between the proximal and distal pouches is usually > 2 vertebrae.
- ✓ CXR finding absence of the gastric bubble.

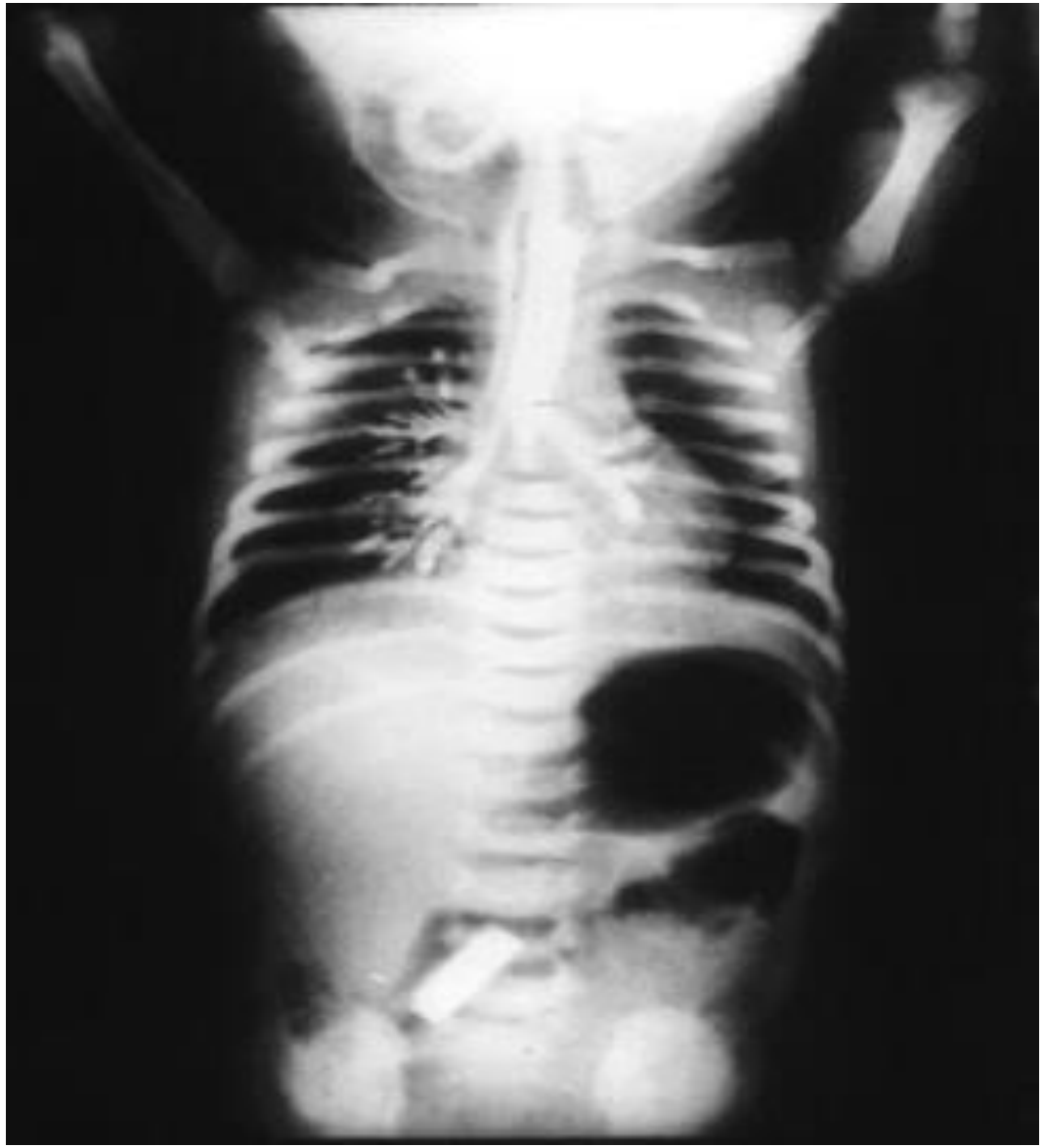


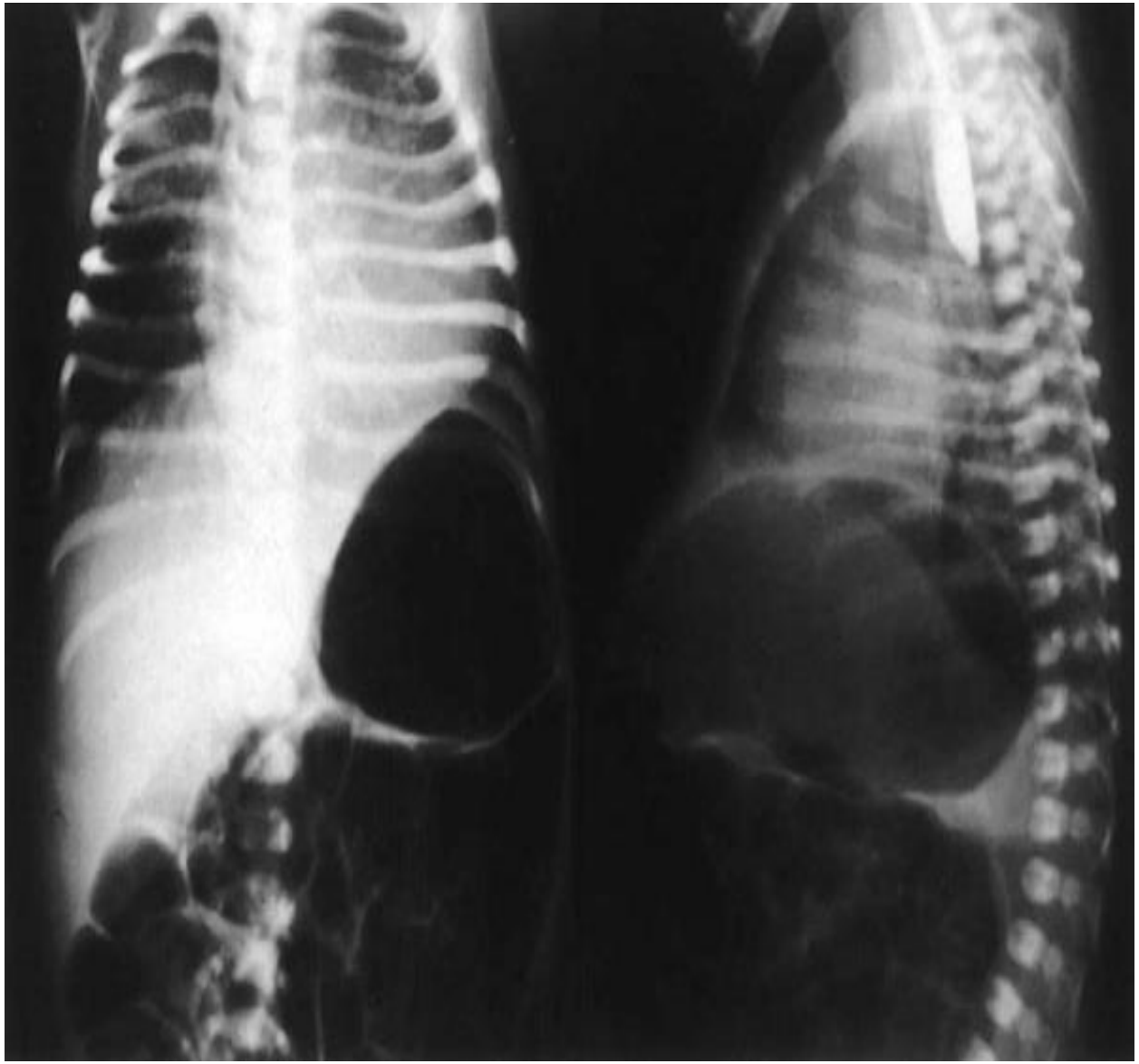
Other Ix

- ✓ Tracheo – bronchoscopy
- ✓ Additional testing: Assess for associated anomalies:
  - ✓ Echocardiography
  - ✓ Renal U/S
  - ✓ Chromosomal analysis

Contrast studies: ***DO NOT ORDER!***

- ✓ These should be performed by an experienced pediatric radiologist or after transfer to the tertiary institution & with the use of a ***small amount (0.5 – 1mL) of water soluble contrast***. ***Care must be taken to avoid aspiration***.





Preoperative treatment

- ✓ ***INITIAL MANAGEMENT***: Resuscitation & stabilization of respiratory status (with avoidance of endotracheal intubation);
- ✓ Nurse in NICU
- ✓ Suction the proximal esophageal pouch every 15 – 30 minutes using an 8F catheter
- ✓ Keep NPO
- ✓ Head elevation (30 – 40^0) or semi – prone positioning to minimize the risk of reflux or aspiration *via* the occult fistula of the distal trachea.

Cont.

- ✓ Monitoring of vital signs
- ✓ Establish vascular access & commence maintenance IVF: 10% Dextrose & hypotonic saline
  - ✓ This is for hydration & provision of caloric intake.
- ✓ Administer BSAs in patients who may have developed LRTIs.
- ✓ For patients known to have pneumonia or other pulmonary problems, a ***gastrostomy*** for gastric decompression may be required to prevent further reflux of gastric contents into the trachea. ***PPIs*** may be helpful.

Cont.

- ✓ Repair is delayed in patients with:
  - ✓ LBW
  - ✓ Pneumonia
  - ✓ Other major anomalies
- ✓ For the above, manage conservatively with parenteral nutrition, gastrotomy & upper pouch suction until they are considered low risk.
- ✓ Infants who have severe RDS may require the use of a ***Fogarty balloon catheter to obliterate the TEF while awaiting surgery***.

Operative repair

- ✓ Considered urgent but not an emergency.
- ✓ Suitable infant is: ***> 2.5kg, stable & without major anomalies***.
- ✓ The type of operative repair depends greatly on the type of anomaly & the presence of concomitant associated anomalies.
  - ✓ Short gap vs. long gap EA
  - ✓ Open vs. Thoracoscopic

Anesthetic considerations

✓ Minimize ventilation through the fistula: place a cuffed ETT distal to the fistula site.

This prevents:

✓ Gastric distention →

- → elevation of diaphragm → decreased tidal volumes → decreased VR → cardiovascular and respiratory compromise
- → aspiration of gastric contents

✓ Gastric perforation

✓ Tension pneumoperitoneum

Short gap EA/TEF

- ✓ Repair is performed via a ***right thoracotomy in the left lateral decubitus/ semi – prone position (with the right arm placed over the head) with the head of table elevated to avoid gastric reflux***.
- ✓ 4 – 5cm long curvilinear incision 1 cm below the angle of the scapula
- ✓ Extra – pleural dissection done postero – superiorly
- ✓ Identify & divide the azygos vein arch to expose the posterior mediastinum, the TEF & the Vagus nerve which are encountered beneath the azygos arch.

Cont.

- ✓ Procedure:
  - ✓ Closure of the fistula with fine non – absorbable sutures.
  - ✓ Correction of the EA:
    - The upper atresic esophageal pouch is identified by downward tension on oro – esophageal tube.
    - A traction suture is placed at the end of the pouch.
    - Blunt & sharp dissection to mobilize the proximal pouch to the level of the thoracic inlet.
    - A single – layer, end – to – end anastomosis with monofilament absorbable sutures

Long gap EA/ TEF

- ***A gap >3 cm or more than the height of 2 vertebral bodies is considered long***. Typically, early primary repair is not possible.
- There are currently no definitive standardized guidelines for the evaluation, management & surgical approach to treatment of LGEA.
- Management depends: on the ***gap length*** & the ***size & quality of the proximal & distal esophageal segments***, and ***any associated anomalies*** such as TEFs, strictures, duplications or cysts ***and vascular anomalies.***

Maneuvers for lengthening the esophagus in LGEA

- ✓ Non operative maneuvers (in combination with delayed primary anastomosis)
  - ✓ Spontaneous growth
  - ✓ Bougienage
    - Proximal
    - Proximal & distal
    - Magnetic

Operative measures using the native esophagus

Upper pouch mobilization

- Upper pouch mobilization → ***myotomy*** of the upper pouch → ***flap lengthening*** of the upper pouch → ***multi – staged extra – thoracic elongation*** of proximal pouch using thoracoscopy.

Lower pouch mobilization

- Lower pouch mobilization → ***myotomy*** of lower pouch → myotomy of the upper & lower pouch → ***traction sutures*** (Thoracoscopic placement) → ***transluminal thread*** with olives (Thoracoscopic assistance) → lower pouch ***hydrostatic distention*** → ***elongation of the lesser curvature***

Operative measures using esophageal replacement

- ✓ Colonic transplantation
- ✓ Stomach
  - ✓ Tube
  - ✓ Transposition (laparoscopy assisted)
- ✓ Jejunum
  - ✓ Pedicle graft
  - ✓ Free graft
- ✓ Ileum

Post – operatively

- ✓ Infant is ***ventilated*** as needed
- ✓ NGT or gastrostomy ***feedings*** are resumed
- ✓ POD 7: ***Contrast swallow radiographic examination*** is performed & If no leak is detected, ***oral feedings are resumed.***
- ✓ POD 21:***Esophagus is dilated*** up to a 24F size in order to prevent future esophageal stenosis.

Post – operative complications

Early:

- ***Pneumonia & atelectasis*** → respiratory failure (MC)
- ***Anastomotic leakage***
- ***Strictures***: common in patients who develop anastomotic leakage.
- ***Pneumothorax***
- ***Tracheal stenosis***

Late

- ***Recurrent fistula***: develop in patients who require continued post – operative intubation; rare.
- ***GERD and associated laryngospasm*** → periodic apneic spells
- ***Tracheomalacia***
- ***Vocal cord paresis/ paralysis***: more often in patients treated with Thoracoscopic repair.
- ***Esophageal motility disorders***

Prognostic classification systems:

Seek to stratify pre-operative risk guiding choice of the type of treatment for each case.

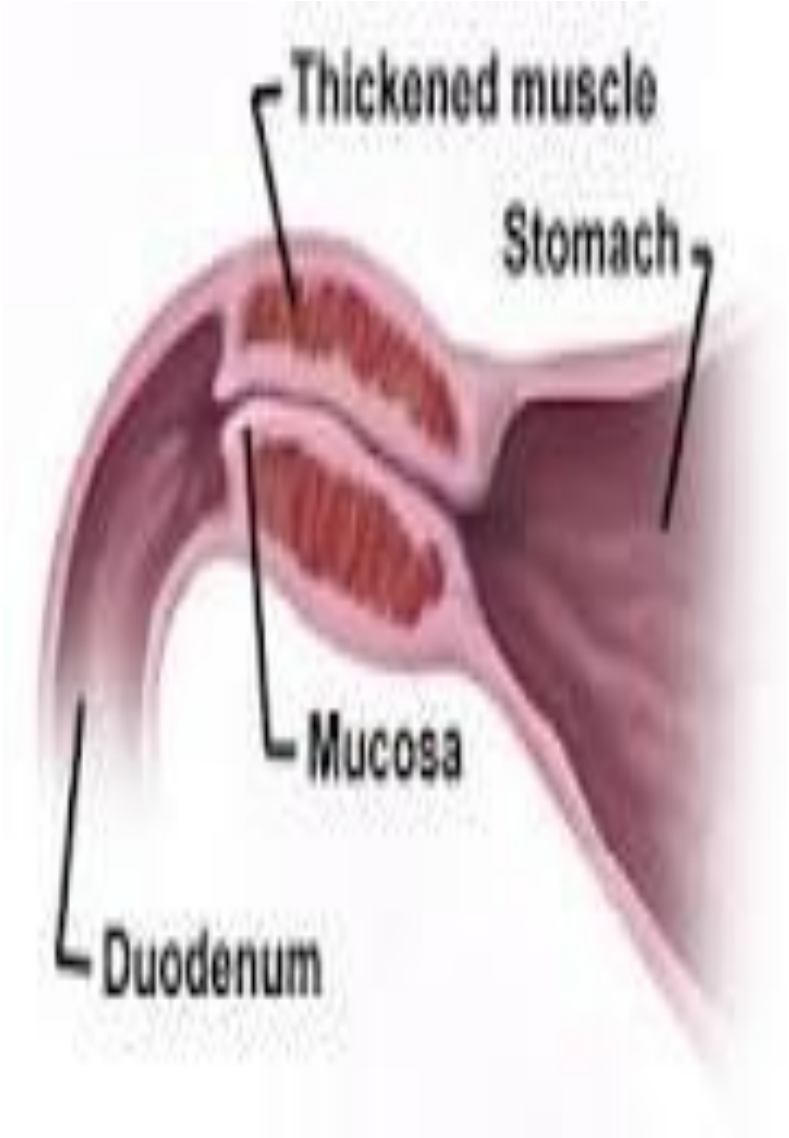
Group	Features	Prognosis
Waterston classification (1969)		
A	Birth weight >2.5kg and no co-morbidity	95% survival
B	Birth weight 1.8-2.5kg, pneumonia and congenital anomaly	68% survival
C	Birth weight <1.8kg, severe pneumonia and congenital anomaly	6% survival
Spitz classification (1994)		
I	Birth weight >1.5kg and no major cardiac anomaly	97% survival
II	Birth weight <1.5kg or major cardiac anomaly	59% survival
III	Birth weight <1.5kg and major cardiac anomaly	22% survival

Outcome

✓ Prone to develop GERD →

Strictures, Barret esophagus →

increased risk of esophageal cancer.



2. INFANTILE HYPERTROPHIC PYLORIC STENOSIS [IHPS]

EDITED BY NAILA KAMADI

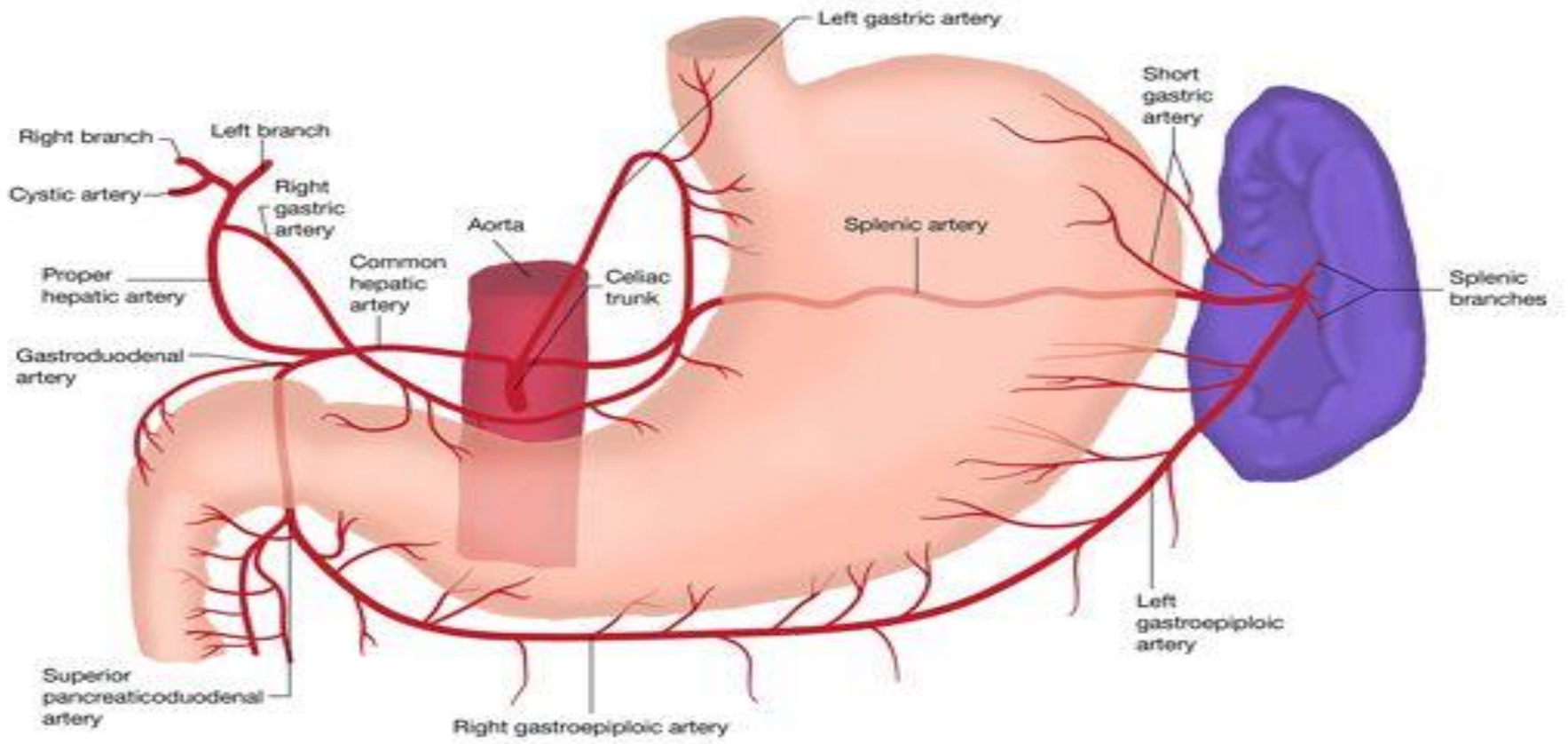
DEFINITION

- ✓ A condition characterized by ***hypertrophy of the 2 circular muscle layers*** of the pylorus resulting in ***constriction & obstruction of the gastric outlet***.

Incidence

- ✓ 1.5 – 4/1000 live births; ***M : F ratio → 4 – 6 : 1***
  - ❖ ~ 30% of cases are ***first born, male babies***.
- ✓ ***More in Caucasian (white, Northern European) infants***: Affects 1/500 Caucasian vs. 1/1000 Asian & African.
- ✓ 4 times MC in ***children whose mothers had pyloric stenosis***.
- ✓ Usually appears between ***3 – 6 weeks of age***: Rare after 3 months.
  - ❖ ***IT DOES NOT OCCUR AT BIRTH!***

Pylorus, “gate guard” (Gr)



Etiology

- ✓ Idiopathic
- ✓ Genetic
- ✓ Rarely autosomal dominant
- ✓ Positive familial history: 15 – 20 fold increase in risk; more common in multiple births.
- ✓ Ethnic origin (more in whites): ***more commonly seen in Caucasians***

Cont.

✓ Environmental:

❖ Drugs →

- ***Erythromycin*** or ***azithromycin*** exposure: Administered for pertussis post – exposure prophylaxis in neonates. They act as motilin agonists & hence result in ***strong, non – propagated contraction***. Pose a seven – fold increase in risk.
- ***IV administration of prostaglandins***

❖ Trans – pyloric feeding of premature babies.

❖ Formula feeding: controversial

Risk factors

- ✓ Environmental
  - ❖ Bottle, formula feeding
  - ❖ Maternal smoking
- ✓ Genetic: Genes encoding for Apolipoprotein 1
- ✓ Macrolide antibiotics

Emerging new theories

- ✓ GI hormones: ***gastrin, substance P*** → chronic *pylorospasm* & stenosis
- ✓ Epidermal growth factor, deficiency of NO (can induce muscle spasm preventing smooth muscle relaxation in the stomach)
- ✓ Muscle layer deficient in: quantity of nerve terminals

Associations

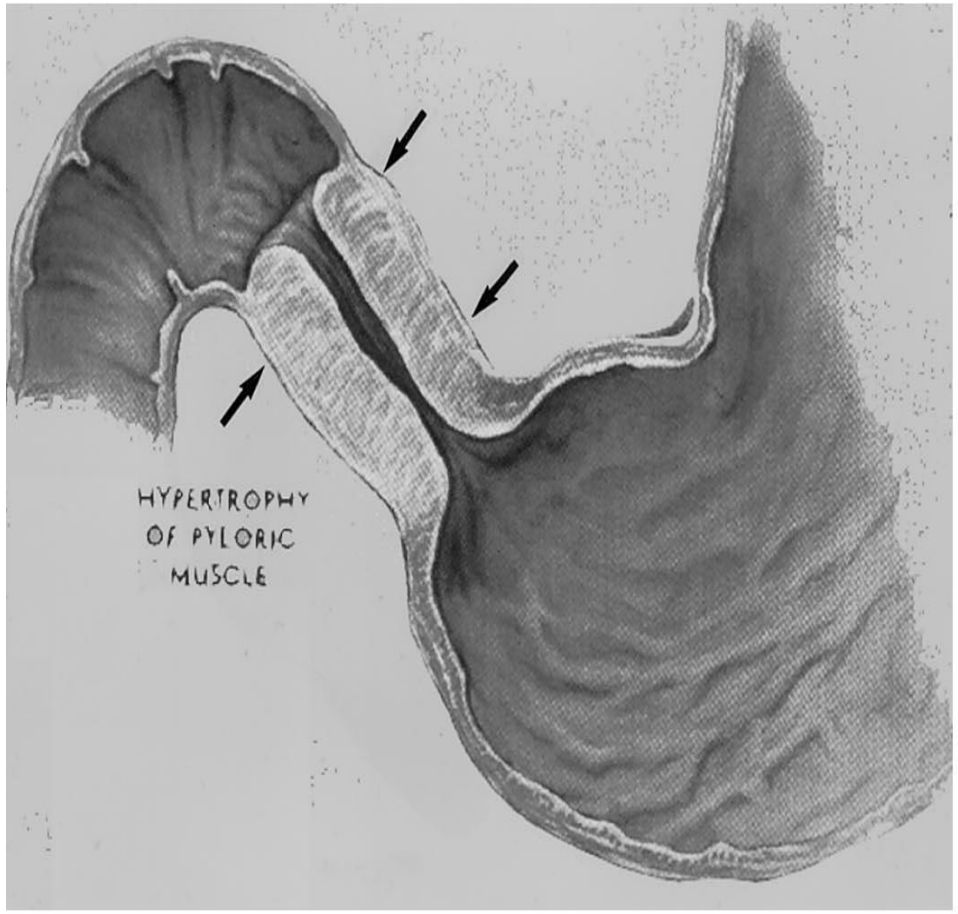
- ✓ Hyperbilirubinemia
- ✓ Midgut malrotation
- ✓ Turner syndrome
- ✓ TEF
- ✓ Esophageal atresia
- ✓ Trisomy 18

Pathology

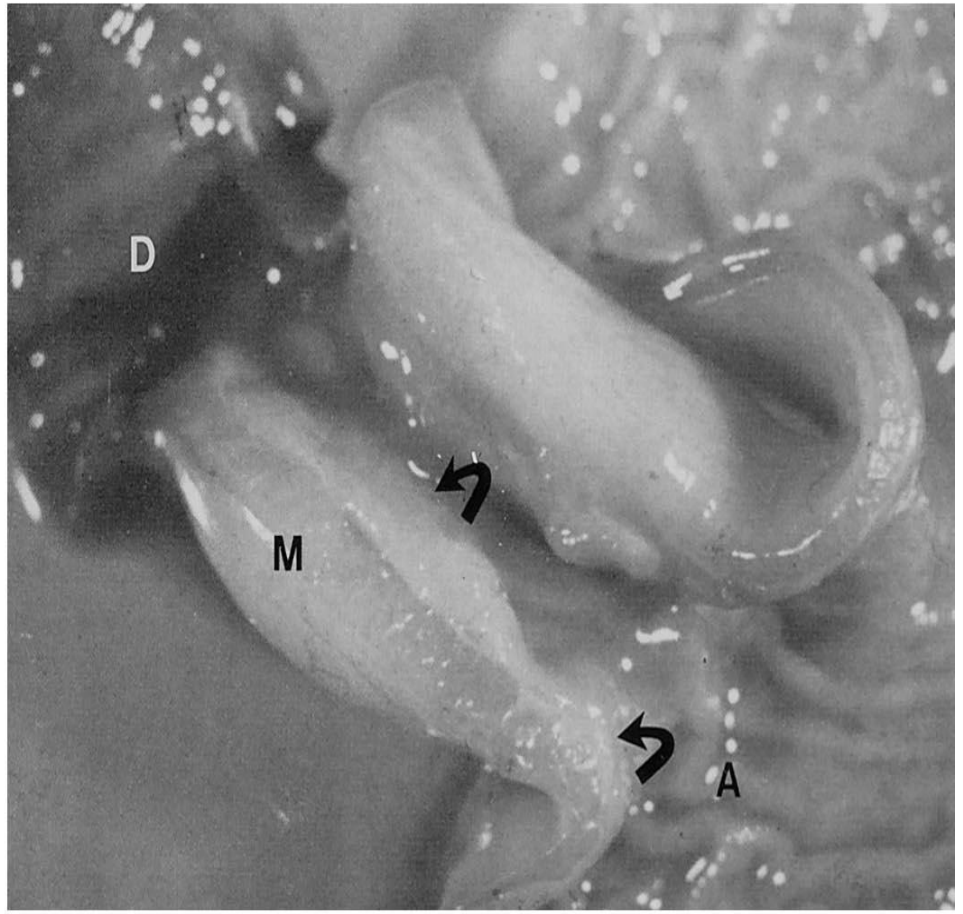
Failure of relaxation:	• Depletion of NANC inhibitory motor neurons: NO, VIP
Hormonal role:	• Insulin like GF – 1, TGF – β, hyper – gastrinemia
Muscle wall hypertrophy:	• Increased amounts of ECM proteins, collagen

Microscopically

- ✓ There is ***progressive hypertrophy of the circular muscle layer*** of the pylorus & an ***increase in chondroitin sulphate within the ECM***.
- ✓ The pylorus becomes elongated & thickened leading to GOO.
- ✓ Grossly, the pylorus is enlarged, ***resembling a tumor approximating the size & shape of an olive (i.e., 2 cm long & 1 cm in diameter)***



a.



b.

Pyloric antrum in IHPS

Note the circumferentially thickened muscle & the lumen is shown as a narrowed canal.

Pathophysiology

- ✓ Hypertrophied muscles block the gastric outlet → GOO → ***Non – bilious projectile vomiting*** → gastric fluid loss → hypo – chloremic, hypokalemic metabolic alkalosis → paradoxical aciduria.

Cont.

✓ Vomiting results in:

- ***Dehydration*** due to loss of fluids → ***hypovolemia*** → secondary hyperaldosteronism → renal sodium retention.
- ***Hypochloremia*** due to loss of chloride ions in HCl
- ***Alkalosis*** due to loss of hydrogen ions in HCl.
  - Impairment of kidney's ability to excrete bicarbonate ions as a compensatory mechanism prevents renal correction of alkalosis.

Cont.

- ✓ Sodium retention to correct intravascular volume depletion occurs along with **concomitant excretion of increased amount of K^+ in urine** → hypokalemia → hypomagnesemia & hypocalcemia.
- ✓ H^+ are excreted in exchange for K^+ leading to ***paradoxical aciduria***
  - ❖ Initial alkalotic urine becomes acidic → paradoxical aciduria
- ✓ Body's compensatory response to metabolic alkalosis is ***hypoventilation*** → increased arterial $PaCO_2$ → 2^0 respiratory acidosis.

Chemistry

- ✓ ↓ Chloride & Potassium
- ✓ ↑ bicarbonate (metabolic alkalosis),

  BUN & creatinine, indirect bilirubin

Why paradoxical aciduria?

- ✓ Gastric fluid loss is associated with the loss of H^+ & Cl^-. Unlike that in vomiting with an open pylorus where one loses gastric, pancreatic, biliary& intestinal fluid.
- ✓ Urinary Na^+ & HCO_3^- losses, which compensate for Cl^- losses, perpetuate this alkalosis.
- ✓ With protracted vomiting, an extracellular volume deficit ensues & urinary excretion of K^+ & H^+ increases in an attempt to preserve Na^+ & volume. The initially alkalotic urine then becomes acidotic hence paradoxical aciduria.

Clinical presentation

- ✓ Onset at 2 – 8 wks. Of age (commonly at around 1 month of age)
- ✓ Vomiting: Projectile; forcible; progressively frequent episodes; non – bilious & coffee ground; 30 to 60 minutes post – prandial.
- ✓ Voracious appetite after vomiting, i.e., "***a hungry vomiter***".
- ✓ Weight loss
- ✓ Lethargy
- ✓ Constipation or hunger diarrhea

Examination

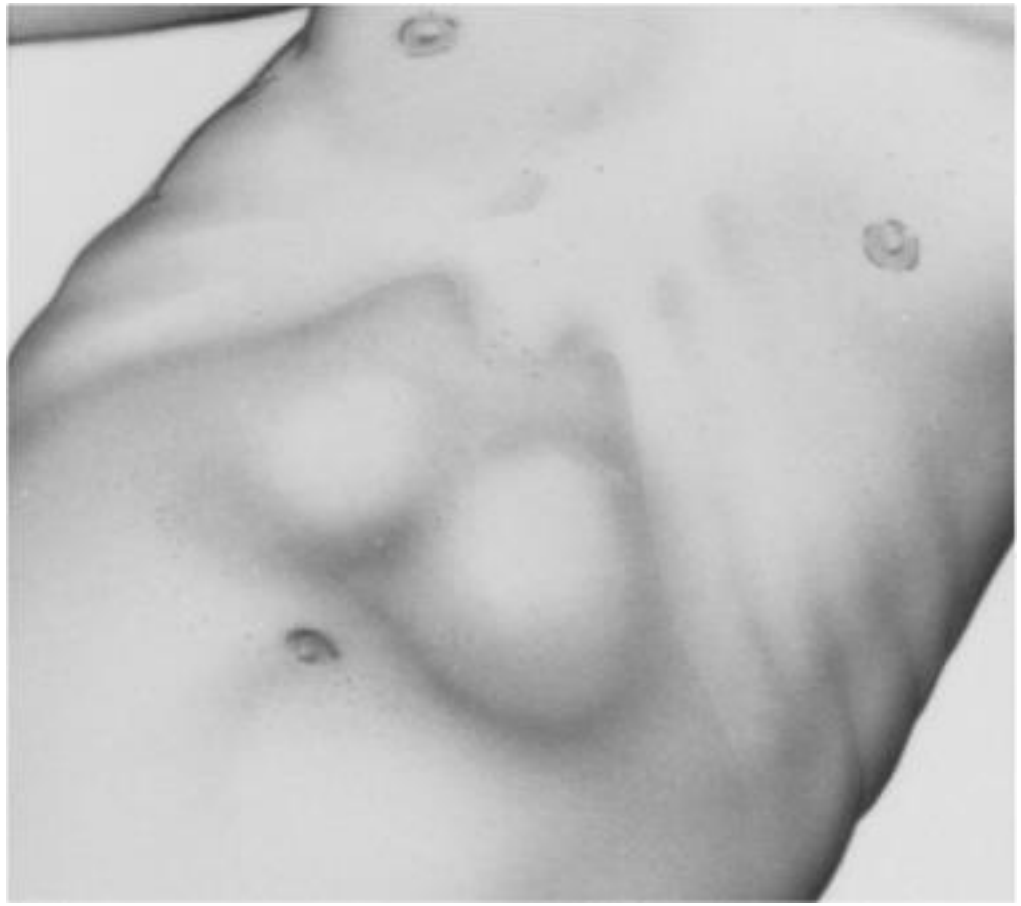
- ✓ General: dehydration, wasting +/- FTT; Jaundice
  - ❖ Indirect hyper – bilirubinemia in 2% of patients due to ***decreased hepatic glucuronyl transferase associated with starvation***.
- ✓ Abdominal examination: visible peristalsis (from left to right; *golf ball waves*) in 75 – 95%; firm, mobile, olive – shaped mass, 1 – 2 cm above umbilicus
  - ❖ Olive – shaped mass: Palpable, mobile, smooth, firm, with all borders well made out, moves with respiration, with impaired resonance on percussion to right of epigastric area (95% of cases).

Cont.

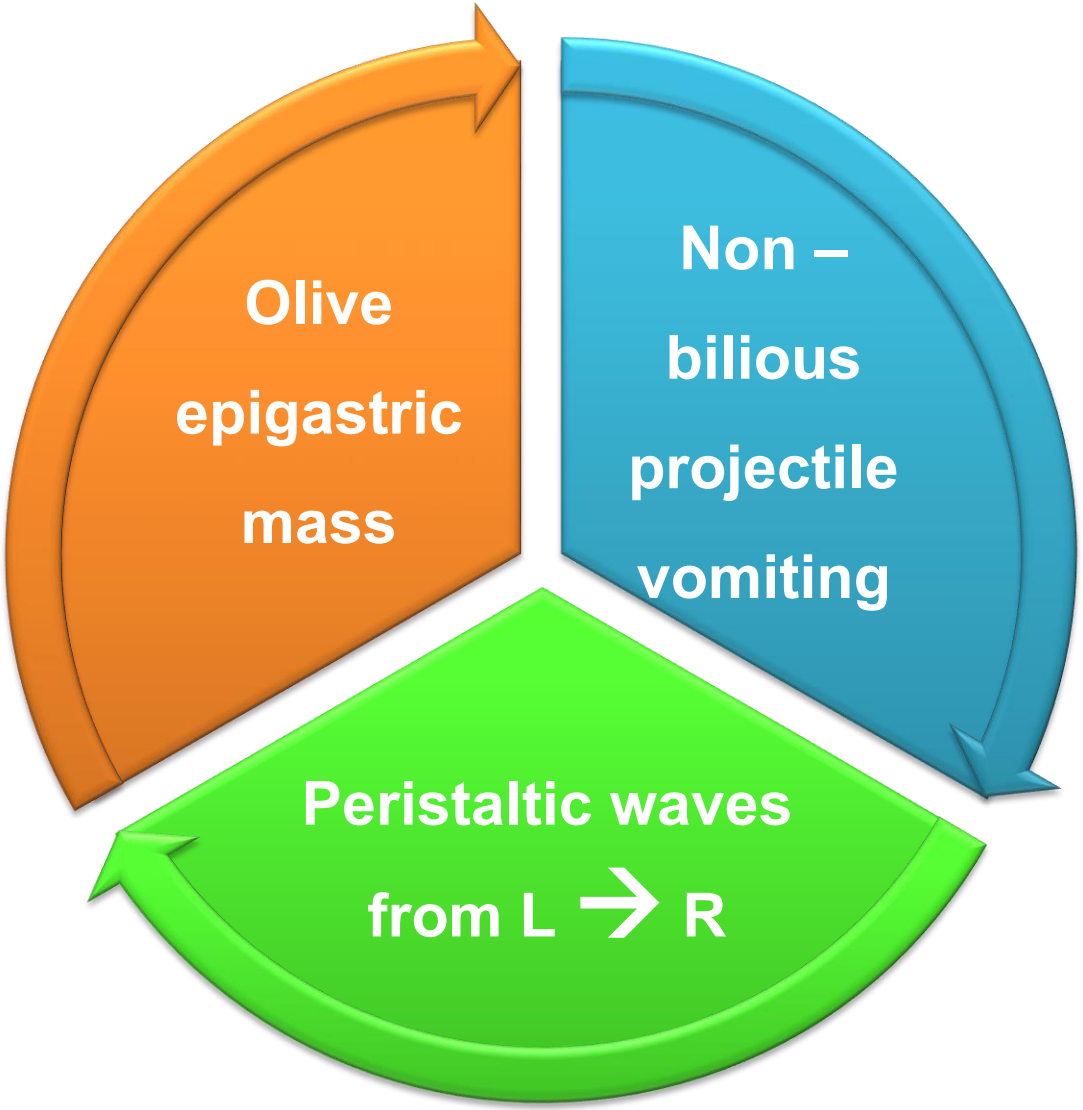
- ✓ Feeding test: allow feeding; after feeding observe for peristaltic waves across the abdomen. After vomiting, abdomen is relaxed & the olive mass is easily palpated.

Gastric peristaltic wave from L → R after feeding

The mass is firm, movable, ~ 2 cm in length, olive shaped, hard, best palpated from the left side, and located above and to the right of the umbilicus in the mid epigastrium beneath the liver edge.



Diagnostic triad (70% accuracy for IHPS)



Diagnosis in premature infants:

- ✓ In premature infants with IHPS ***vomiting may be less forceful***; ***voracious appetite & exaggerated gastric peristalsis may be lacking***, & ***U/S criteria for diagnosis may not apply***.
- ✓ In hospitalized premature infants, non – projectile vomiting, weight loss & lethargy may initially be attributed to sepsis; ***negative cultures, rapid improvement with IVFs & metabolic alkalosis (rather than acidosis) in such infants should prompt consideration of IHPS***.
- ✓ The diagnosis of IHPS should be considered in ***young infants with repeated non – bilious vomiting, hypochloremic alkalosis, and/or rapid clinical improvement after rehydration***, even in the absence of projectile vomiting or epigastric mass.

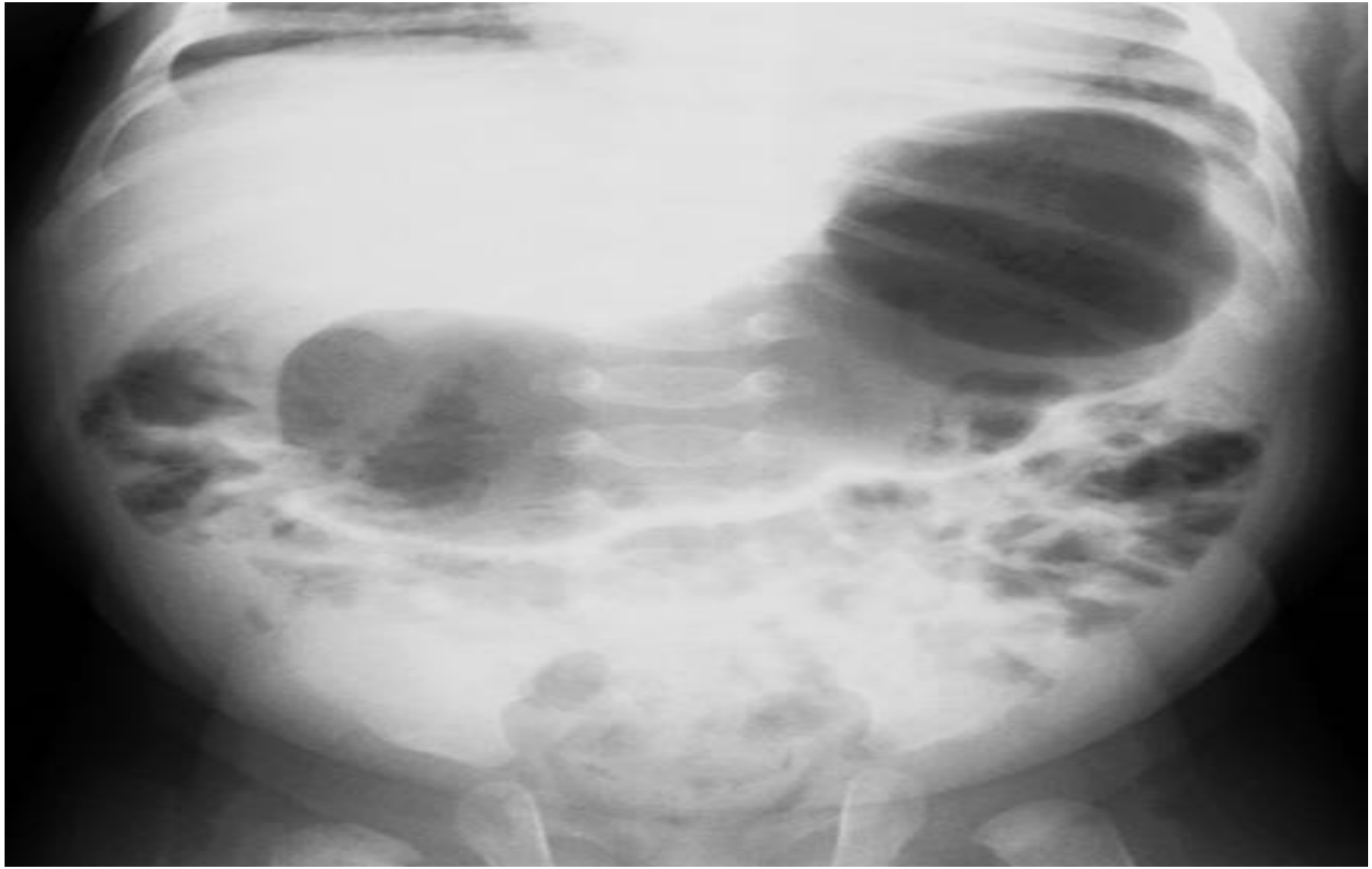
Lab investigations

- ✓ FHG
- ✓ U/E/Cr
- ✓ LFTs
- ✓ Urinalysis

Imaging

- ✓ Abdominal X ray (erect posture)
  - ❖ Upper abdominal gas bubble in stomach

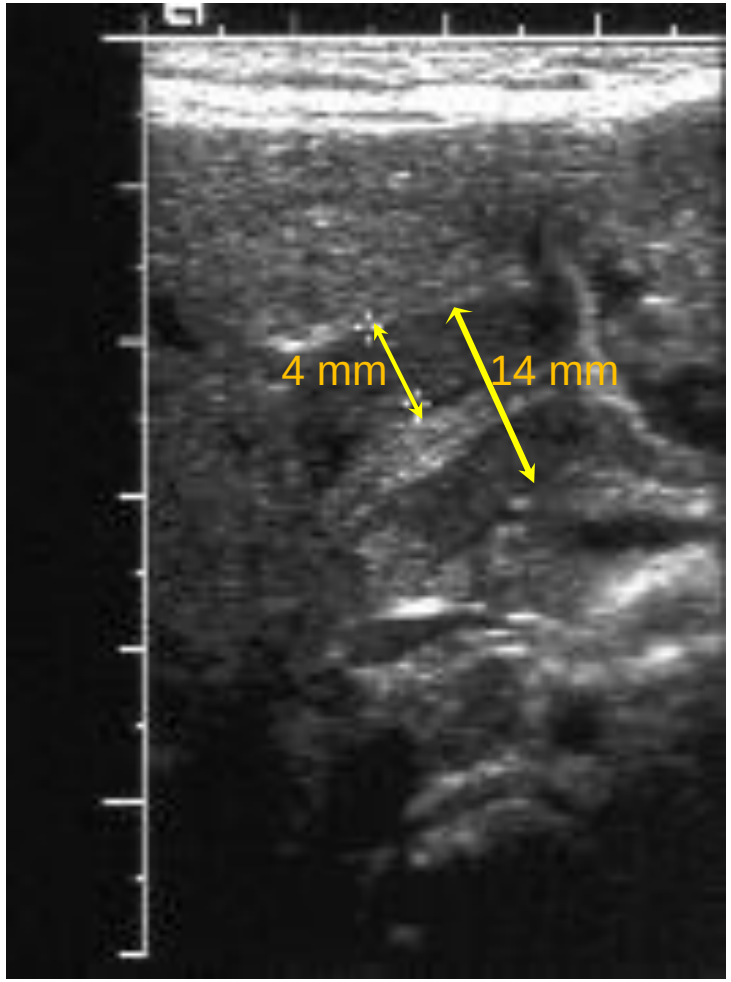
Supine radiograph in an infant showing the ‘caterpillar sign’ (markedly dilated stomach with exaggerated incisura)

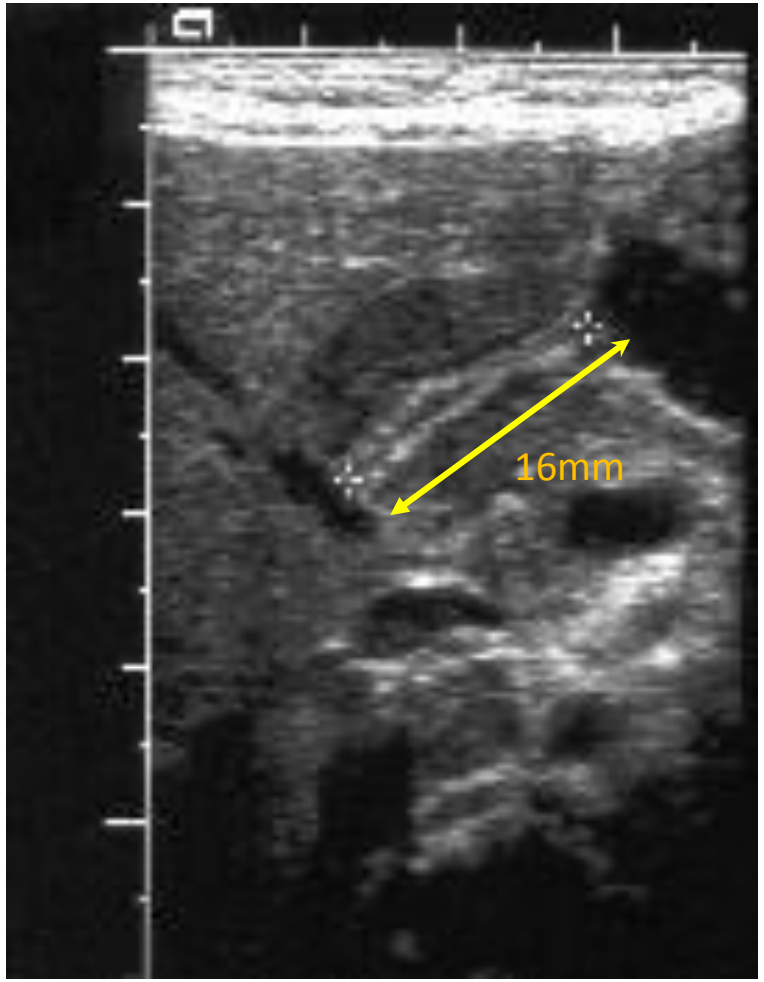


Ultrasound

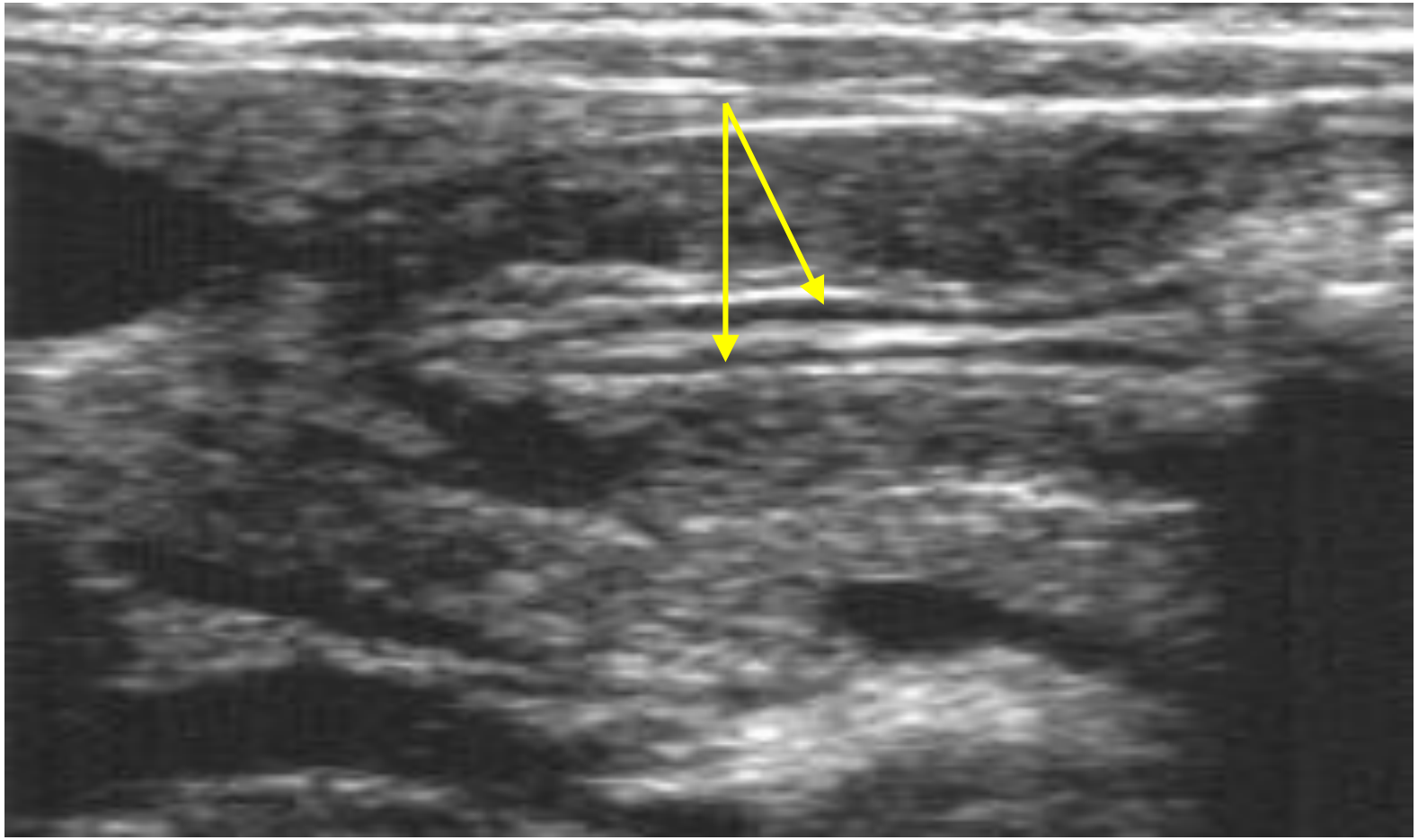
- ✓ **GOLD STANDARD at present: doughnut sign or cervical pyloric sign**
- ✓ Sensitivity → 98%; Specificity → 100%; PPV → 100%; NPV → 90%.
- ✓ Diagnostic criteria includes:

1. Pyloric channel length > 17mm in presence of functional GOO
2. Trans – pylorus diameter > 14mm
3. Pylorus muscle wall thickness > 3mm
4. Pyloric volume > 1.5cm^3

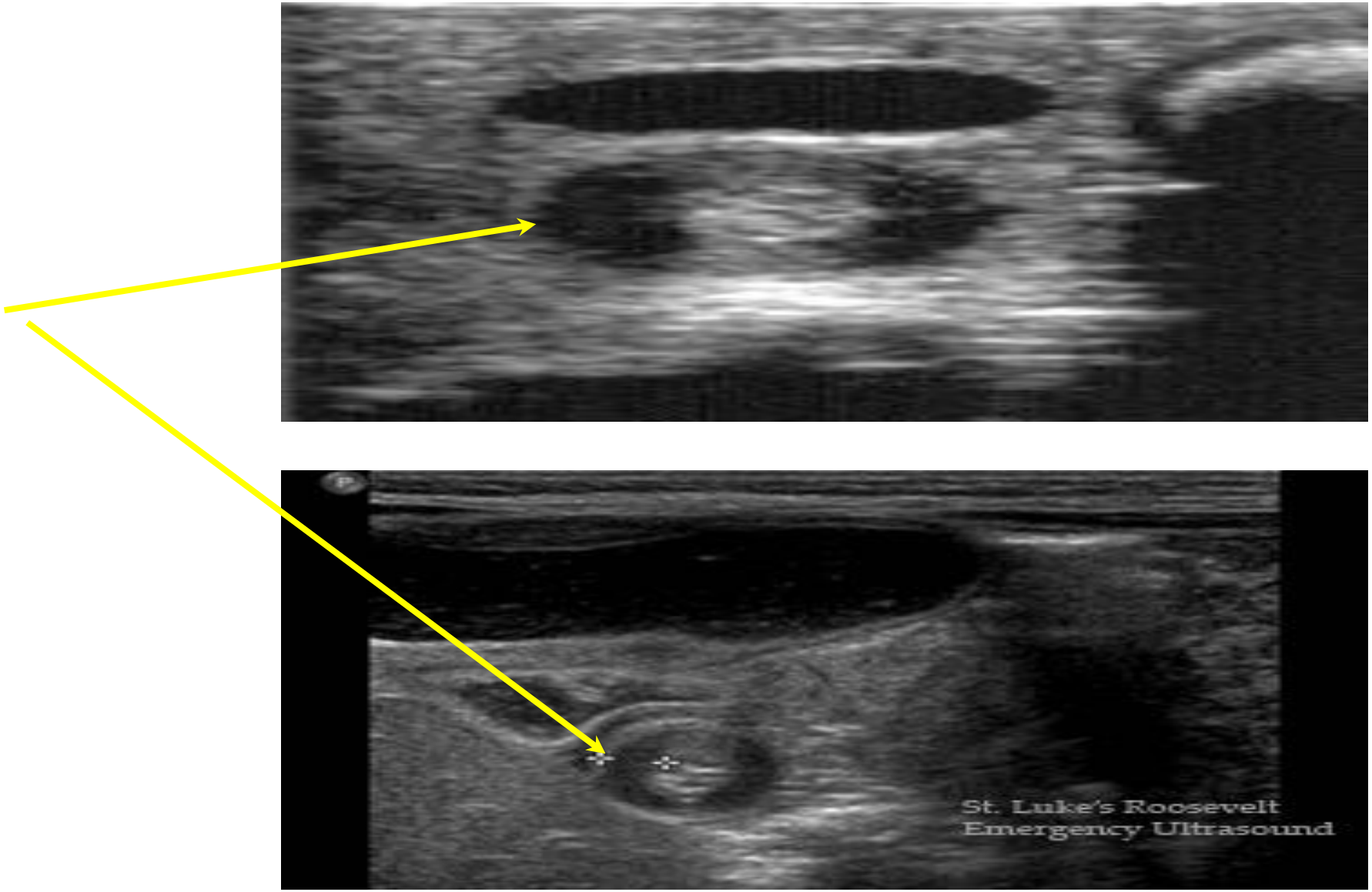

4 mm
14 mm


16mm

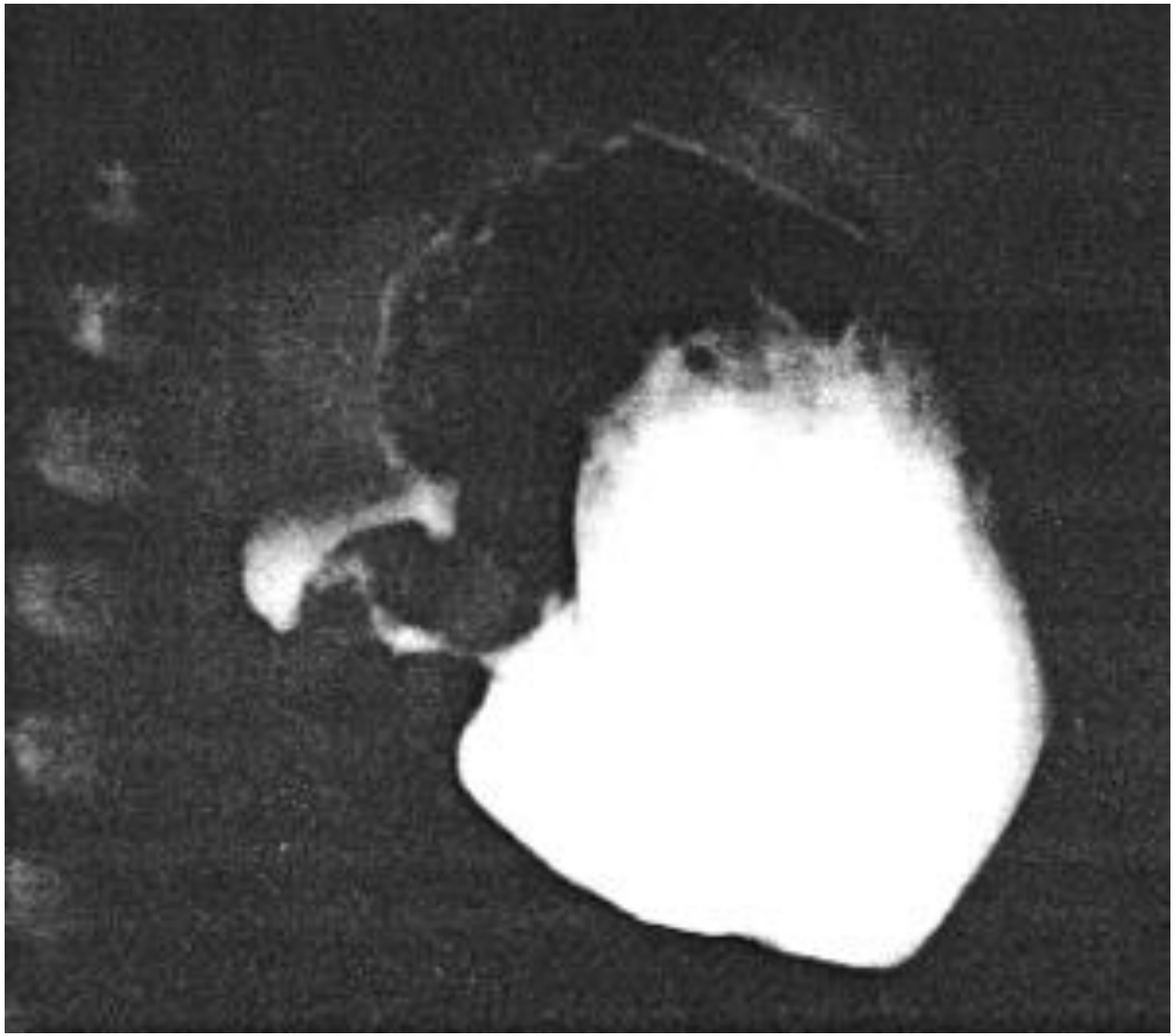
Double track sign: redundant mucosa in the narrowed lumen creating 2 mucosal outlines



Target sign & heterogeneous echo – texture of the muscular layer in a transverse sonographic image in a patient with IHPS



Upper GI Series: string sign or shouldering of the hypertrophied pyloric muscles bulging into the gastric lumen



Barium meal/ fluoroscopy

- ✓ Peristaltic waves: caterpillar sign
- ✓ Delayed gastric emptying
- ✓ Elongated & narrowing pyloric canal – string sign/ rail road track sign
- ✓ The pylorus indents the contrast filled antrum (shoulder sign) or base of the duodenal stump (mushroom sign)

Biochemical changes

- ✓ Dehydration
- ✓ Malnutrition
- ✓ Hypochloremic, hypokalemic metabolic alkalosis
- ✓ Paradoxical aciduria
- ✓ Hyperbilirubinemia
- ✓ ABG: ↓ serum K^+ & Cl^- & ↑ blood pH, HCO_3^- & BUN.

DDX

- ✓ Sepsis
- ✓ GERD
- ✓ Cow milk protein intolerance: presents when children are being weaned.
- ✓ Adrenal crisis
- ✓ IO, Hirschsprung’s disease
- ✓ Liver disease

Management

- ✓ In 1937, Sir Lancelot Barrington – Ward wrote, “under no circumstance should a case of pyloric stenosis be considered an emergency requiring immediate operation ***WITHOUT PROPER PREPARATION***. The more feeble & dehydrated the infant, the greater the necessity to restore its fluid & chemical deficiency before subjecting it to the ordeal of an operation.

1. Preoperative resuscitation: [ABCs]

✓ Resuscitation (***IHPS is a medical rather than a surgical emergency***)

- ❖ Insert NGT, IV access points
- ❖ Initiate fluid therapy: NS (***can be bolused without lethal altering of K^+ levels***)
  - Resuscitation fluid
  - Maintenance fluid

Correct the following

- ✓ Metabolic alkalosis
- ✓ Hypokalemia
- ✓ Hyponatremia
- ✓ Hypochloremia

Cont.

- ✓ For infants presenting with normal electrolyte values & mild dehydration:
  - ❖ Bolus as per requirement based on G/E, UOP & Chloride levels
  - ❖ IVFs at **1.5 times maintenance** with **[5% dextrose + 0.45% NaCl + 2 mEq/ 100mL or 20mmol/L of KCl]** given at **160mL/kg/d**

Cont.

- ✓ For infants with moderate dehydration or severely impaired circulation
  - ❖ Bolus: 20ml/Kg NS or Hartman's fluid as per WHO guidelines
  - ❖ Maintenance IVFs @ 1.5 times maintenance with [***5% dextrose + 0.45% NaCl + 20mEq]*** at a rate of ***120 – 150ml/kg/24h over 24 – 48h***
- ✓ Ensure NGT drainage to prevent aspiration of vomited secretions.

CAUTION!

1. ***In the more severely dehydrated infants, ensure adequate kidney function prior to administering KCl.***
2. ***The correction of alkalosis prior to surgery is imperative as there is an increased risk of post – operative apnea.***

Surgery should take place when

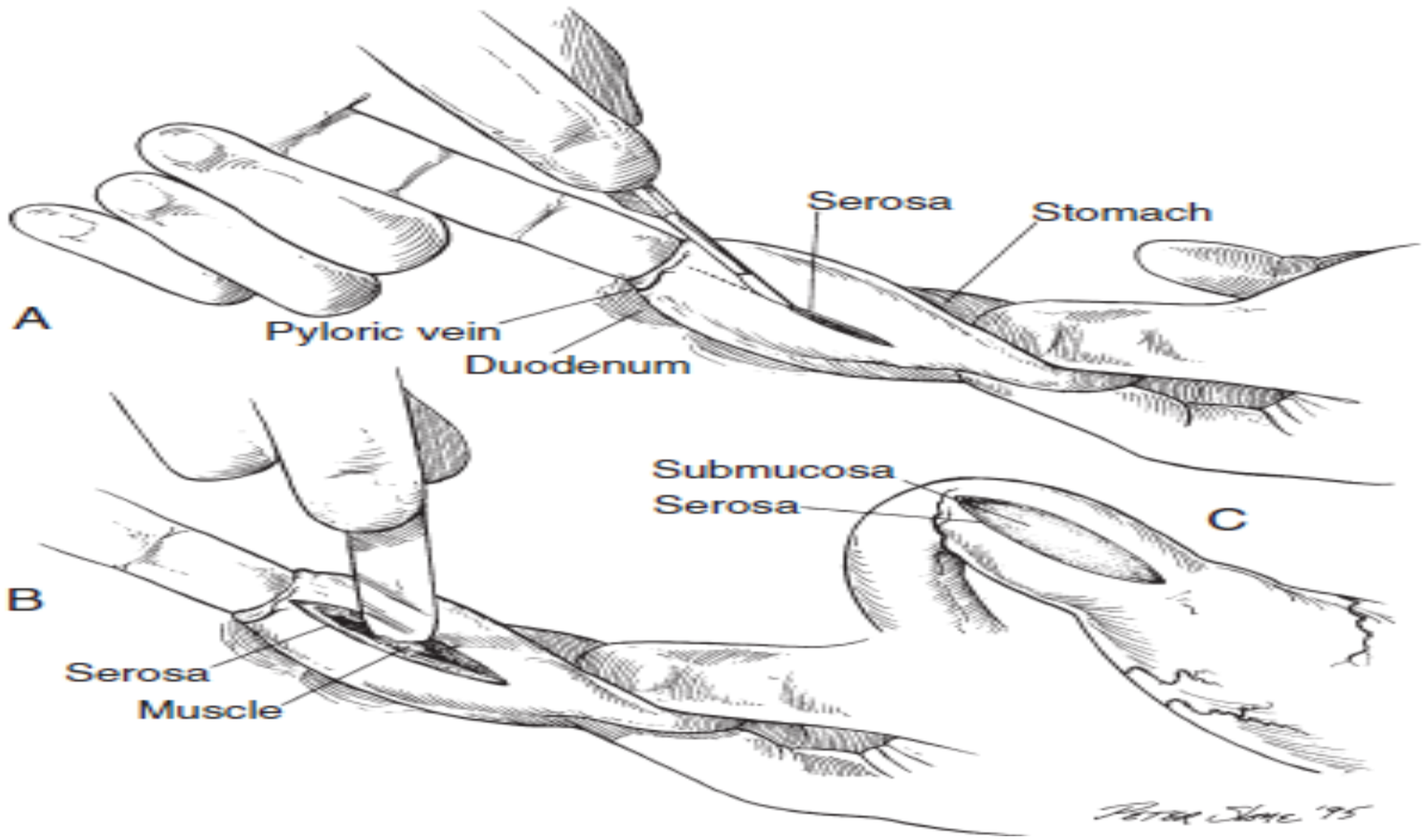
- ✓ Intravascular volume is optimized (Dehydration is corrected)
- ✓ Electrolyte optimization:
  - ❖ Normal serum Na^+ & K^+
  - ❖ Chloride ion > 90mmol/L
  - ❖ Bicarbonate < 28mmol/L

2. Non – operative strategies

- ✓ ***IV atropine sulfate*** to relax the pylorus muscle
  - ❖ Initial dose of ***0.4mg/kg/d*** at increments of 0.1mg/kg/d over 8 days or until vomiting ceases then maintain on ***PO atropine methyl nitrate*** for 2 weeks.
  - ❖ Requires prolonged hospitalization & IV nutrition.
  - ❖ Rarely definitive; a temporizing measure.
- ✓ Endoscopic ***balloon dilatation***
- ✓ Endoscopic pyloromyotomy using an electroscopic needle knife or sphincteretomy.

3. Surgery: open/ laparoscopic pyloromyotomy

The **Ramstedt's pyloromyotomy**, whether performed through a RUQ incision, through an umbilical incision, or via laparoscopy, **remains the standard operation for pyloric stenosis today.**



Cont.

- ✓ ***Fredet Ramstedt's pyloromyotomy*** → ***conventional open procedure***
- ✓ Laparoscopic pyloromyotomy
- ✓ Double – Y pyloromyotomy

Cont.

- ✓ The **Trans – pyloric plane**, (**Addison's Plane)** is an upper transverse line, located halfway between the jugular notch & the upper border of the pubic symphysis. Roughly a hand's breadth beneath the xiphoid process of the human sternum.
- ✓ The plane in most cases cuts through the pylorus of the stomach, the tips of the ninth costal cartilages and the lower border of the first lumbar vertebra. Intraoperatively, the surgeon should *ID* the serosal demarcation between the duodenum & the pylorus. The pre – pyloric vein, or Mayo vein, is located at this junction. The risk of duodenal perforation is prevented by stopping the extent of the myotomy 1 – 2 mm proximal to this point.

Journal of Pediatric Surgery (2007) **42**, 692–698

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Journal of Pediatric Surgery

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Laparoscopic pyloromyotomy for hypertrophic pyloric stenosis: a prospective, randomized controlled trial

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Abstract

Background: Several authors have reported on laparoscopic pyloromyotomy (LP) since the technique was originally described in 1990, but its benefits remain unproven. We performed a randomized controlled trial comparing LP to open circumumbilical pyloromyotomy (OP) for hypertrophic pyloric stenosis.

Methods: In a prospective study, 102 infants with pyloric stenosis were randomly assigned to either LP (n = 50) or OP (n = 52). The primary outcome measure was the incidence of postoperative vomiting; the secondary parameters were the durations of surgery and anesthesia, surgical complications, and postoperative pain. All infants were managed according to standardized procedures regarding general anesthesia, surgical technique, postoperative analgesia, and feeding regimen. Parents, carers, and assessors responsible for the postoperative evaluation were blinded for the technique used.

Results: There was no difference in the incidence of postoperative vomiting between the 2 groups. The overall incidence of complications was similar, but the durations of surgery and general anesthesia were significantly longer in the LP group than in the OP group ($P = 10^{-4}$ and $P = .02$, respectively). There were 3 cases of incomplete pyloromyotomy after laparoscopy, requiring a repeat procedure.

Conclusions: Laparoscopic pyloromyotomy does not decrease the incidence of postoperative vomiting, has a similar complication rate compared with the open umbilical approach, but may expose patients to a risk of inadequate pyloromyotomy.

Post – operatively

- ✓ Post – operative emesis after pyloromyotomy may occur. May be due to ***pyloric edema*** & ***ileus***. Often subsides within a week.
- ✓ Introduce oral feeding at least ***6 – 8hrs post op***. Initially low volume (15 cc).
- ✓ ***Pedialyte or glucose water*** increased gradually as tolerates 60 – 90cc of breast milk twice without significant vomiting.
- ✓ Most infants can be discharged within 24 – 36h post op.

Journal of Pediatric Surgery (2013) **48**, 2175–2179

ELSEVIER

Journal of
Pediatric
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A review of postoperative feeding regimens in infantile hypertrophic pyloric stenosis

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[b] *Indiana University School of Medicine, Indianapolis, IN, USA*

Cont.

- ✓ Aim: to determine an optimal feeding pattern that influences post – operative emesis
- ✓ Method:
  - ❖ Review of relevant literature
  - ❖ Identified 4 broad feeding regimens:
    - Immediate vs. delayed
      - —Immediate: ad lib vs. standardized
      - —Delayed: ad lib vs. standardized

Results:

Immediate vs. delayed

- Many studies advocated to ***delay feeding for 4h postoperatively*** in light of gastric dysmotility & increased severity of emesis. Many studies showed no shorter time to discharge with immediate feeding regimens.

Incremental vs. ad lib

- Majority of the studies found no increase in the incidence of postoperative emesis in ad lib regimes despite reaching full strength feeds more quickly, thereby decreasing time to discharge.

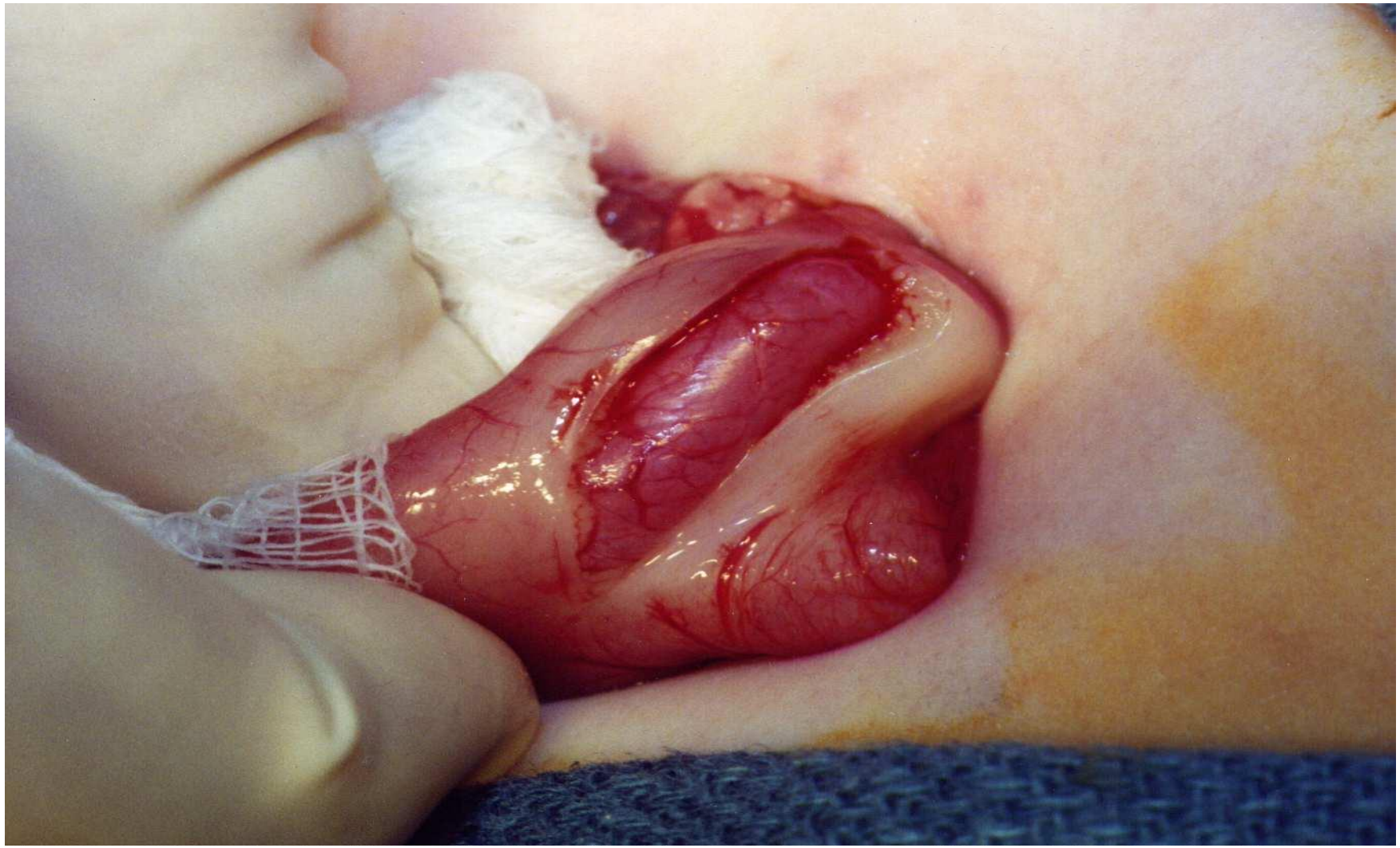
Outcome of operation

- ✓ Yoshizawa et al demonstrated in ultrasonography studies that after pyloromyotomy, ***the pylorus changes significantly within 3 days postoperatively & returns to normal within 5 months***.

Surgical complications

- ✓ Intraoperative
  - ❖ Duodenal perforation (3 – 4%)
  - ❖ Mucosal perforation
- ✓ Post – operative
  - ❖ Transient persistence of vomiting due to inadequate pyloromyotomy or foveolar cell hyperplasia (3 – 19%)
  - ❖ Wound infection (1 – 4%)
  - ❖ Wound dehiscence (1.4%)
  - ❖ Continued post – operative bleeding

Pyloromyotomy completed



Conclusion

- ✓ Adequately performed pyloromyotomy is curative for IHPS & has minimal complications.
- ✓ Post – operative emesis is common but resolves over a period of 2 weeks. ***If emesis persists beyond 2 weeks, concern should be raised for GERD or inadequate pyloromyotomy***.

Case scenario

- ✓ 5wk old male infant with ***persistent vomiting for 2 weeks***. Mom says ***baby throws up almost every feed – getting worse & more forceful***, emesis looks like formula she feeds him
- ✓ On Gaviscon for reflux diagnosed 1 wk. ago
- ✓ Using rice cereal to thicken feeds but no improvement
- ✓ Not wetting as many diapers.
- ✓ Diagnosis: **INFANTILE HYPERTROPHIC PYLORIC STENOSIS**

3. PEDIATRIC ABDOMINAL SURGICAL EMERGENCIES

LEVEL VI

2019

COMPILED BY

NAILA KAMADI

Outline

1. Bowel obstruction – 111
    a. Intestinal atresias – 114
    b. Hirschsprung's Disease – 140
    c. Malrotation – 210
    d. Volvulus – 240
    e. Intussusception – 251
    f. Meconium conditions – 279
2. Acute appendicitis – 284
3. Meckel's diverticulum – 297
4. Necrotizing enterocolitis (NEC) – 306

Objectives

- Understand the age distribution of common pediatric abdominal emergencies.
- Describe the MC clinical presentation of these emergencies
- Choose an appropriate imaging technique
- Interpret the MC imaging findings.

Pathophysiology of pain

- Visceral pain
  - Mechanical – stretching
  - Chemical – mucosa
  - Aching and dull, poorly localized
- Parietal pain: sharp, well localized.
- Referred pain: somatic and visceral afferent fibers enter the spinal cord close to each other.

History

- Usual: quality, location, severity, associated symptoms, aggravating/alleviating factors
- Kids cannot give a history
- Dangerous signs given by parents

The red flags

- Duration – acute vs. chronic
- Fever – inflammation, infection
- Vomiting – stasis, obstruction, dehydration
- Urine output – volume depletion
- Diarrhea – bloody

Examination

- Usual: **inspection, auscultation, percussion, palpation**.
- Rectal – Retrocecal appendicitis, occult blood
- Scrotal - torsion

Investigation

- Chemistry – electrolyte abnormality, BUN/creatinine, LFT
- CBC – infection, bleeding
- Plain AXR – free air, obstruction
- Urinalysis – pyuria, hematuria

Fluid management in surgical emergencies

1. Resuscitation fluids: **20ml/kg of NORMAL SALINE** (don't give IVFs containing potassium); Bolus over 30m – 1 hr.
2. Maintenance fluid: what any living thing needs to survive
   - The best fluid is one most similar to plasma; **Hartman's** is most practical
   - Best for neonates & younger children is a mixture of **5% dextrose PLUS 0.45% saline PLUS 20mmol/L KCl**
3. Replacement for ongoing loses
   - Replace ALL gastric loses with saline ml for ml; Replace ALL other loses above "normal" loses:
     - Urine above 4ml/kg/hr; Stool and stoma loses above 20ml/kg/day; All loses from drains

Initial management of bowel obstruction

- Fluid resuscitation
- Antibiotics if perforation is suspected
- Please remember: "***NOT ALL CENTRES CAN ADEQUATELY MANAGE CHILDREN WITH SURGICAL EMERGENCIES***"
- Refer these patients, after adequate resuscitation, to the nearest centre that can handle these delicate patients!
- Sometimes even KNH may not be the best centre for best outcomes
  - 0% survival for gastroschisis, 10% for oesophageal atresia & 33% for other atresia.

TABLE 2. COMMON CAUSES OF ABDOMINAL PAIN BY AGE GROUP

Infants	3-11 Year Old	Adolescent
Intussusception	Appendicitis	Appendicitis
Hirschsprung's enterocolitis	Trauma	Pelvic inflammatory disease
Strangulated hernia	Meckel's diverticulitis	Mittelschmerz
Trauma (child abuse)	Pneumonia	Crohn's disease
Meckel's diverticulitis	Bacterial enterocolitis	Enterocolitis
Bacterial enterocolitis	Yersinia	Peptic Ulcer Disease
Pneumonitis	Campylobacter	Cholecystitis
Pyelonephritis	Salmonella	Pneumonia
Mesenteric cysts	Shigella	Trauma
Testicular torsion	Crohn's disease	Ectopic pregnancy
Pancreatitis or pseudocyst	Pancreatitis	Hematocolpos
Intestinal obstruction/ volvulus	Infected mesenteric cyst	Psychosomatic
	Ruptured tumors	
	Pyelonephritis	

Pediatric abdominal emergencies

Neonates:

- Malrotation with volvulus
- NEC
- Hirschsprung's Disease (HSD)
- Omphalitis
- ARM

Infants/ Children

- IHPS
- Incarcerated inguinal hernia
- Intussusception
- Acute Appendicitis

1. BOWEL OBSTRUCTION

1. BOWEL OBSTRUCTION

- Diagnosis is often ***age specific***.
- ***Bilious/ feculent vomiting in the infant & child is a surgical emergency until proven otherwise*****.** It is indicative of obstruction beyond the 2nd part of the duodenum.
- Regarding volvulus in children:
  - It is difficult to tell when it is present; Isolated volvulus is an unusual finding as it is often associated with other conditions such as ***Meckel's diverticulum & intestinal atresias***.
- Children will look surprisingly well until it's too late. They have very good compensatory mechanisms esp. in terms of hemodynamics. This necessitates vigilant monitoring.

Age – specific presentation

- 1st week: Atresias
- 3 – 6 weeks: IHPS
- 4 – 6 months: Intussusception
- 6 months – 1.5 yrs.: Malrotation, adhesion bands

a. Intestinal Atresia

a. Intestinal Atresia

- This is failure of development of a bowel segment.
- May involve any part of the gut, from the pharynx to the anus.
- Usually presents in the ***first few days of life*** depending on the site. ***The lower the atresia is, the later the children tend to present***.
  - The child may feed well for a day or two with distal atresia.

Pathophysiology

- Atresias are thought to be due to ***vascular accidents after the bowel is properly formed***.
- The nature of arterial supply to the bowel is radial from the mesentery.
- Interference of specific blood supply to specific segments of bowel results in dying off of the segment hence atresia.

Types of intestinal atresia

I. Hollow tube with ***simple, thin, intra – luminal membrane*** (that can't be easily visualized). Proximal dilatation of gut with sudden collapse of the distal part. (20% of atresias)

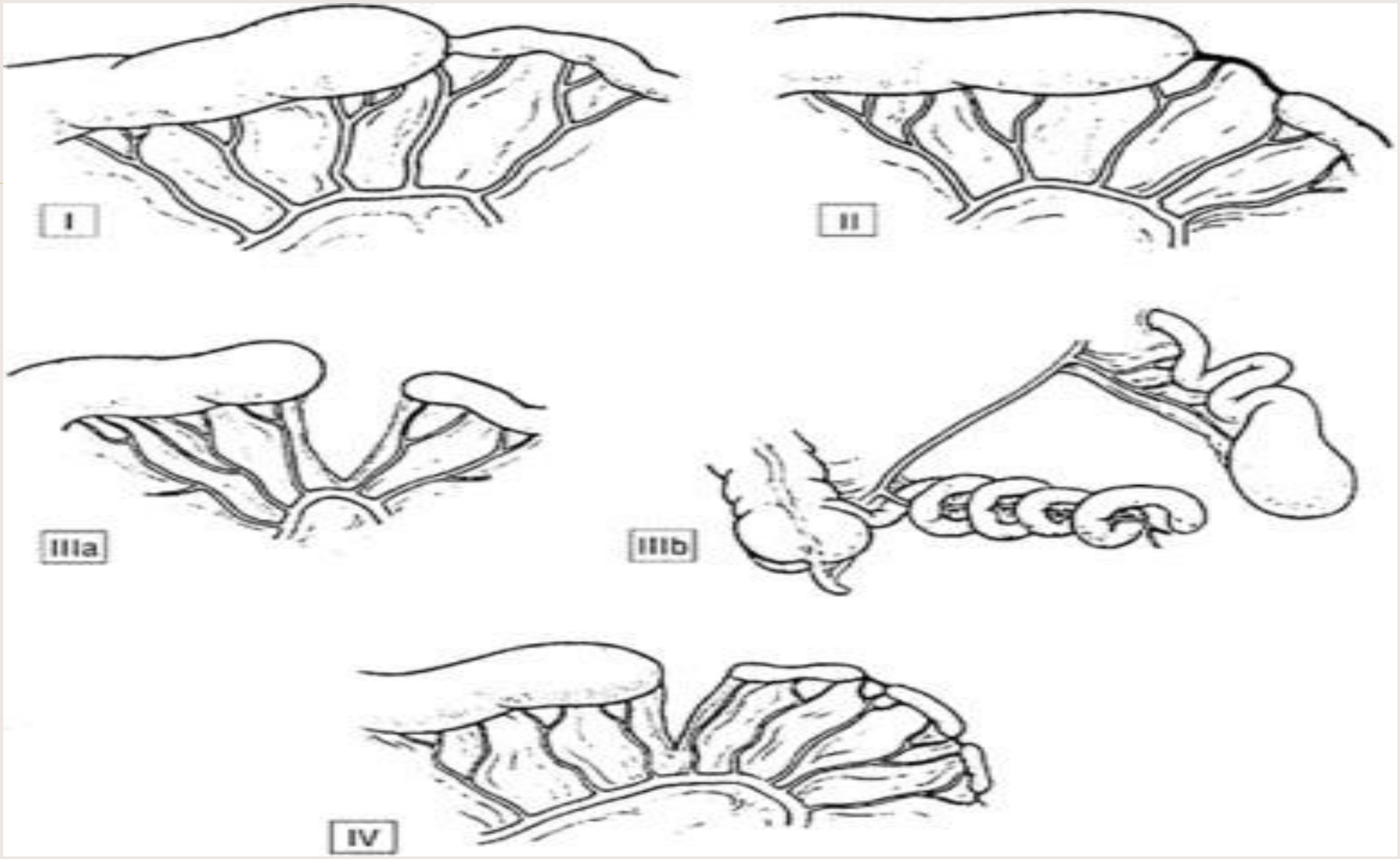
II. Proximal & distal blind loops with ***a gap in between comprising a small, fibrous, solid band***. The ***mesentery is intact***. Proximal blind loop is dilated & the distal loop is collapsed.

III. Like type II but with ***a small V – shaped defect on the mesentery***

   I. **IIIa.** Both ends of the bowel end in blind loops accompanied by a small V – shaped mesenteric defect.

   II. **IIIb.** Extensive mesenteric defect; loss of normal blood supply to distal bowel. Distal ileum coils around the ileocolic artery from which it derives its entire blood supply, producing an 'apple peel' appearance. Associated with prematurity, an unusually short distal ileum and significant foreshortening of the bowel.

IV. Multiple, segment atresias: Carry a poor prognosis


I
II
IIIa
IIIb
IV

Esophageal atresia

- Esophageal atresias present at birth with ***respiratory symptomatology*** as the proximal pouch will be filled with saliva that is regurgitated & aspirated into the resp. system resulting in ***aspiration pneumonia***.
- They also have ***drooling of saliva***, ***choking on breastfeeding***.

Management

- Confirmation: insert a radio – opaque NG tube & take images → the tube will coil.
- Stop feeds & prop up
- Put low pressure, continuous suction (***syringe attached to feeding tube***) to avoid aspiration
- Keep babies warm
- Refer for definitive management.
  - Right sided thoracostomy → anastomosis of proximal & distal esophageal pouches.

Duodenal atresias

- Incidence: 1 in 5,000 – 10,000 live births; 75% of stenosis & 40% of atresias are found in the duodenum. Usually involve ***2nd part of duodenum***. Multiple atresias are found in 15% of cases. 50% of patients are LBW & premature. 75% are associated with polyhydramnios.

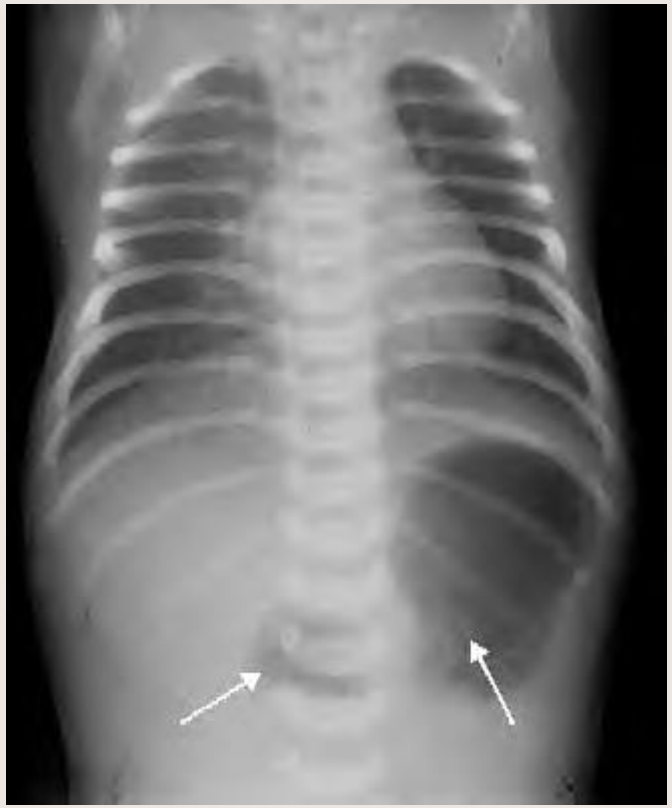
Anomalies associated with duodenal atresia

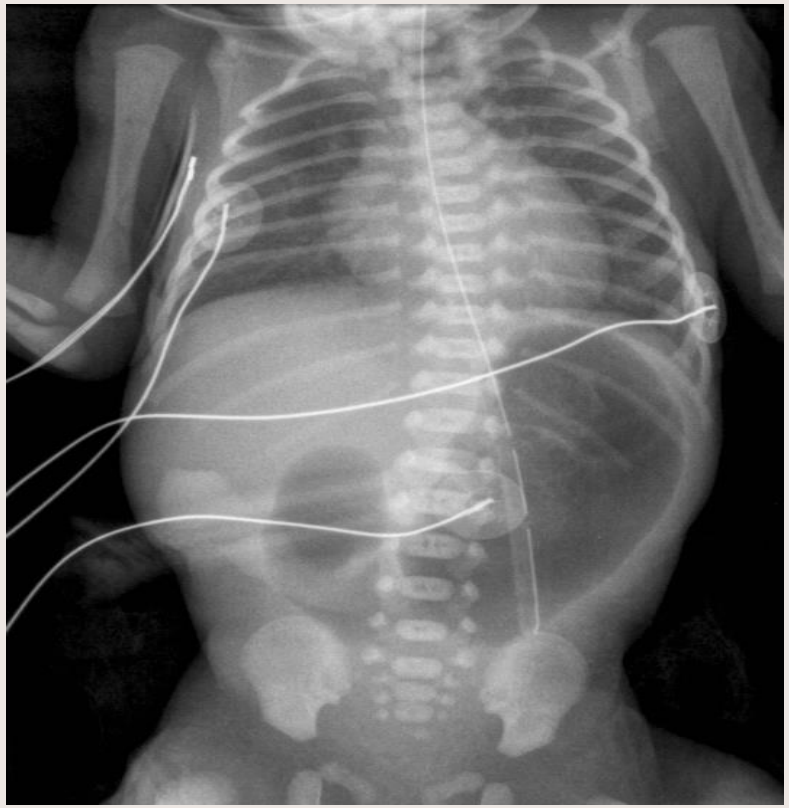
- Down’s syndrome (30%)
- Malrotation
- Congenital heart disease
- Esophageal atresia
- Urinary tract malformations
- ARMs
- VACTERL malformations

Cont.

- Present in 24 – 48 hrs. with ***bilious vomiting with a little epigastric distention***
- Plain X – ray/ KUB: ***double bubble sign*** (stomach & duodenum) with absence of distal gas.
  - DDX: annular pancreas
- ***Duodenal atresia*** is often diagnosed on antenatal U/S.

Double Bubble Sign of duodenal atresia





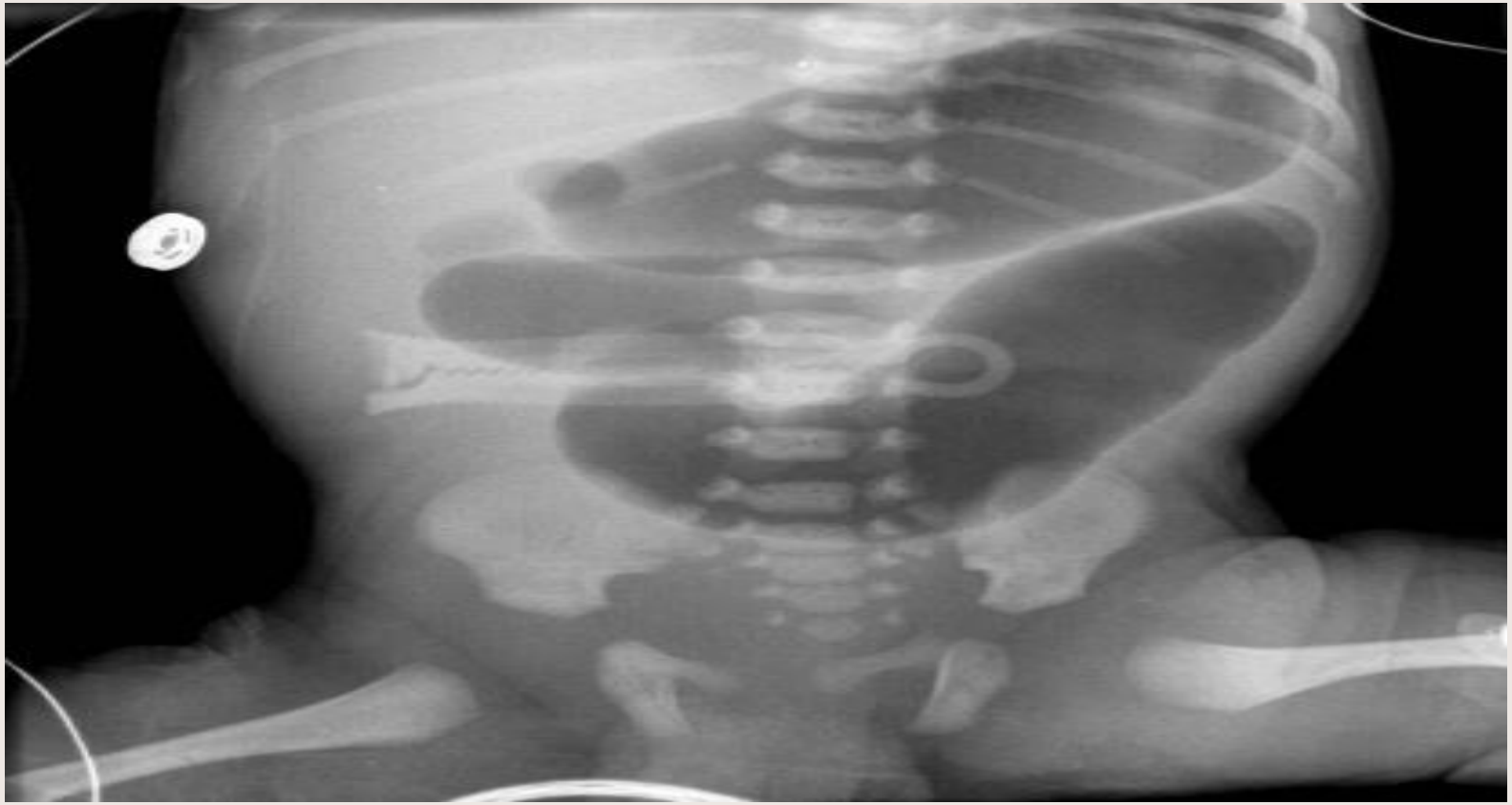
Treatment of duodenal atresia

- NGT decompression
- Hydration
- Surgery
  - Double diamond duodeno – duodenostomy
  - Continuous prolonged NG decompression, sometimes for > 2 weeks.

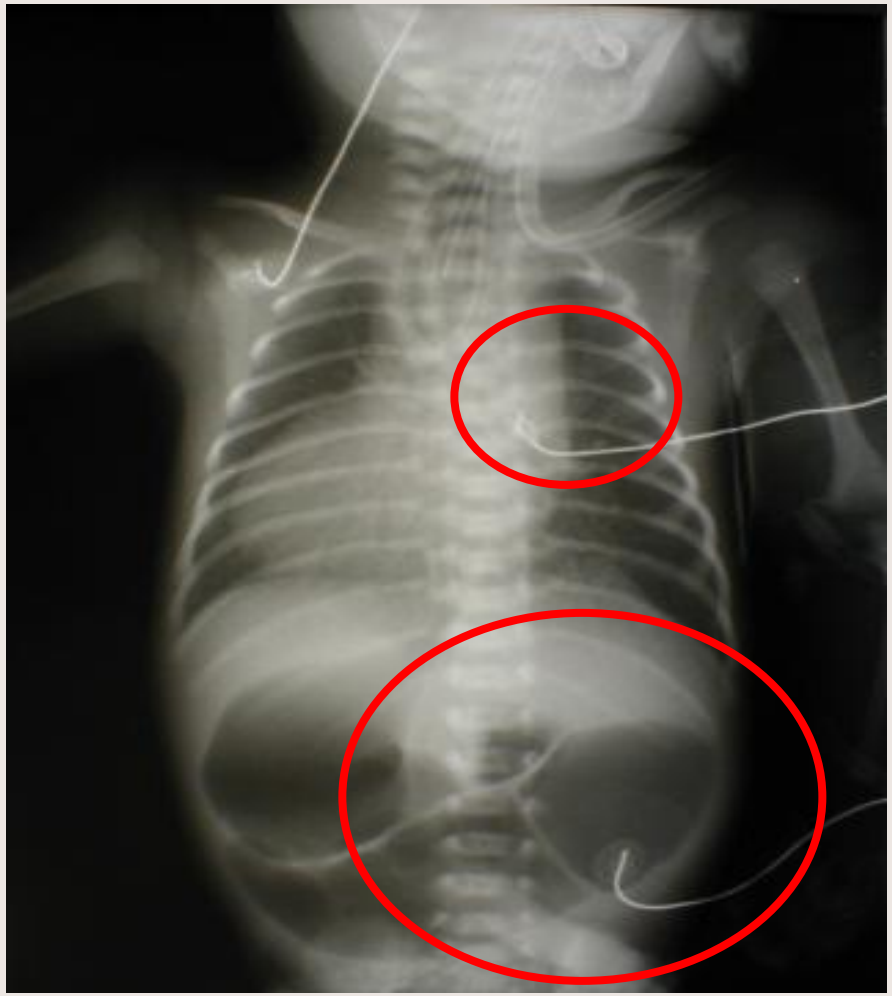
Jejunal atresia

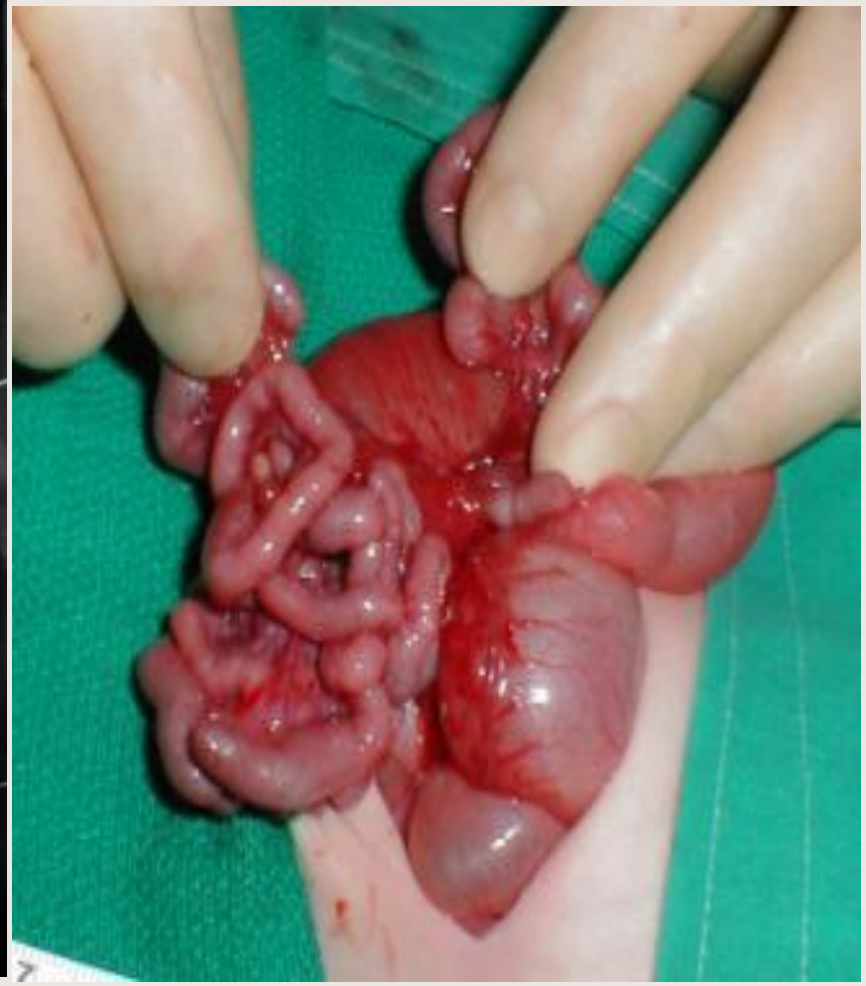
- MC of small bowel atresia; incidence in about 1 per 2,000 live births.
- Atresias in small bowel are due to in – utero occlusion of all or part of the radial blood supply to the bowel.
- Usually presents in the 1st 24 – 72 hrs. with bilious emesis, abdominal distention & failure to pass meconium (70%).
- Large dilated proximal loop or loops
- Plain X ray: 3 air bubbles.

High obstruction of Jejunal atresia



Atresias may be multiple
Jejunal atresia: 3 air bubbles

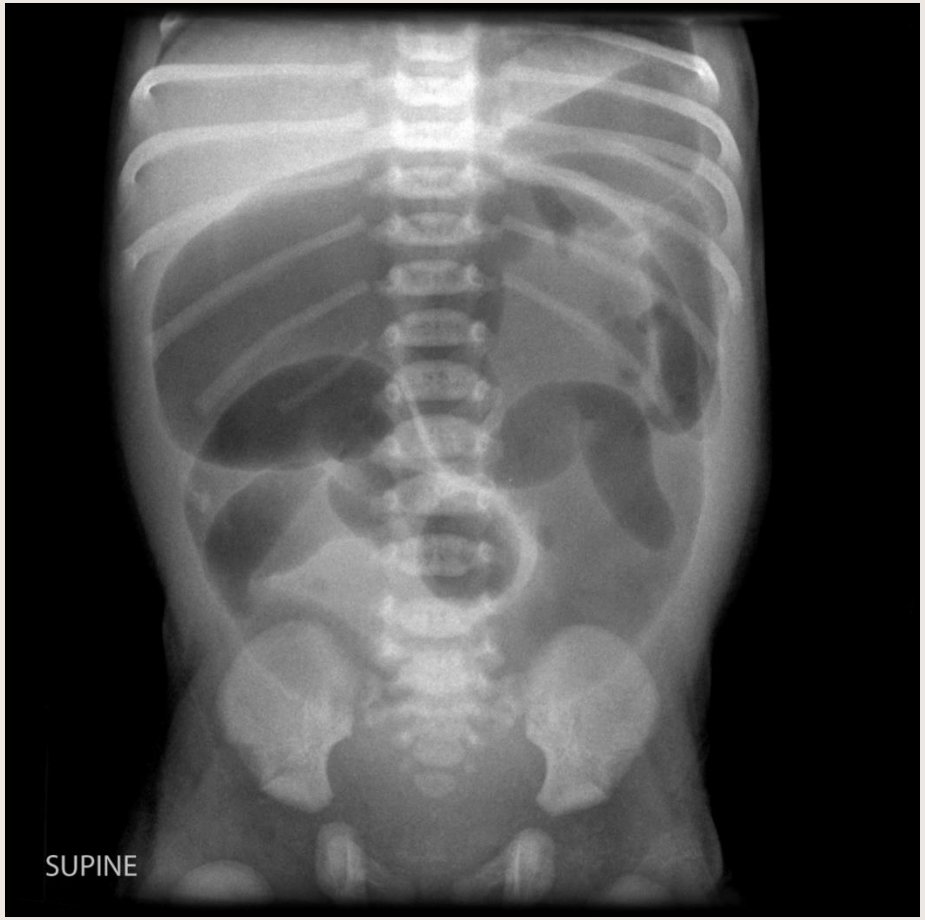




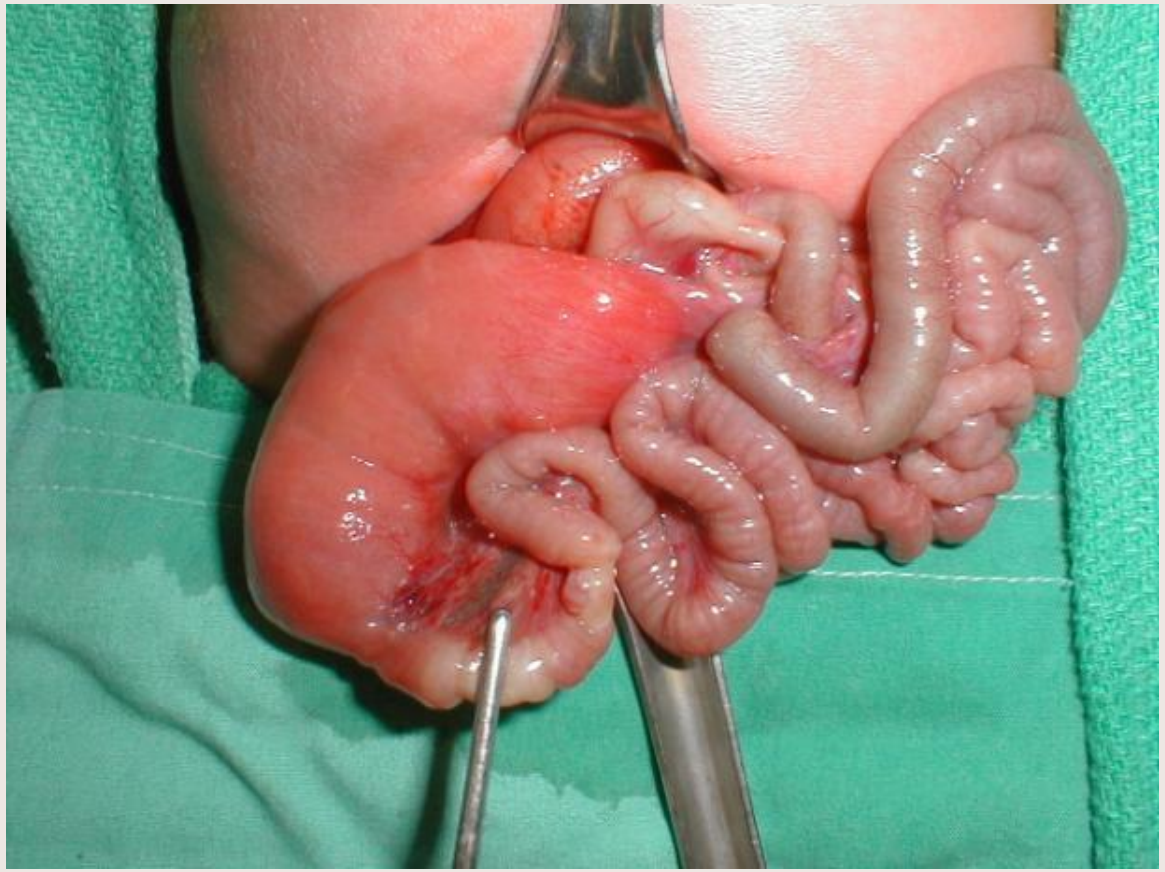
Ileal atresia

- Multiple air bubbles.
- May present in 24 – 48 hrs. with bilious emesis.

Small bowel atresias



Type I atresia
(may be Jejunal or ileal)



Anomalies associated with small bowel atresias

- Other atresias
- Hirschsprung's disease
- Biliary atresia
- Polysplenia syndrome (situs inversus, cardiac anomalies, atresias)
- CF (10%)

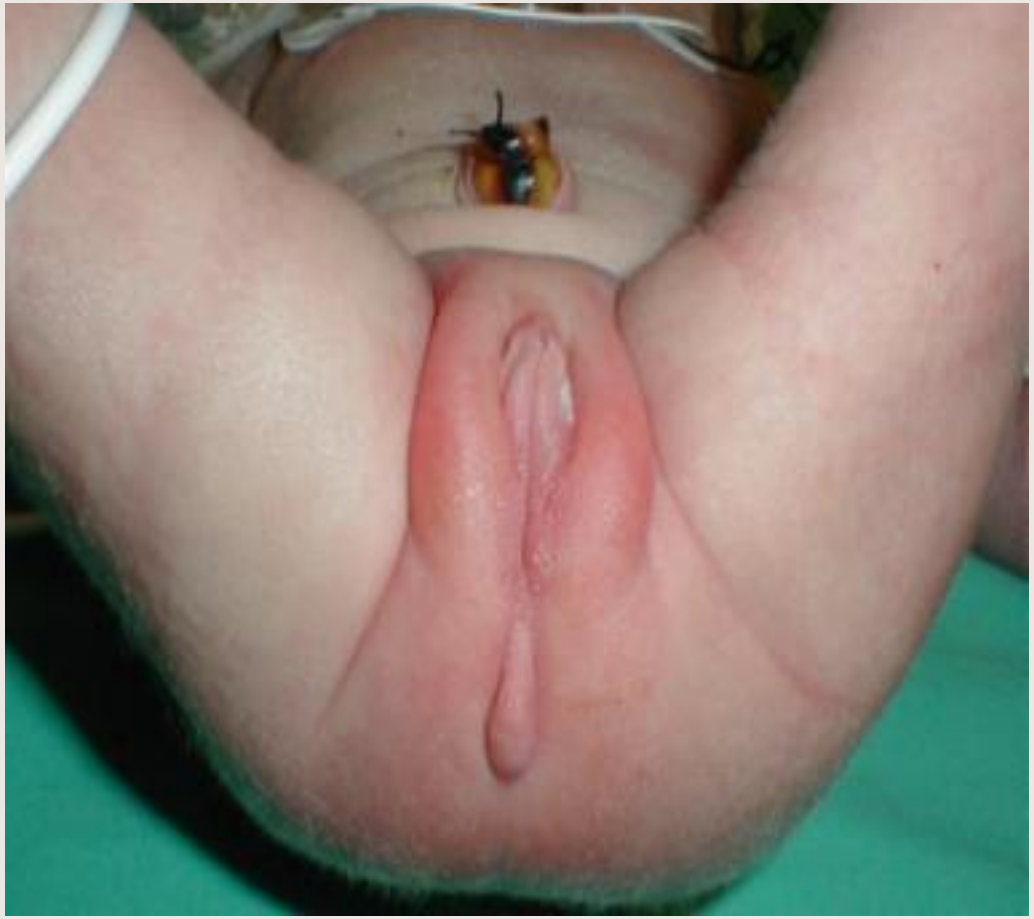
Colonic atresia

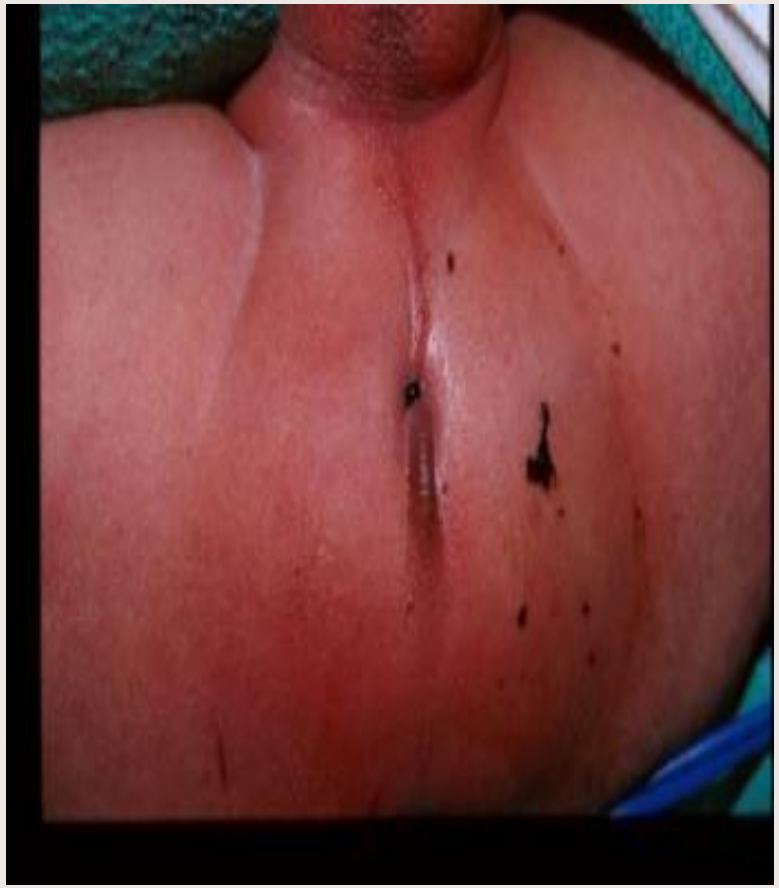
- Rare
- Multiple air bubbles
- Bilious emesis after 2 – 3 days.

Anal atresia

- Should be diagnosed at birth, often a perineal fistula is labeled normal.

Imperforate anus: anal atresia





Diagnosis of atresias

- Plain films: ***dilated loops of small bowel***
- Contrast enema: ***small, unused colon***
- UGI/ SBFT: ***failure of contrast to pass beyond atretic part***

Management

- Stop all feeds
- Start on IVFs
- Usually start on antibiotics esp. in esophageal atresia because of pneumonia.
- Keep the baby warm.

Cont.

- Refer in an ambulance to centers where they can receive definitive management.
- Definitively, correct the defect by laparotomy, intestinal resection & tapered primary anastomosis; other atresias/ associated anomalies are looked for.
- Post – op, place on ***TPN for at least 14 days***.
- Complications in post – op period:
  - Coagulopathy due to deficiency of vitamins A, D, E, K absorbed at the terminal ileum → administer FFP before surgery.
- Stomas may be fashioned if the baby is very sick.

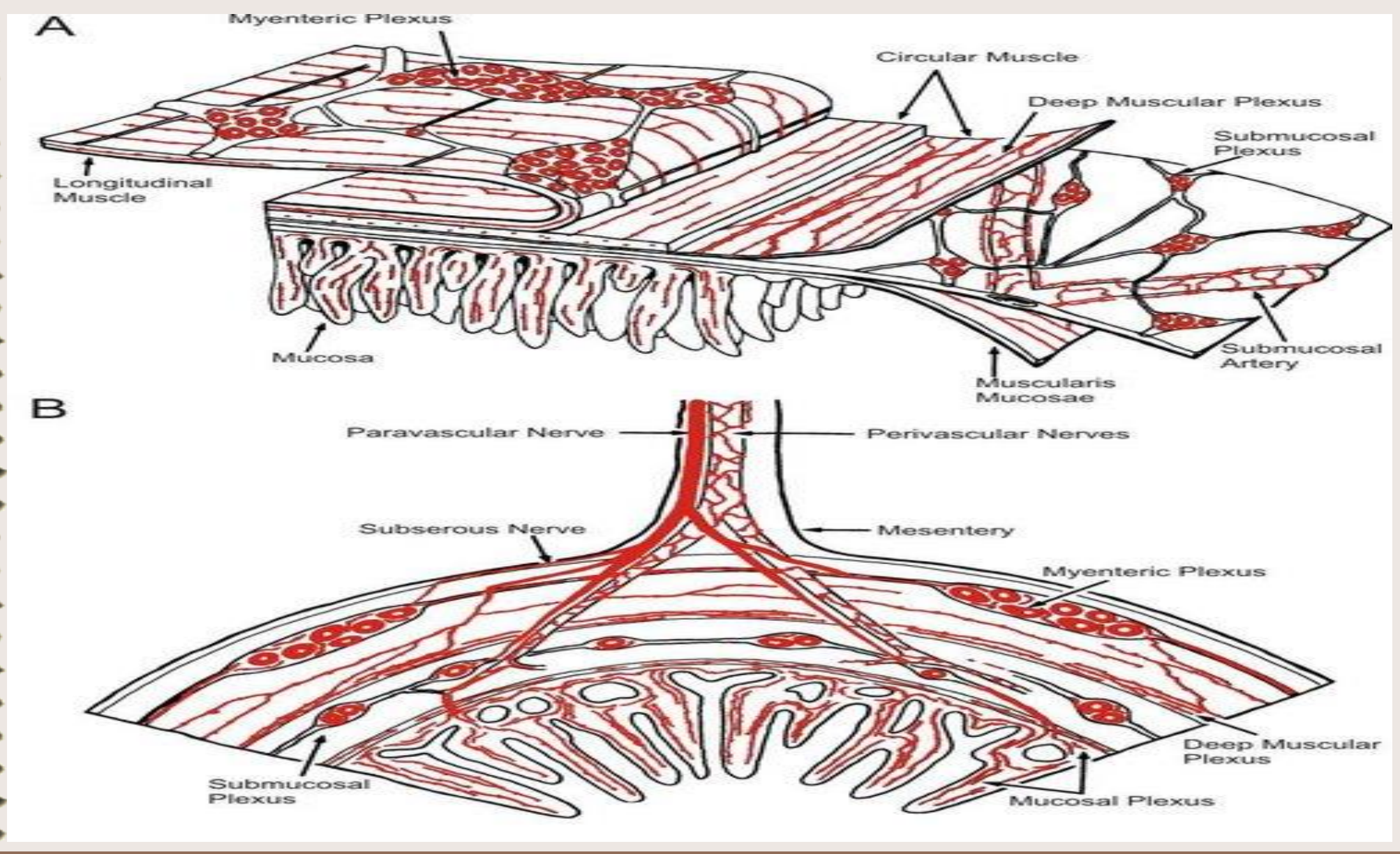
Case

- One day old infant with vomiting, poor oral intake, abdominal distension
- Mother had polyhydramnios
- Has not passed stools
- Vomiting initially non – bilious but now yellowish
- Upper abdominal distension
- Diagnosis: gut atresia

b. Hirschsprung's Disease/ Congenital aganglionic megacolon

Outline

- Introduction
- Epidemiology
- Etiology
- Pathology & pathophysiology
- Clinical presentation
- Hirschsprung’s Associated Enterocolitis
- Investigations
- Treatment


A
Myenteric Plexus
Circular Muscle
Deep Muscular Plexus
Submucosal Plexus
Longitudinal Muscle
Mucosa
Submucosal Artery
Muscularis Mucosae
B
Paravascular Nerve
Perivascular Nerves
Subserous Nerve
Mesentery
Myenteric Plexus
Deep Muscular Plexus
Submucosal Plexus
Mucosal Plexus

Introduction

- These is a developmental disorder of the intrinsic component of the ENS characterized by:
  - ***Congenital absence of intramural parasympathetic ganglion cells (aganglionosis) in the myenteric & submucosal plexi of the distal intestine*** &
  - The presence of ***hypertrophic nerves in the distal large bowel resulting in a functional obstruction***
- The resulting aganglionic segment of the colon fails to relax, causing a functional obstruction.

The aganglionosis

- Is restricted to the rectum & sigmoid colon in 75% of cases i.e. ***short segment disease***
- Involves the proximal colon in 15% of cases i.e. ***long segment disease***
- Affects the entire colon & a portion of the terminal ileum in 10% of cases i.e. ***total colonic aganglionosis***

Cont.

- May present in the 1st few days to weeks of life if the segment of aganglionosis is long.
- Short – segment disease is often tolerated for months e.g. just above the dentate line.
- Starts at anus & extends proximally a variable distance.

Epidemiology

- Incidence: 1 in 5000 live births.
  - Highest incidence: Caucasians > Hispanics > Africans > Asians
- Short segment (rectosigmoid) disease is more common in males (M:F → 4:1); Longer segment disease shows a decreased sex ratio & it approaches 1:1 in total colonic aganglionosis.
- It ***is uncommon in premature infants***.
- ~ 20% of cases are familial; it most commonly presents with a sporadic occurrence (multifactorial complex pattern). Currently, ***approximately 90% of patient's are diagnosed in the newborn period***.

Approximately 30% of patients show an association with:

- ***Down syndrome (3%)***
- ***Neurocristopathy syndromes***: Waardenberg – Shah syndrome, Yemenite deaf – blind – hypopigmentation, Piebaldism, Other hypopigmentation syndromes
- ***Goldberg – Shprintzen syndrome***
- ***Smith – Lemli – Opitz syndrome***
- ***Multiple endocrine neoplasia 2***
- ***Congenital central hypoventilation syndrome (Ondine's curse)***
- ***Isolated congenital anomalies***: congenital heart disease, malrotation, urinary tract anomalies, CNS anomalies

Etiological theories

- ***Embryologic basis***: arrest of Craniocaudal/ aboral migration of neuroblasts originating from the vagal neural crest into the developing bowel that occurs during the 1st 8 – 12 wks. of gestation.
  - Commonest point of arrest is the ***rectosigmoid area*** hence gut contents cannot be propelled beyond the point of arrest.
  - A transition zone exists between the ***dilated, proximal, normally innervated*** bowel & the ***narrow, distal aganglionic segment.***

Cont.

- ***Failure of differentiation of neuroblasts into ganglion cells***
- ***Accelerated ganglion cell destruction within the intestine may also contribute to the disorder.***
  - The enteric microenvironment may become inhospitable for colonization by crest – derived cells if EDN3 is deficient or if the EDRNB is lacking.

Genes

- ✓ At least 8 genetic mutations have been identified in patients with HSD. The predominant gene affected is the ***RET proto – oncogene***. RET malfunction accounts for at least 50% of familial & 20% of sporadic cases & is especially seen in patients with long segment disease.
  - ✓ Deficiency causes migration arrest of neural crest cells in mice
  - ✓ Mutation found in 17 – 38% of short segment & 70 – 80% of long segment disease
- ✓ Other genes include: endothelin 3 (EDN3), endothelin receptor B (EDNRB), endothelin converting enzyme (ECE1), the gene encoding the SRY – related transcription factor (SOX10), the PHOX2B gene.

Cont.

- Majority of HSD cases are sporadic (80 – 90%): mutations of RET gene account 15 – 20% of sporadic cases of HSD; mode of inheritance is a multifactorial complex pattern.
- 10 – 20% are familial:
  - 50% from RET gene at chromosome 10 (AD)
  - Endothelium Receptor B gene at chromosome 13 (AR)
  - Endothelium 3 gene at chromosome 20 (AR)

Pathophysiology

- 3 nerve plexi innervate the intestine:
  1. Submucosal ***Meissner's plexus***
  2. Myenteric ***Auerbach plexus*** (between the longitudinal and circular muscle layers)
  3. Smaller mucosal plexus
- Normal motility is primarily under control of intrinsic neurons (i.e., the enteric nervous system/ 2nd brain)
- Extrinsic neural afferents to the ENS contain cholinergic & adrenergic fibers. The cholinergic fibers cause contraction whereas the adrenergic fibers mainly cause inhibition.

Cont.

- Neural crest – derived cells first appear in the developing esophagus at the 5^{th} week of gestation.
- Craniocaudal migration to the rectum starts by the 12^{th} week of gestation.
- Neural crest cells first form the myenteric plexus.
- After the Craniocaudal migration has ended, the submucosal plexus is formed by the neuroblasts which migrate from the myenteric plexus across the circular muscle layer & into the submucosa.
- This progresses in a Craniocaudal direction during the 12^{th} to 16^{th} weeks of gestation.

Cont.

- In Hirschsprung's disease, failure of migration of vagal neural crest cells results in absence of ganglion cells in ***both the myenteric & submucosal plexi***.
- ***The internal anal sphincter is invariably affected (distention of proximal gut doesn't cause its relaxation)*** & aganglionosis proceeds proximally for a variable distance. The earlier the arrest of migration, the longer the aganglionic segment is.
- In the absence of ENS reflexes, control of intestinal smooth muscle is overwhelmingly extrinsic. The cholinergic (excitatory system) predominates over the adrenergic (inhibitory) system leading to unopposed increase in smooth muscle tone → imbalance of smooth muscle contractility → uncoordinated peristalsis & a functional obstruction.

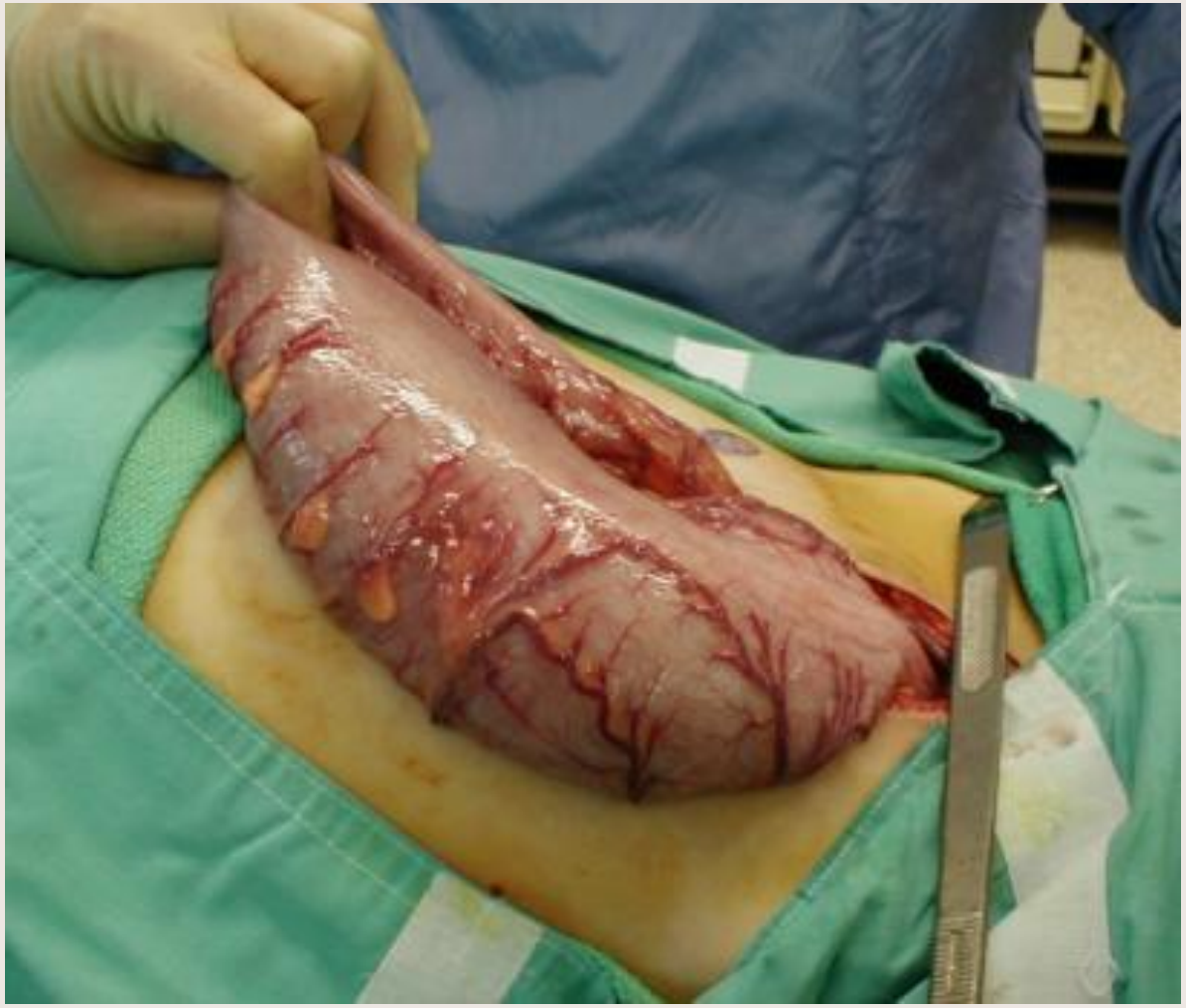
Classification

- Ultra – short segment
- Short segment: confined below the rectosigmoid junction (70 – 80%) → has a male preponderance
- Long segment: below the splenic flexure (20%): M = F
- Total colonic aganglionosis: appendix is involved (9%)
- Total intestinal involvement (2 – 5%)

Macroscopic features

- Narrow aganglionic segment
- A transitional zone
- Dilated proximal portion with a thickened bowel wall as a result of hypertrophy of the muscular wall of the intestine.

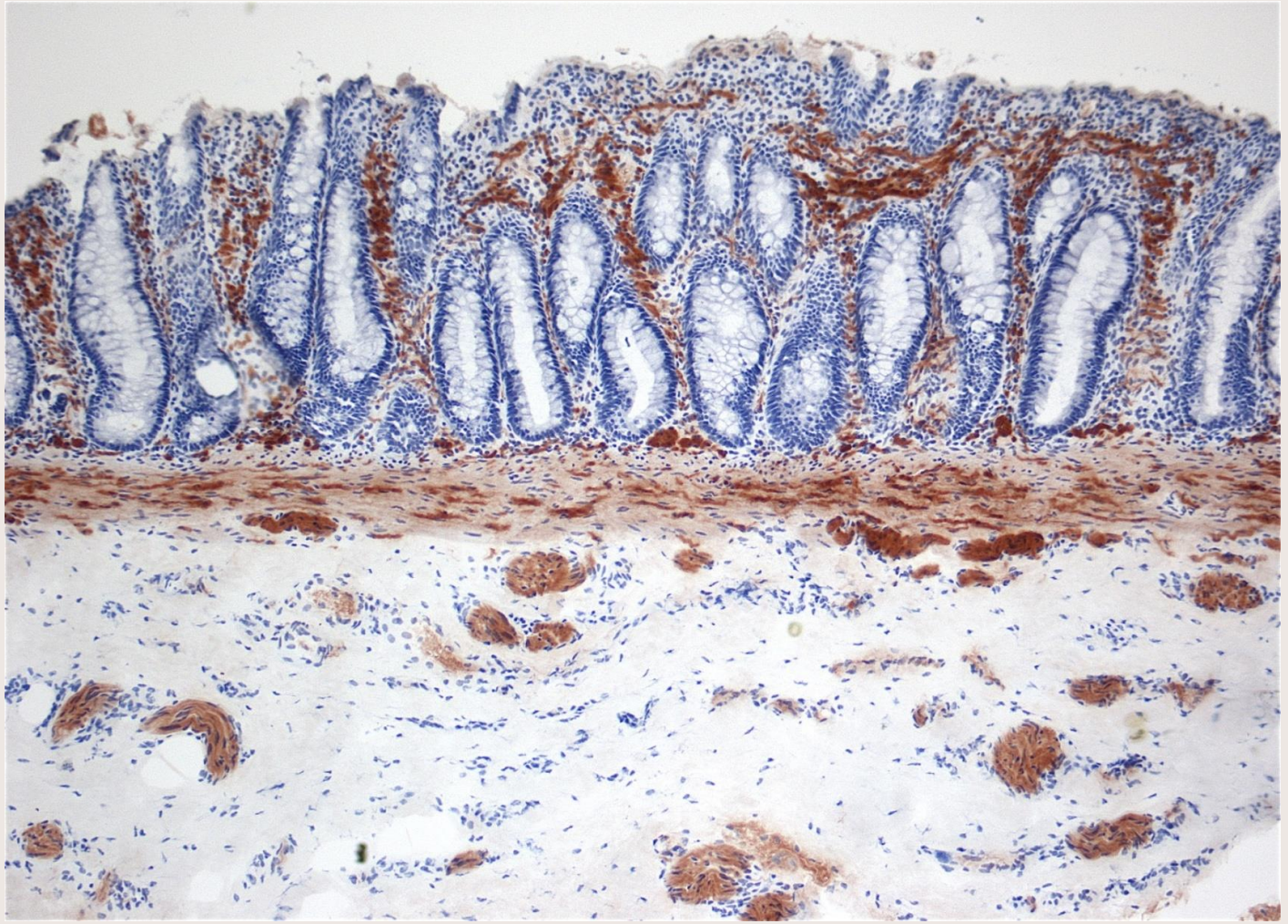
Hirschsprung's in an 8 yr. old



Microscopic features

- Absence of ganglion cells in the IM myenteric/ Auerbach's & the submucosal Meissner's plexi.
- Proliferation of peripheral nerves may also be seen in the affected bowel.

Histopathology: enzyme histochemistry showing aberrant AChE – positive fibers (brown) in the lamina propria



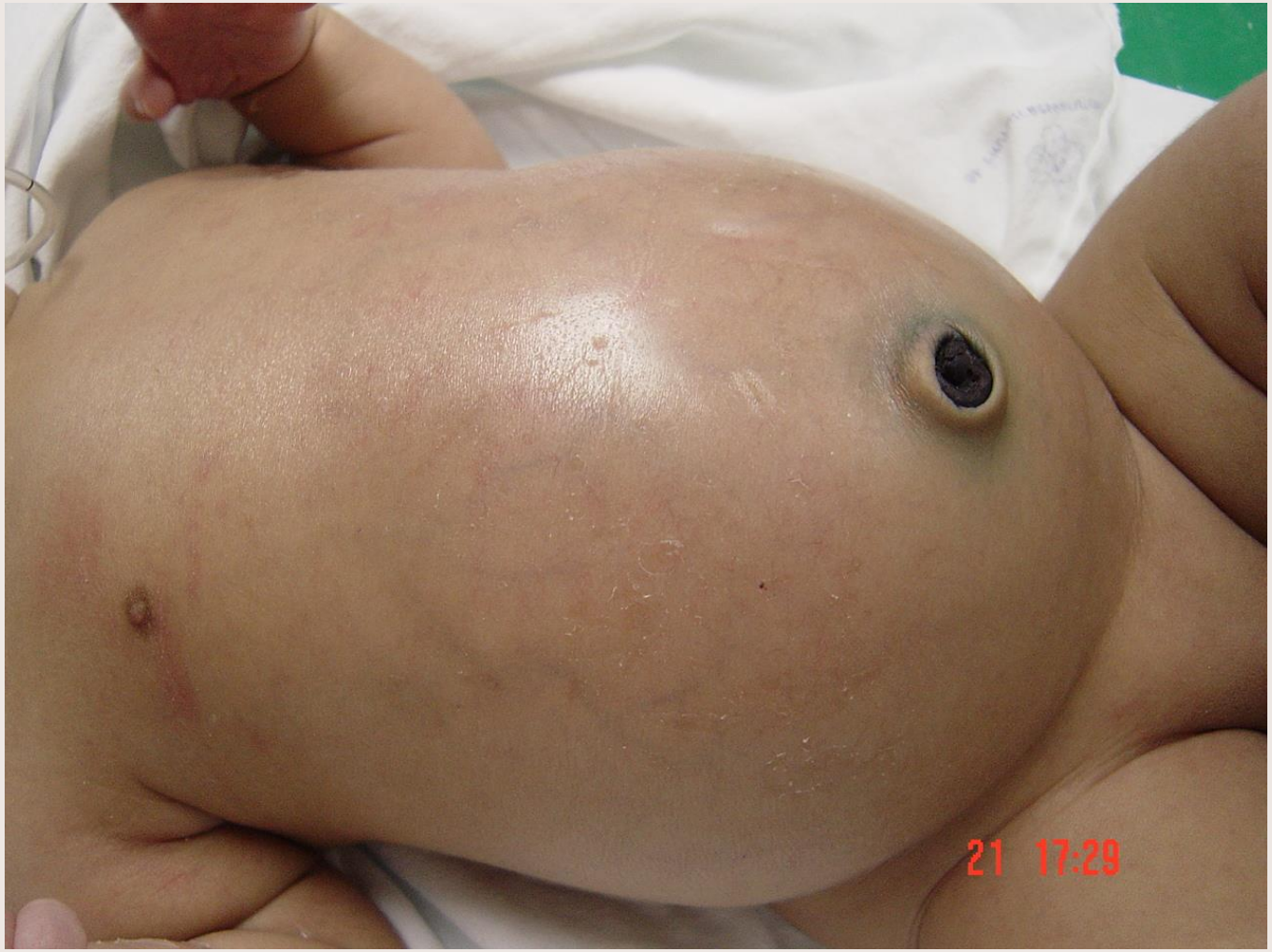
Clinical presentation

- Depends on extent & age of patient.
- DRE is important to R/O ARMs.
- Delayed meconium passage:
  - 95% of normal children pass meconium in 24hrs.
  - 10% of children with HSD pass meconium within 24 hrs.

In infants

- It typically presents in the neonatal period with ***delayed (>24h – 72h) passage of meconium, abdominal distention & bilious vomiting***.
- Decreased defecation frequency: constipation
- Requires digital stimulation/ suppositories
- Decreased appetite
- FTT
- Intermittent episodes of obstruction & diarrhea (enterocolitis)

Cont.



Childhood

- Severe, chronic constipation: failure to open bowel for ≥ 4 days.
- Inability to open bowels without aid (enema) esp. in children with long segment disease.
- Gross abdominal distention with a visible outline of bowel loops.
- Bacterial overgrowth & toxin production → massive inflammation of gut epithelium resulting in malabsorption syndromes → Malnutrition/ FTT
- Large fecal masses
- Do not soil, bleed or have anal pain (more consistent with functional constipation)
- Often have extensive enema programs.

On exam

a. Wasted, dehydrated, sometimes febrile.
b. Abdomen distended with palpable multiple fecal filled loops.
c. Rectal examination reveals hypertonic sphincter with an empty rectum.
d. Rectum examination: is often followed by an explosive discharge of fluid or by foul smelling gas, stools and mucous and is indicative of some degree of enterocolitis.

Hirschsprung's Associated Enterocolitis (HAEC)/ Toxic Megacolon

- Severe enterocolitis is a potentially fatal complication of Hirschsprung's disease. It is very rare to get a enterocolitis with idiopathic constipation & it is usually only seen with Hirschsprung’s disease or ulcerative colitis.
- Mortality: 20 – 30% in some studies.

Etiology

- Mechanisms underlying HAEC are not fully understood.
- Theories:
  - Stasis & reduced production of protective mucin → luminal bacterial overgrowth in bowel proximal to involved segment.
  - Commensal bacteria (e.g. ***bifidobacterium*** & ***lactobacillus***) are reduced & pathogenic bacteria (e.g. ***C. difficile – MC***) are isolated. ***Rotavirus*** & ***S. aureus*** may also have a role

Cont.

- The bowel wall in the affected area is subsequently invaded by colonic organisms which can lead to intestinal perforation, peritonitis, systemic sepsis, shock & death.
- The incidence of HAEC ranges from 24 – 34% in different series of patients.
- HAEC can occur prior to surgical intervention, in the immediate post – operative period or even yrs. after definitive repair.

Cont.

- Post – operative HAEC usually occurs soon after the definitive pull – through repair, ranging from 3 wks. – 20 months post – op.

Risk factors

- Delay in diagnosis of HSD (> 1 wk. of age)
- Increased length of aganglionic segment
- Trisomy 21 (possibly caused by an underlying immune deficiency in these children)
- Presence of other associated anomalies.

Cont.

- Post – op risk factors:
  - Coexisting intestinal neuronal dysplasia (IND) or hypoganglionosis proximal to the resected aganglionic segment.
  - Presence of an anastomotic stricture or leak, or any form of intestinal obstruction has been associated with up to a 3.5 – fold increase in the incidence of HAEC.
  - Children who have had HAEC before their definitive surgery.

Clinical presentation

- Fever, lethargy, anorexia, spurious vomiting, abdominal distention & foul smelling diarrhea.
- Explosion of gas and liquid stool may occur following DRE.

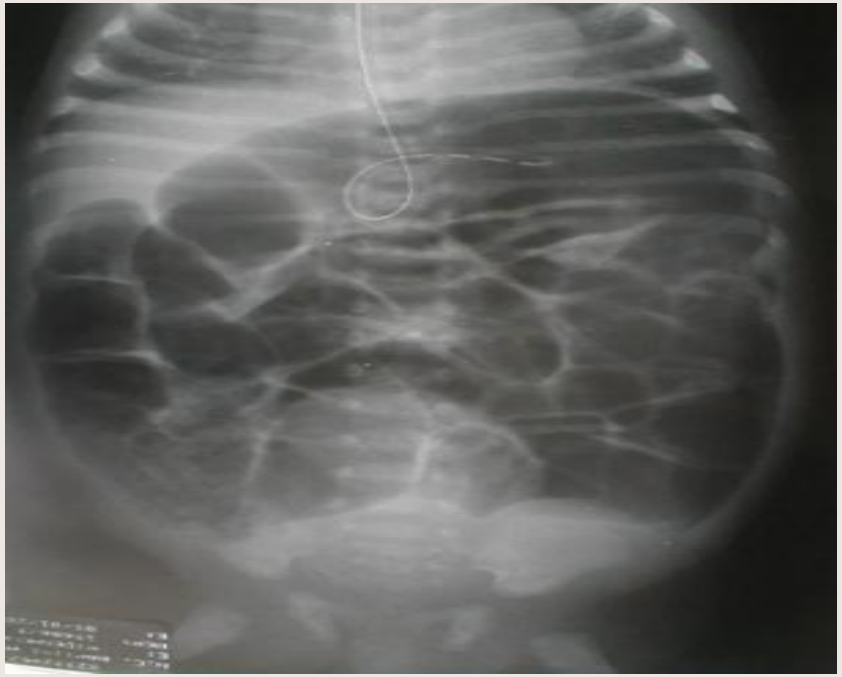
Clinical grading system for HAEC

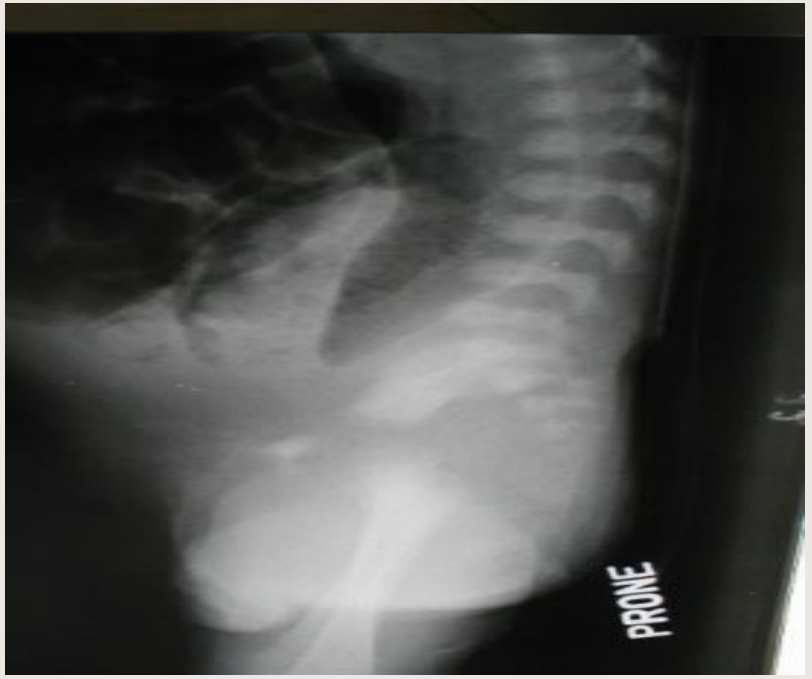
Grade	Features
I	Mild explosive diarrhea; mild or moderate abdominal distention; mild systemic symptoms
II	Moderate explosive diarrhea; moderate to severe abdominal distention; mild systemic symptoms
III	Severe explosive diarrhea; marked abdominal distention; shock or impending shock

Plain Abdominal X – ray for HAEC

- Obstructive picture with air – fluid levels & dilated bowel loops.
- Findings specifically associated with HAEC include: distention of the proximal colon & the ‘cut – off sign’ or absence of air in the distal rectosigmoid colon, with an abrupt cut – off at the level of the pelvic brim.
- Pneumatosis intestinalis is occasionally seen.

Cont.





- Fecal loading (mottling) with distended loops of bowel; Very many gaseous shadows
- Empty pelvis

Management of HAEC

- Volume resuscitation
- IV BSA Abs covering aerobic & anaerobic organisms.
- Repeated rectal irrigation through rectal tube with saline; this decompresses the colon & often decreases the severity of disease.
- Diverting colostomy if no improvement from above.

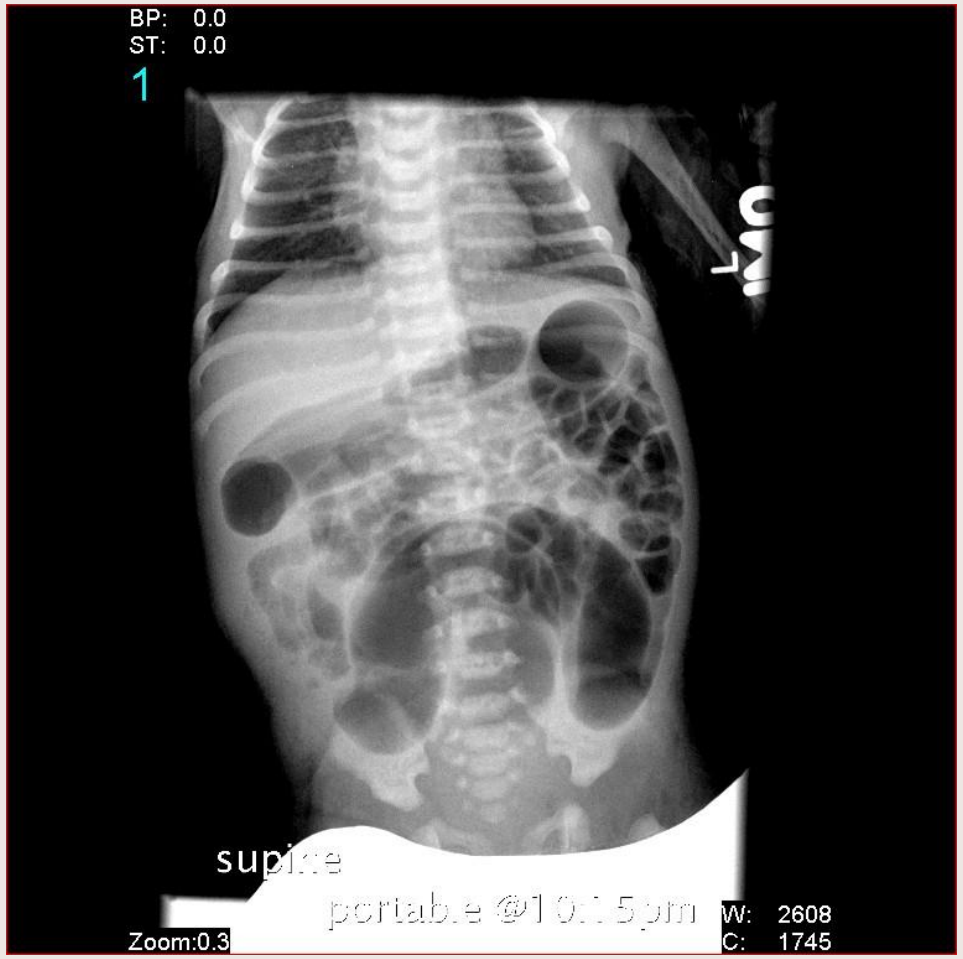
Diagnosis of Hirschsprung's disease

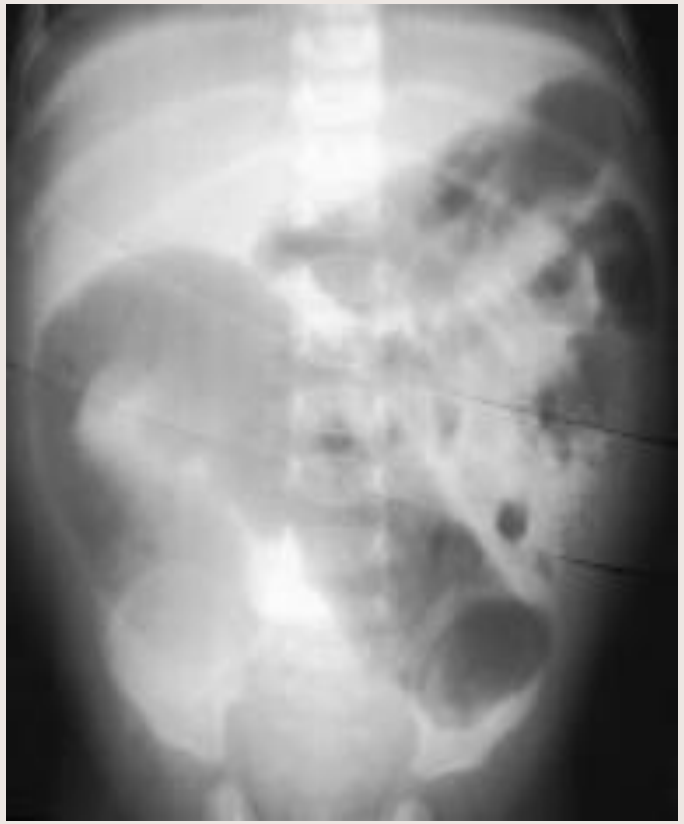
- History
- Clinical Examination
- Investigations:
  - Radiology: Plain AXR, contrast studies
  - Manometry (Barium or Gastrografin), histopathology

Radiology: AXR

- Diagnostic accuracy: 52%
- Look for signs of low IO & distended bowel loops of different calibers
- Erect films can demonstrate air – fluid levels
- A lateral view may demonstrate the ***narrow rectum with absence of gas in the rectum***.

Frontal abdominal radiograph showing marked dilatation of the bowel with no gas in the rectum.





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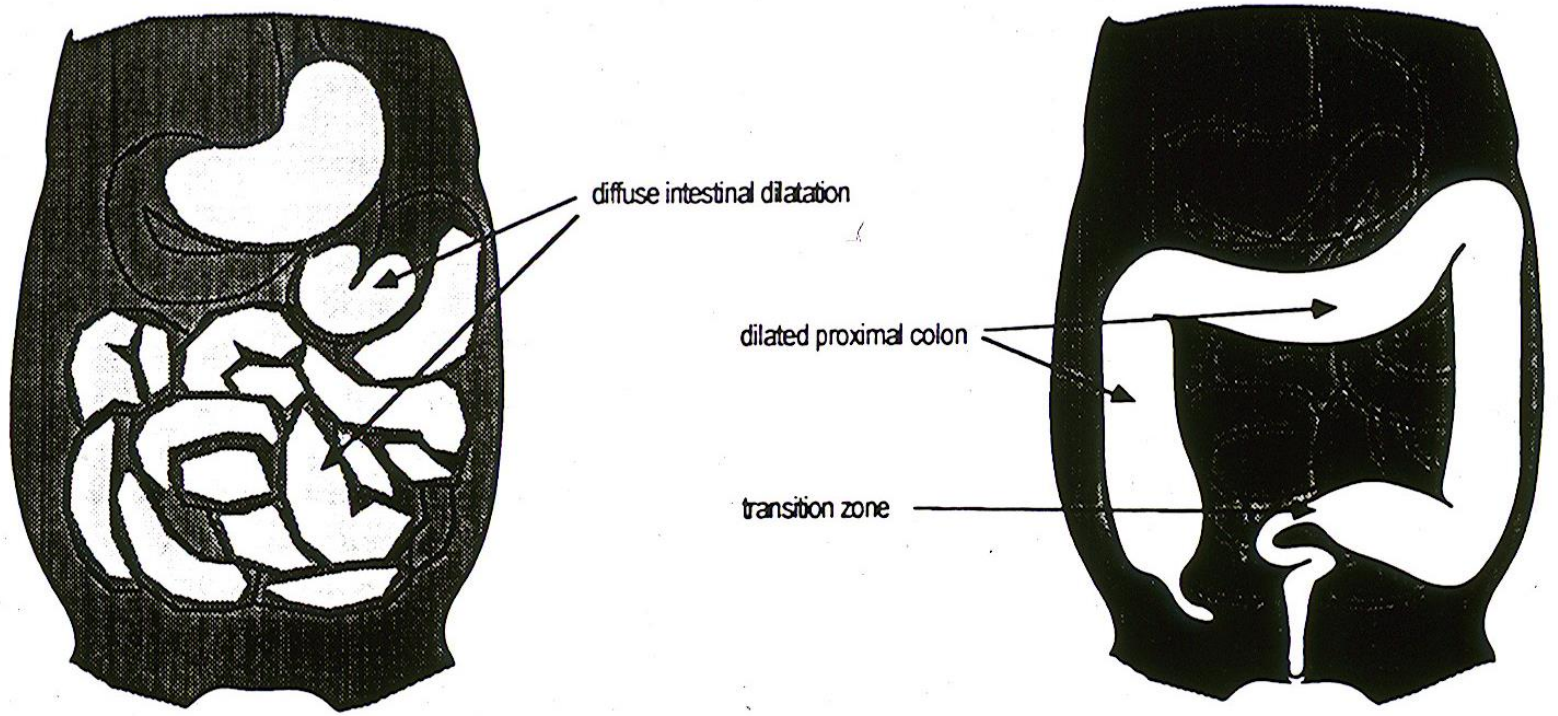


Figure 2. Radiological features of Hirschsprung's disease on plain film (left) and contrast enema (right).

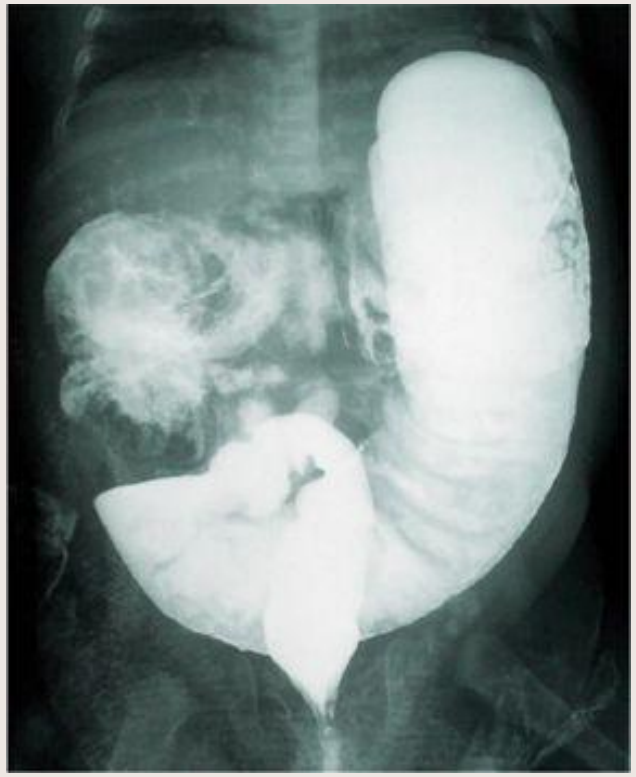
Barium enema

- They are not useful in neonates but they are useful in older children.
- Most important view is lateral projection, in which a rectal transition zone will be most evident.
  - R/O an obstructing lesion e.g. meconium plugs in a neonate
  - Rectal caliber ≤ calibre of rest of colon
  - Hallmark: conical transition from proximal dilated to distal narrowed colon
  - ~ 10% of neonates may not have a demonstrable radiographic transition zone.
  - Small colonic calibre with rounded flexures suggests aganglionosis.

Cont.

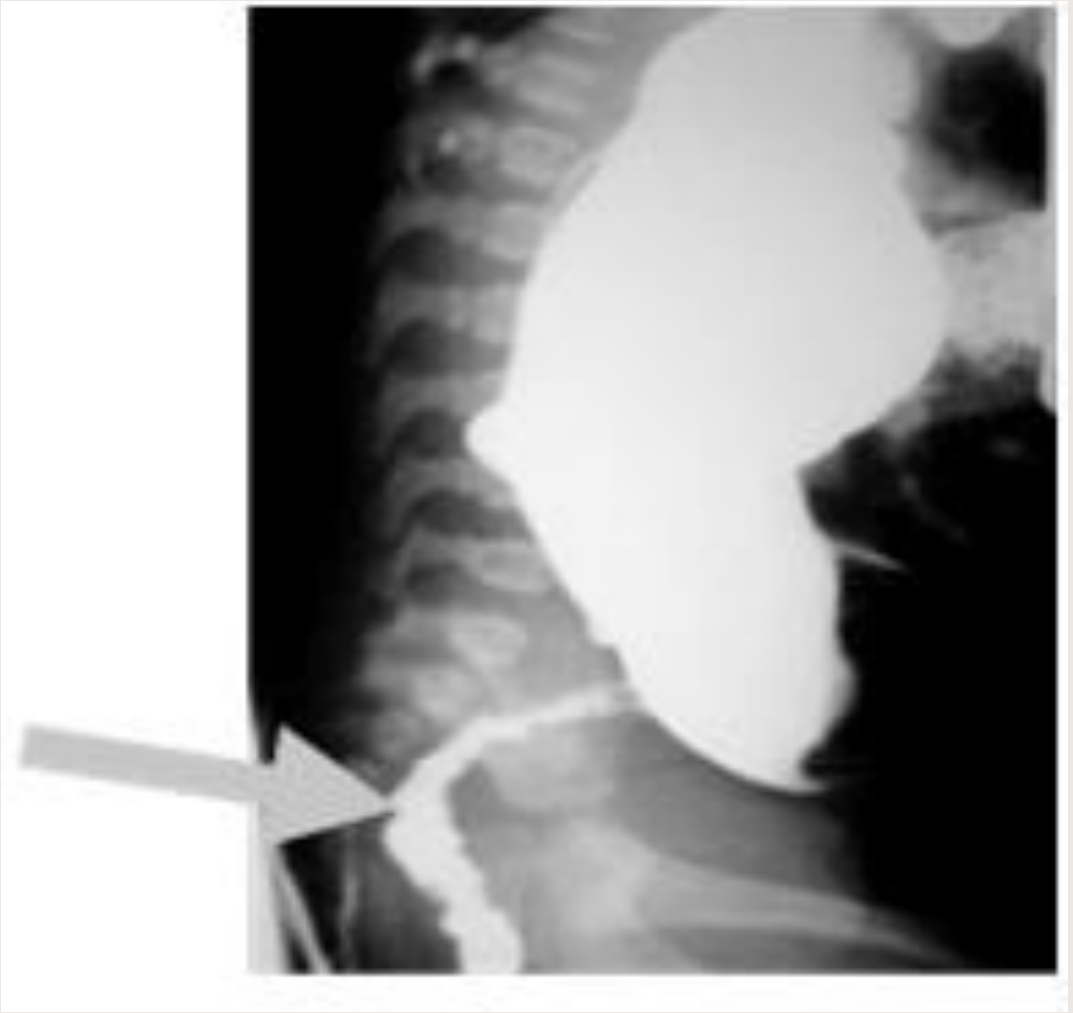
- Irrigation of the colon before the contrast must not be performed as it may result in possible decompression of the megacolon or the distended bowel.
- Care must be taken not to apply pressure in neonates as the bowel can easily be distended.

Lateral view from a barium enema examination depicting the reduced diameter of the rectum & sigmoid.



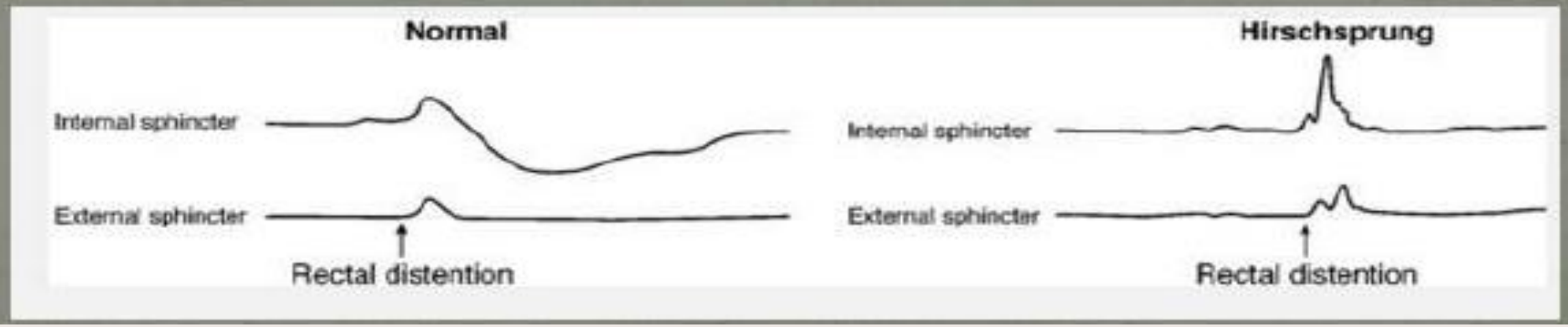
Cont.

- The aganglionic segment may be irregular, demonstrating a ***sawtooth appearance***, probably as a result of muscular fasciculation.
- A delay in the clearing of contrast (barium sulphate) within 24 hrs. is also a reliable sign & a follow up X – ray should be performed the following day.



Ano – rectal manometry

- Rectal manometry:
  - Helpful In older children who present with chronic constipation and an atypical history.
  - Children with HD fail to demonstrate reflex relaxation of the internal anal sphincter in response to inflation of a rectal balloon.



Full thickness or suction rectal/ colonic biopsy

- Gold standard of diagnosis.
  - Suction rectal biopsy is done in neonates at the bedside & full thickness rectal biopsies is done in older children in theater.
- Assesses the plexi. It is important to get a biopsy 2 – 3 cm (3 – 5 cm in older children) above the dentate line.
- Disadvantage: requires GA, presacral abscess may form.
- Alternatives: punch biopsy, suction biopsy.

Stains used

- Hematoxylin & Eosin:
  - Identifies ganglion cells
  - Difficulties in identifying immature ganglion cells from plasma cells or lymphocytes
- AChE staining
  - Shows an increased activity in the parasympathetic nerves of the affected zones as well as neurofibrils within the lamina propria & muscularis mucosa.

Histological findings in HSD

1. Absence of ganglion cells in the submucosal Meissner's & inter – myenteric Auerbach's plexus.
2. Presence of the enlarged peripheral nerve trunks in the submucosa: ***due to hypersecretion of Acetylcholine in an attempt to stimulate an unresponsive bowel.***
3. Increased AChE staining: proliferation of neurofibrils in the lamina propria and the muscularis mucosa (absent in normally innervated intestine). Not done routinely.

Differential diagnoses

- Neonates:
  - Small left colon, meconium plug, meconium ileus, megacystis – microacolon – intestinal hypoperistalsis syndrome (MMIHS), intestinal dysmotility, hollow viscus myopathy, chronic intestinal pseudo – obstruction
- Infants & Children:
  - Trauma, spina bifida, anal fissures, IND, DM, Hypothyroidism, psychosocial.

Management of HSD

- Initial aim: confirm diagnosis
- Subsequent aim: correct problem (i.e. relieve the IO) by means of definitive surgery; removal of non – functional intestines establishment of intestinal continuity.
- Emergency management is tailor made according to the mode of presentation.

Supportive management

- Rehydration: IVFs
- Decompression:
  - NGT
  - Rectal wash – out enemas
  - Laxatives
- IV antibiotics

Principles of surgical management

- A Defunctioning colostomy followed by a definitive ***pull through procedure,*** i.e. staged treatment (standard treatment in this country)
  - Surgery aims to remove the aganglionic segment & 'pull – through' ganglionic bowel to the anus.
- Bowel irrigation techniques allows for early one – stage surgery to be performed at a much earlier age (mostly still within the neonatal period).
- Colostomy may still be necessary if nursing care is unreliable. Site of colostomy depends on the length of involvement. At time of colostomy, full thickness rectal biopsies are taken to confirm the diagnosis initially made & serial biopsies are taken to establish the length of involvement.

Surgical procedures

- Swenson procedure
- Duhamel procedure
- Soave/ endo – rectal procedure
- Laparoscopic pull – through
- Trans – anal (perineal) pull through → one stage
  - Indication: short & ultrashort segment disease.
- Posterior myectomy +/- lateral sphincteretomy

Swenson procedure

- Sharp extra – rectal dissection down to just above the anal canal.
- Aganglionic colonic segment resected.
- End – to – end anastomosis of normal proximal colon to anal canal.
- Completely removes defective aganglionic colon.
- Most commonly used.

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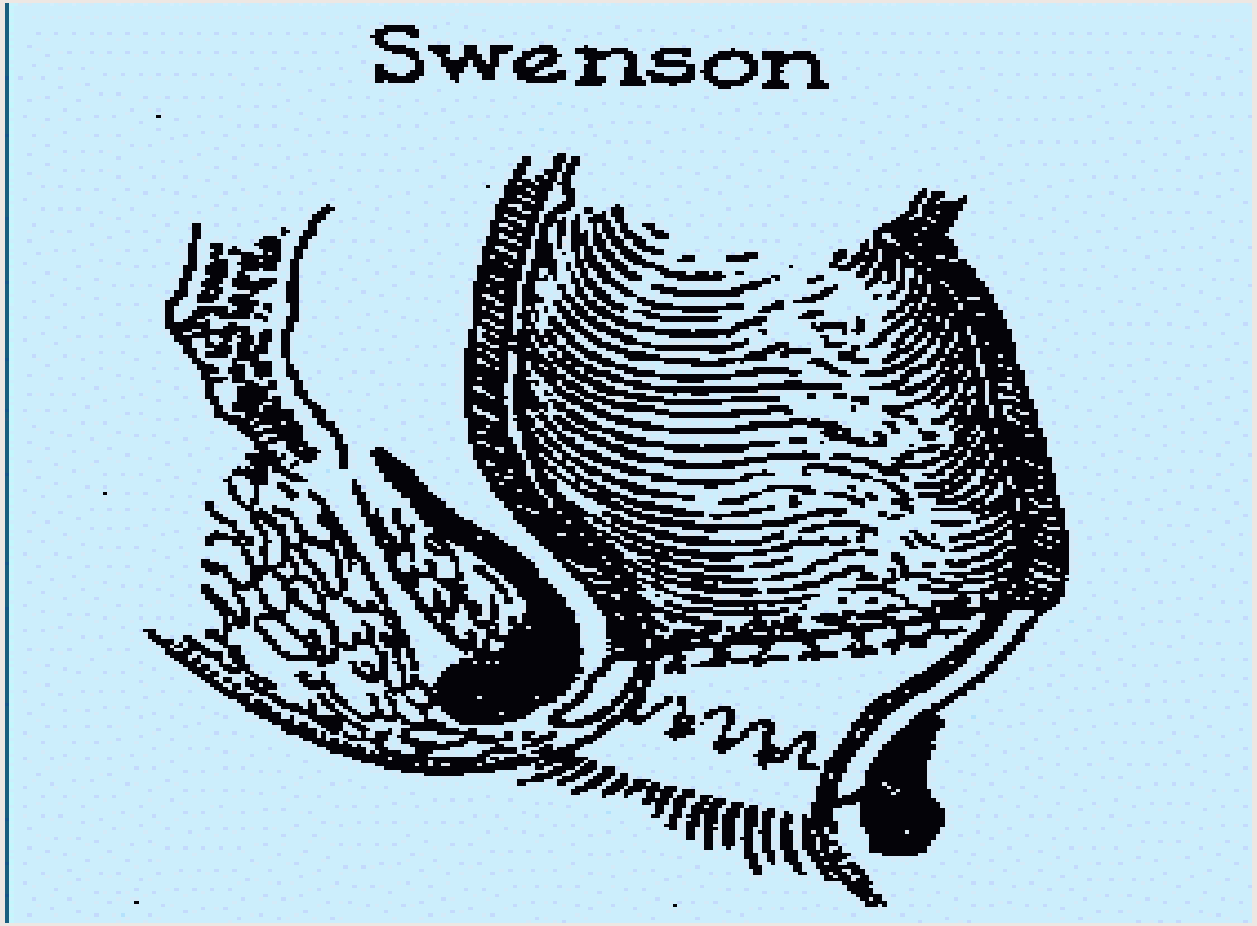

Soave/ Endorectal procedure

- Circumferential cut through the muscular coat of colon at peritoneal reflection.
- Mucosa is separated from the muscular coat down to the anal canal.
- Proximal normal colon is pulled through retained muscular sleeve.
- Telescoping anastomosis of normal colon to anal canal.
- Commonly used. (Soave & Swenson are the major operations used)

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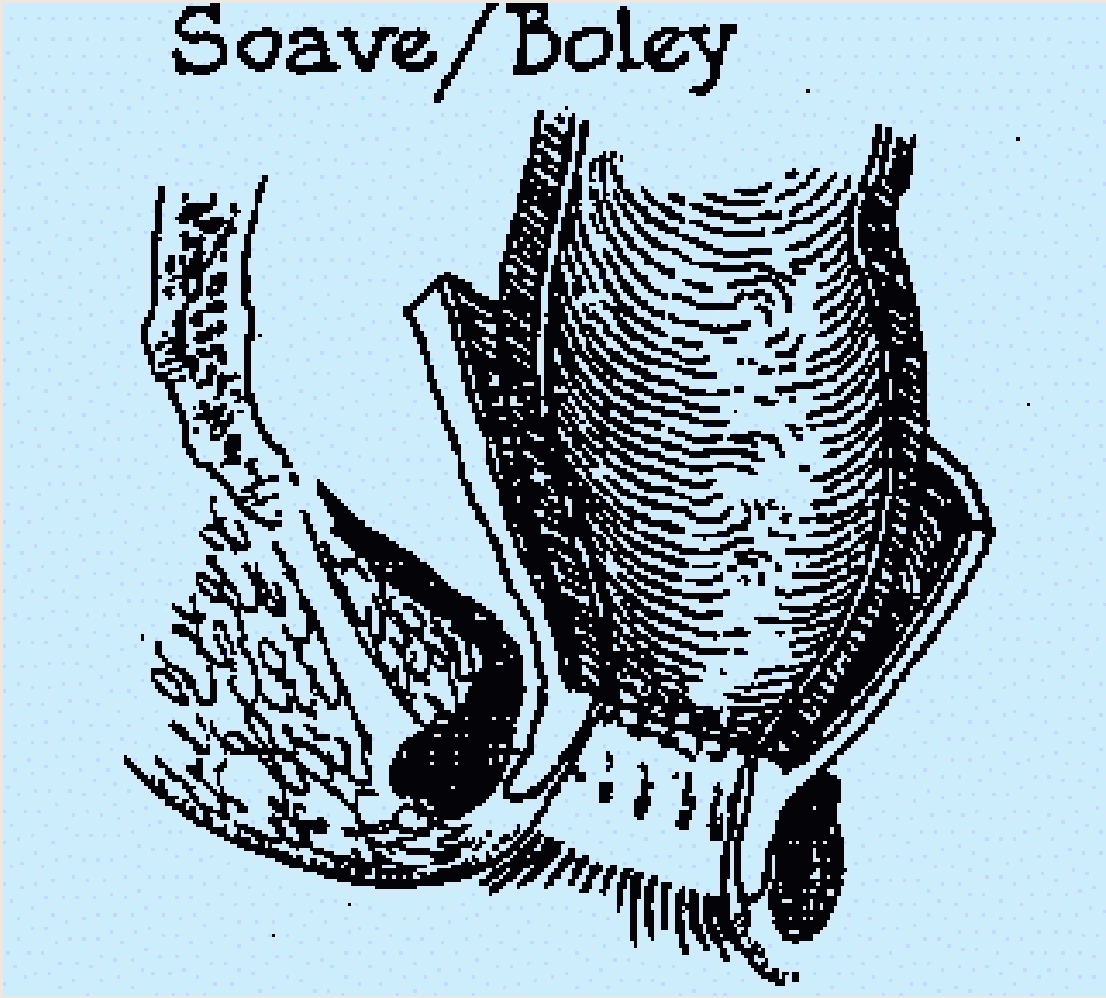

Cont.

- Advantage: rectal intramural dissection ensures no damage to pelvic neural structures.
- Disadvantage:
  - –Higher rates of enterocolitis & diarrhea.
  - –Cuff abscesses may form.

Duhamel procedure

- Portion of defective colon segment is resected → end – to – side anastomosis to left over portion of rectum.
- ***Constipation is a major problem due to remaining aganglionic tissue.***
- Simpler operation as it involves less dissection (pelvic dissection is avoided)

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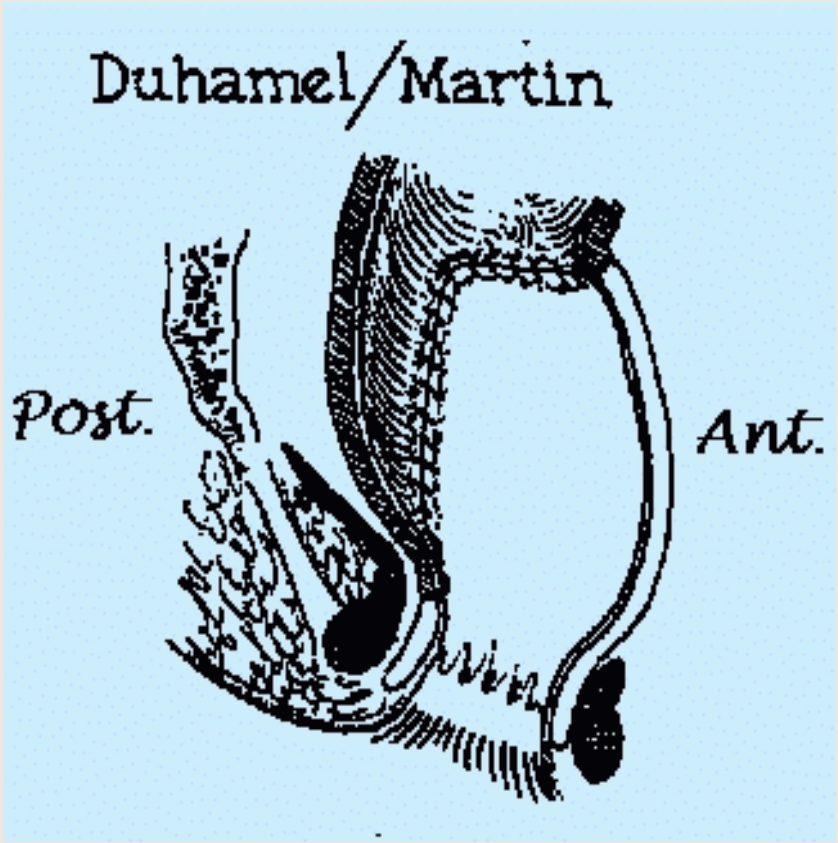
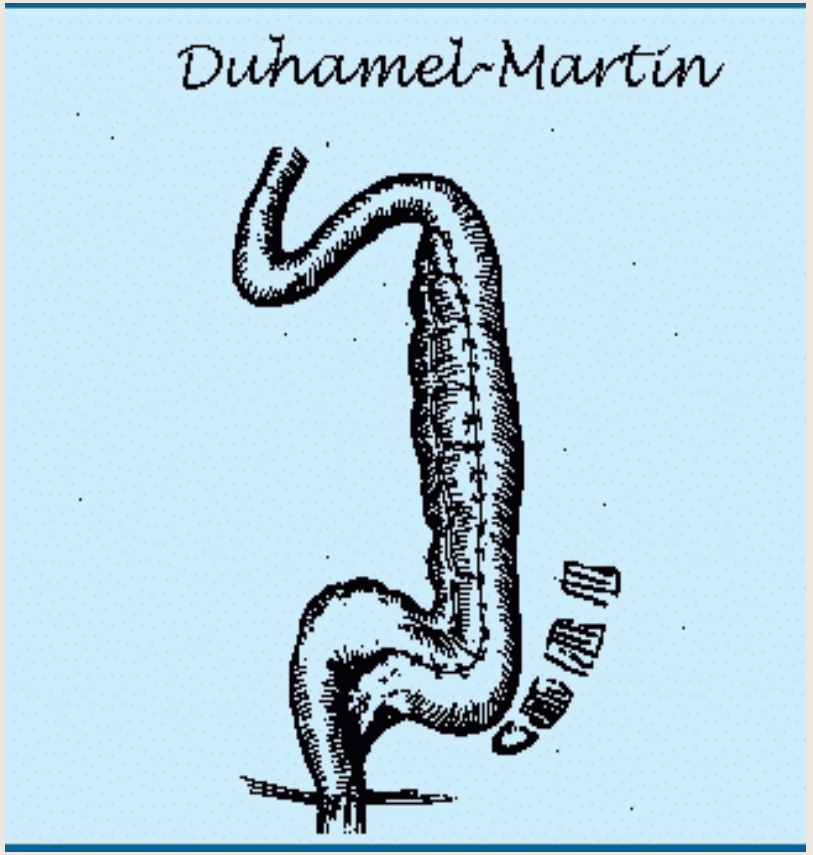




Others

- Rehben:
  - –For long segment disease
  - –Anterior resection
- Martin’s procedure
  - –Total removal of the colon

Operative complications

- Constipation
- At anastomosis sites:
  - Leak at anastomosis (5 – 7%); Bowel retraction; Cuff abscess
- Post – op enterocolitis: 19 – 27%
- Rectal stenosis
- Stricture formation
- Incontinence
- Rectal achalasia due to incomplete resection (8%)

One vs. two stage procedure

- Historically, a 2 stage procedure was performed as follows: preliminary colostomy then completion pull through.
- This was done because the delicate muscular sphincters of the newborn may be injured.
- In the 1980s, a 1 stage procedure became more popular.

Cont.

- Early complications: there is no difference in incidence of anastomotic leak, pelvic infection, prolonged ileus, wound infection or wound dehiscence in between the 2 procedure types.
- Late complications: there is no difference in incidence of anastomotic stricture, late obstruction, incontinence, urgency.
- Post – op enterocolitis is higher in the 1 stage procedure (42% vs. 22%)

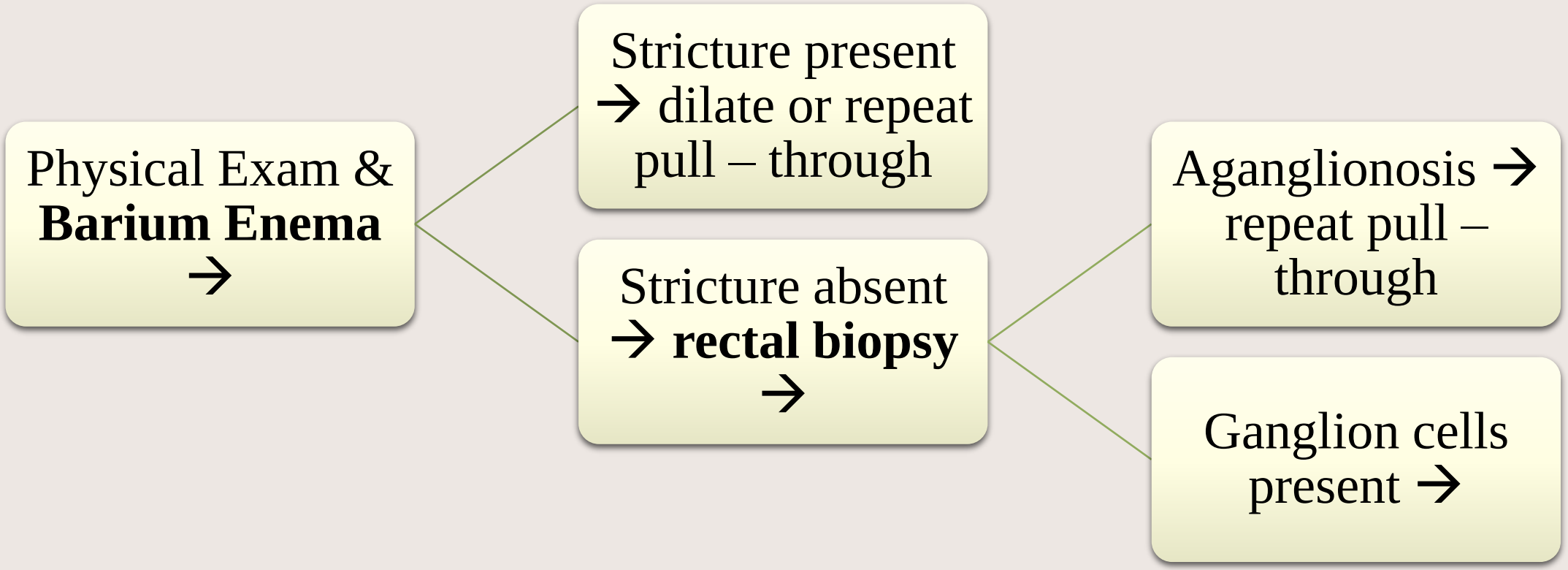
Laparoscopic techniques

- Small studies of laparoscopic pull through procedures
- Excised aganglionic tissues removed through anal canal, no abdominal incision
- Better results in terms of pain, return of bowel function, hospital stay
- Similar incidence of leaks, pelvic abscesses, enterocolitis, postop bowel function

Follow up

- Obstructive symptoms may suggest
  - Mechanical obstruction: stricture
  - Recurrent or acquired aganglionosis
  - Disordered motility in the proximal colon or small bowel
  - Internal sphincter achalasia
  - Functional megacolon caused by stool – holding behavior.

Algorithm for the investigation & management of the child with obstructive symptoms following a pull – through



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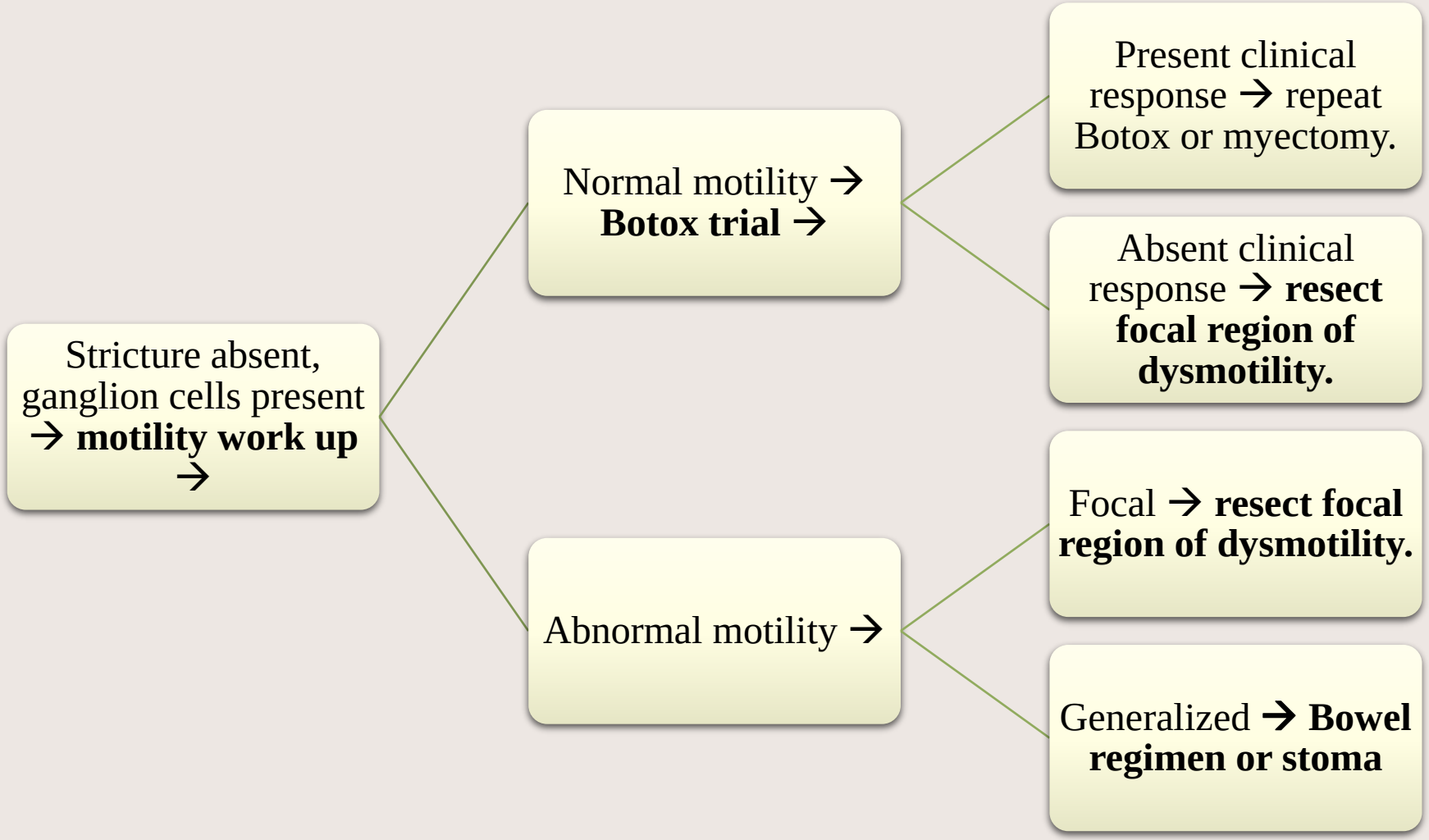

Stricture absent, ganglion cells present → **motility work up** →
Normal motility → **Botox trial** →
Present clinical response → repeat Botox or myectomy.
Absent clinical response → **resect focal region of dysmotility.**
Abnormal motility →
Focal → **resect focal region of dysmotility.**
Generalized → **Bowel regimen or stoma**

Prognosis

- Most (> 90%) patients achieve good bowel control but a significant minority experience ***residual constipation*** &/or ***fecal incontinence (~1%)*** or further ***enterocolitis***.
- Factors associated with poor clinical outcome:
  - Total colonic aganglionosis → 33% of patients experience persistent incontinence & 14% require permanent ileostomy.
  - Associated chromosomal abnormalities

Case 5

- 4 day old female presents to A&E with lethargy, abdominal distension, vomiting
- 37 wk. gestation, Twin A
- Small ASD, no other medical problems
- Mom says patient not making as many diapers as her twin sister and not eating as much
- PE: abdominal distension, rectal exam (make sure you stand to the side)

c. Malrotation

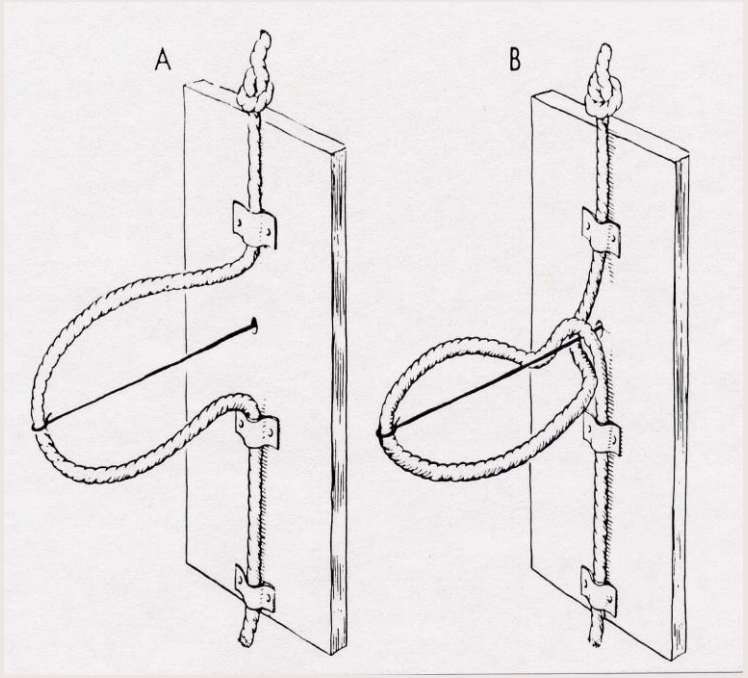
Introduction

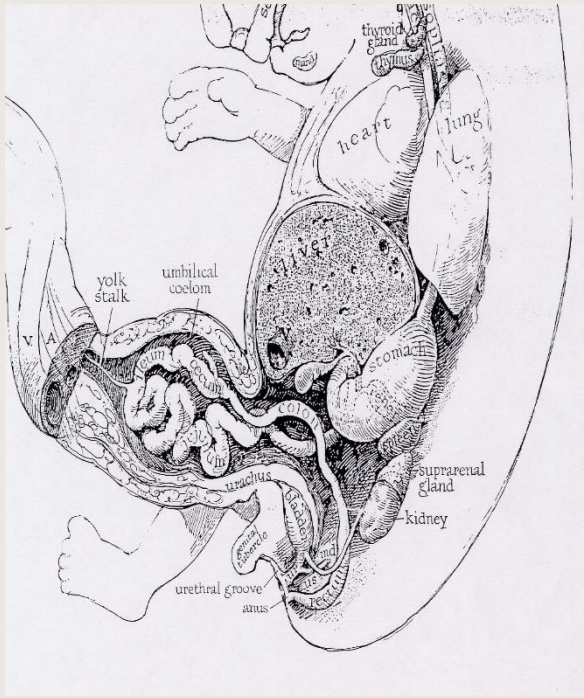
- Malrotation: group of congenital anomalies resulting in *aberrant intestinal rotation & fixation*.
- Takes place during the *1st 3 months of gestation*.
- First reported by William Ladd in 1932.
- This is an important clinical entity as *it leads to volvulus*.

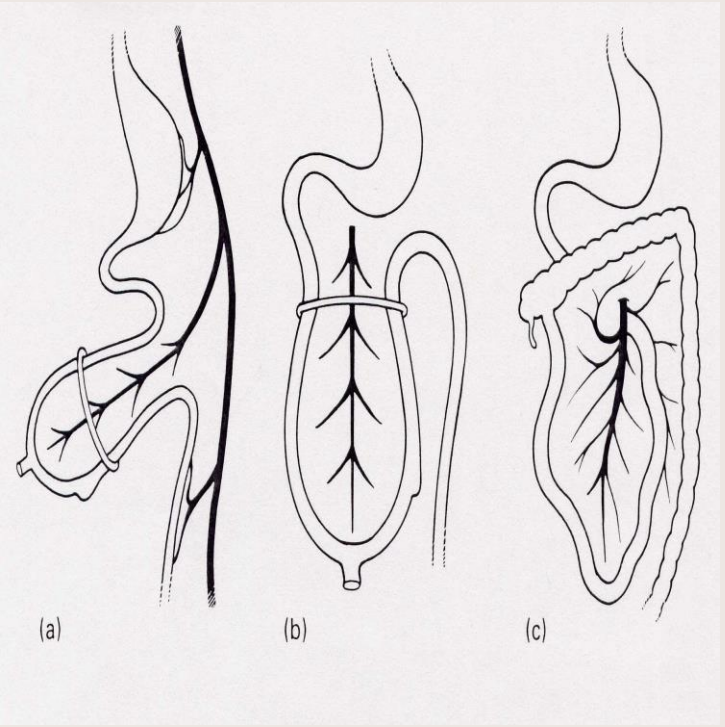
Cont.

- Malrotation is defined by:
  - Diameter of small bowel mesentery
  - The presence of Ladd's bands (adhesion bands between small bowel & the area around the duodenum resulting in partial obstruction of this area)
- Children tend to present with frequent regurgitation.

Embryology


A
B

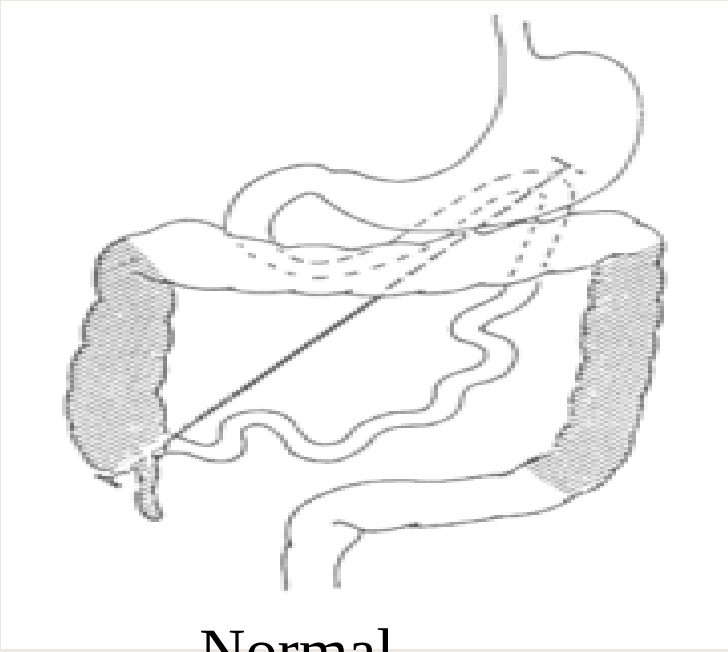

thyroid gland
thymus
heart
lung
liver
yolk stalk
umbilical coelom
stomach
suprarenal gland
kidney
urachus
urethral groove
anus


(a)
(b)
(c)

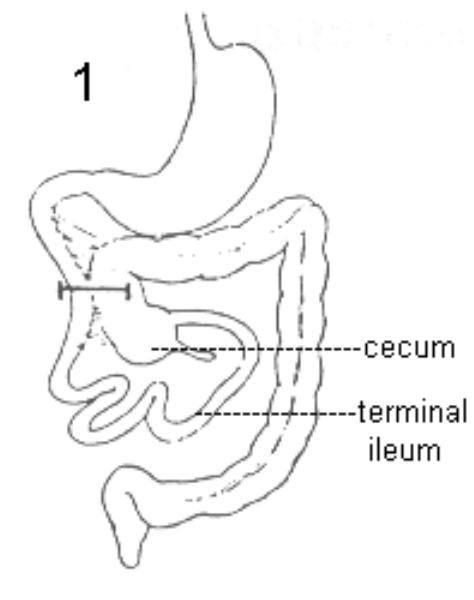
Key points in embryology

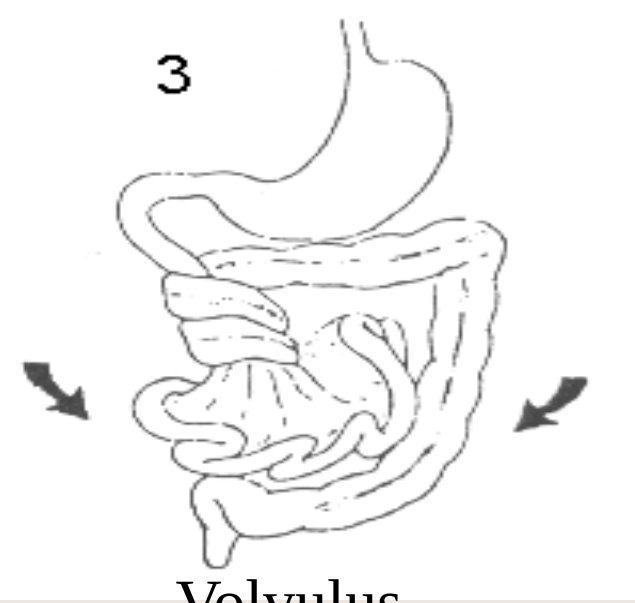
- Intestinal rotation starts at 5^{th} week.
- Midgut: SMA
  - Rotation takes place *around the SMA axis*
- 270^{0} counterclockwise rotation of pre – arterial & post – arterial limb
- Ladds bands attach to the cecum irrespective of its position at the end of rotation from right para colic region.

Cont.



Normal





Volvulus

Shortened mesentery diameter

Stage I

- 5 – 10 wks.
- Gut herniates
- 180^0 duodenojejunal loop
- 90^0 cecocolic loop
- Aberration in the above results in: *volvulus*.

Stage II

- Gut returns into abdomen.
- 90^0 duodenojejunal loop.
- 18^0 cecocolic loop.
- Aberration in the above results in: *volvulus, duodenal obstruction, mesenteric hernia*

Stage III

- Cecum descends
- Fixation of the mesenteries
- Aberration in the above results in: *volvulus, hernia*

Non – rotation

- Neither colon or duodenum undergo rotation. They remain in the state they are at 5 wks.
- MC form of malrotation.
- M:F → 2:1

Cont.

- Duodenum not posterior to SMA hence susceptible to obstruction.
- Ligament of Treitz fails to reach its normal position in the RUQ.
- Midgut mesentery is narrow & highly mobile.
- May cause:
  - Duodenal obstruction by abnormal peritoneal (Ladds) bands
  - Acute midgut volvulus

Incomplete rotation

- Counter clockwise rotation of only 180^0.
- Cecum in epigastrium overlying 3^{rd} part of duodenum.
- MC form of surgically treated malrotation.

Reverse rotation

- Rotates clockwise
- DJ loop anterior to SMA and transverse colon posterior to SMA
  causes:
  - Compression of colon by SMA → obstruction
  - Ileocecal volvulus

Stringer's classification

Type	Defect	Clinical effect
IA	No rotation	Volvulus of middle intestine
IIA	No duodenal rotation; normal colon rotation	Duodenal obstruction due to bands
IIB	Inverse rotation of duodenum and colon	Transverse colon obstruction due to duodenal mesentery
IIC	Inverse duodenal rotation; normal colon rotation	Right mesenteric sac (obstruction)
IIIA	Normal duodenal rotation; no colon rotation	Volvulus of middle intestine
IIIB	Incomplete fixation of the hepatic angle of colon	Obstruction due to Ladd bands
IIIC	Incomplete fixation of the cecum and its mesentery	Volvulus of the cecum; invagination (Waugh's syndrome)
IIID	Internal hernias	Paraduodenal hernia

Edited from: Jamieson and Stringer[11] and Bill.[15]

Epidemiology

- Incidence: 1/6000 live births
- Most present < 1 month
- Incidence in general population: 0.2 to 0.5%
- No sex/ race predilection.

Associated anomalies: (seen in 30 – 60%)

- Duodenal atresia
- Duodenal stenosis
- Duodenal web
- Congenital diaphragmatic hernia
- Gastroschisis
- Omphalocele
- Choanal atresia
- Polysplenia/ asplenia
- Congenital megacolon

Heterotaxy syndromes/ situs ambiguous/ isomerisms

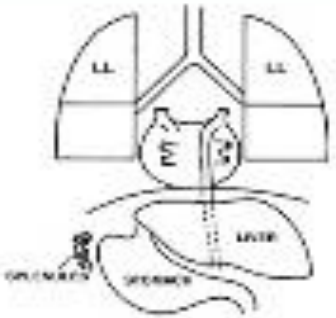
- A variation from normal that involves the heart and other organs.
- Include:
  - Situs solitus (normal)
  - Situs inversus
  - Asplenia
  - Polysplenia

{ X , _ , _ }

Visceroatrial Situs

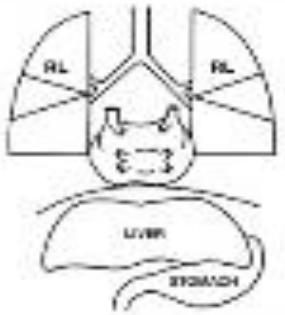
SITUS AMBIGUUS (HETEROTAXY SYNDROME)

With Polysplenia



- Left isomerism
- Bilateral bilobed lung, hyparterial bronchus with absent minor fissure
- Bilateral morphologic LA
- Interruption of IVC with azygos or hemiazygos continuation
- Multiple spleens
- Pulmonary veins drains into both RA and LA

With Asplenia



- Right isomerism
- Bilateral trilobed lung, eparterial bronchus and bilateral minor fissures
- Bilateral morphologic RA
- Large symmetric, centrally located liver
- Absent spleens
- Frequent total anomaly of pulmonary venous return (TAPVR)

Cont.

- Asymptomatic if total malrotation.
- Most often presents during the 1st few months of life (usually 6 months)
- Partial may present as: mid gut volvulus, mesocolic hernias, duodenojejunal obstruction & colonic obstruction.
- P/C: Infant with acute onset of bilious emesis.
- Malrotation is a surgical urgency due to the possibility of volvulus. ***Volvulus is a surgical emergency***.

In children

- Acute mid gut volvulus
- Chronic midgut volvulus
- Acute duodenal obstruction
- Chronic duodenal obstruction
- Internal hernia

In adults

- Intermittent cramping or persistent aching pain
- Severe abdominal cramping following by diarrhea – chronic volvulus
- Vomiting: bilious/ non – bilious, variable in duration and frequency
- Malabsorption – diarrhea, nutritional deficiencies
- Rare: obstructive jaundice, chylous ascites & SMV thrombosis

Plain radiograph

- No pathognomonic signs
- Right sided Jejunal markings
- Absence of colonic shadowing in RIF
- Features of complications:
  - Dilated bowel loops
  - Air fluid levels
  - Pneumoperitoneum

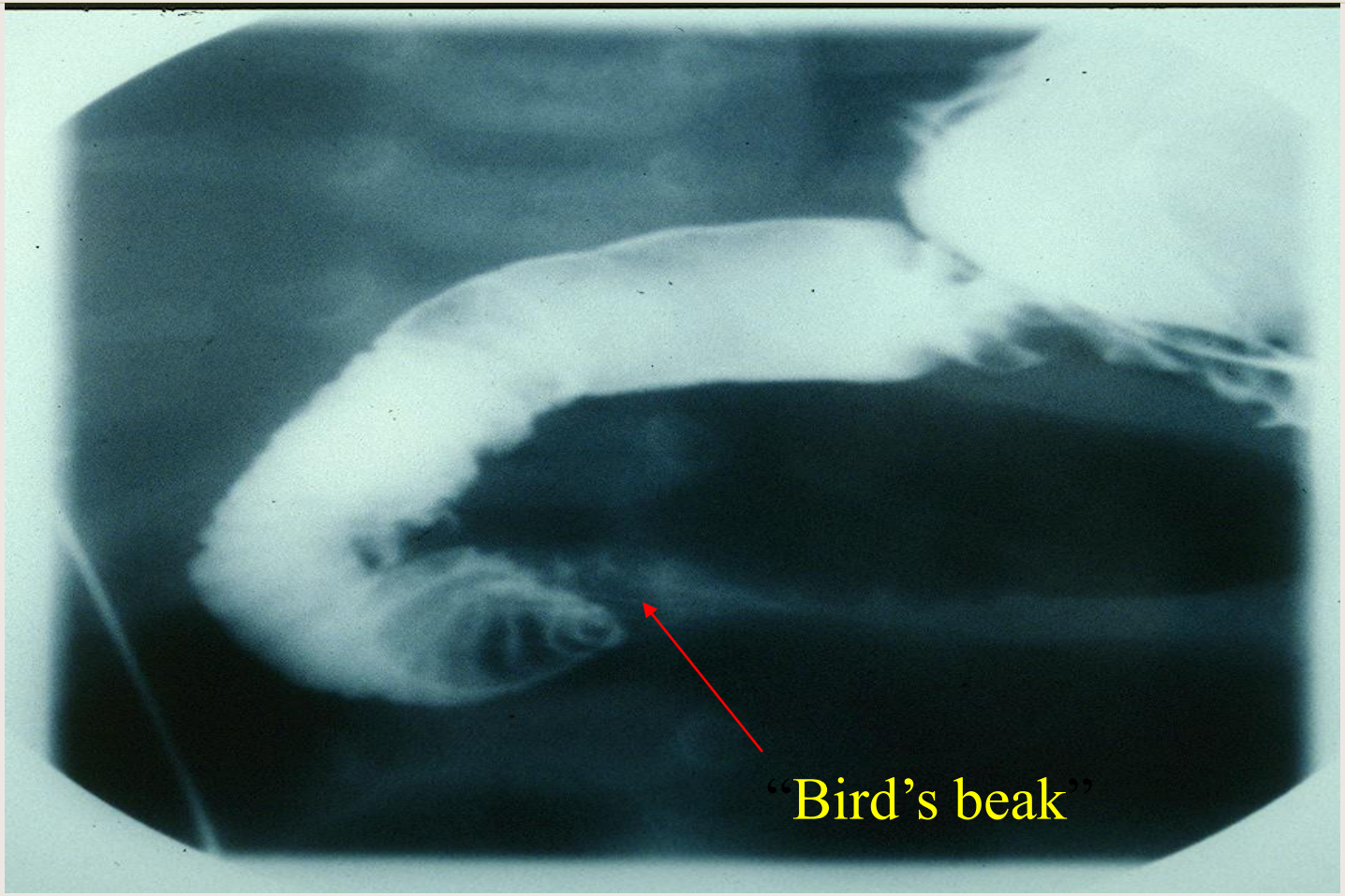
Ultrasound

- Reversal of the normal anatomic relationship between SMA & duodenum
- “whirlpool” sign → midgut volvulus
- “bird beak” appearance → duodenal obstruction
- FP rates: __ (not very diagnostic)

UGI studies

- Gold – standard
- Upper GI studies with water – soluble radio – opaque contrast.
  - S – appearance is diagnostic. (instead of C appearance)
  - Cork – screw sign: spiral appearance of the distal duodenum and proximal jejunum seen in midgut volvulus.
  - Incomplete Duodenal Obstruction usually in the 3rd portion
  - Ligament of Treitz is not to the left of the midline or at the level of the gastric antrum

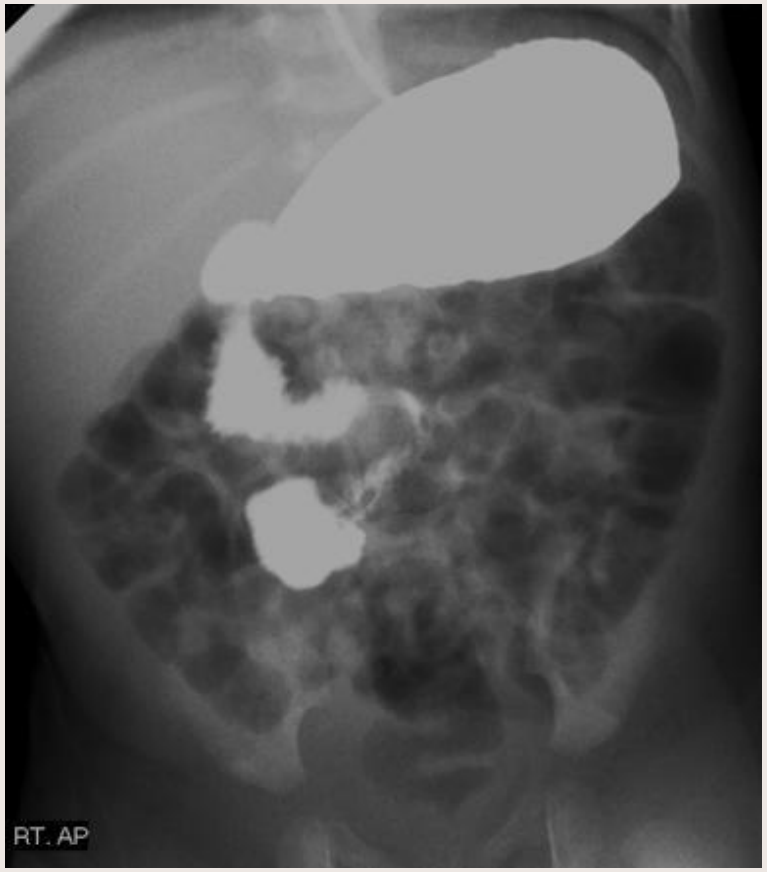
Bird's Beak Appearance



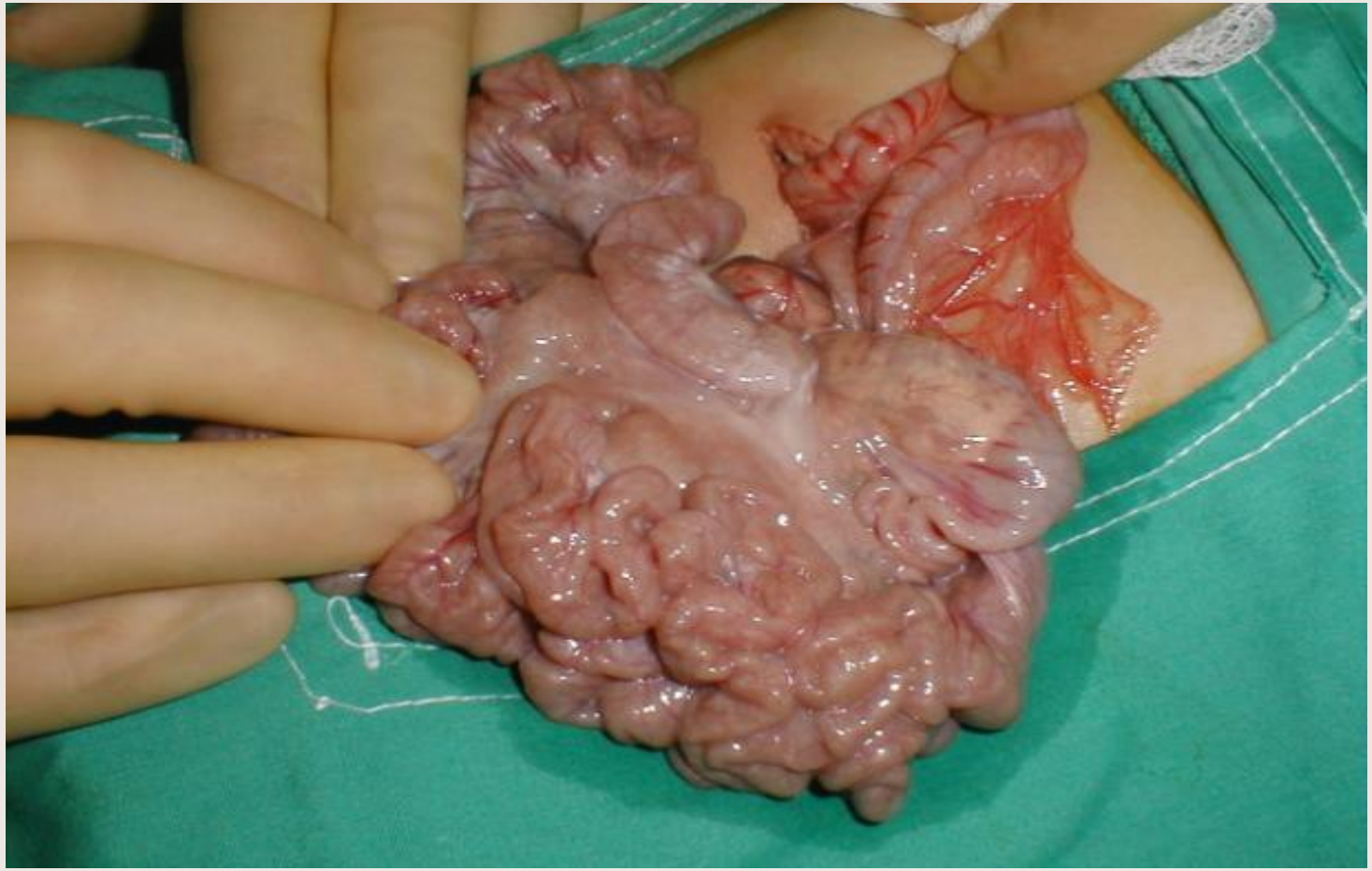
CT abdomen

- Anatomic location of small bowel on right & colon on left
- Relationship of SM vessels
- Aplasia of the uncinated process
- Gangrene

Malrotation



Malrotation



Management

- Supportive management
- Surgical management: *Ladds procedure* (should be done in 6h. Of volvulus or the child may lose a significant portion of bowl → short bowel syndrome or their life)
  - Laparotomy → evisceration → counterclockwise untwisting of volvulus → division of Ladd's bands → widening of mesenteric base → relief of duodenal obstruction → appendectomm
  - Release of adhesion & Ladds bands
- Recurrence is 10% after Ladd's procedure.

Post – op care

- NG decompression
- TPN until return of bowel function
- Mortality from midgut volvulus: with severe bowel compromise may > 30%
- Long term complications
  - Adhesive SBO (10%)
  - Recurrent volvulus
  - Short gut syndrome

d. Volvulus

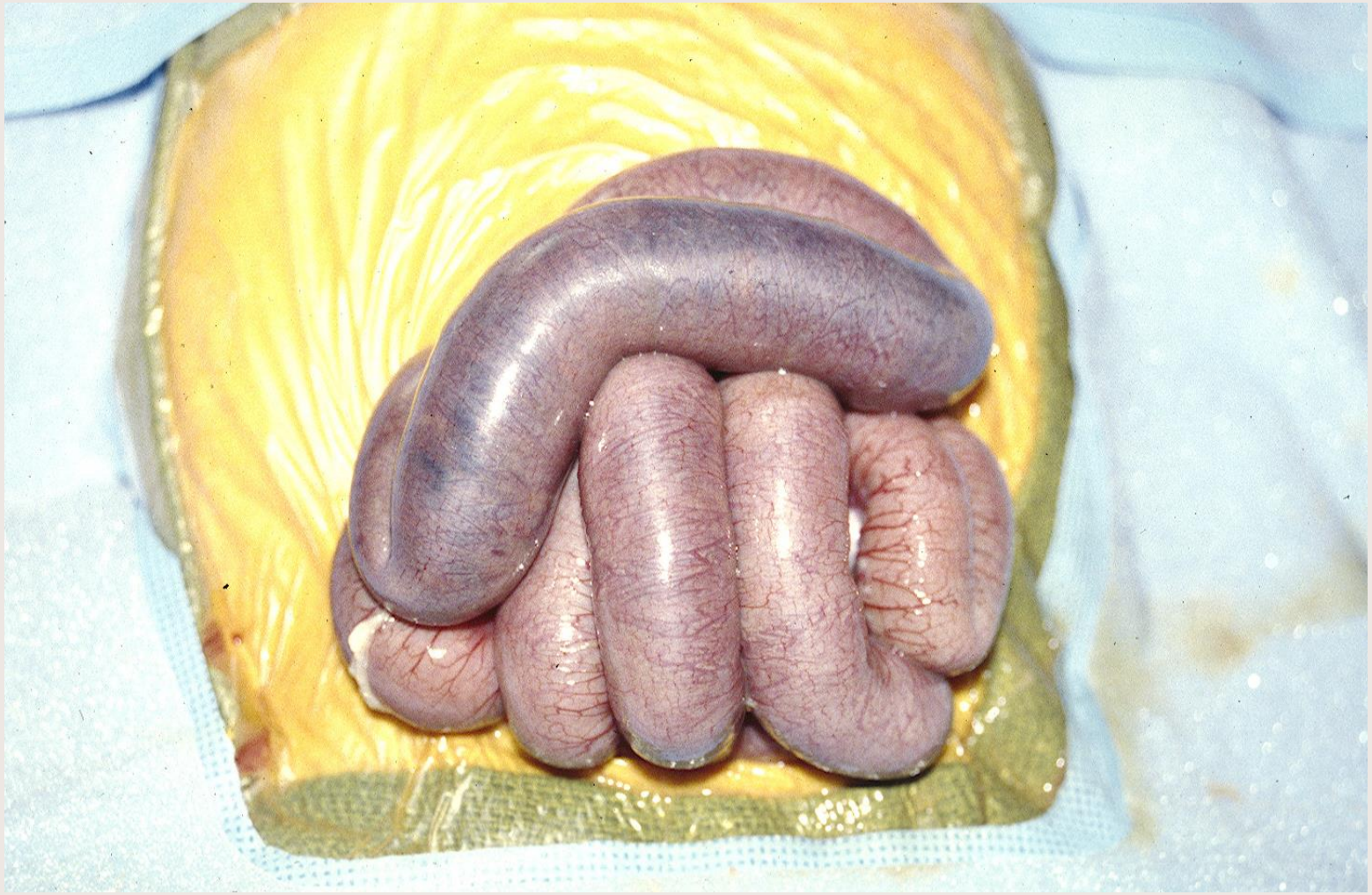
Introduction

- 75% First month of life, 90% first year.
- Malrotation is the risk for volvulus
  - Small & large bowel are not fixed
  - Narrow mesentery → more likely to turn around itself
- Malrotation can cause or present with:
  - Volvulus (this is dangerous)
  - Acute obstruction
  - Chronic intermittent obstruction

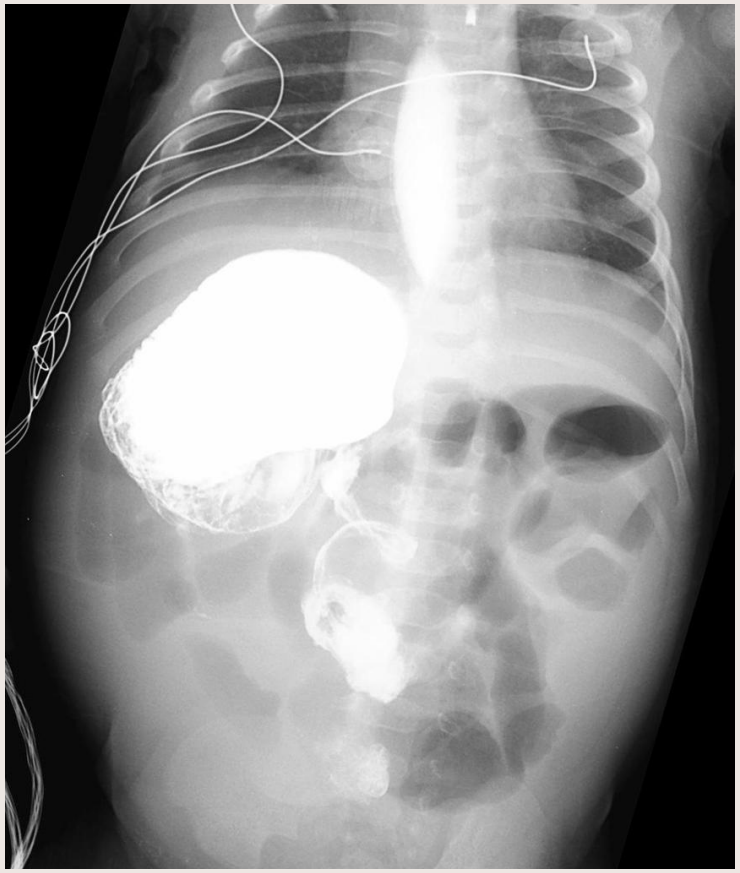
Cont.

- One can have volvulus with normal rotation in the following conditions:
  - Omphalomesenteric remnant
  - Internal hernia
  - Duplication
  - Adhesive SBO

Volvulus & Ischemia



Malrotation with volvulus



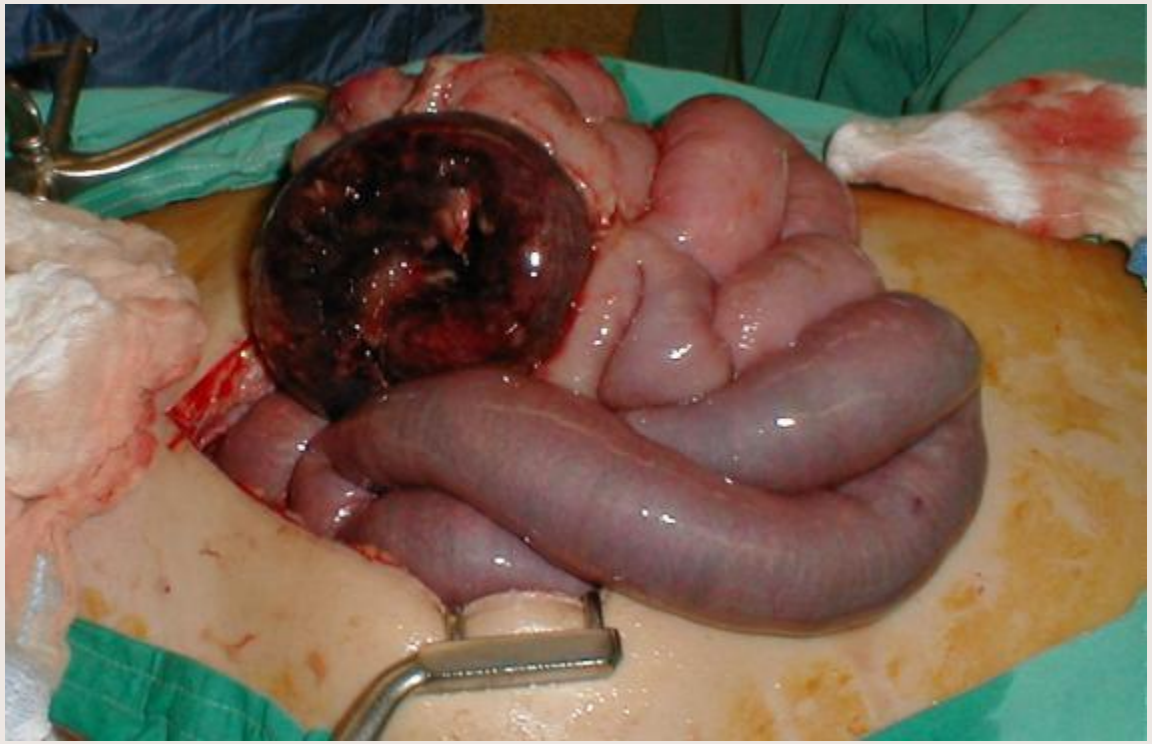
Cont.

- Malrotation → mid – gut volvulus → midgut intestinal death → surgery (resected) → short gut syndrome → death
- P/C
  - Most in infant (1st year of life)
  - Bilious vomiting
    - With/ without pain
      - If with pain (irritable) → likely volvulus + ischemia
      - If without pain (calm) → malrotation + obstruction

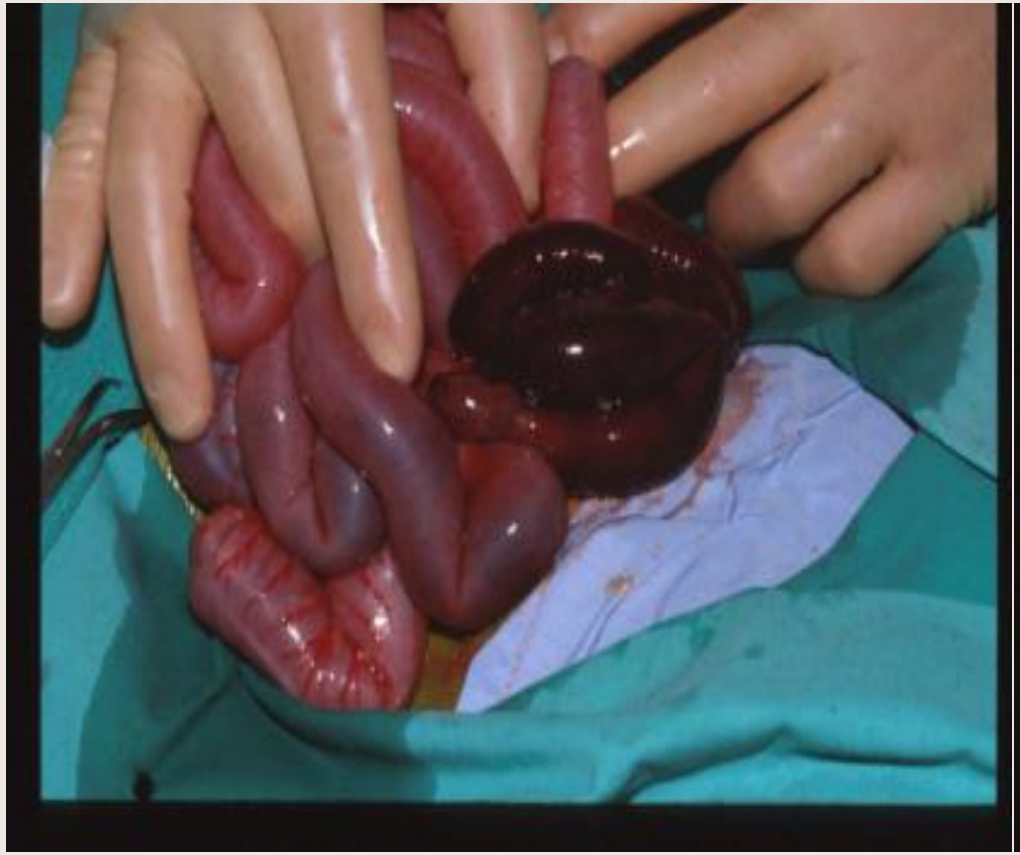
Management

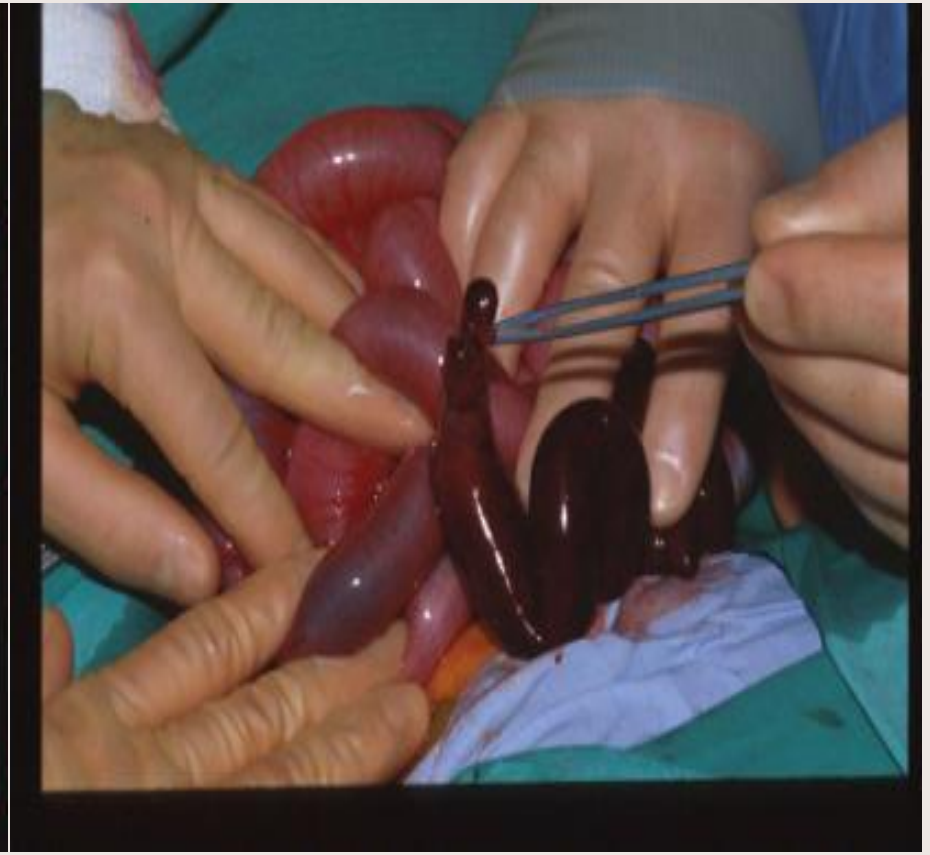
- Patient should go directly for surgery if:
  - Can’t do investigation immediately
  - Patient is sick with bilious vomiting.
- Surgery: (‘NB: in volvulus, rotation is clockwise’)
  - Untwist (counter – clockwise) → assess viability
  - If extensive ischemia → close & do a 2nd look 24 – 48 hrs.
  - Viable small bowel → close and observe
  - Ladd’s procedure
    - Cut Ladd’s band; broaden mid – gut mesentery; place SB on the right & colon on the left.
    - Do appendectomy.

SBO



Meckel's diverticulum





Case

- 6mo infant with vomiting, poor oral intake, abdominal distension.
- Mom says not tolerating his bottle today. Began having ***green vomitus***, has not had a wet diaper today.
- Baby looks ill, not very reactive on examination
- P/A: distended, tense, tender

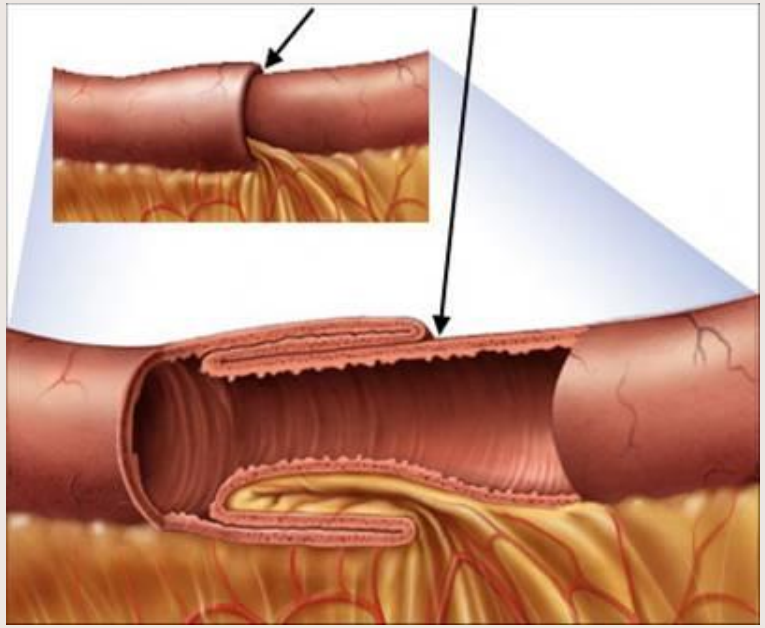
NOTE!

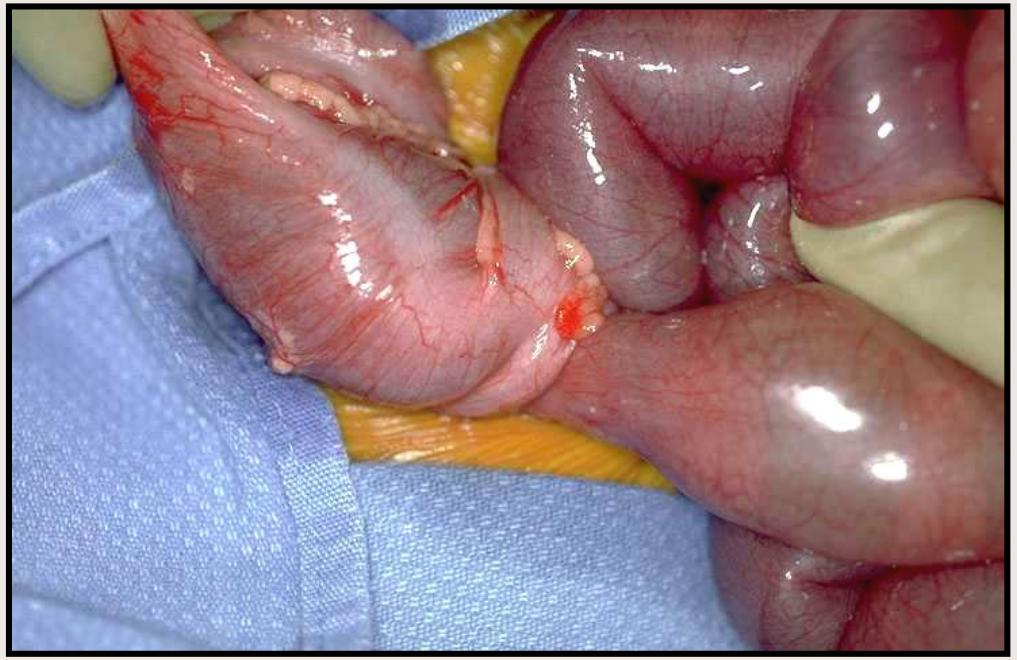
Bilious Emesis is BAD
It is Malrotation with Volvulus
Until Proven Otherwise

e. Intussusception

Definition

- This is bowel obstruction caused by ***invagination/ telescoping of the proximal bowel into the distal bowel***.
- ***Intussusceptum***: The proximal bowel
- ***Intussuscepiens:*** The distal bowel





Epidemiology

- Age: 3 – 36 months; peak at 6 – 9 months.
  - –If present later in age → likely to find PLP
- More prevalent in males.
- Associated with viruses: ***rotaviruses***, ***adenoviruses*** which happen mostly in the cold season.

Classification

- Anatomic:
  - Ileo – ileal (5%, post operative)
  - Colo – colic (2%, adults)
  - ***Ileo – colic (MC)***
  - Ileo – ileo – colic (12%)
  - Multiple
  - Reverse (retrograde)

Pathological classification:

1. Pathologic: There is a PLP
2. Non – pathologic: No specific PLP

Pathophysiology

- Idiopathic in 90%
- NPLP
  - Hypertrophied inflamed (due to a ***viral infection***) Peyer's patches (submucosal lymphoid tissue) that are circumferentially distributed → MC in children esp. ileocolic
- PLP (common in children >2yrs. & adults) may be:
  1. Meckel's diverticulum (MC)
  2. Tumors in adults: intestinal lymphoma, FB, polyp, hemangioma
  3. Appendix
  4. Duplication cysts

Predisposing factors

- Bleeding Disorders: Henoch Schonlein purpura, Hemophilia, Leukemia
- Trauma: Blunt abdominal trauma, Major Retroperitoneal abdominal procedure
- Other:
  - Cystic fibrosis
  - *Ascaris lumbricoides* worm infestation

Clinical presentation

- Abdominal pain
  - Irritability: infant is calm in between attacks of pain
- Abdominal distension
- Red currant jelly stool
- Rectal mass
- ± Bilious vomiting: Often a late feature
- History of URTI (***often associated with a preceding viral illness***)

Classic triad (present in 20 – 50% of cases)

Progressively increasing, intermittent colicky, abdominal pain. Relieved by assuming the fetal position (*pulling knees up to the abdomen*)

Abdominal mass (intussusceptum): curved, sausage – shaped, palpated in the peri – umbilical area (65%)

Red currant jelly (bloody) stools (60%)

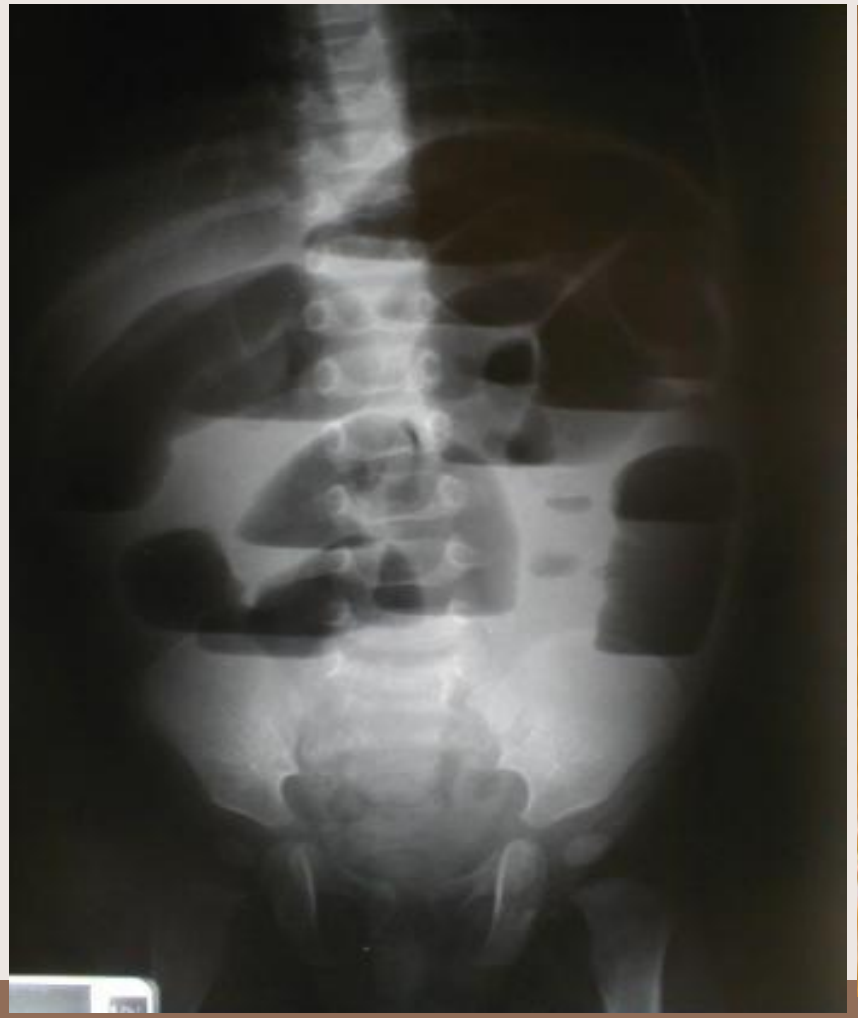
Cont.

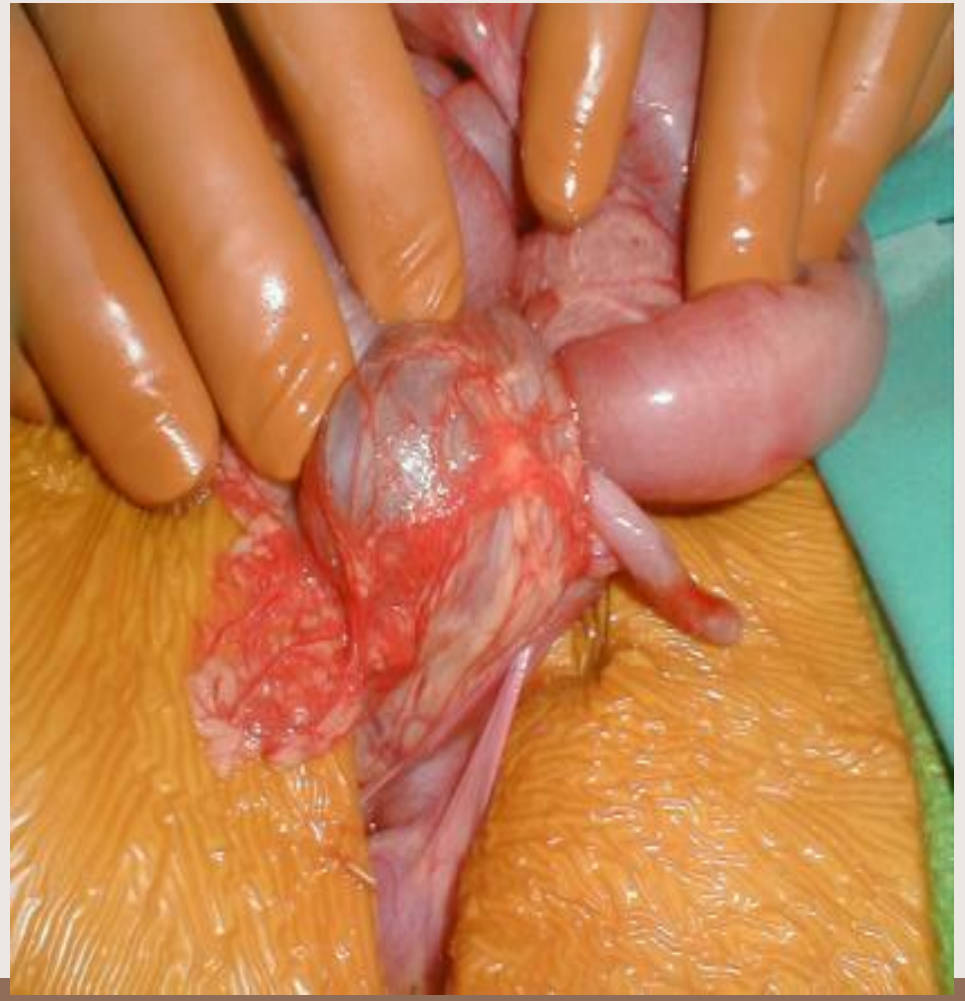
- Colicky pain:
  - Increased peristalsis → intermittent pain → intussusceptum progresses further into the intussuscipiens → carries with it mesentery containing blood vessels → with time ischemic pain supervenes.
- Red – currant Jelly Stool:
  - Ischemia → sloughing off of mucosa → resulting in exposing of submucosal vessels coupled with hypersecretion of mucus → red currant jelly (bloody) stools

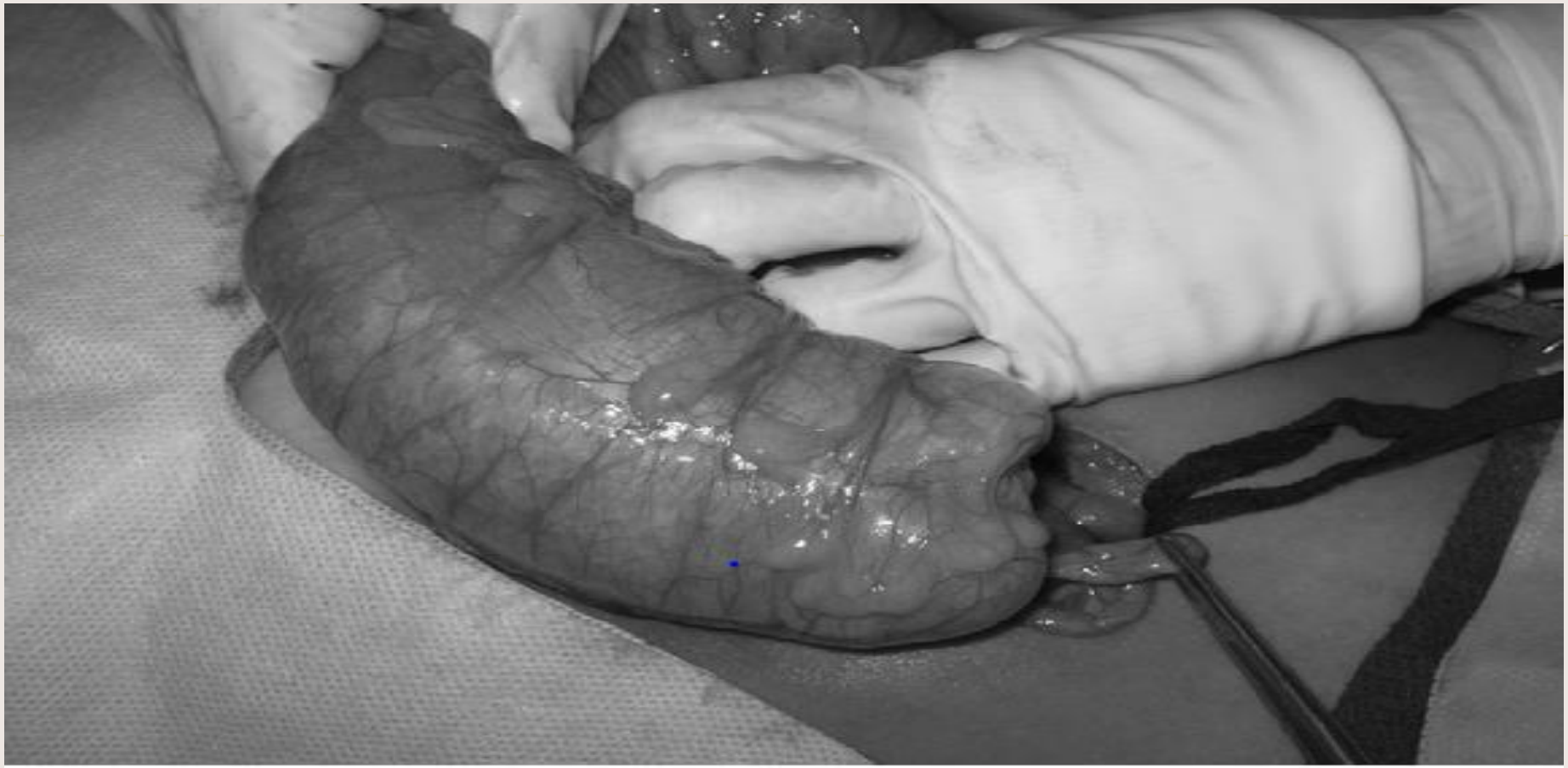
Physical Examination

- General Exam
  - Generally sturdy
  - Well – developed & well nourished infant/ child
  - +/- sick looking, lethargic, dehydrated & pale
- Abdominal exam: ***Dance sign*** (peri – umbilical mass with RIF emptiness)
- +/- palpable rectal mass (5%); red, mucoid stool, hemorrhage, prolapsed intussusceptum.

Cont.







Ileocolic intussusception. The hand is milking the most distal intussusceptum retrograde. Note the forceps on the appendix and the pneumatosis of the cecal wall.

Investigations

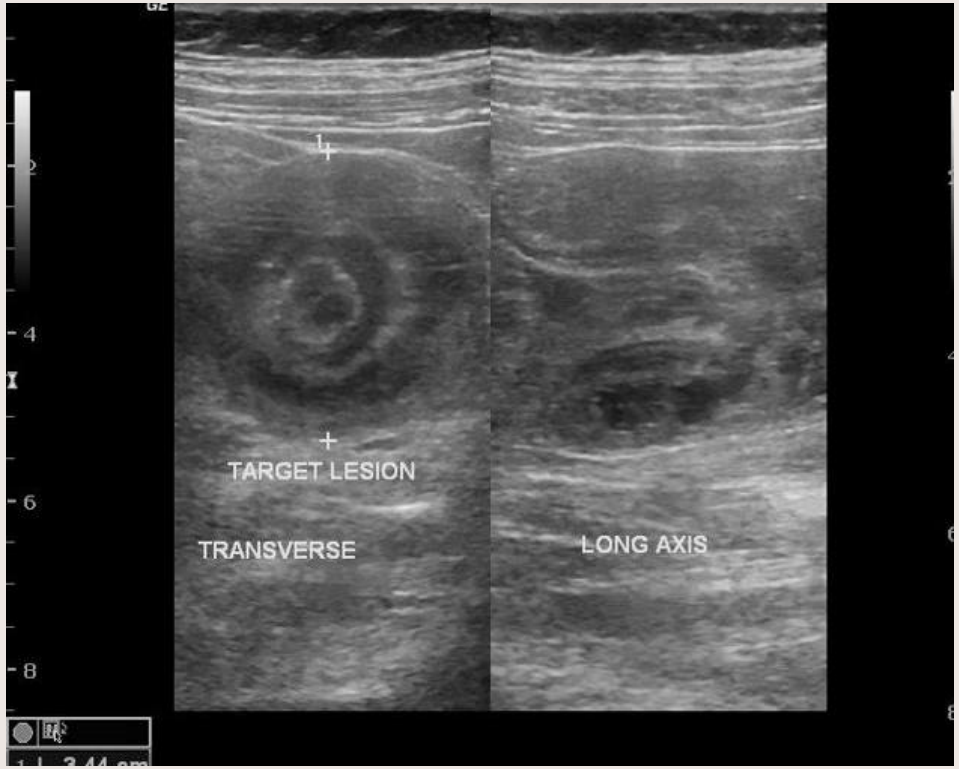
- FHG: RBC count, WBC count
- U/E/Cr: Effects of dehydration & 3rd spacing
- BGAs: Tends to lean towards lactic acidosis due to hypovolemic shock

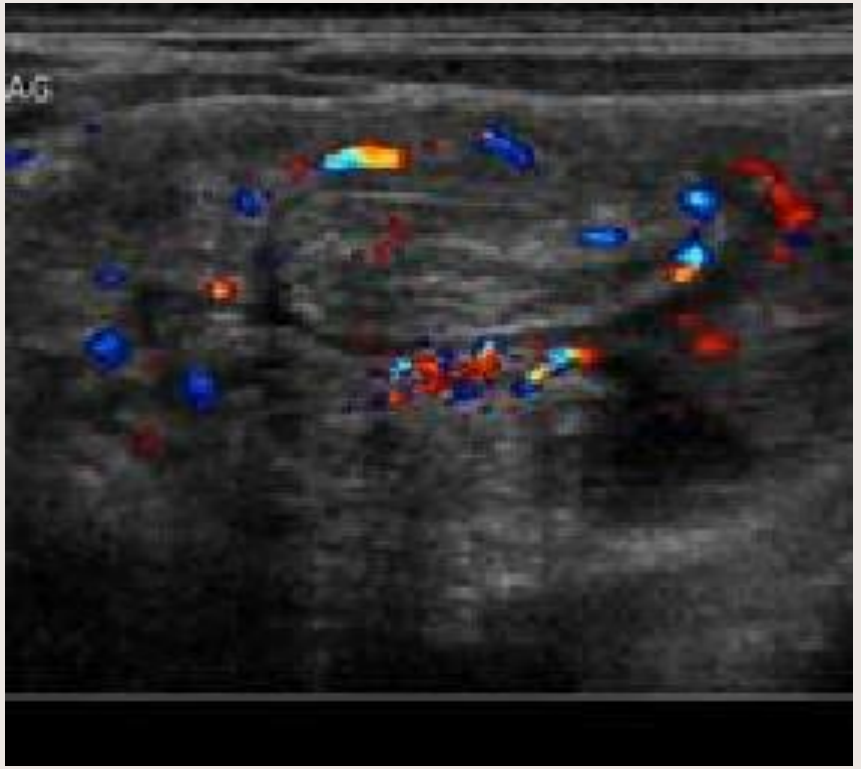
Imaging

- Abdominal U/S
- Plain abdominal radiograph:
  - –Supine views
  - –Dorsal decubitus in children
- Contrast enema: can be both ***diagnostic*** and ***therapeutic***

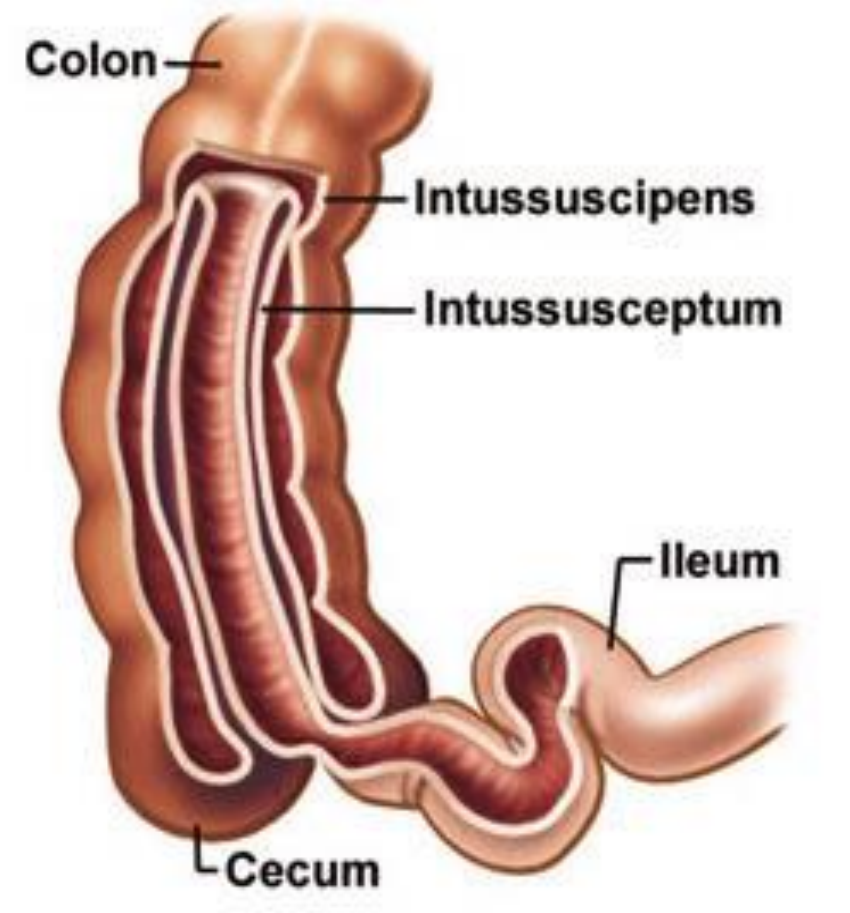
Ultrasound

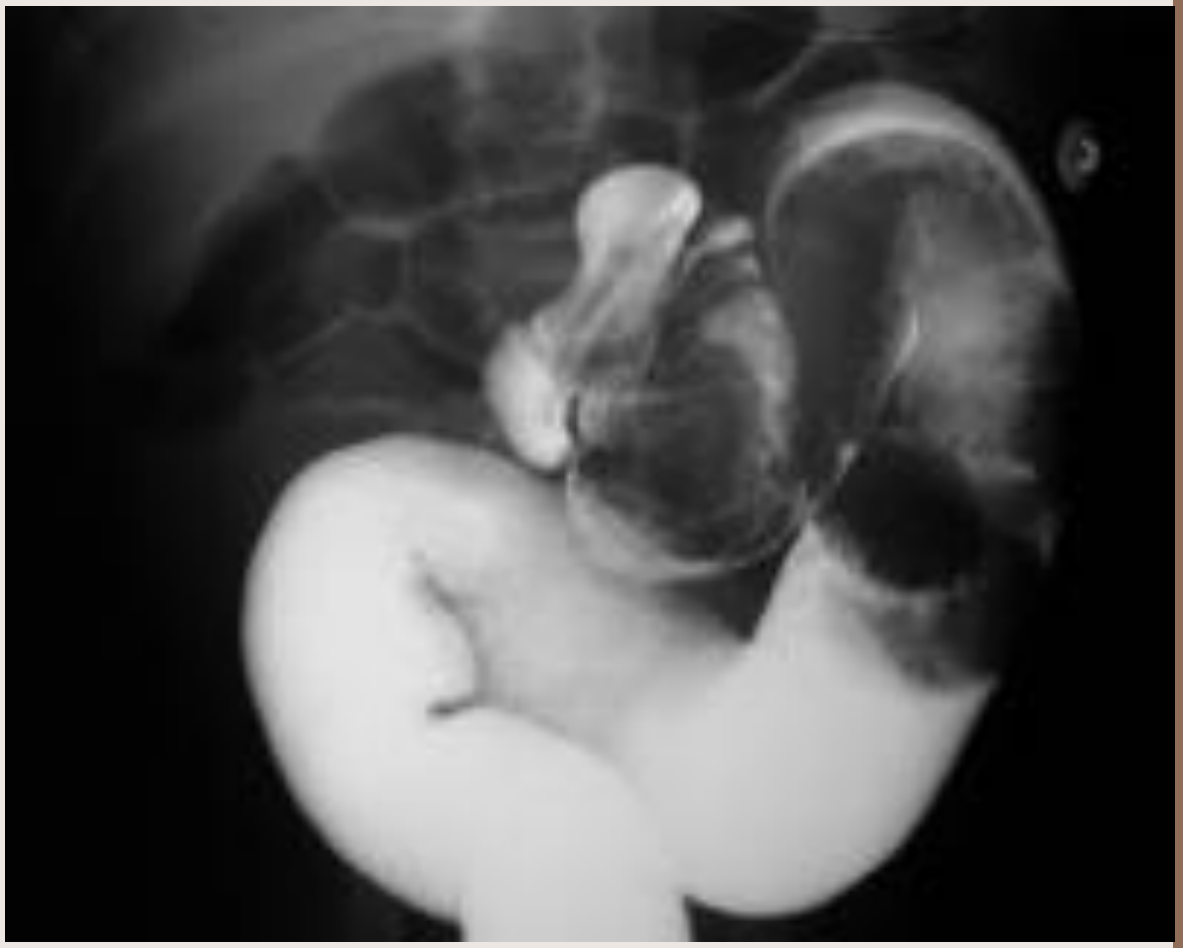
- In the hands of an experienced examiner, ***U/S can have 100% accuracy for the diagnosis of intussusception***. Most sensitive & specific test. U/S features:
  - Doughnut sign
  - Target sign
  - Pseudo – kidney sign
  - Target/ doughnut sign: 95% accurate
- Gut viability can be evaluated by ***Duplex U/S***.


TARGET LESION
TRANSVERSE
LONG AXIS



Contrast enema





Differential diagnosis

- Rectal prolapse: also has red stool & rectal mass.
- Dysenteric GE

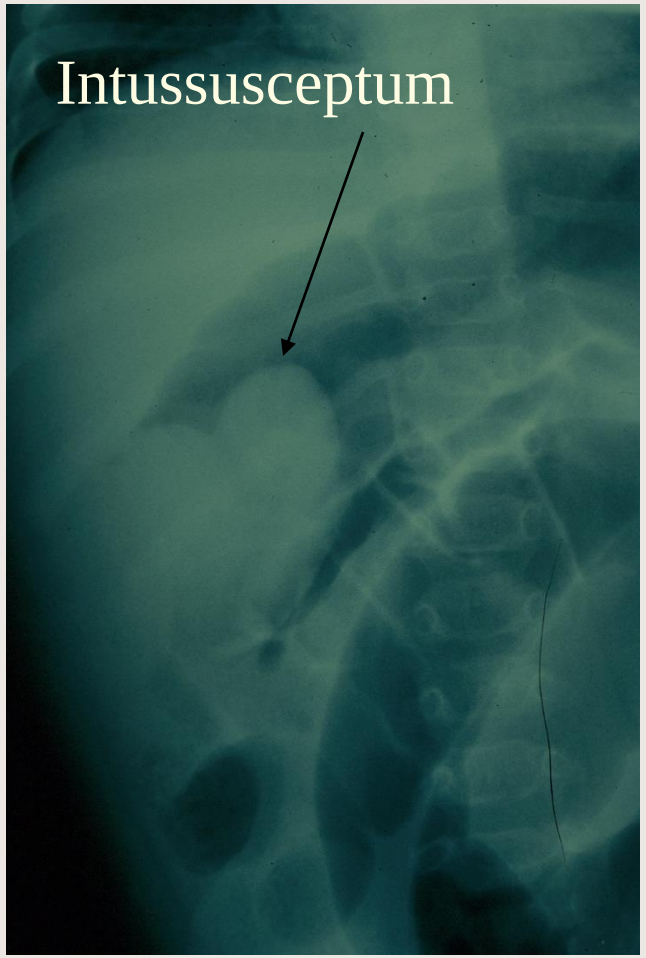
Supportive management (*3 tubes, 3 observations, 3 medications*)

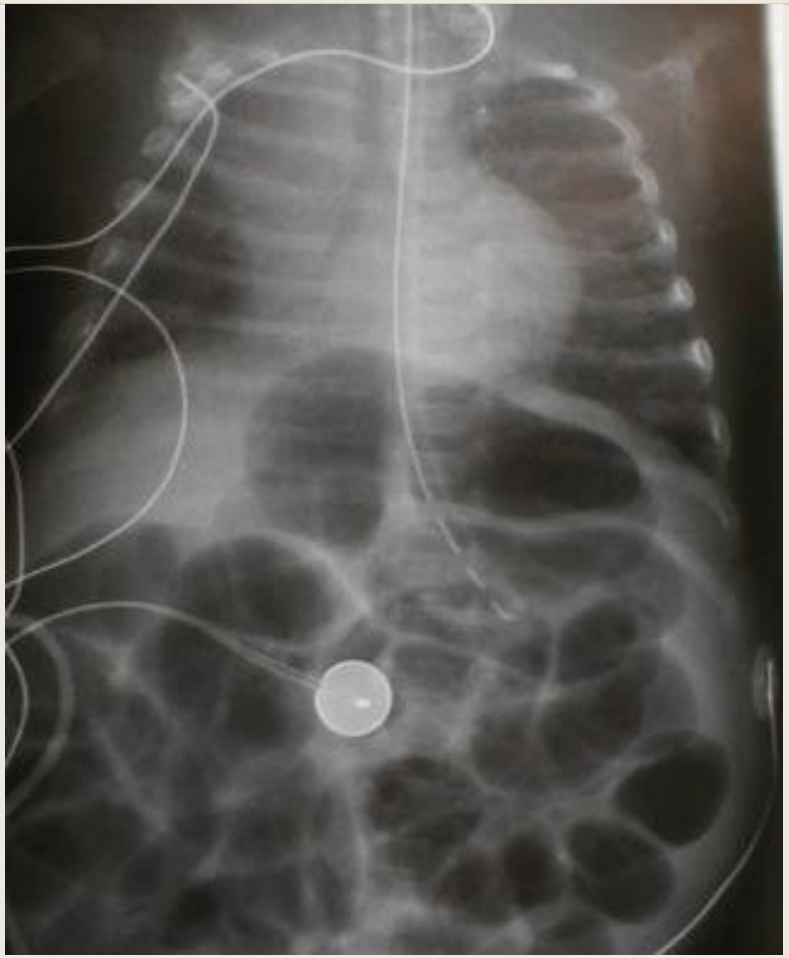
1. Tubes:
   - Decompression: NGT, Keep NPO
   - IV access: rehydrate
   - Urinary catheter
2. Observations: Monitor I/O, vitals, frequent clinical assessment
3. Medications: analgesia, antibiotics, IVFs (boluses & maintenance)

Definitive management: reduction

1. Pressure reduction:
   - Hydrostatic reduction: Barium enema, water
   - Pneumatic reduction: MC, less complications
2. Manual reduction:
   - Failed pressure reduction → surgical reduction → if impossible (likely PLP) → resection.

Pneumatic reduction





Complications of surgery

- Immediate:
- Early:
  - Infection
  - Wound dehiscence
- Late:
  - Recurrence

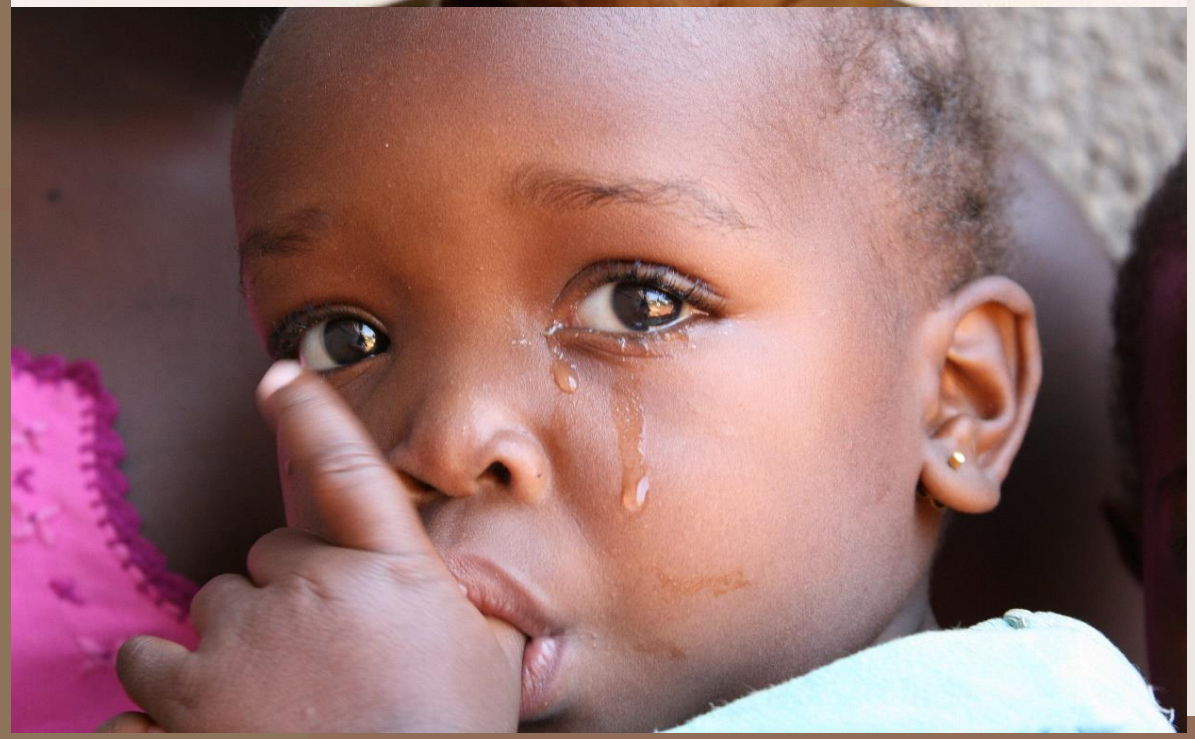
Prognosis

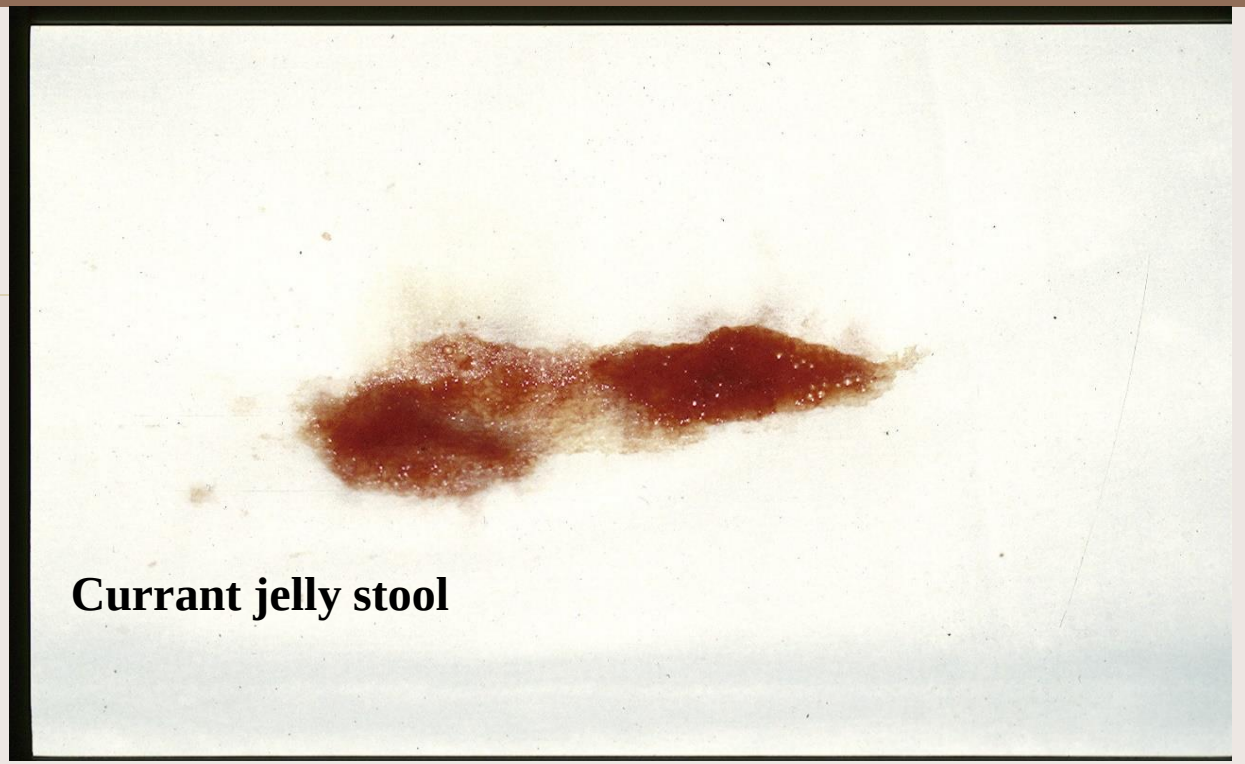
- Depends on prognostic factors:
  - –Early presentation & intervention improves mortality.
  - –Comorbidities; intestinal gangrene upon reduction worsens prognosis.

Case

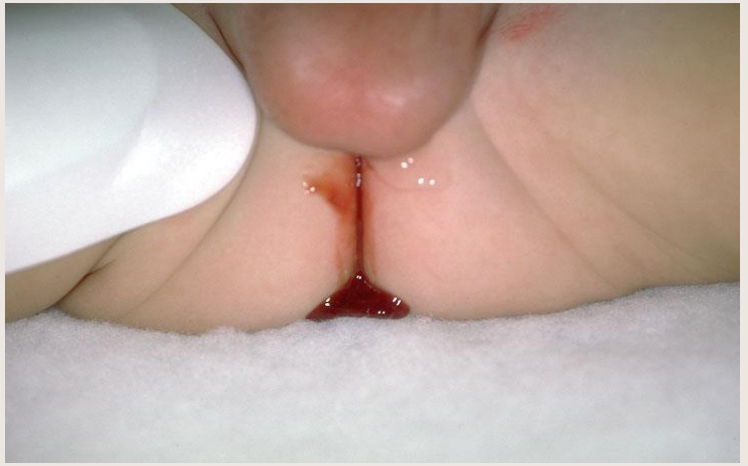
- 6mo infant with vomiting, poor oral intake, abdominal distension.
- Otherwise healthy infant, no previous feeding intolerance
- Looks well, mom says intermittent crying.
- Mom says patient passed reddish, thick – mucous stool.
- Classical presentation of: intusussception

Looks well intermittent crying





Currant jelly stool



f. Meconium conditions

f. MECONIUM CONDITIONS

Meconium Ileus

- Obstruction of bowel by ***thick tenacious meconium***
- 30% of intestinal obstruction cases in neonates
- Frequent cause of meconium peritonitis
- Most are associated ***with cystic fibrosis*** (but only 15% of infants with CF will have meconium ileus)
- ***Abdominal distention is typically present at birth***

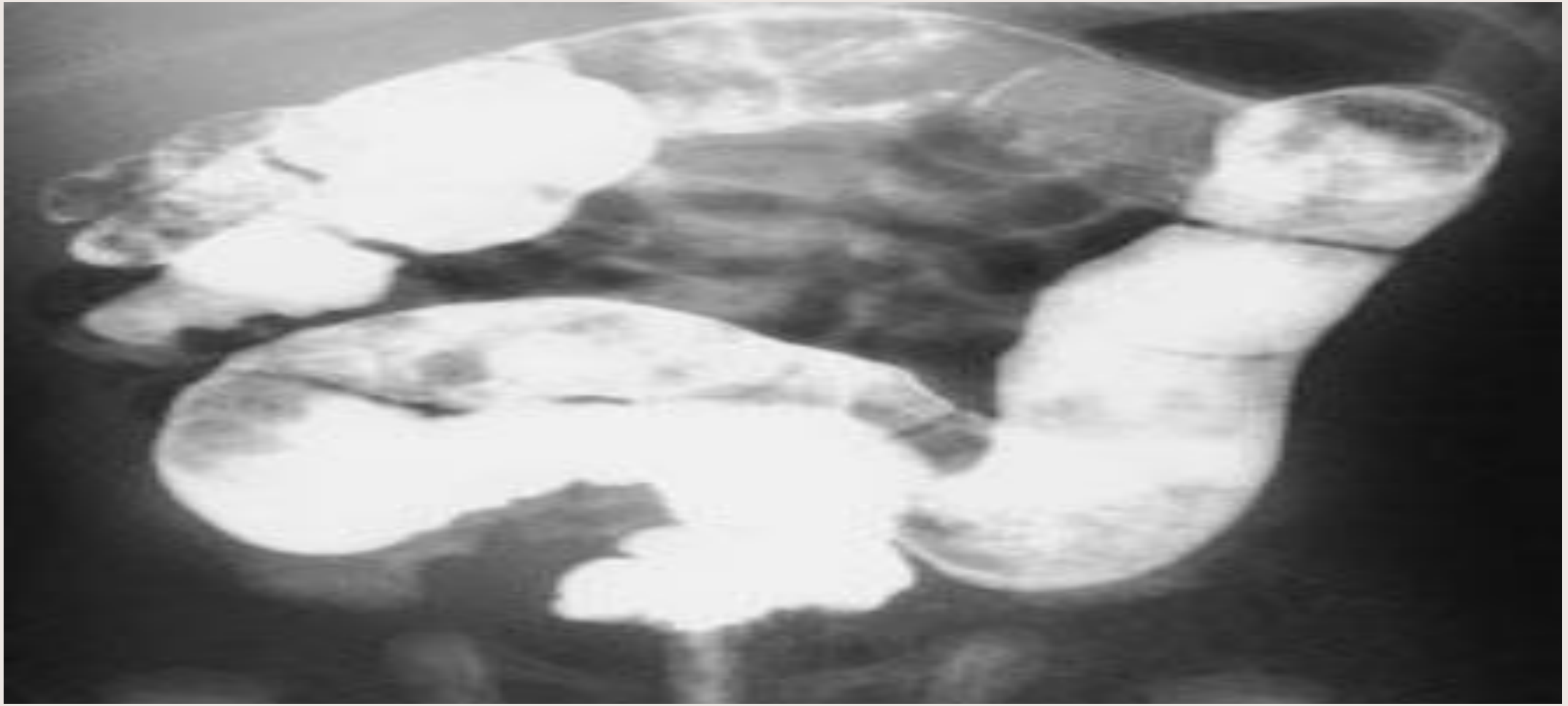
Cont.

- Diagnosis made with contrast enema
- ***Gastrograffin enema*** with aggressive hydration can be used to treat some cases.
- Operative evacuation of meconium
- May require ostomy
- Proximal bowel dilated & distal bowel may be very small (micro – colon) and require time to dilate with use

Meconium plug syndrome

- Mildest & MC form of functional obstruction in the newborn.
  - Difference between meconium ileus & meconium plug is site and severity of obstruction
- Risk factors: Preterm infants, infants of diabetic mothers, IUGR babies, otherwise ill babies
- Treatment with glycerin suppositories and warm saline enemas
- May require contrast enema to make diagnosis
- Normal stooling pattern should follow evacuation of plug

Meconium plugs



2. ACUTE APPENDICITIS

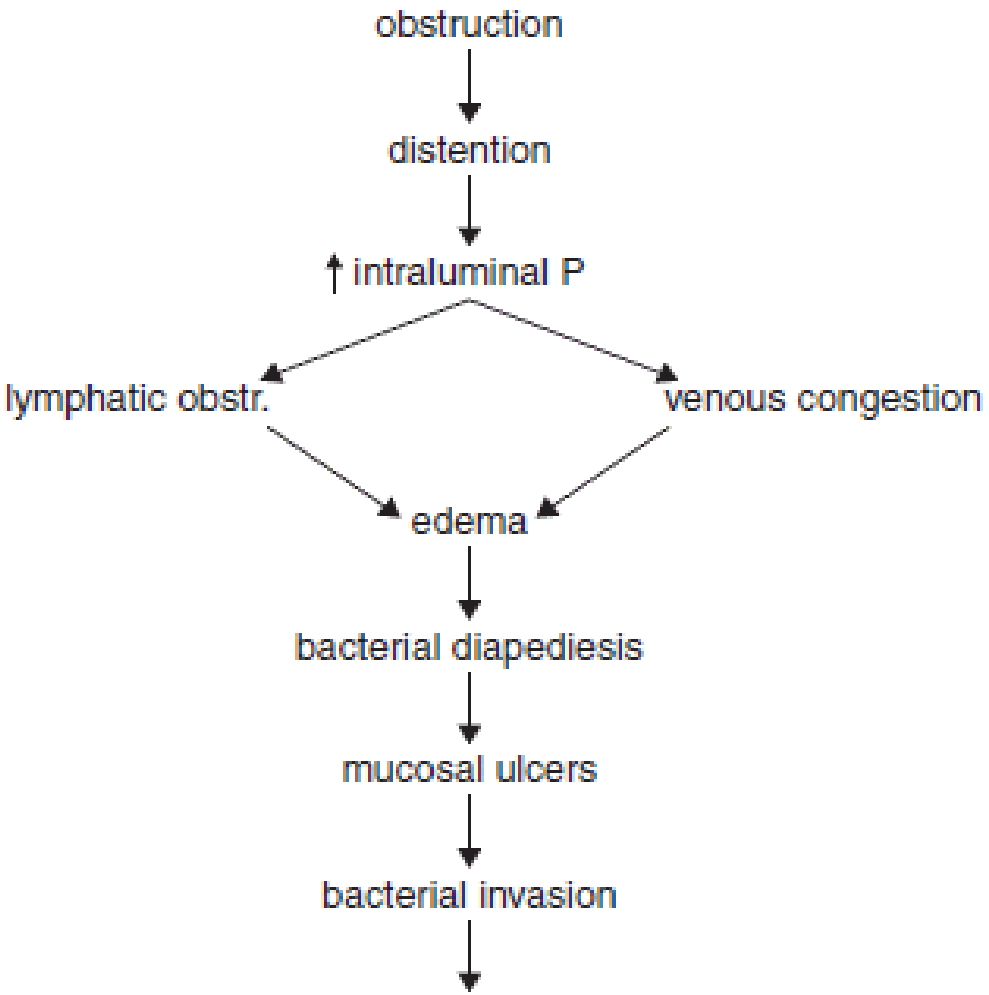
2. ACUTE APPENDICITIS

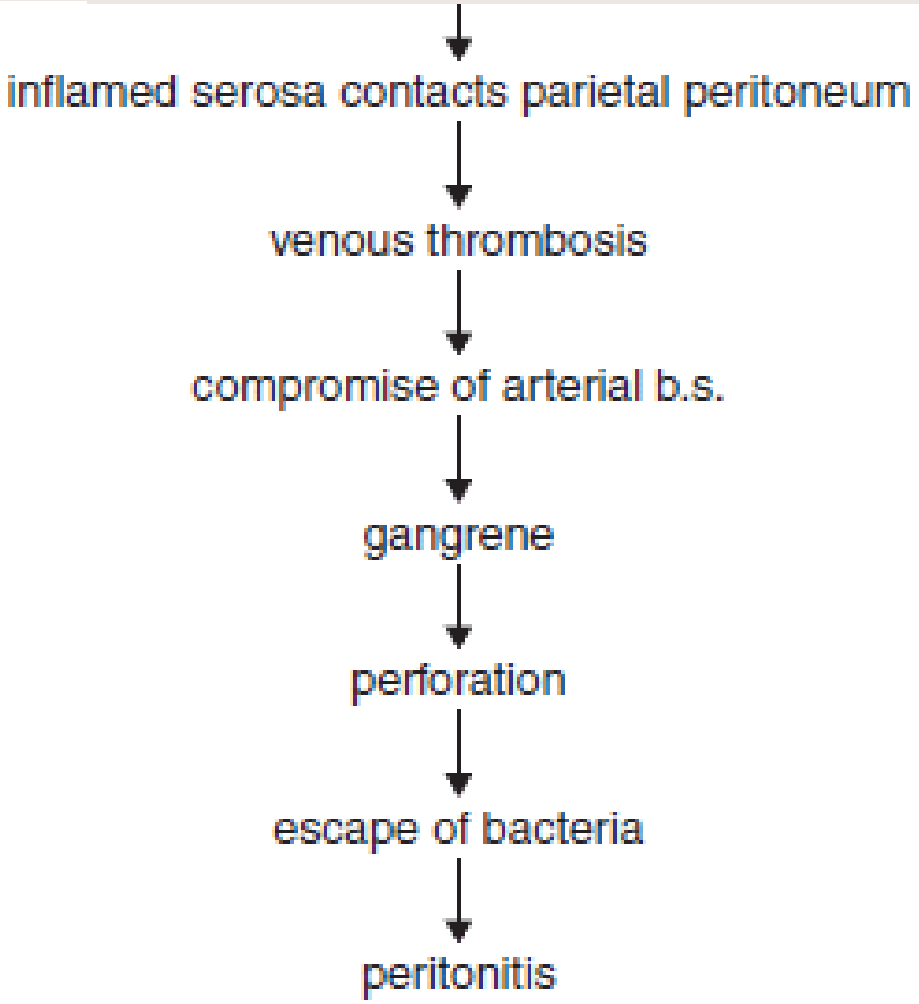
- This is the MCC of abdominal, surgical emergencies in children.
- 2 peaks:
  - Neonates
  - Adolescents
- Diagnosis is mainly clinical: typical history, P/E, CBC + WBC differential

Cont.

- In children < 3 yrs. esp. in infants, it may be difficult to diagnose hence there is a tendency to:
  - Early rupture
  - Sepsis: fever, high WBC count
  - Vomiting: ileus or abscess

Pathogenesis: begins with a luminal fecolith, lymphoid tissue, parasites, FB or tumor →





Alvarado score

Clinical Variable	Alvarado Score	PAS
Migration of pain	1	1
Anorexia	1	1
Nausea or vomiting	1	1
Right lower quadrant tenderness	2	2
Rebound pain	1	–
Elevated temperature*	1	1
Leukocytosis (≥10,000/μL)	2	1
Shift of WBC count to the left (≥75% polymorphonucleocytes)	1	1
Cough/percussion/hopping cause pain in the RLQ	–	2
Total	10	10

NEJM Journal Watch

- 0 – 3: discharge
- 4 – 6 (equivocal): imaging
- > 7 (definitive): manage +/- surgery.

Laboratory Investigations

1. Total WBC count: Leukocytosis 10,000 to 18,000/cu.mm (***If >18,000/cu.mm, consider perforation with or without abscess***)
2. Urine analysis: Several WBCs & RBCs may be found in appendicitis 2^0 to bladder irritation from inflamed appendix.
3. Elevated CRP: perforated appendix has higher levels, sensitivity of 58 – 90%

Ultrasound findings

- Sensitivity → 78 – 94%; Specificity → 89 – 98%
- Findings supportive of the diagnosis of appendicitis include :
  - Aperistaltic, non – compressible dilated appendix (> 6mm in diameter)
  - Appendicolith
  - Distinct appendiceal wall layers
  - Echogenic peri – cecal fat
  - Peri – appendiceal fluid collection
  - Target appearance on axial section
  - Sonographic tenderness

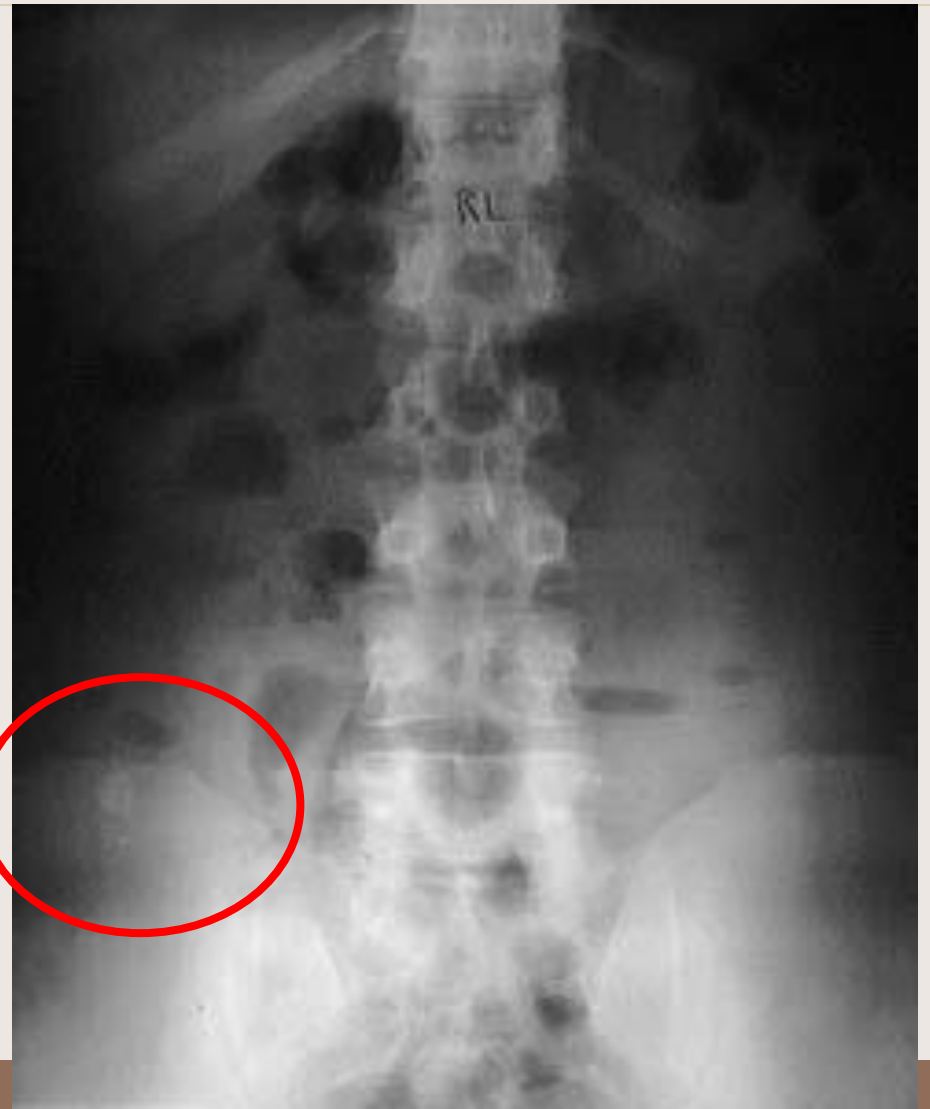
Treatment

- Medical:
- Surgical:
  - Open Appendectomy
  - Laparoscopic Appendectomy

Case

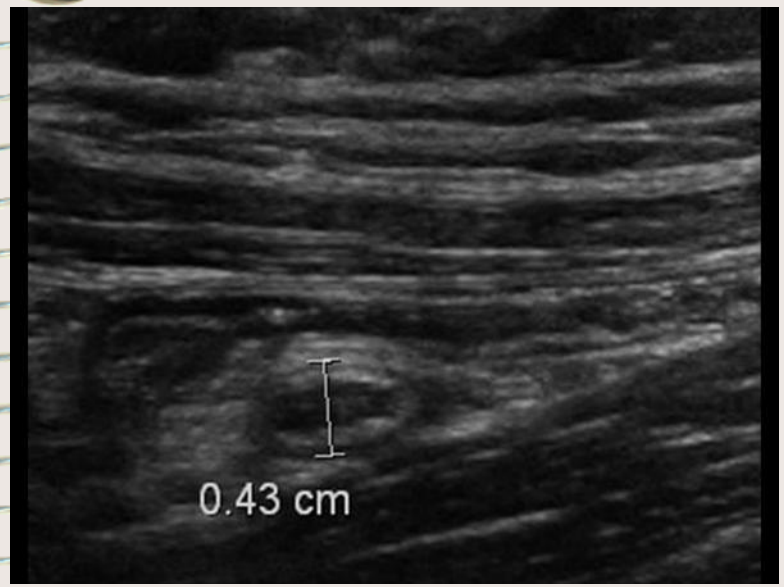
- 6yo male, otherwise healthy, presents to pediatrician with abdominal pain and nausea.
- Dad says patient started complaining about abdominal pain yesterday after school (1st day of school)
- Ate dinner but then woke up around midnight c/o pain again
- Vomited once this am
- Walks hunched over
- H/O occasional constipation

KUB

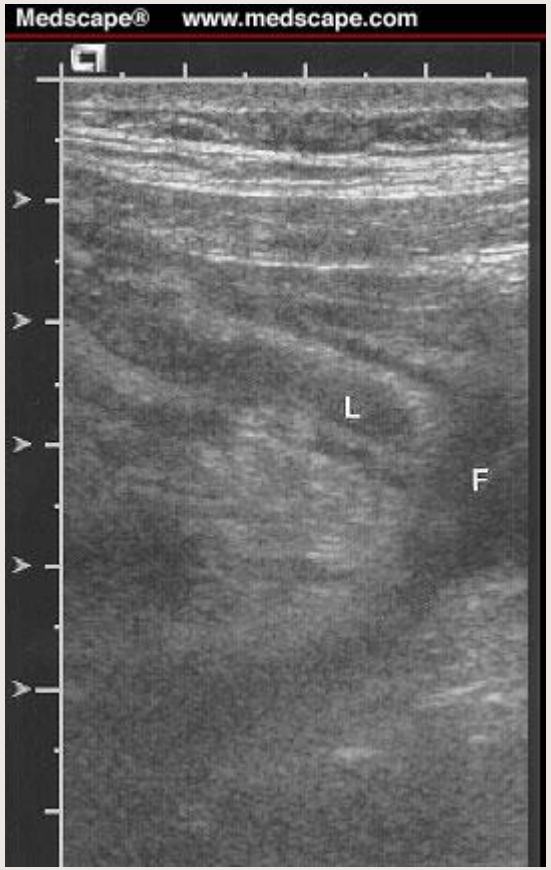


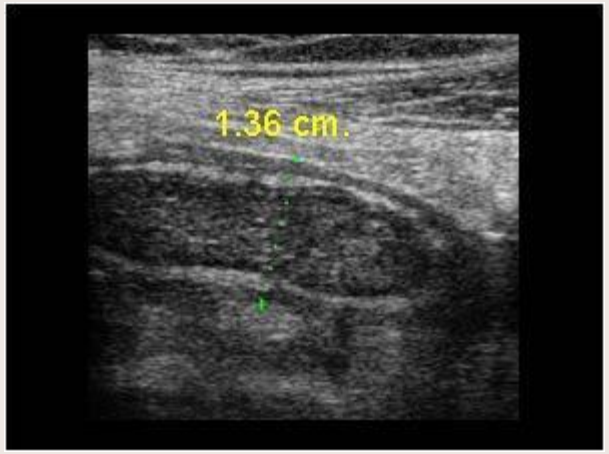
Appendicolith/ fecolith

Cont.

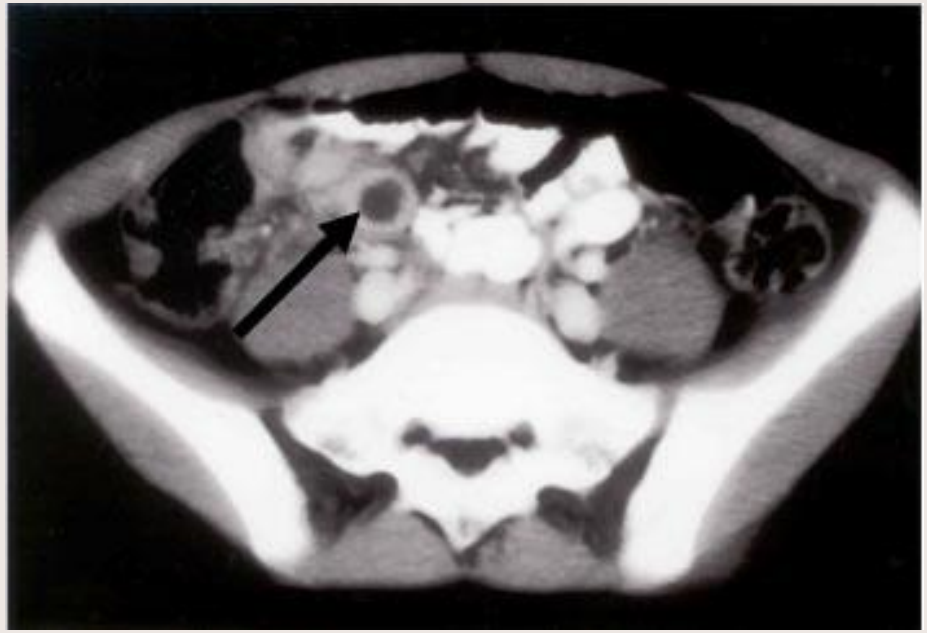


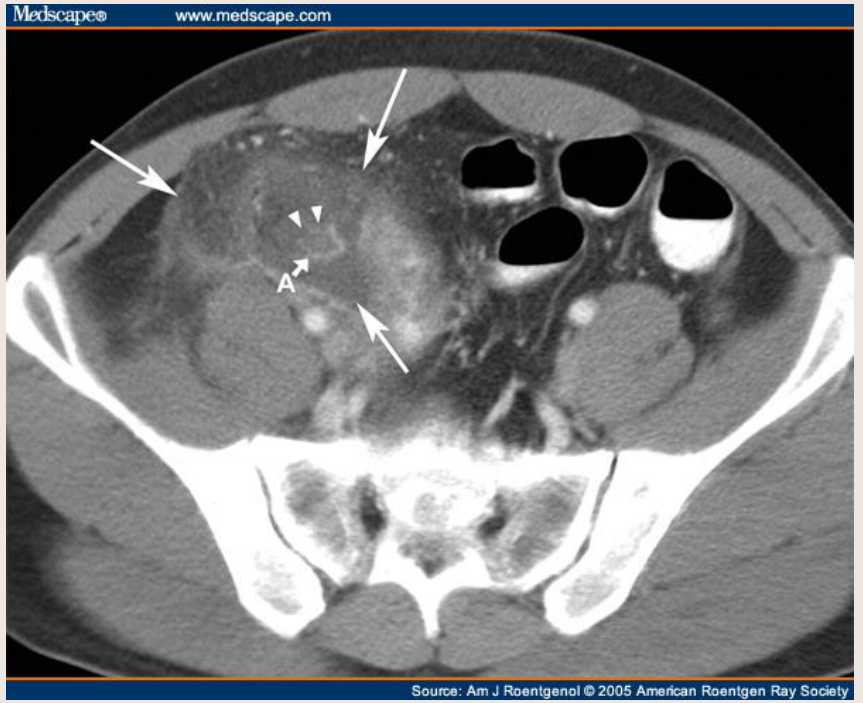
U/S: should be <0.8 cm; if more → appendicitis



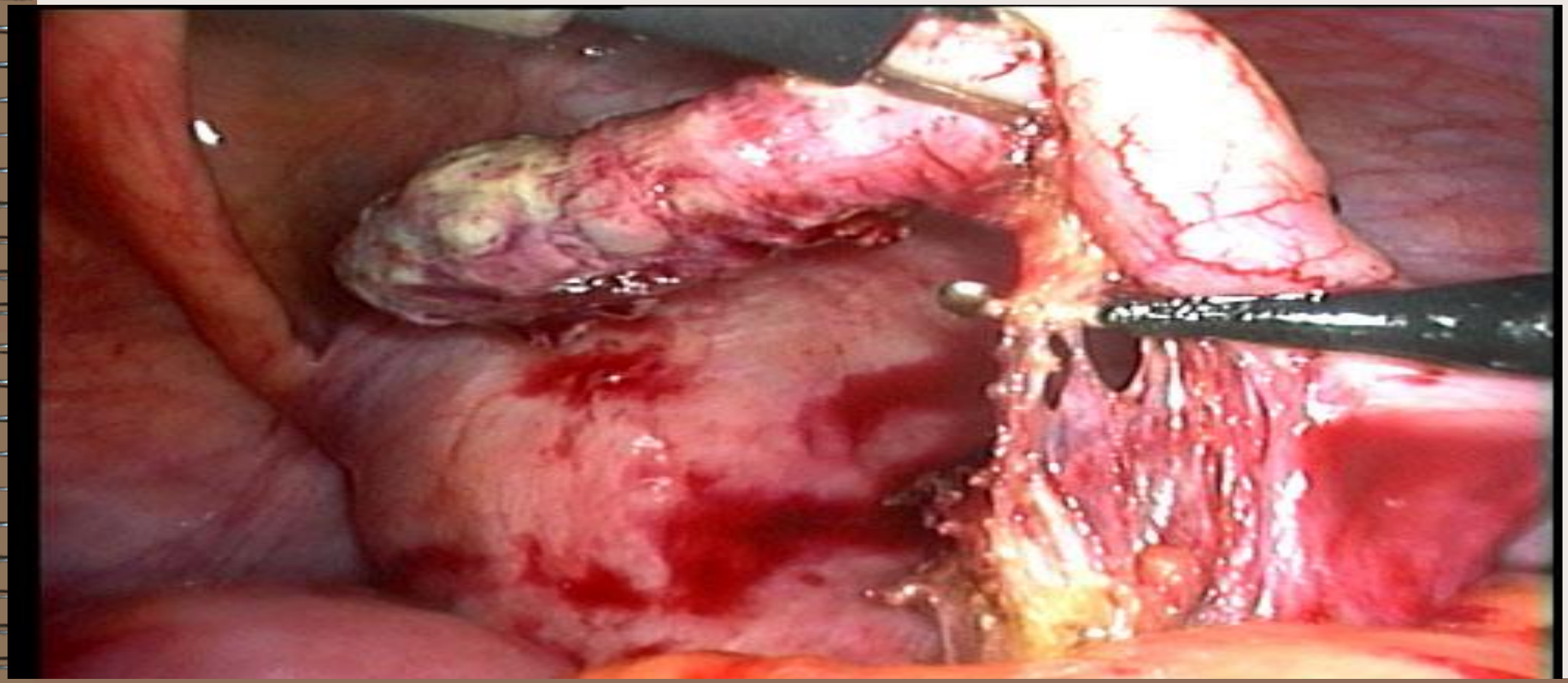


Abdominal CT





Laparoscopic Appendectomy



3. MECKEL'S DIVERTICULUM

3. MECKEL'S DIVERTICULUM

- Remnant of ***vitelline duct*** that connects the lumen of the developing gut to the yolk sac.
- In newborns & infants: presents as ***bowel obstruction*** (*volvulus*, *intussusception*)
- In children: MC presents as ***bleeding*** since it has gastric mucosa that produces gastric acid → peptic ulceration.

Common presentations of MD

- ***LGIB***: Ulcer from ectopic gastric mucosa. Can cause severe bleeding requiring transfusion
- ***Diverticulitis***: like appendicitis (non – shifting pain)
- ***Intussusception***: acts as a pathological lead point
- ***Obstruction***: fibrous band remnant
- ***Littre hernia:*** when the Meckel's diverticulum is constricted in an inguinal hernia.

Age – specific categorization

- Children < 3 years → difficult to diagnose
  - Atypical presentation
  - Don't complain of pain (cry, irritable, poor feeding)
  - Late → septic (lethargic, Non-responsive, vomiting)
- Children > 3 yrs.: Similar to adult Symptom &Signs
- Girls 12 – 16 yrs.
  - DDX ovarian pathology (rupture cyst, torsion)
  - U/S is helpful

Rule of 2s of MD

- Occurs in ***2% of pediatric population***
- Most often ***symptomatic by 2 yrs.*** (only 4% are symptomatic)
- ***2 times*** more common in males
- Presents ***within 2 ft. (60cm) of the ileo – cecal valve***
- ***~ 2 in (5cm) long***

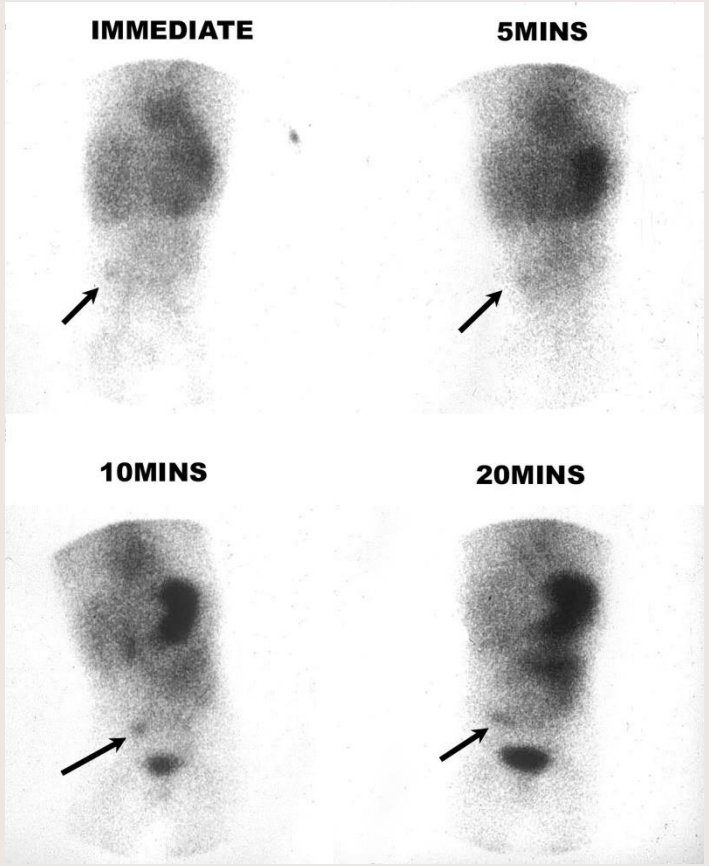
Investigations

- LGIB
  - Meckel's Tc99 scan: uptake by gastric mucosa in Meckel's diverticulum
- Laparoscopy or laparotomy

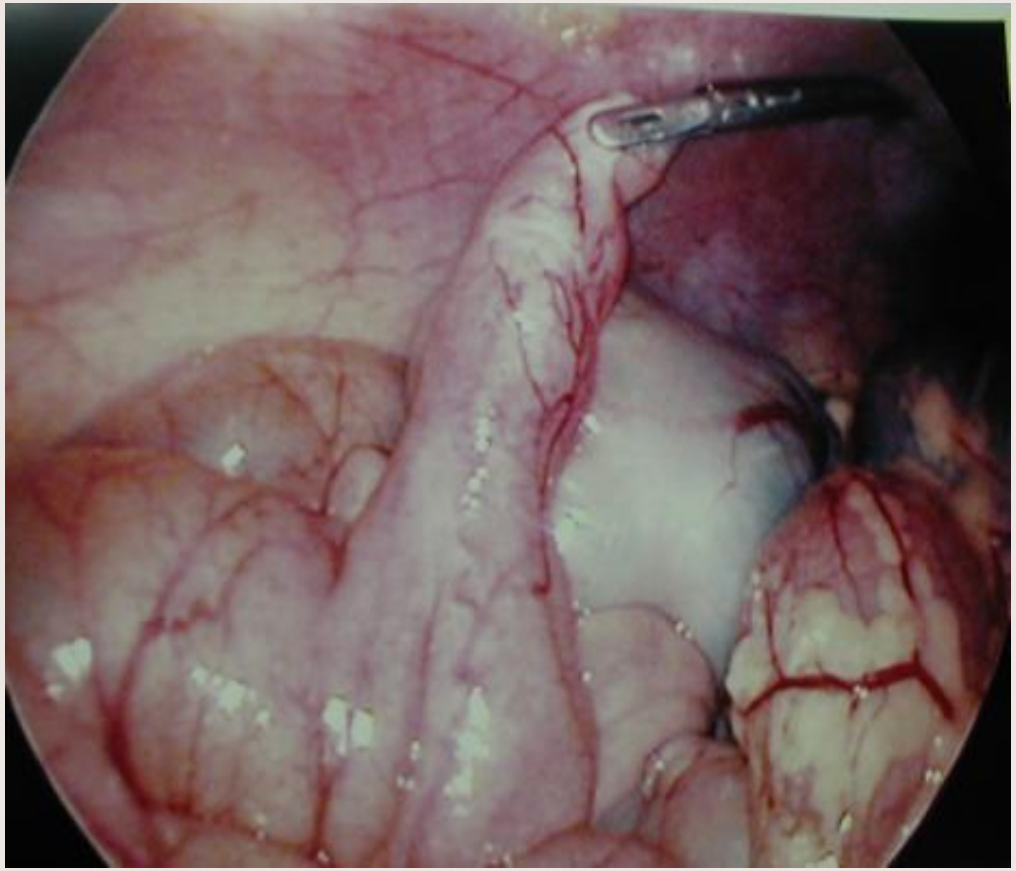
Cont.

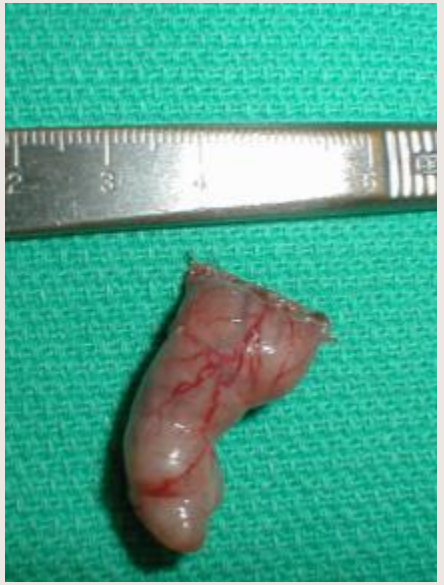
- Can be diagnosed with a Technetium scan
- Pre – treatment with cimetidine enhances uptake of tracer and improves sensitivity
- Often have to repeat scan more than once
- If a 1 – 3 yr. old has 2 significant LGI bleeds requiring transfusion, exploration warranted even if scan is negative
  - DDx: rectal polyps usually don't need transfusion

Tech 99



Meckel's diverticulum





4. NECROTIZING ENTEROCOLITIS

4. NECROTIZING ENTEROCOLITIS (NEC)

- Incidence: 25,000 per year, 10 – 70% mortality
- MC serious GI disease of LBW infants
- Etiology is unknown; thought to be triggered by ***enteral feeding in immature gut***.
- MC in ***terminal ileum*** & ***colon***
  - Pan necrosis involves 75% of gut and occurs in 19% of patients; mortality approaches 100%

Pathophysiology

- Likely multifactorial: involving a combination of:
  - Mucosal compromise (bowel immaturity)
  - Immune system immaturity
  - Pathogenic bacterial proliferation &
  - Substrate: early enteral feedings

Cont.

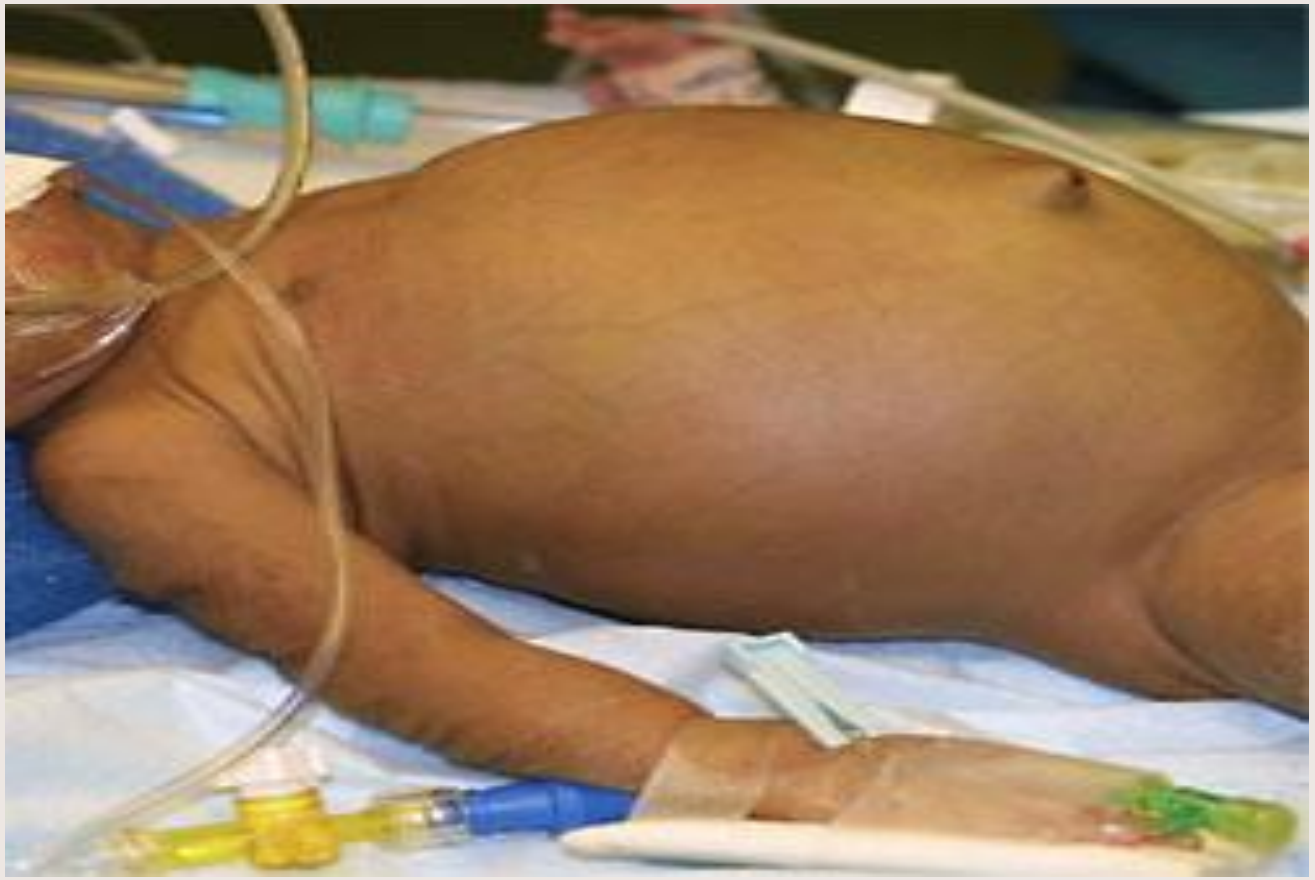
- In a susceptible host, the above result in bowel injury & an inflammatory cascade.
- The ***terminal ileum*** & ***right colon*** are involved in the majority of cases, local vascular component?, low perfusion area?

Presentation

- ***Abdominal distention*** is the MC finding
- ***Feeding intolerance*** (earliest sign) with bilious NG aspirate
- Palpable bowel loops & crepitus
- Edema & erythema of abdominal wall → peritonitis
- Rectal bleeding (***due to sloughing of bowel mucosa and disruption of small arterioles that supply the bowel***) is a common finding: gross &/or occult

Cont.

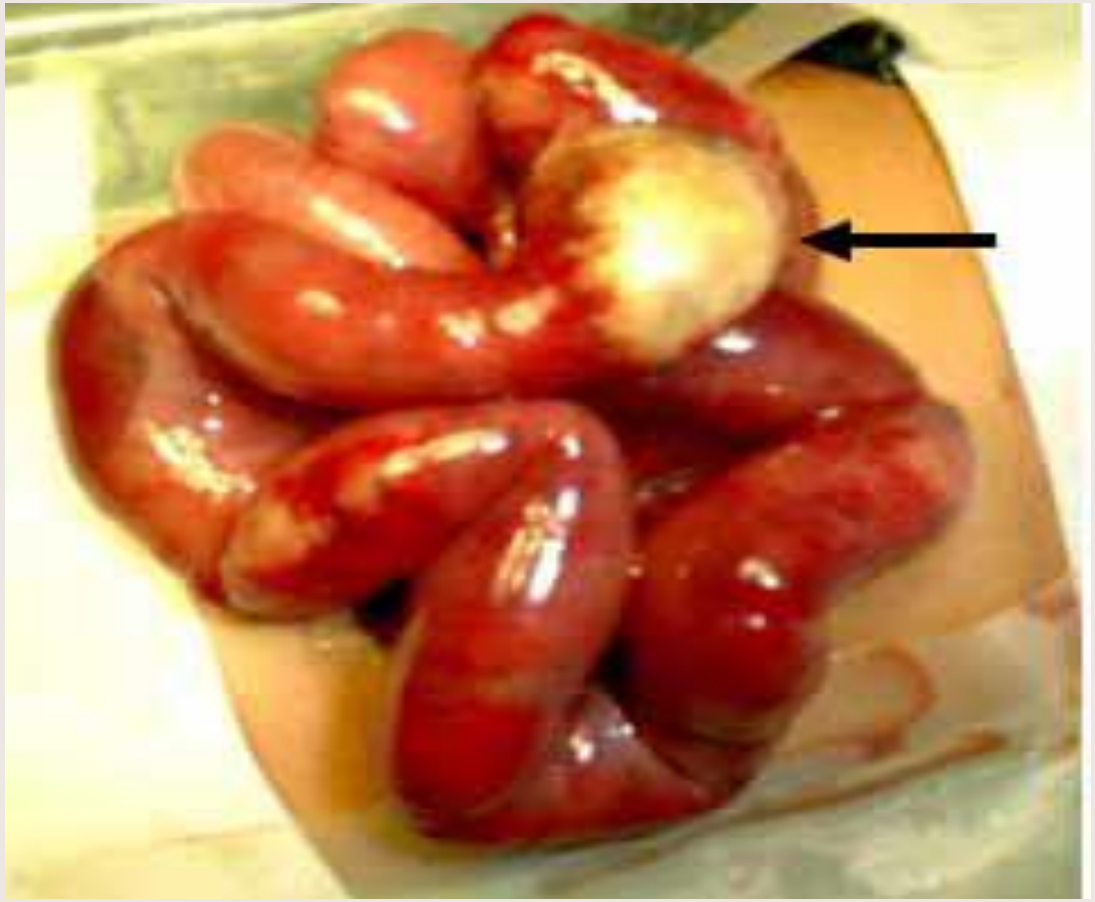
- Distention, tachycardia/ bradycardia, lethargy, bilious output, heme +ve stools, oliguria
- Diagnosis
  - Clinical
  - KUB may show pneumatosis, fixed loop, free air, portal venous gas, ascites

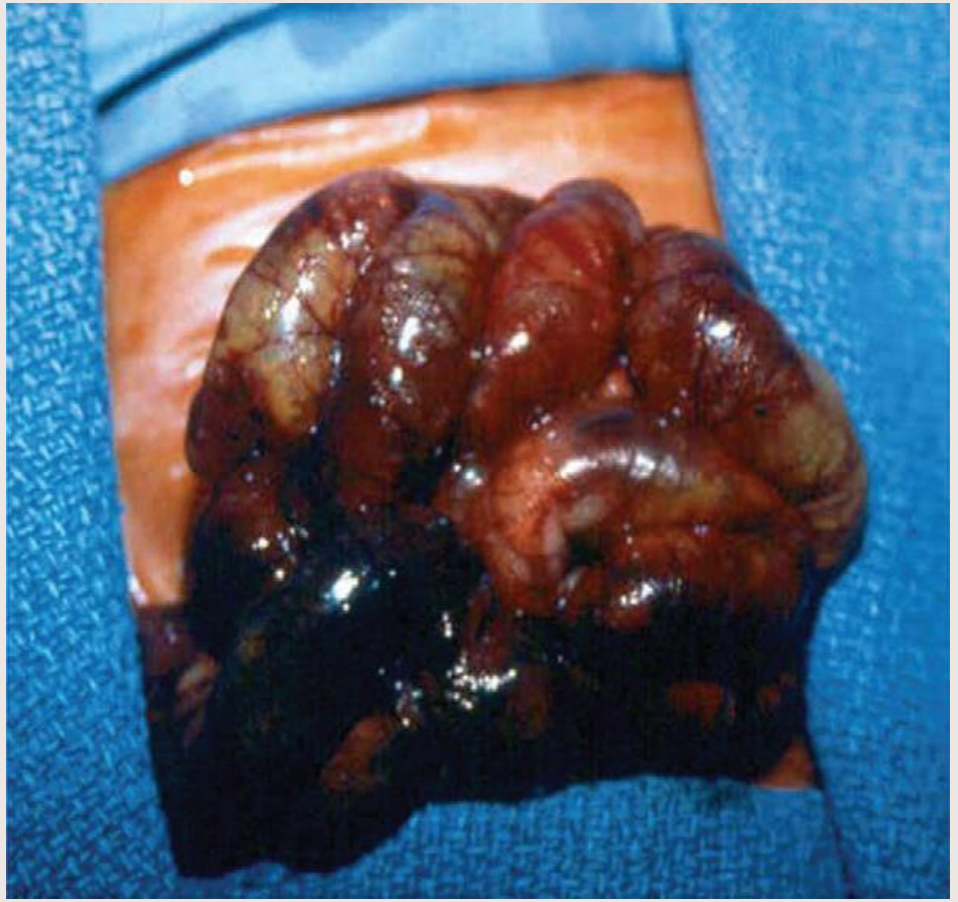


Gross findings

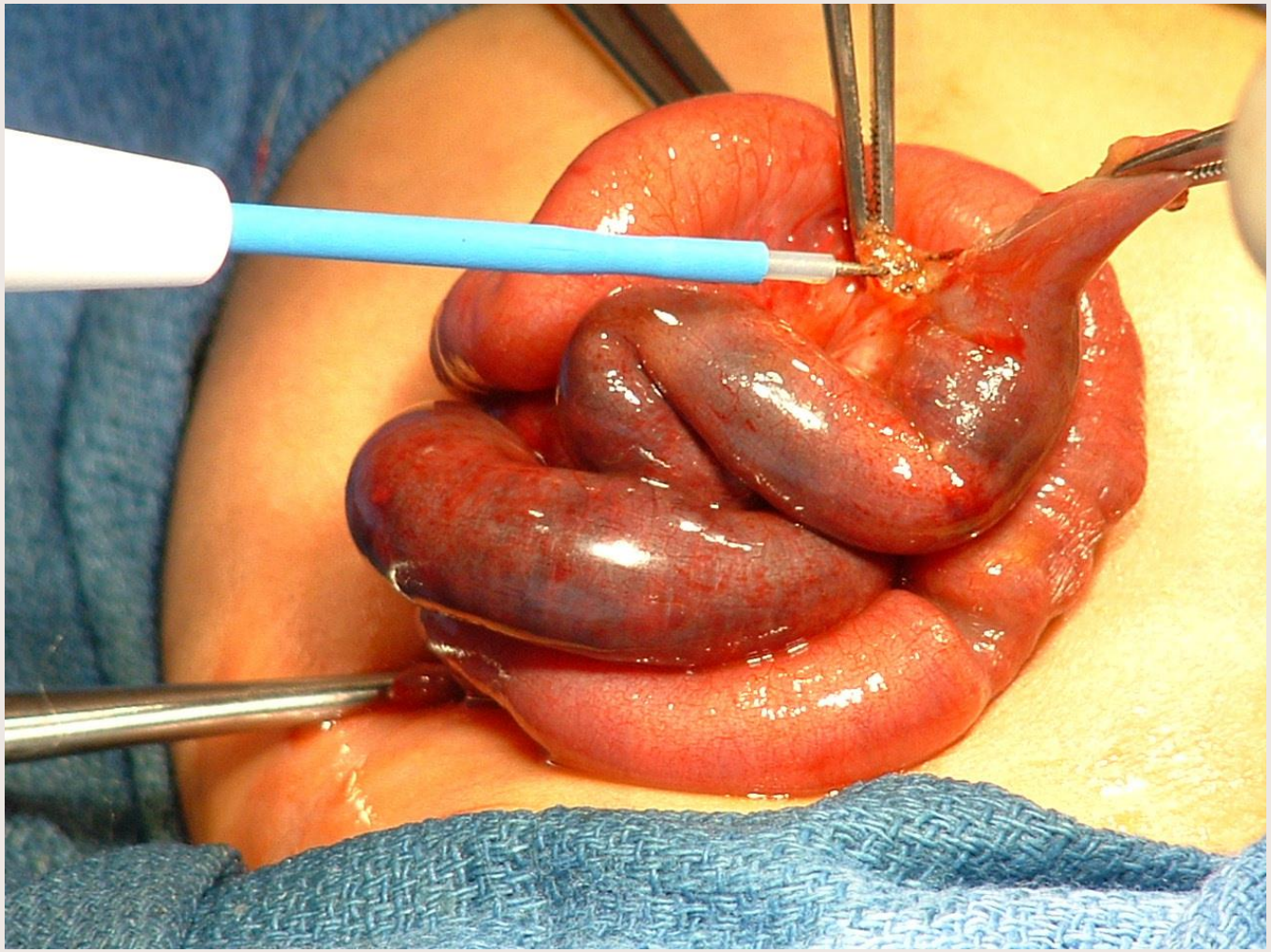
- Bowel appears:
  - Distended & hemorrhagic
  - Sub – serosal collections of gas occasionally are present along the mesenteric border
- Gangrenous necrosis occurs on the anti – mesenteric border & perforation may be present (bland infarcts)
- The affected bowel frequently extends beyond the macroscopic disease seen at surgery.

Cont.

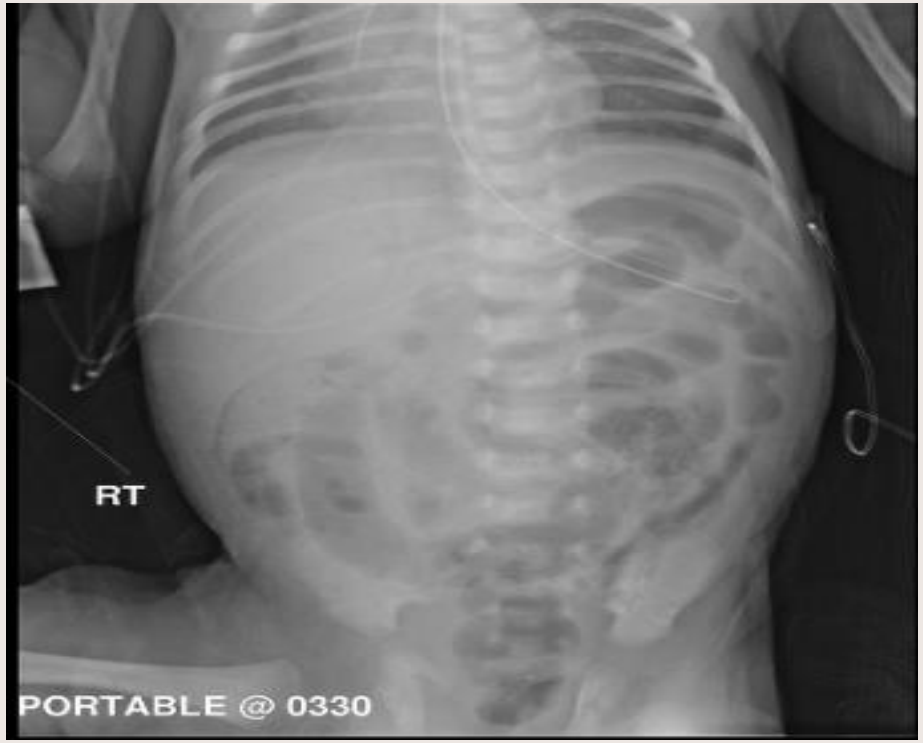


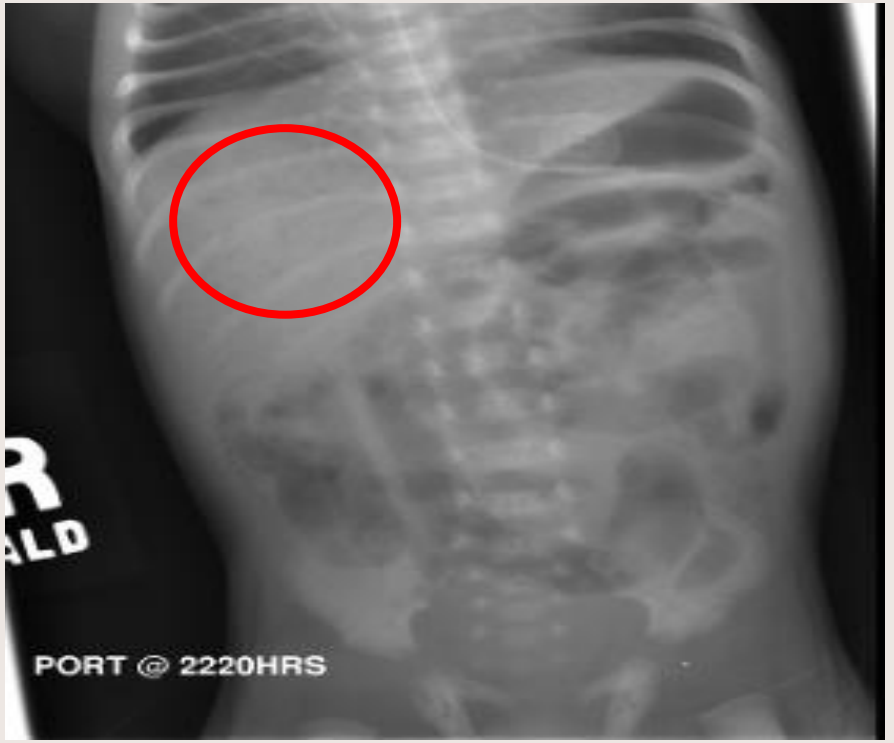


Necrotic segment ileum



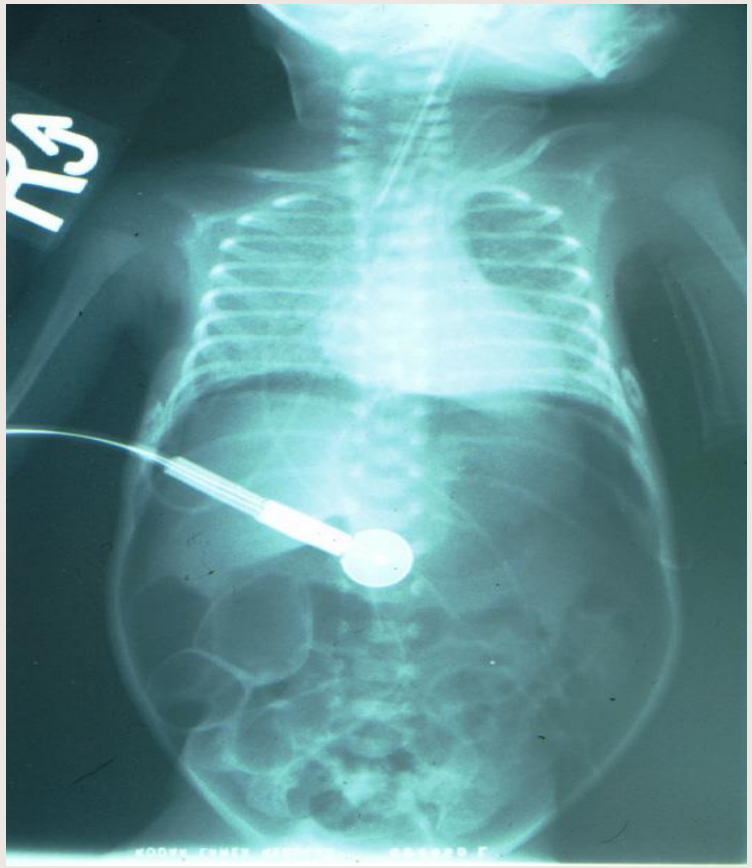
Abdominal films

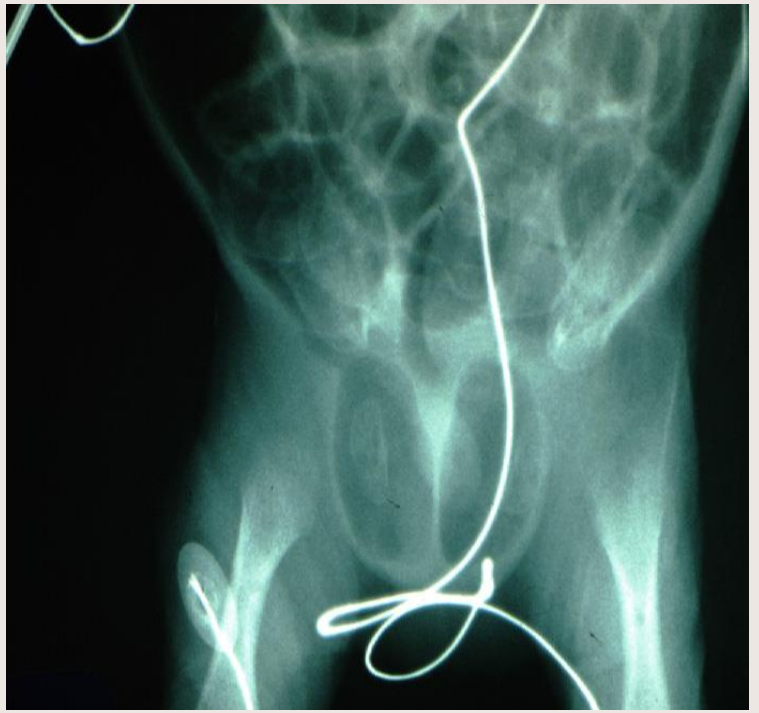




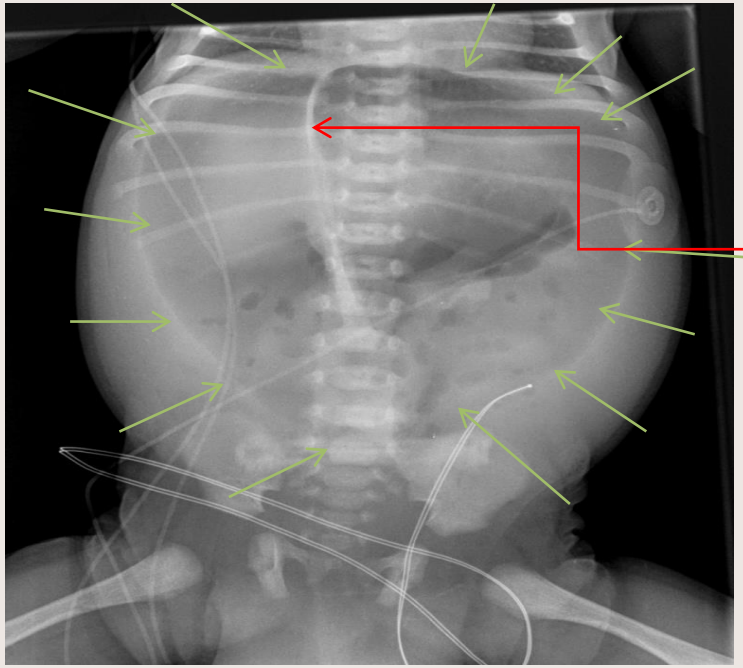
Pneumatoceles: air within bowel wall

Pneumoperitoneum

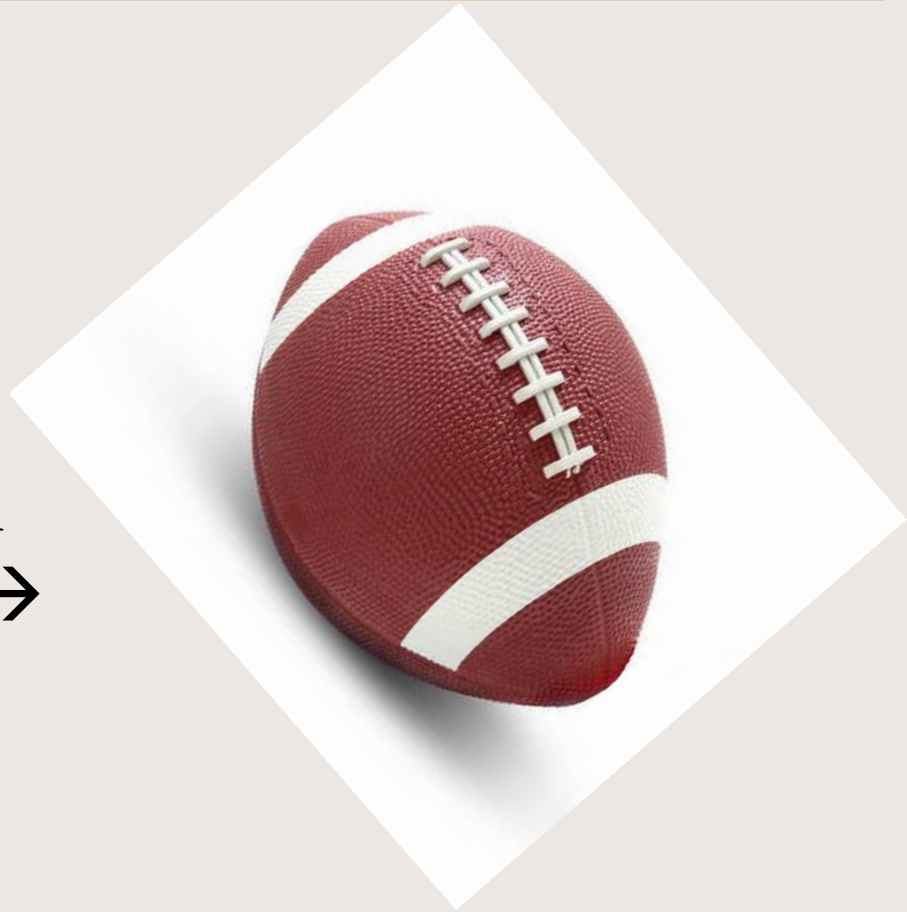




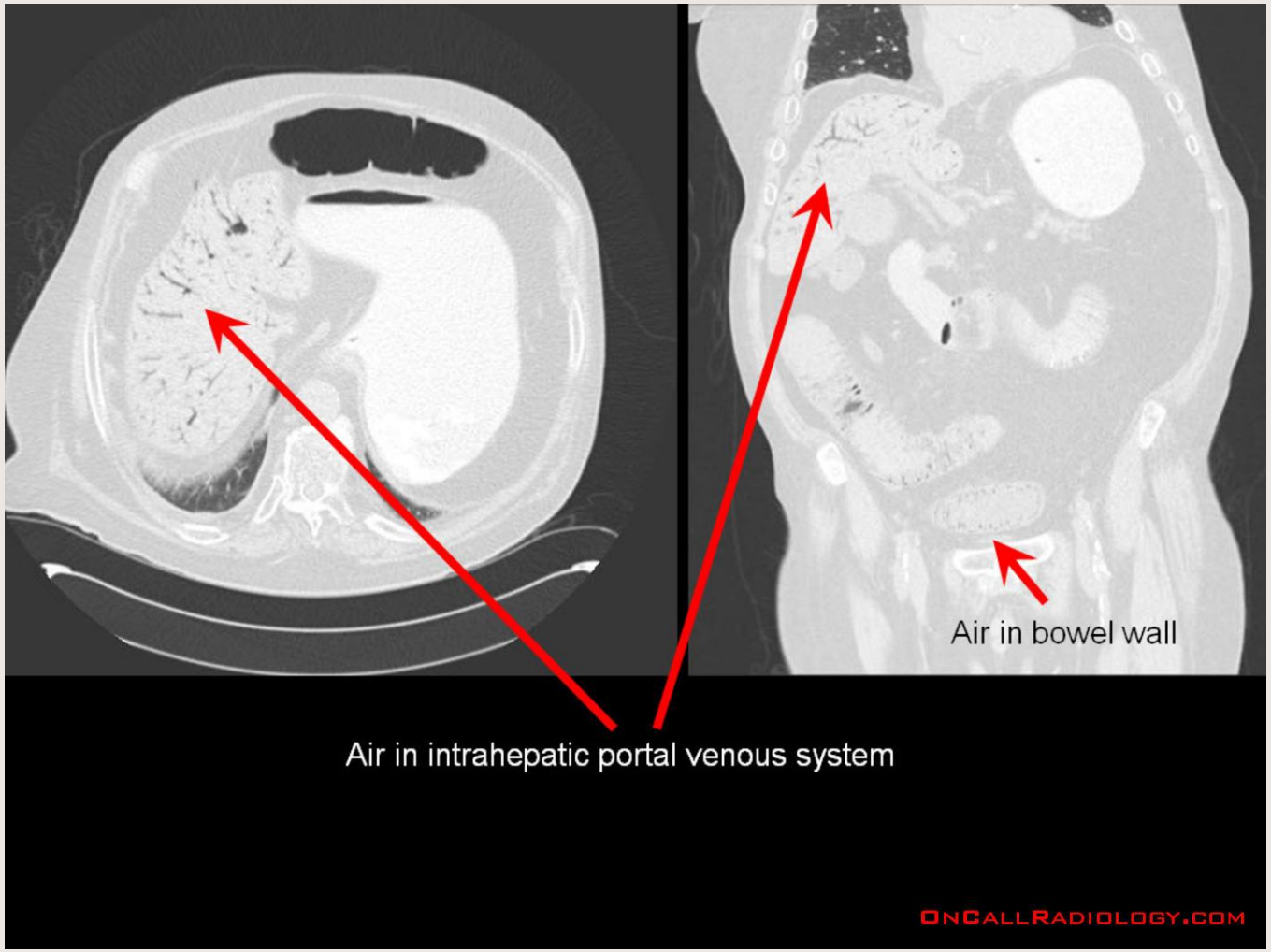
Football sign → large pneumoperitoneum
Falciform ligament → laces



Falciform Ligament → laces



Portal venous Gas



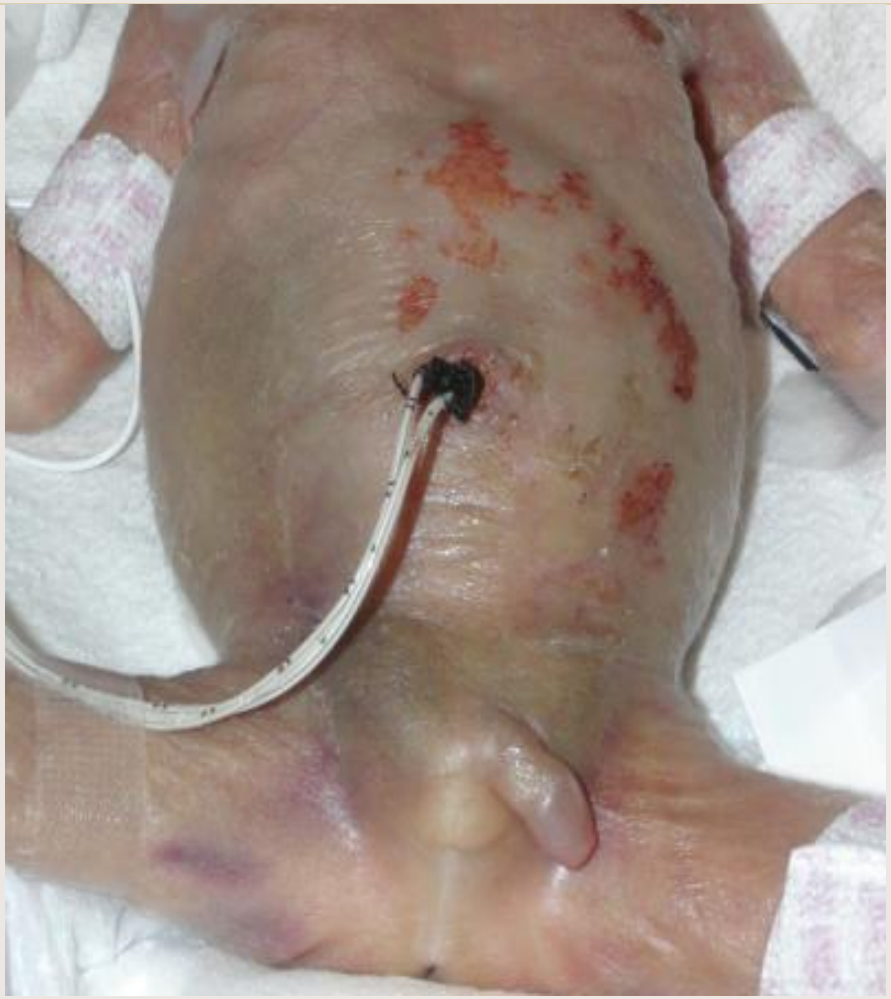
Management

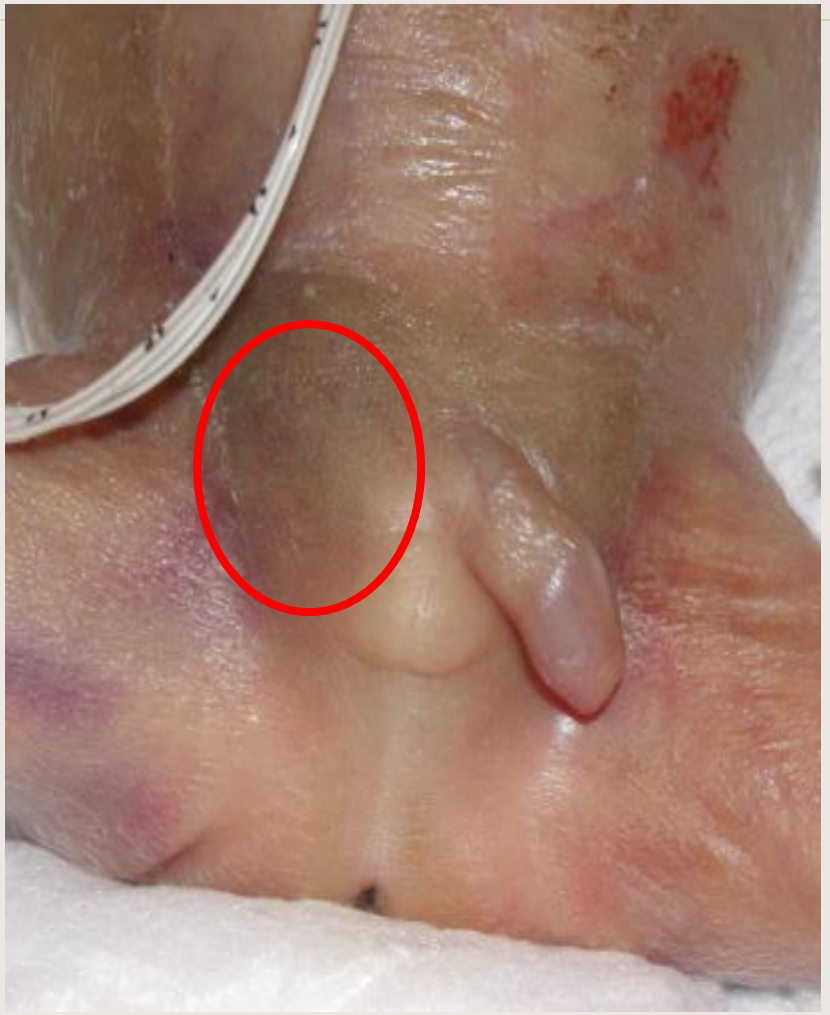
- ***Initial medical management unless evidence of necrosis or perforation.***
- Manage in NICU (*may require mechanical ventilation*)
- NG decompression
- BSAs
- NPO, TPN, fluid resuscitation
- Abdominal film surveillance
- Serial labs: CBS with platelets, ABGs, CRP

Cont.

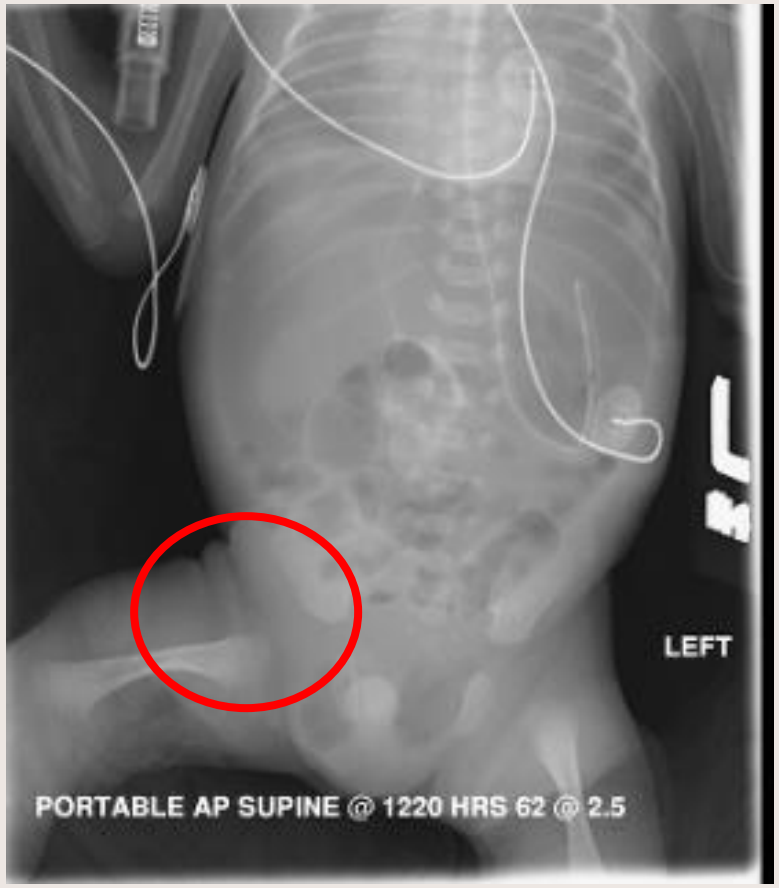
- Surgical management:
  - –Primary peritoneal drainage
  - –Resection and anastomosis
  - –Stomas

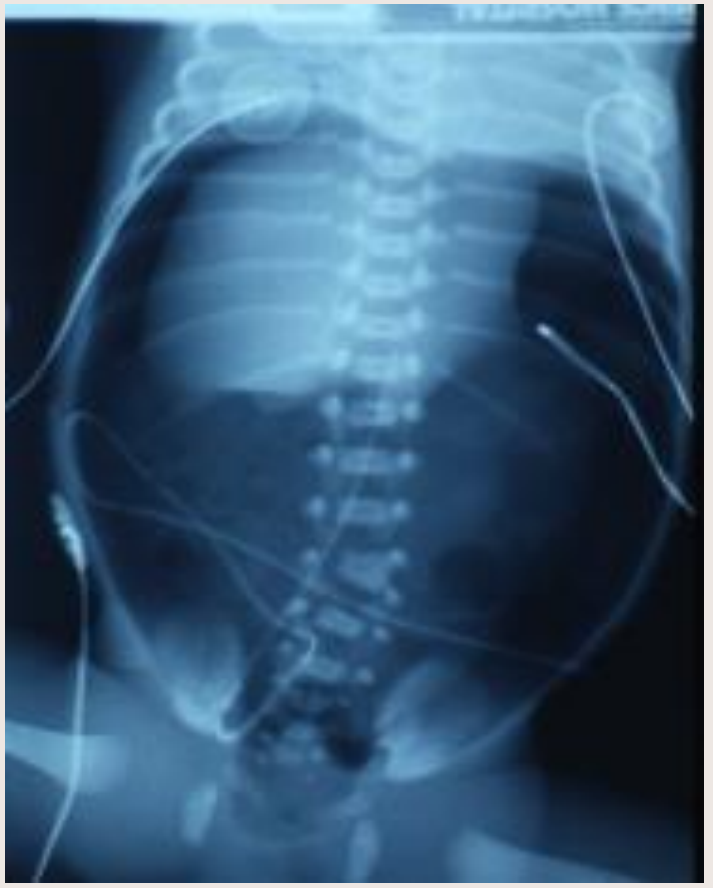
NEC Abdomen



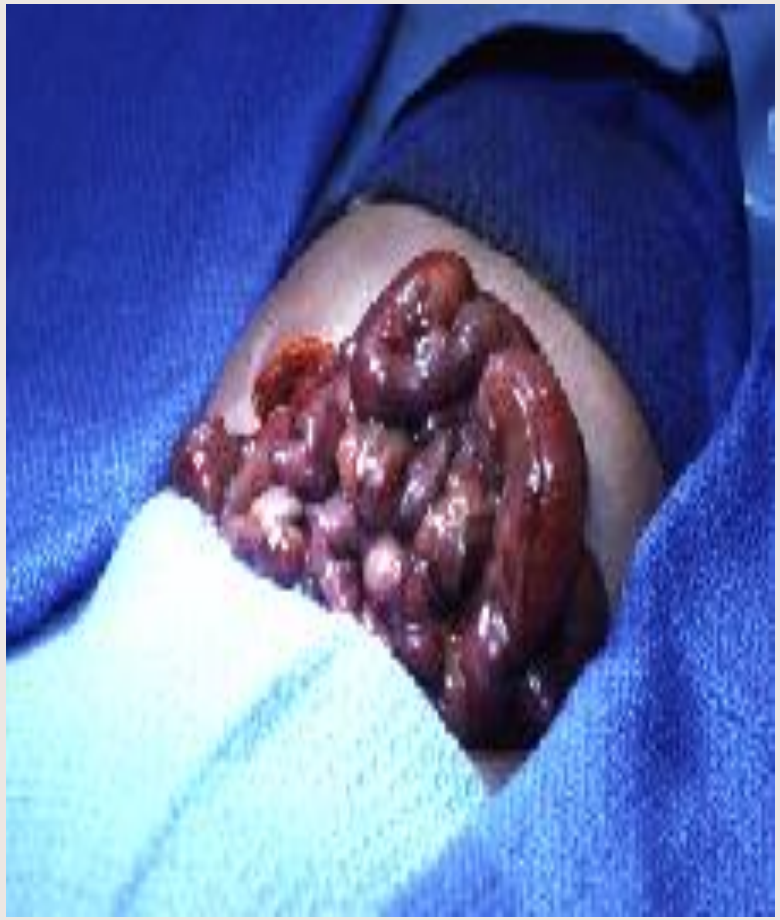


NEC Pneumoperitoneum





NEC totalis



- Best management: ***Put a drain to remove all peritonitic debris***
- Prognosis is generally poor
- ***Desist form surgery.***

Case 1

- A two week old in NICU born at 34 weeks. Develops increasing gastric residue, abdominal distension and bradycardia.
- Mother had PROM
- Has been on formula feeds
- Has temperature irregularity
- Abdomen distended and mottled
- Has been passing a little brownish stools

OTHERS

The acute groin

Table 70-3 Causes of the Acute Scrotum

PATHOLOGY	FREQUENCY	AGE AT PRESENTATION
Extravaginal torsion of the testis	Uncommon	Perinatal
Intravaginal torsion of the testis	Common	Any time, peak at 13–16 years
Torsion of testicular appendage	Very common	Any time, peak at 11 years
Epididymitis	Rare	0–6 months
Mumps orchitis	Uncommon	Only after puberty
Idiopathic scrotal edema	Uncommon	0–5 years
Fat necrosis of scrotum	Rare	5–15 years

Testicular torsion

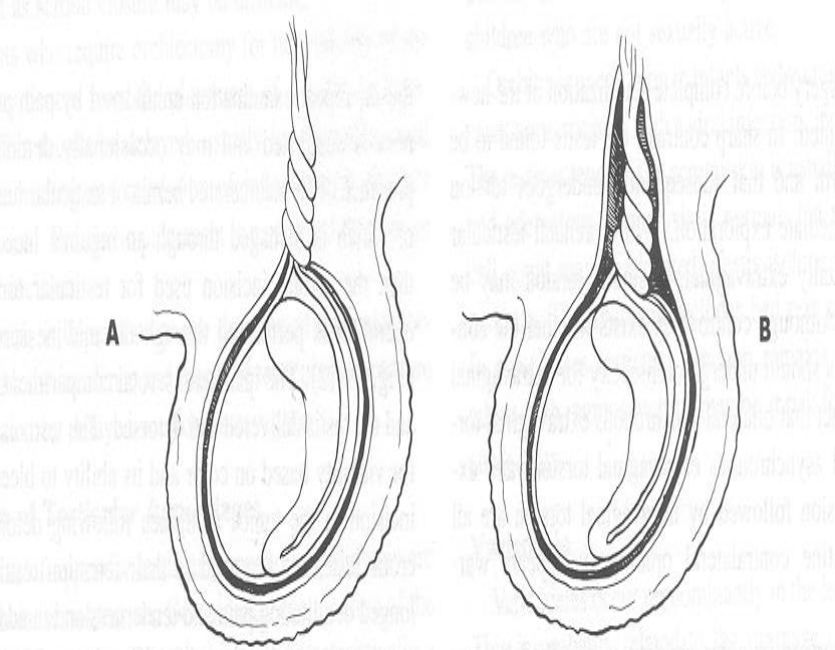
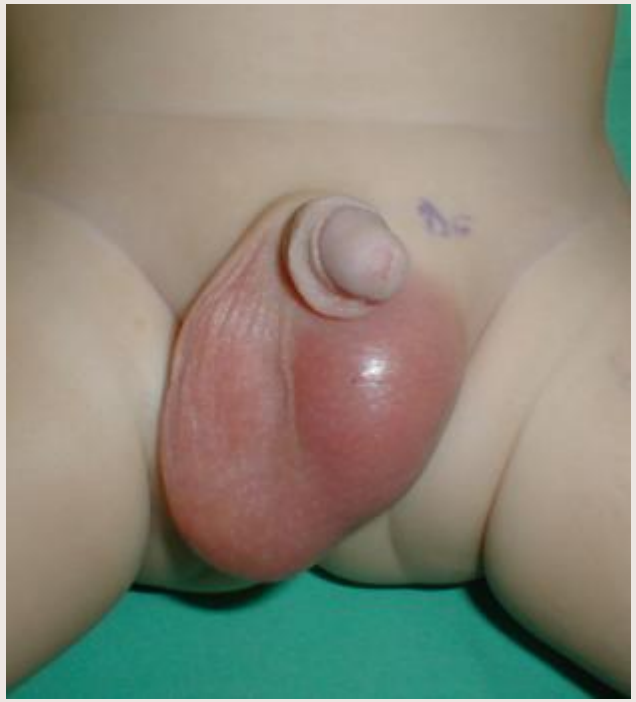
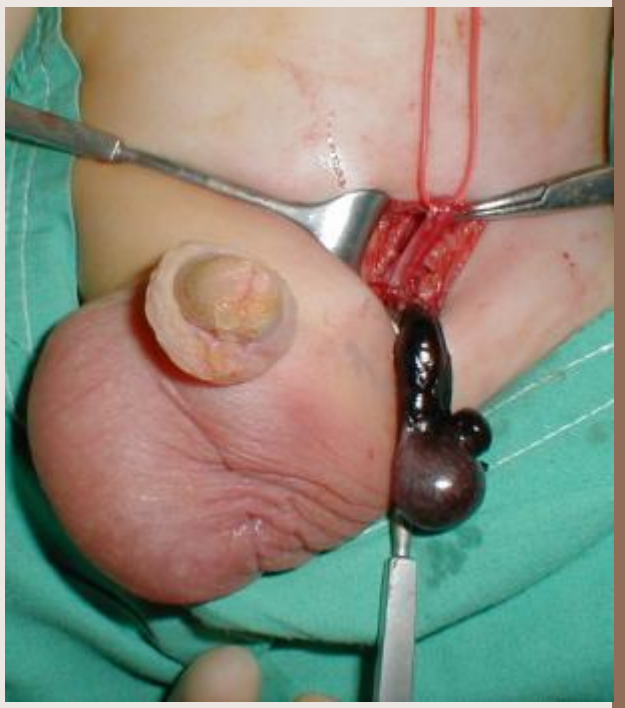
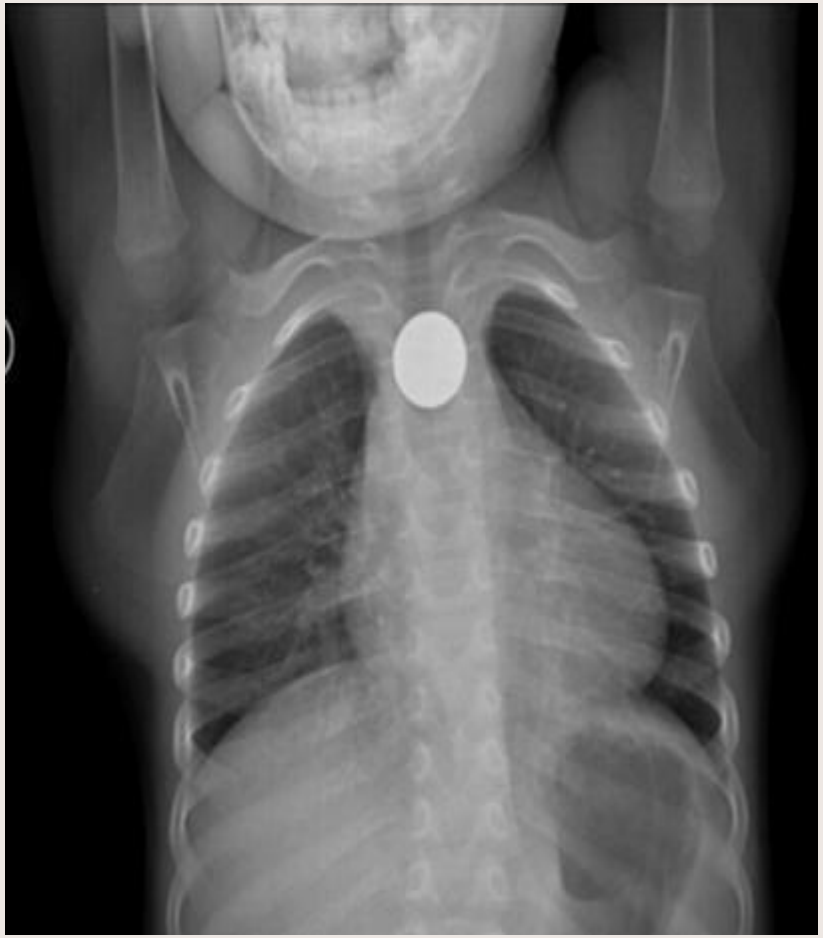


Fig. 49-9 Classification of testicular torsion. A, Extravaginal. B, Intravaginal.





Esophageal coin
Beware of coin battery

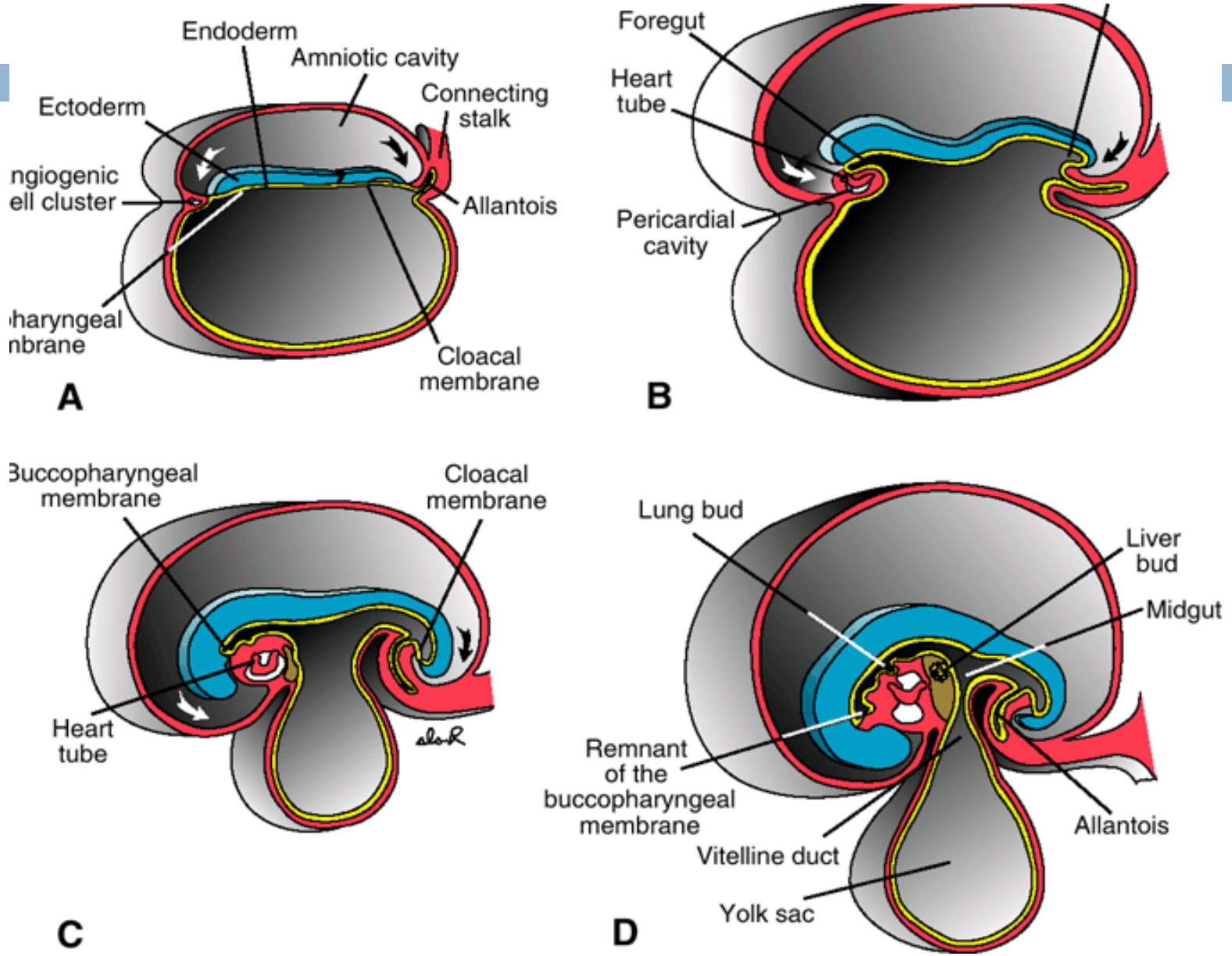


Summary

- Bilious vomiting is surgical unless proven otherwise. Non – bilious vomiting can at times be surgical.
- Resuscitation of children is paramount to good outcomes.
- In most cases, definitive management comes AFTER adequate fluid and electrolyte resuscitation
- Clinical diagnosis more useful than doing battery of tests that take long to come back
- Timely diagnosis is of prime importance

4. DISORDERS OF THE UMBILICUS & ANTERIOR ABDOMINAL WALL

BY: DR. MWIKA M. P.
CONSULTANT PEDIATRIC SURGEON


Endoderm
Amniotic cavity
Ectoderm
Connecting stalk
ngiogenic ell cluster
Allantois
haryngeal nbrane
Cloacal membrane
A
Foregut
Heart tube
Pericardial cavity
B
Buccopharyngeal membrane
Cloacal membrane
Heart tube
C
Lung bud
Liver bud
Midgut
Remnant of the buccopharyngeal membrane
Allantois
Vitelline duct
Yolk sac
D

Abdominal wall development

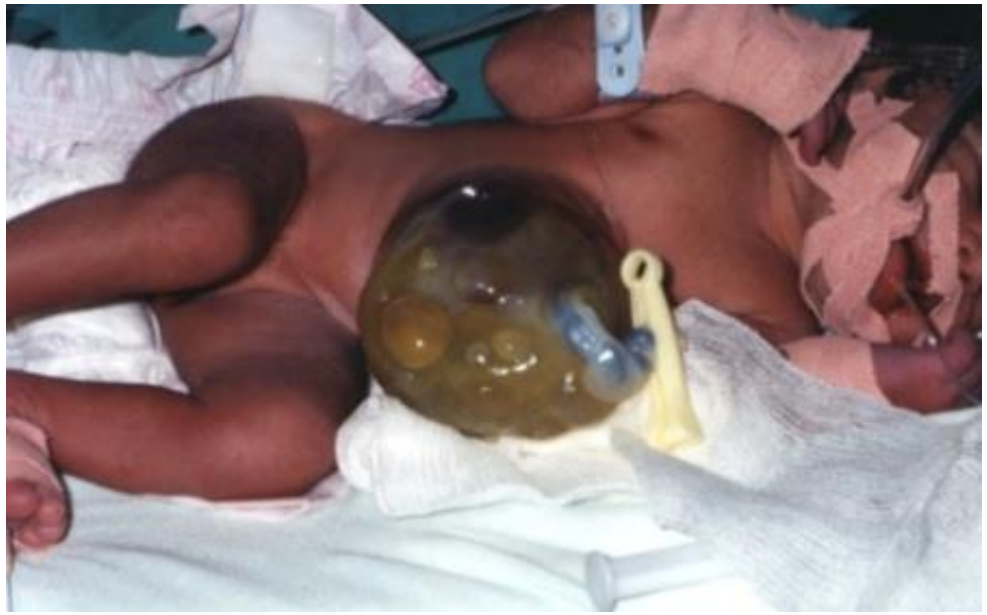
- Formed by infolding of the **cranial, caudal,** and **two lateral** embryonic folds.

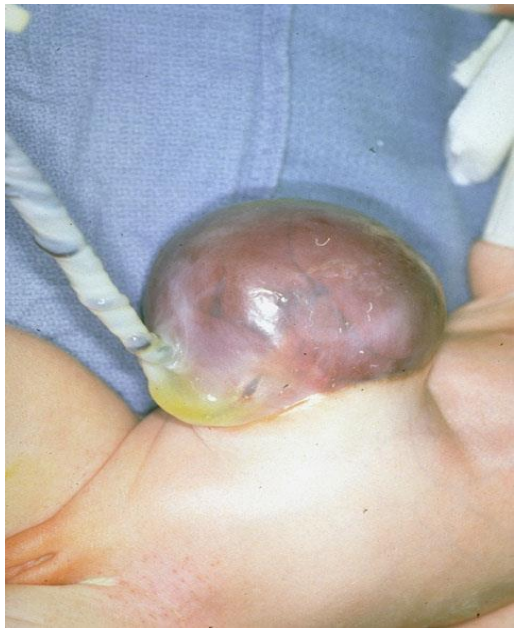
- Rapid growth of the intestinal tract leads to its migration outside the abdominal cavity through the umbilical ring and into the umbilical cord during the sixth week of gestation.

- By the 10th to 12th week, the abdominal wall is well formed and the intestine returns to the abdominal cavity.

Exomphalos (omphalocele)

- CONGENITAL Anterior abdominal wall defect at the base of the umbilical cord with herniation of the abdominal contents



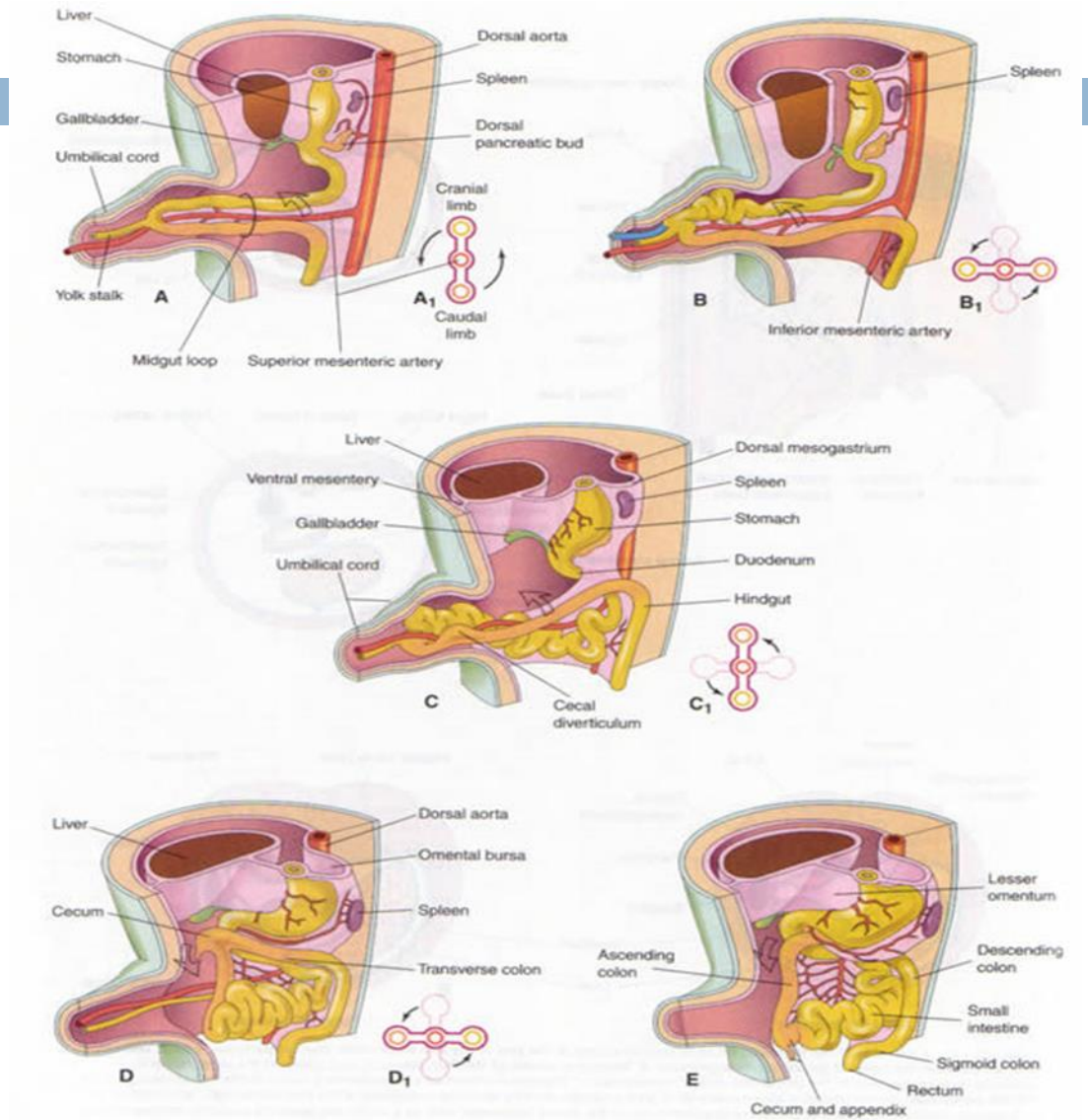


Etiology: Omphalocele

- Etiology not known.

- Theories:

1) Failure of bowel to return into the developing abdomen by 10 – 12 weeks. (1)
2) Failure of lateral mesodermal body folds to migrate centrally


Liver
Stomach
Gallbladder
Umbilical cord
Yolk stalk
A
Midgut loop
Superior mesenteric artery
Dorsal aorta
Spleen
Dorsal pancreatic bud
Cranial limb
A1
Caudal limb
Spleen
B
B1
Inferior mesenteric artery
Liver
Ventral mesentery
Gallbladder
Umbilical cord
C
Cecal diverticulum
Dorsal mesogastrium
Spleen
Stomach
Duodenum
Hindgut
C1
Liver
Cecum
D
Dorsal aorta
Omental bursa
Spleen
Transverse colon
D1
Ascending colon
E
Lesser omentum
Descending colon
Small intestine
Sigmoid colon
Rectum
Cecum and appendix

Contin…

- Cranial fold deficits predominately result in epigastric omphaloceles that may be associated with additional cranial fold abnormalities;

  - diaphragmatic hernia,
  - sternal clefts,
  - pericardial defects, and
  - Cardiac defects.
  - **Pentalogy of Cantrell**

Contin…

- When the infolding deficit involves the **caudal fold**, the omphalocele may be associated with **bladder** or **cloacal exstrophy**

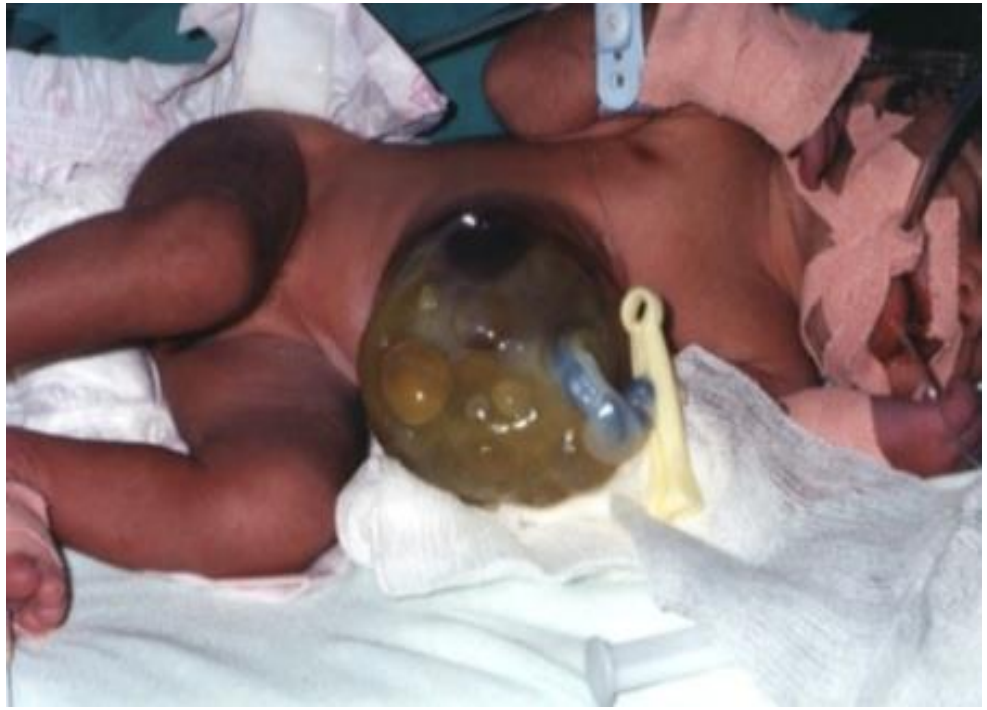
Epidemiology

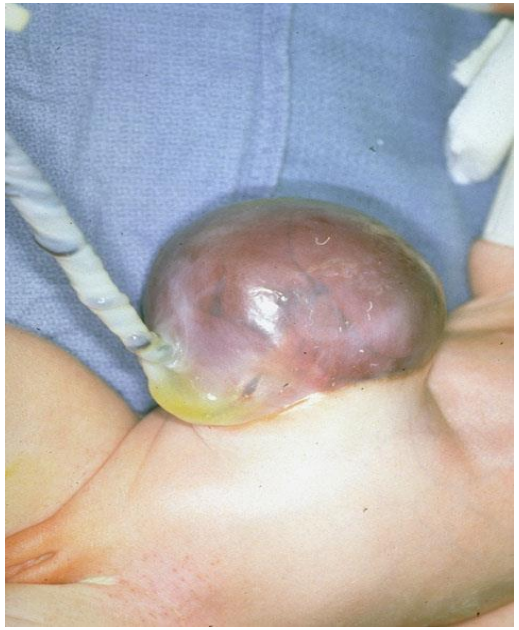
- Incidence of omphalocele ranges between 1.5 and 3 per 10,000 births

Clinical Findings

- Central defect of the abdominal wall beneath the umbilical ring.
- Defect may vary from 2-10 cm
- **Always** covered by sac which may be intact or ruptured
- Sac is composed of **amnion**, **Wharton's jelly** and **peritoneum**
- The umbilical cord inserts directly into the sac in an apical or occasionally lateral position.
- Sac contains intestinal loops, liver, spleen and bladder , testes/ovary
- >50% have associated defects

Exomphalos (omphalocele)





Associated defects

- Chromosome anomalies; trisomy 13, 14, 15, 18, and 21, are present in up to 30% of cases.
- Cardiac defects are present in 30% to 50% of cases.

- Multiple anomalies are frequent and may be clustered in syndromic patterns.

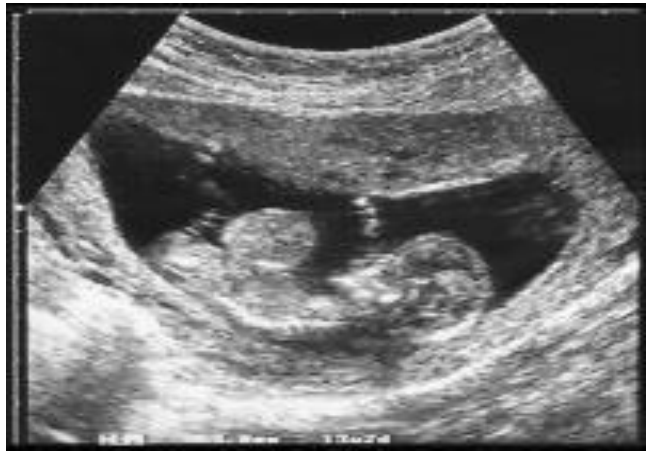
e.g **Beckwith-Wiedemann syndrome** that may be present in up to 10% of cases

- *size of the abdominal wall defect in omphalocele does not directly correlate with the presence of other anomalies,*

Prenatal diagnosis

- Elevated maternal serum alpha fetoprotein (AFP)

- Ultrasound



13 wk embryo

Newborn management

- starts with the ABCs of resuscitation
- warm environment to minimise heat loss
- check and mentain normal glucose levels
- Gastric decompression

GASTROSCHISIS

- Defect of the anterior abdominal wall just lateral to the umbilicus

Incidence

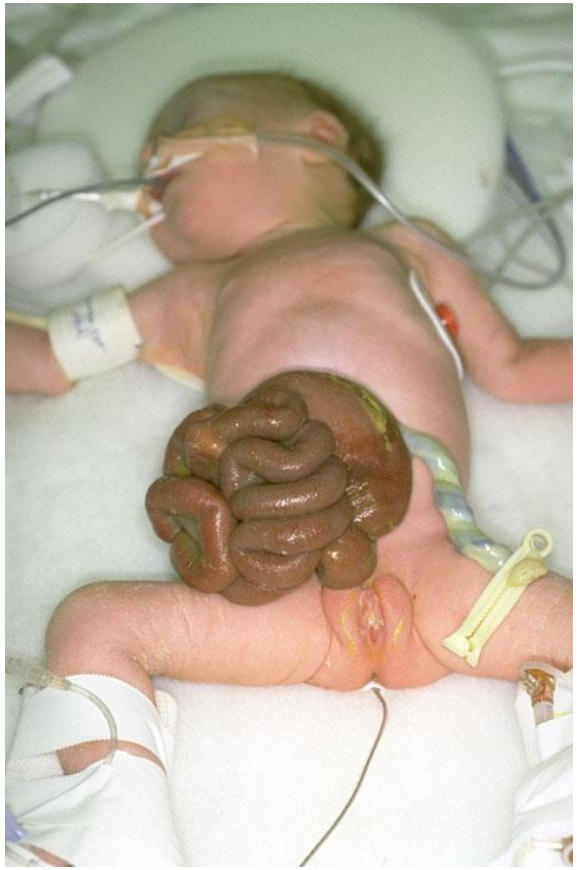
- 1:20,000-30,000
- Sex ratio 1:1
- 10-15% have associated anomalies
- 40% are premature/SGA

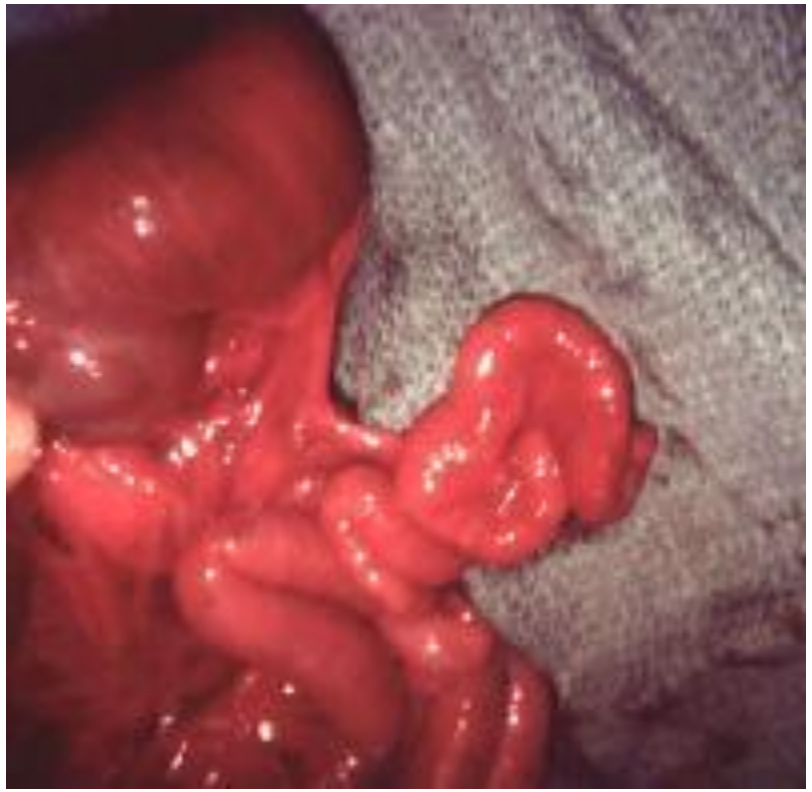
Pathophysiology

- Abnormal involution of right umbilical vein
- Rupture of a small omphalocoele
- Failure of migration and fusion of the lateral folds of the embryonic disc on the 3rd-4th week of gestation

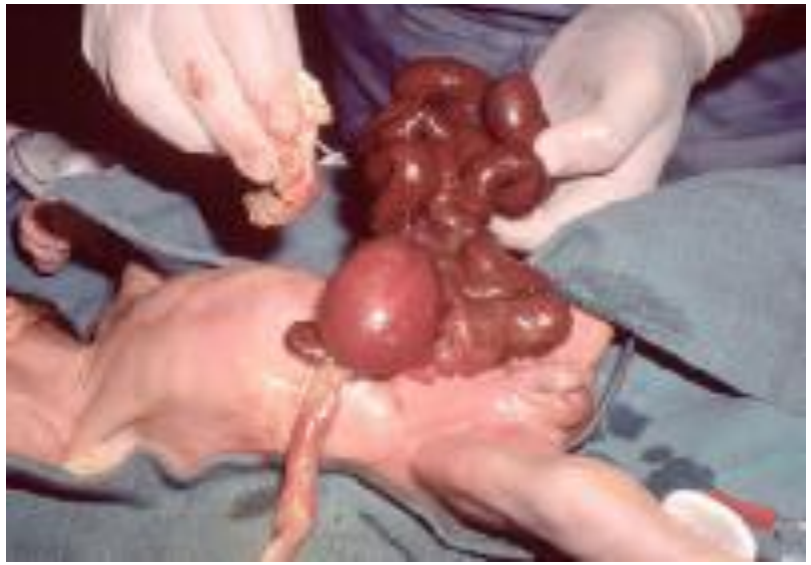
Clinical Findings

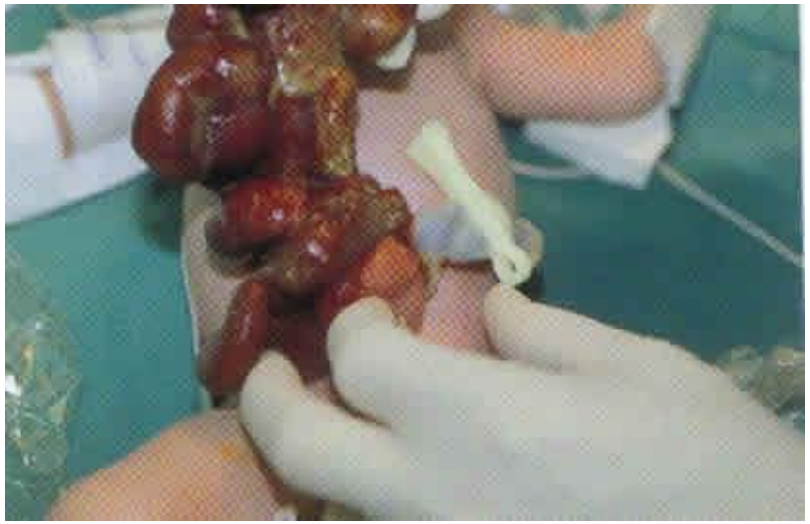
- Defect to the right of an intact umbilical cord allowing extrusion of abdominal content
- Opening ≅ 5 cm
- No covering sac





- Bowels often thickened, matted and edematous
- 10-15% with intestinal atresia





MANAGEMENT

- ABC
- Heat Management
  - Sterile wrap or sterile bowel bag
  - Radiant warmer
- Fluid Management
  - IV bolus 20 ml/kg LR/NS
  - D10¼NS 2-3 maintenance rate

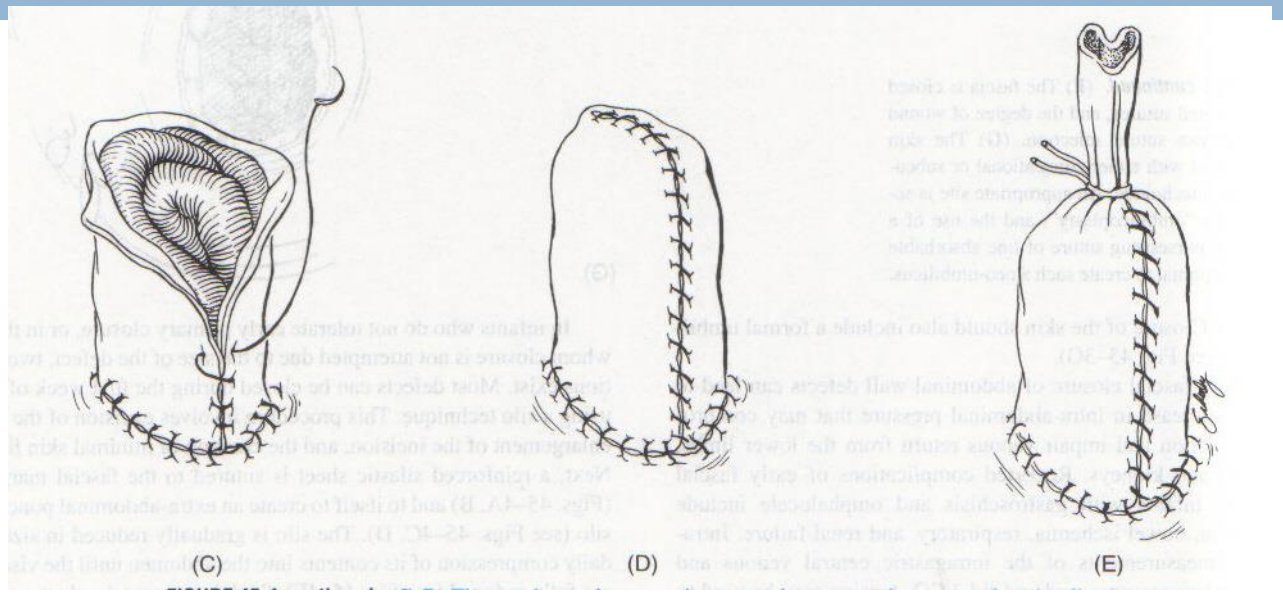
- Nutrition
  - NPO and TPN
- Gastric Distention
  - OG/NG tube
- Infection Control
  - Ampicillin and Gentamicin
- Associated Defects

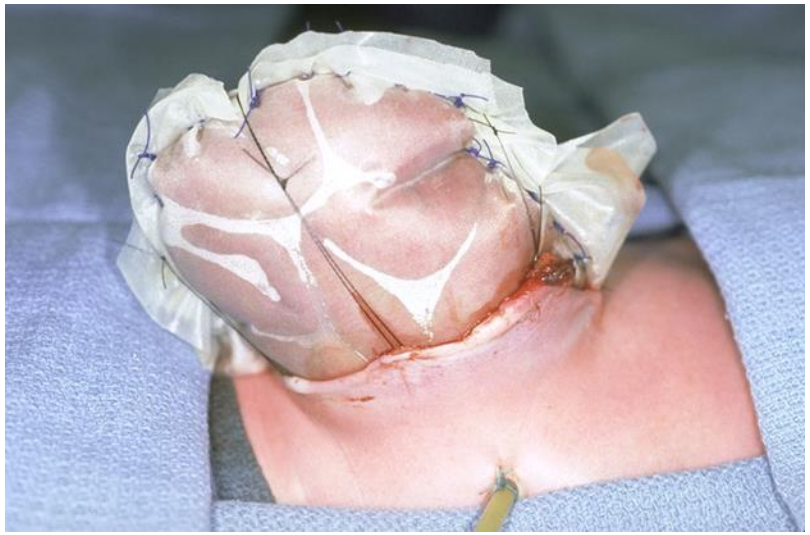
Management: omphalocele

- Conservative treatment

- Painting sac with escharotic agent
  - 0.25% Silver nitrate
  - 0.25% Merbromin (Mercurochrome)

mx. Ruptured ompholecele



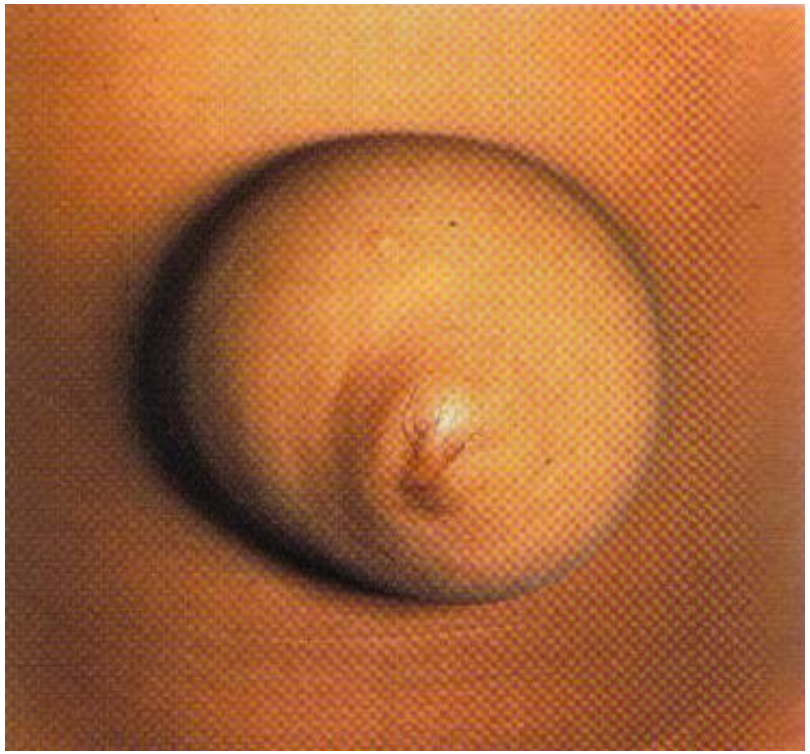


	Omphalocoele	Gastroschisis
Incidence	1:6,000-10,000	1:20,000-30,000
Delivery	Vaginal or CS	CS
Covering Sac	Present	Absent
Size of Defect	Small or large	Small
Cord Location	Onto the sac	On abdominal wall
Bowel	Normal	Edematous, matted

	Omphalocoele	Gastroschisis
Other Organs	Liver often out	Rare
Prematurity	10-20%	50-60%
IUGR	Less common	Common
NEC	If sac is ruptured	18%
Associated Anomalies	>50%	10-15%
Treatment	Staged/primary	Often staged
Prognosis	20%-70%	70-90%

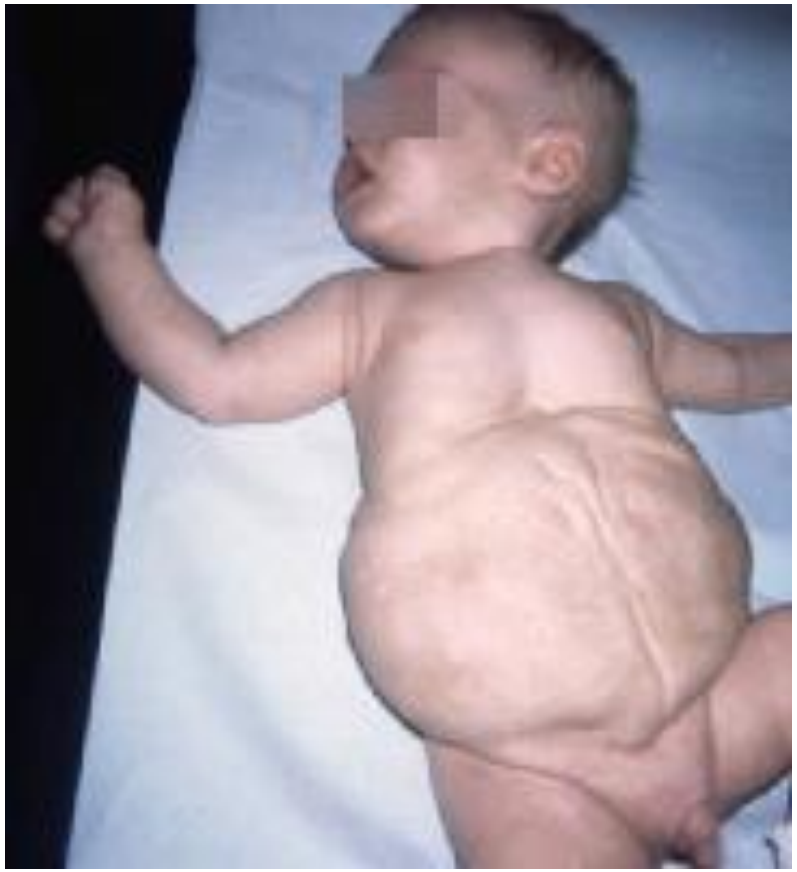
UMBILICAL HERNIA

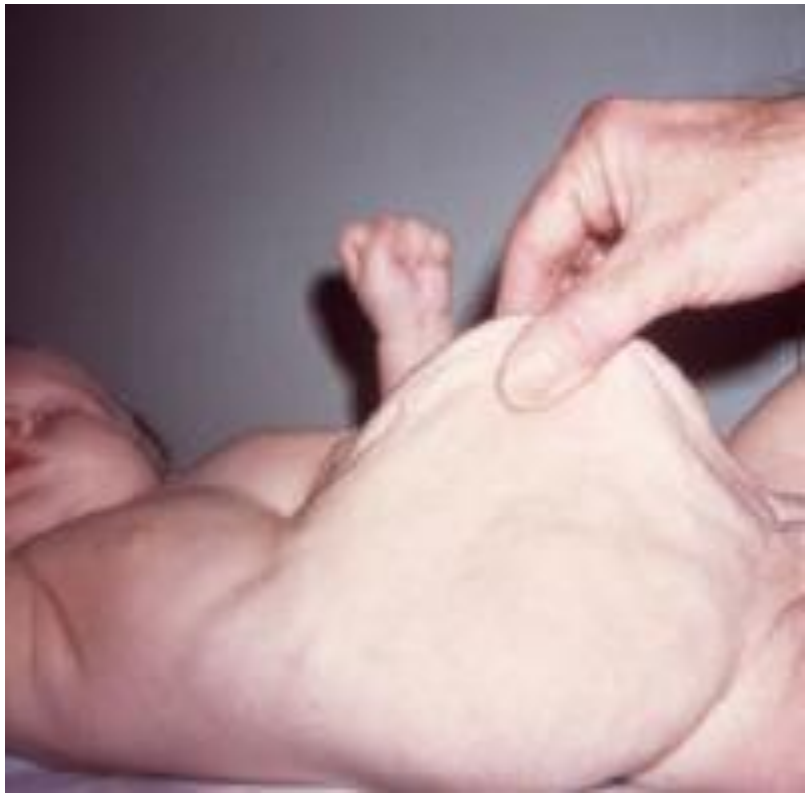
- Defect in linea alba, subcutaneous tissue and skin covering the protruding bowel
- Frequent in premature infants



PRUNE BELLY SYNDROME

- Thin, flaccid abdominal wall
- Dilation of bladder, ureter & renal collecting system
- 1:30,000-50,000
- 95% are male





5. PEDIATRIC SURGICAL JAUNDICE 3RD/5/2019

BY: DR. KURIA

TYPED BY NAILA KAMADI

DEFINITION

- Jaundice: yellowish discoloration of mucosal membranes with elevated bilirubin levels >50 μmol/L.
- Normal values: 3 – 20 μmol/L
  - Normal Conjugated bilirubin levels: < 5 μmol/L (<20% of total).
- Can be classified as:
  - Medical: can be treated by medical means
  - Surgical: can be treated surgically

CONT.

- This is a physical finding that is examined in the following areas:
  - Upper part of the sclera
  - Buccal mucosa
  - Palms
  - Skin (in light individuals)

BILIRUBIN METABOLISM

- Senescence of RBCs → spleen sequestration & destruction by splenic macrophages → hemoglobin
- Breakdown of hemoglobin → heme & globin
- Globin → amino acid pool
- Heme oxygenase breaks down heme to biliverdin
- Biliverdin reductase breaks down biliverdin to bilirubin

CONT.

- Bilirubin – albumin complex is taken up by the hepatocyte actively
- Bilirubin glucuronidase carries out glucuronidation.
- Bile is then secreted into the bile canaliculi → hepatic duct → CBD → intestines via ampulla of vater → bacterial degradation in intestines into urobilinogen & stercobilinogen → EHC of some of the urobilinogen (@ terminal ileum) & excretion of the rest.

UNCONJUGATED HYPERBILIRUBINEMIA

- Physiologic (2 – 10 days)
  - Immature liver enzymes
  - Increased destruction of red blood cells due to a shortened life span
- ABO & Rh incompatibility
- Breast milk jaundice
- Congenital enzyme defects

CONJUGATED HYPERBILIRUBINEMIA

Medical causes of conjugated hyperbilirubinemia

- ***Genetic/ metabolic***: A1 antitrypsin deficiency, Tyrosinemia, galactosemia, hypothyroidism, Cystic Fibrosis (CF), Pan hypopituitarism, Gaucher's disease, iron storage diseases
- ***Hepatocellular***: Giant cell hepatitis, Idiopathic, CMV, HIV
- Infections: TORCHES, UTI, sepsis, syphilis
- ***Toxic***: parenteral nutrition
- ***Alagille's syndrome***: biliary hypoplasia, abnormal vertebrae, pulmonary stenosis, odd facies, JAGGED 1 mutation

CONT.

Surgical causes:

- MC: Biliary atresia, Choledochal cyst
- Spontaneous bile perforation
- Inspissated bile syndrome (e.g. in CF)
- Others:

CLINCAL PRESENTATION

- Icterus
- Acholic stools (pale, white, chalky)
- Hepatomegaly
- Bleeding: due to hypoprothrombinemia
  - Always administer IM vitamin K
- Pruritus is not pronounced in infants.
- Dark urine: may or may not be there
- Contrast with medical jaundice which presents from birth & the infants are usually LBW.

BILIARY ATRESIA & SPLENIC MALFORMATION (BASM)

- ***C***ardiac anomalies
- ***H***eterotaxy/ dextrocardia & situs inversus
- ***A***bsent vena cava
- ***M***alrotation
- ***P***re – duodenal portal vein
- ***P***olysplenia/ asplenia

INVESTIGATION OF SURGICAL JAUNDICE

- Laboratory tests:
  - LFTS: direct/ conjugated bilirubin >20% is indicative of surgical/ cholestatic/ obstructive jaundice
  - α – I antitrypsin levels: MS2 alleles
  - Liver biopsy
- Radiological:
  - USS
  - ***HIDA***, IDA, DISIDA: ^{99m}Tc
  - Cholangiogram: done intra – operatively in children
  - ERCP
  - MRCP

1. BILIARY ATRESIA

- Neonatal disease characterized by ***inflammatory & sclerotic obliteration*** of part or all of extra – hepatic biliary tree with varied involvement of intra – hepatic biliary ducts.

INCIDENCE

- 1:15000 in UK; 1:9000 in Japan
- Etiology:
  - Unknown, mostly
  - Perinatal viral infection: ***reovirus type III*** (infections that happen in – utero or shortly after birth), rotavirus, CMV
  - Aberrant, autoimmune response to infected cells.
  - Aberrant early bile development
  - Abnormal bile acids
  - Leakage of pancreatic juices

TYPES

- **Type I BA**: CBD has disappeared: 5%
- **Type II (cystic BA)**: common hepatic ducts, CBD is patent
  - 3 – 5%
  - Better prognosis
  - Disordered intrahepatic ducts
- **Type III**: > 90% with porta hepatis; moderate prognosis: whole system is affected

CLINICAL PRESENTATION

- Normal baby at birth
- Progressive jaundice > 2 – 3 weeks
- Dark urine, Acholic stools
- Firm hepatomegaly
- FTT
  splenomegaly
- Bleeding varices: due to increased portal vein pressure after the development of 2^0 biliary cirrhosis
- Successful outcome depends on ***age at presentation/ diagnosis***.

LABORATORY TESTS

- Both direct (> 20%) & indirect bilirubin elevated.
- Elevated transaminases
- Markedly elevated ALP, GGT
- TORCH screen
- Inborn error screen: Guthrie scan (CF, A1AT deficiency)
- Urinalysis: urobilinogen will be decreased due to decreased EHC

RADIOLOGY

- USS
  - Small or absent GB
  - Extrahepatic ducts not visualized
  - Cone shaped fibrotic portal plate
  - BASM
- Intraoperative cholangiogram
- HIDA

DIFFERENTIALS

- Choledochal malformation
- Spontaneous bile duct perforation
- Inspissated bile syndrome
- Liver hemangioendothelioma/ harmatoma compressing the duct

TREATMENT

- Kasai Porto – enterostomy: better prognosis when done in ≤ 60 days.
  - Excision of sclerosed biliary tree tissue up until the porta hepatis.
  - Forma Roux – En Y loop where part of intestine is anastomosed with the proximal stump of the biliary tree.
- Liver transplantation

II. CHOLEDOCHAL MALFORMATION ("CYST")

- Term refers to a spectrum of congenital biliary tract disorder previously grouped under the name ***idiopathic dilatation of the CBD.***
- Described by Abraham Vater (German anatomist) in 1723.
- F:M → 4:1.
- High incidence in Japan/ China
- Can be: prenatal or up to 4 yrs. post – natal.
- It is ***not a true cyst*** with a cover/ capsule & free fluid inside.

TODANI MODIFICATION OF ALONSO – LEJ CLASSIFICATION OF CHOLEDOCHAL CYSTS

Type	Imaging description		Incidence (%)
I	Dilatation of ***extra – hepatic bile duct*** only	IA: cystic	80 – 90
		IB: Saccular	
		IC: Fusiform	
II	***Diverticulum*** protruding from the extrahepatic CBD		2
III	Choledococele involving the ***intra duodenal portion of the hepatic CBD*** (near the ampulla of Vater)		4 – 5
IV	***Multiple*** cystic dilatation	IVA: cysts involving the intra & extra – hepatic ducts.	10 (IVA > IVB)
		IVB: cysts involving only the extrahepatic duct.	
V (Caroli's disease)	***Multiple dilatations limited to the intra – hepatic bile duct***		Rare

CLASSIFICATION OF CHOLEDOCHAL CYSTS -

Mnemonic : ONE , 2D , 3D , Fourm , FIVE
= medicowesome
2014

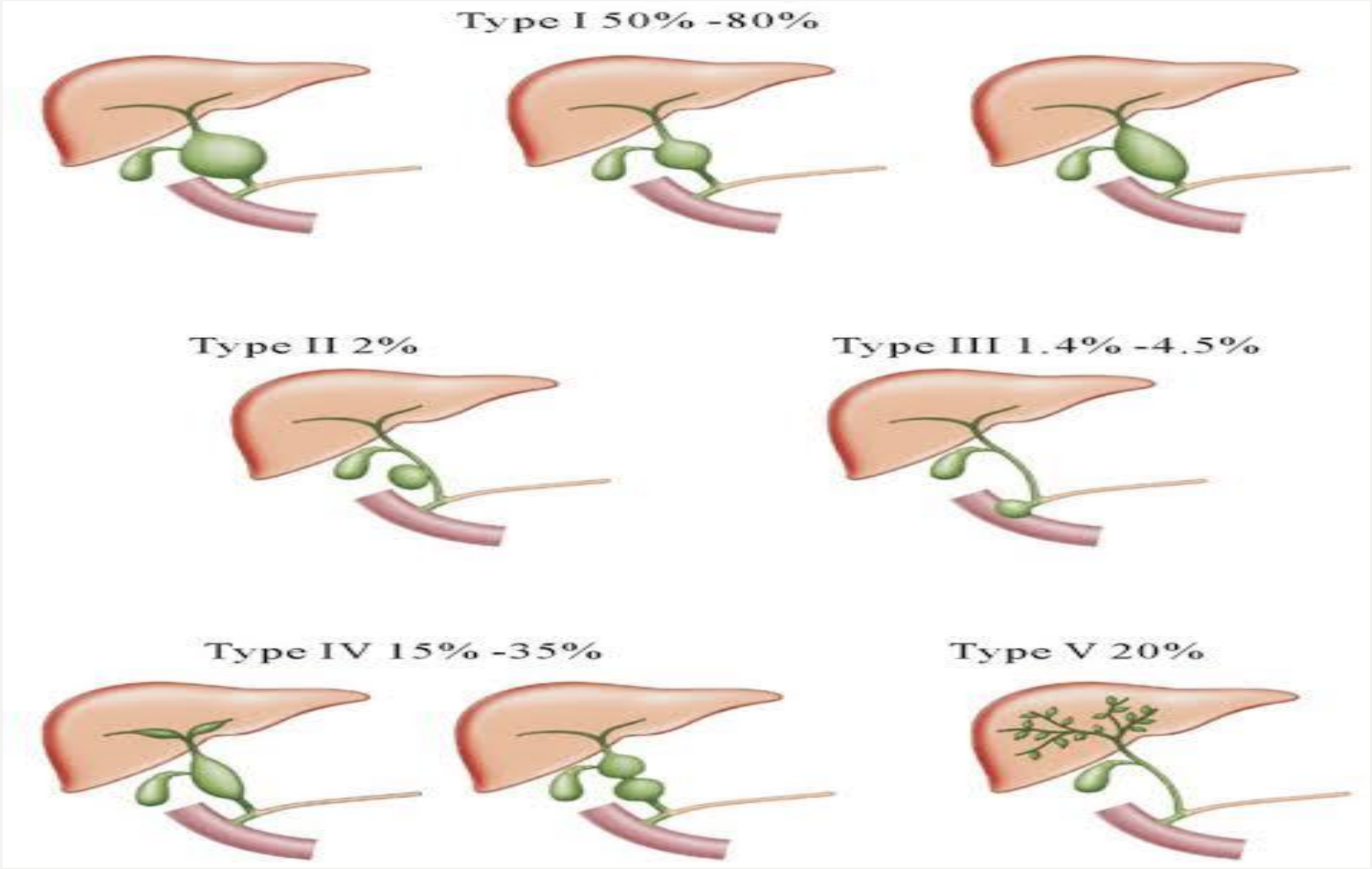
Type one E : Extrahepatic
Subtypes : CSF
IA : Cystic
IB : Saccular
IC : Fusiform

Type Two : Diverticuli (2D)

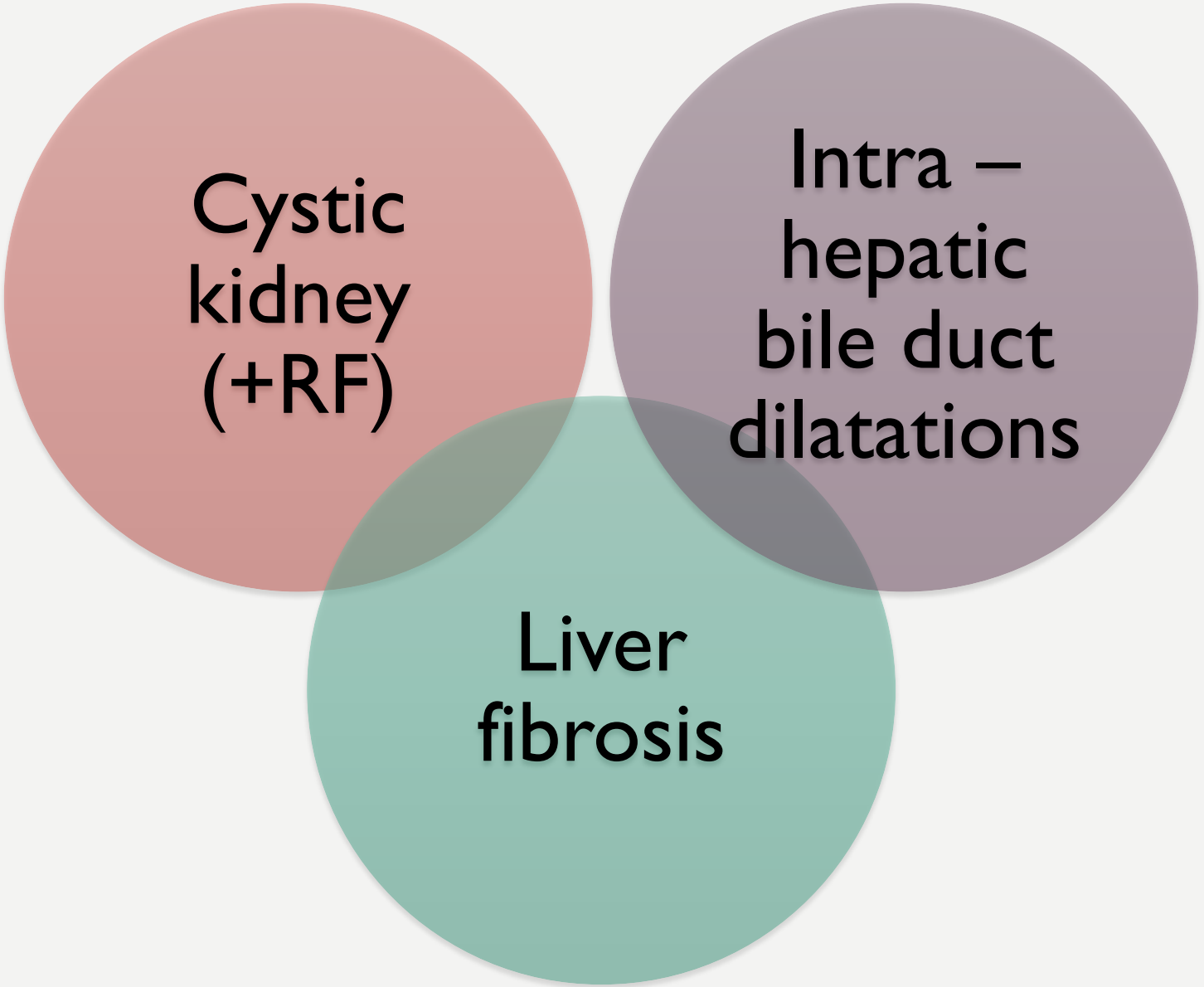
Type Three : Distal CBD (Duodenal) (3D)

Type Forur : Multipe (Forum?)
IV A : Extra And Intra
IV B : Extra only

Type FIVE : Intrahepatic


Type I 50% -80%
Type II 2%
Type III 1.4% -4.5%
Type IV 15% -35%
Type V 20%

CAROLI'S DISEASE



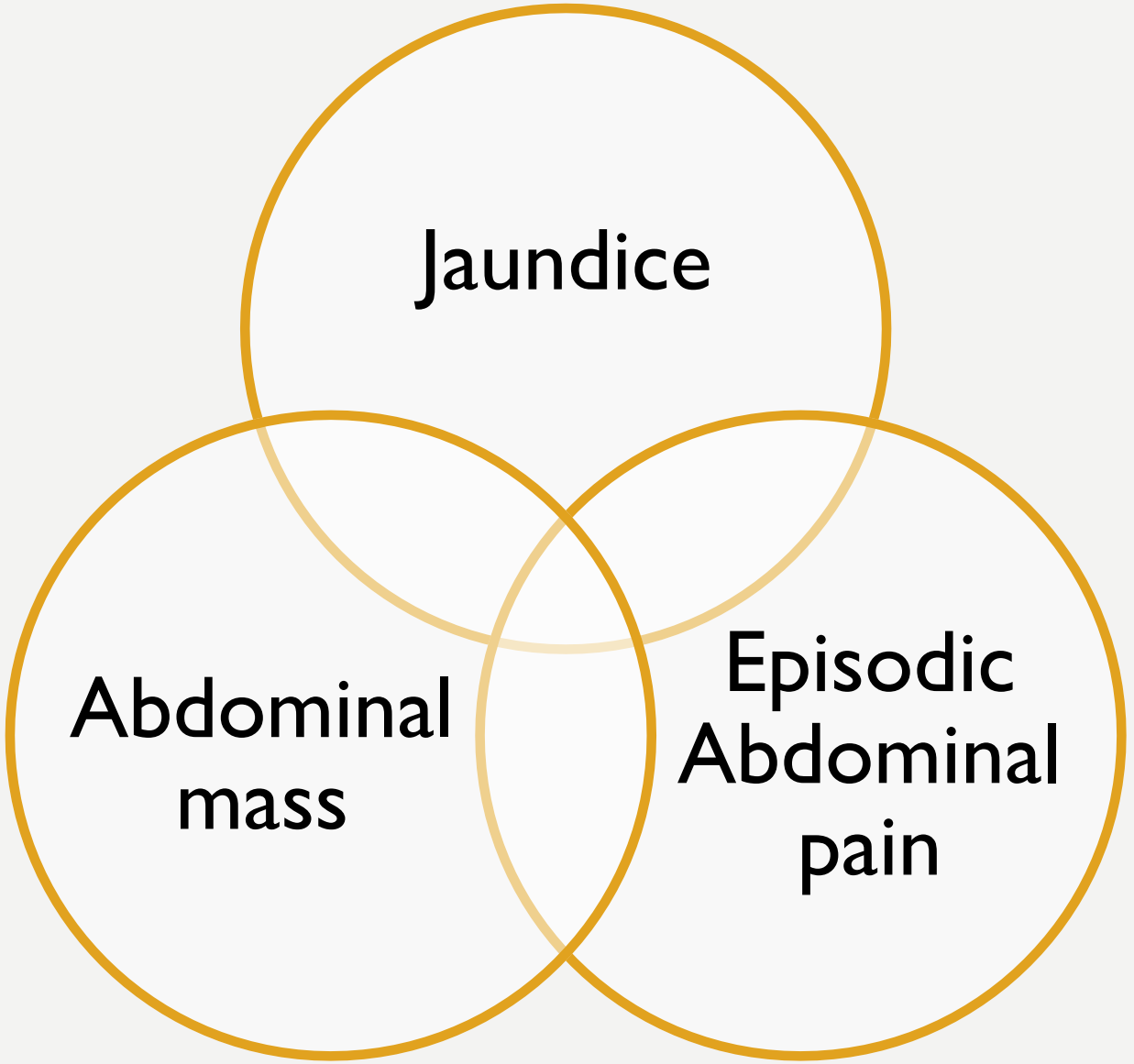
CONT.

- Caroli's disease is a hereditary disorder that carries a poor prognosis from ***recurrent cholangitis*** & the development of ***intrahepatic stones, liver abscess formation*** & eventually ***cirrhosis.***

ETIOLOGY

- Frequently (90%) associated with Choledochal cyst is an anomalous junction between the pancreatic & CBD.
- Babbit proposed an abnormal pancreatic & biliary duct junction → common channel → pancreatic enzymes are secreted & they reflux into the bile duct → weakening of bile duct wall due to gradual enzymatic destruction → dilatation, inflammation & cyst formation. There is a risk for malignancy.
  - However, not all patients with Choledochal cysts demonstrate an anatomic common channel.

TRIAD OF CHOLEDOCHAL CYST



CLINICAL PRESENTATION

- Triad only in <50%
- Commonly presents with episodic abdominal pain with minimal jaundice (can escape detection)
- If left undiagnosed patients may develop:
  - Cholangitis: may lead to development of cirrhosis & portal HTN
  - Pancreatitis
- Neonatal period: presents like biliary atresia with an abdominal mass.

INVESTIGATIONS

- Prenatal: screening U/S
- Additional to investigations of obstructive jaundice, perform:
  - Abdominal U/S: cystic structure from biliary tree.
  - Abdominal CT scan is confirmatory
  - MRCP
  - ERCP

MANAGEMENT

- Surgical excision of the whole cyst (?biliary tree) then →
- Anastomose proximal bile duct to intestinal tract typically via a Roux – en Y jejunal limb.

PROGNOSIS

- Excellent
- Complications (that may develop long after surgery) include:
  - Anastomotic stricture
  - Cholangitis
  - Intrahepatic stone formation

REFERENCE

- Schwartz Principles of Surgery, 10ED (Page 1630)

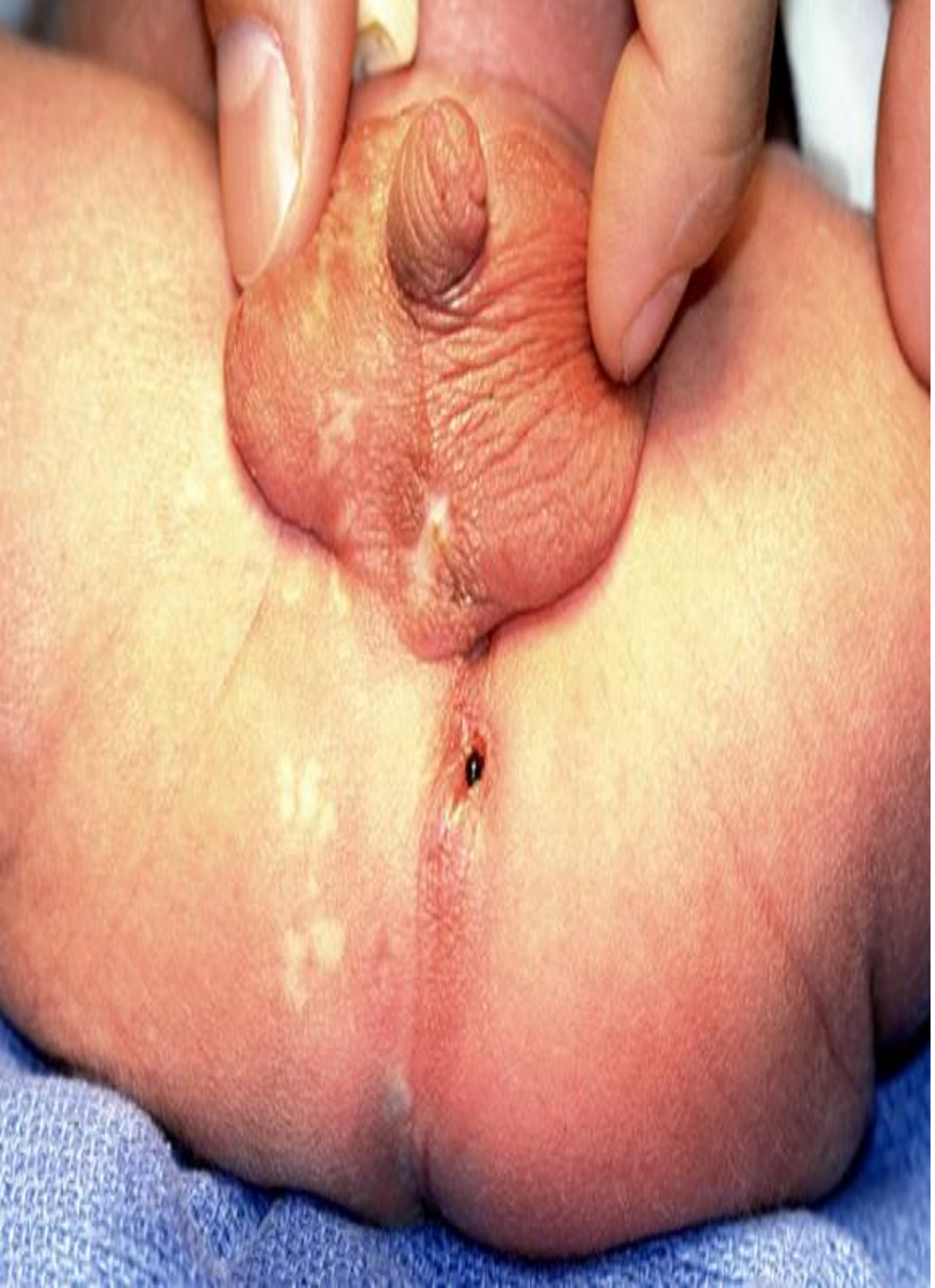
TYPED BY NAILA KAMADI

There is no need to be perfect to inspire others.

Leave that to Jesus Christ.

Let people be inspired by ***how you deal with your imperfections***.

#Jesus_Is_The_Light



6. ANO – RECTAL MALFORMATION

BY: DR. MWIKA M. P.

CONSULTANT

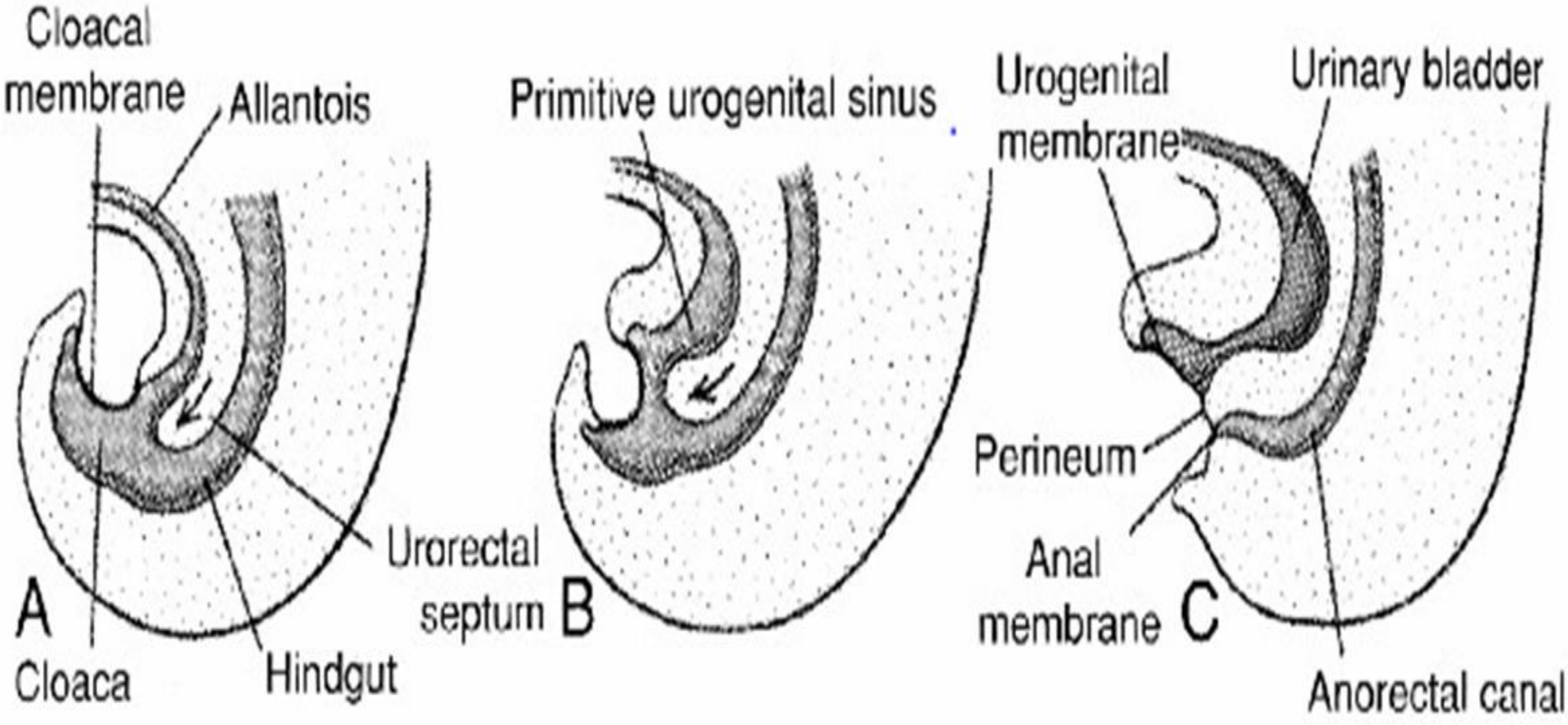
PEDIATRIC SURGEON

EDITED BY NAILA KAMADI

Definition

- ✓ ARM: these include a wide spectrum of defects in the development of the lowest portion of the intestinal & urogenital tracts.
- ✓ The term imperforate anus may accurately describe a child's outward appearance but it often belies the true complexity of the malformation beneath.

Embryology of the ano – rectum



Cont.

✓ Development of the ano – rectum is complete by the ***9th week*** of intrauterine life & consists of:

1. ***Cloaca*** formation.
2. Partitioning of the cloaca by the ***urorectal septum*** into the urogenital sinus anteriorly & anorectal canal posteriorly.
3. Fusion of the ***urorectal septum with the cloacal membrane*** at the future site of the gross anatomical ***perineal body***

Cont.

- ✓ The lower anal canal develops from the ***proctodeum*** which is an invagination of surface ectoderm by a proliferation of mesoderm surrounding the anal membrane.
- ✓ The junction between the upper & lower canals is indicated by the ***pectinate line*** which marks the site of the former ***anal membrane***.
- ✓ In the 9^{th} week the anal membrane ruptures opening the canal to the outside.

Cont.

- ✓ Abnormality at any point in the above process leads to the various ARMs.
  - ✓ The embryologic basis is failure of descent of the urorectal septum. The level to which this septum descends determines the type of anomaly that is present, which subsequently influences the surgical approach.
- ✓ The external anal sphincter derived from ***external mesoderm*** is usually present but has varying degrees of formation ranging from robust musculature to minimal.

Cont.

- ✓ In patients with imperforate anus, the rectum fails to descend through the external sphincter complex & it ends ‘blindly’ in the pelvis, above or below the levator ani muscle.
- ✓ In most cases, the blind rectal pouch communicates more distally with the genitourinary system or with the perineum through a fistulous tract.

Incidence

- ✓ 1/ 5000 live births.
- ✓ A ***slight male preponderance*** exists: 55% to 70% in favor of boys.
- ✓ ***Familial (genetic) predisposition*** exists ARMs occurring in succeeding generations.

Cont.

- ✓ ***95% of ARM pathology have associated fistula***. Close to 50% of those with no fistula have ***Down syndrome***.
- ✓ The MC defect in females is ***recto – vestibular fistula***, whereas in males is ***recto – urethral fistula***.

Genetics in ARM

Common chromosomal anomalies	Association with ARM in comparison with neonates in the general population
Down Syndrome	15 times higher
Trisomy 18	90 times higher
Trisomy 13	30 times higher

Common genetic syndromes include:

- ✓ Currarino syndrome
- ✓ VACTERL association
- ✓ Pallister – Hall syndrome
- ✓ Sirenomelia
- ✓ Caudal regression syndrome

Associated anomalies in order of decreasing frequency

- ✓ Urogenital anomalies → 45%
- ✓ Skeletal anomalies → 30%
- ✓ GIT → 20%
- ✓ CVS → 15%
- ✓ Chromosomal conditions e.g. Down's syndrome → 10%
- ✓ CNS → 5%
- ✓ Others → 15%

Cont.

SYSTEM	TYPE OF ANOMALY	FREQUENCY
Urinary	Vesico-ureteric reflux Hydronephrosis Renal agenesis Renal dysplasia	50%
Genital	Vaginal septum	50%
	Uterine didelphys/Bicornuate uterus	35%
	Cryptorchidism	3-19%
	Vaginal duplication/vaginal agenesis/absent ovary	
Vertebral [15]	Lumbosacral anomalies Tethered cord Cord lipomas Syringohydromyelia	30-35%

Cont.

SYSTEM	TYPE OF ANOMALY	FREQUENCY
Cardiovascular	VSD Tetralogy of Fallot Transposition of great vessels Hypoplastic left heart syndrome	12-22%
Gastrointestinal [16]	Tracheo-esophageal fistula	10%
	Duodenal obstruction	
	Malrotation	
	Hirschsprung's disease	
Curarino triad [17,18]	- Sacral defect + presacral mass + imperforate anus	> 350 cases reported in literature
Other anomalies	As listed in Pena's classification	Rare

VACTERL Anomalies

- ✓ Vertebral anomalies: ***sacral agenesis*** is the MC
- ✓ ARM
- ✓ Cardiac anomalies: ***TOF*** (MC), ***ASD, VSD.***
- ✓ Tracheoesophageal malformations
- ✓ Renal (Genito – urinary) malformations: ***renal agenesis*** is the MC; others include hypospadias etc.
- ✓ Limb malformations: ***radial anomalies*** are the MC

Genitourinary anomalies

- ✓ Incidence:
  - ✓ 25 – 60% of ARM
  - ✓ The higher the ARM defect, the greater the incidence
  - ✓ < 10% incidence in low ARM
- ✓ The common anomalies include: ***renal agenesis, VUR, mega – ureter, ectopic ureter or ureterocele.***

Sacral & Vertebral anomalies

- ✓ The ***sacrum is the most frequently affected bony structure***. Assessment of the hypo development of the sacrum correlates with the patient's future prognosis.
- ✓ Rough assessment of post – op continence can be done on the basis of number of sacral vertebra:
  - ✓ > 3 sacral vertebrae → continence expected
  - ✓ 3 sacral vertebrae → intermediate continence
  - ✓ < 3 sacral vertebrae → incontinence expected

Cardiovascular anomalies

- ✓ Present in approximately 1/3 of patients, but only 10% of these require treatment.
- ✓ The MC: **ASD** & **PDA**, followed by **TOF** & **VSD**

GI anomalies

- ✓ Tracheoesophageal abnormalities occur in about 10% of cases. ***Esophageal atresia is the MC.***
- ✓ Duodenal obstruction caused by atresia or malrotation has been reported to have an incidence of 1% to 2%.

Classification

- ✓ Can be classified as:
  - ✓ **High type**: rectum ends above the levator ani muscle.
  - ✓ **Low type**: rectum partially descends through the levator ani muscle.

Pena classification based on the type of fistula present

- ✓ Proposed by Pena in 1995.
- ✓ He distinguished between:
  - ✓ *Perineal*, *vestibular*, *bulbar*, *prostatic* & *bladder neck* fistulas
  - ✓ Imperforate anus without fistula
  - ✓ Vaginal fistulas
  - ✓ Cloacal fistulas
  - ✓ Rectal atresia or stenosis.

Pena classification

<table>
<tr><th colspan="2">Female ARMs</th><th colspan="2">Male ARMs</th></tr>
<tr><td colspan="2">Cutaneous perineal fistula (low type)</td><td colspan="2">Cutaneous perineal fistula (low type)</td></tr>
<tr><td colspan="2" rowspan="2">Vestibular fistula (MC)</td><td rowspan="2">Recto – urethral fistula (high type; MC)</td><td>Bulbar</td></tr>
<tr><td>Prostatic</td></tr>
<tr><td rowspan="2">Persistent cloaca (most severe; high type)</td><td>< 3cm common channel</td><td colspan="2" rowspan="2">Recto – vesical fistula (most severe): fistula communicates with the bladder neck</td></tr>
<tr><td>> 3cm common channel</td></tr>
<tr><td colspan="4">Imperforate anus without fistula</td></tr>
<tr><td colspan="4">Rectal atresia or severe stenosis</td></tr>
</table>

Cont.

- ✓ In males:
  - ✓ The most frequent defect ***is imperforate anus with recto – urethral fistula > recto – perineal fistula > recto – vesical fistula***.
- ✓ In females:
  - ✓ The most frequent defect is ***recto – vestibular fistula > cutaneous perineal fistula > persistent cloaca*** (wide spectrum of malformations where rectum, vagina and urinary tract meet and fuse into a single common channel/ typically, external genitalia is hypoplastic)

Clinical features at history taking

- ✓ Clinical features depend on the type of ARM. Those with ***no fistula present within 24h. with distention & a history of failure to pass meconium***. Those ***with fistula have a delayed presentation*** that can be discovered at weaning or by the mother while bathing the infant.
- ✓ In general boys with ARMs present with ***IO*** (*abdominal distention, failure to pass meconium, vomiting*) in the newborn period. Girls present with a h/o ***passing stool from an abnormal opening***.
- ✓ Patients with imperforate anus are usually stable and the diagnosis is readily apparent. Despite the obstruction, the abdomen is initially not distended & there is rarely any urgency to intervene.

Cont.

- ***Muconuria*** ***(passage of meconium in urine)*** may be present in recto – vesical/ recto – bulbar fistula.
- ***Abdominal distention*** & ***non – passage of stool per anus***.
- ***Constipation*** in older patients as *solidification of stool occurs by 3 – 4 months*.

General examination

- ✓ Screening for associated anomalies (*full body exam*)
  - ✓ Examination of abdomen, spine
  - ✓ Passage of rigid NG tube: R/O TEF
  - ✓ Central cyanosis: points towards a cardiac condition
  - ✓ Limb anomalies
- ✓ Examination of genitalia

Examination of perineum

- ✓ Check all orifices (anus, vagina, urethra)
- ✓ Meconium from the perineum: points towards a fistula
- ✓ Associated anomalies
- ✓ Look for an opening at the tip of the urethra & check for meconium
  - ✓ If present → severe ARM

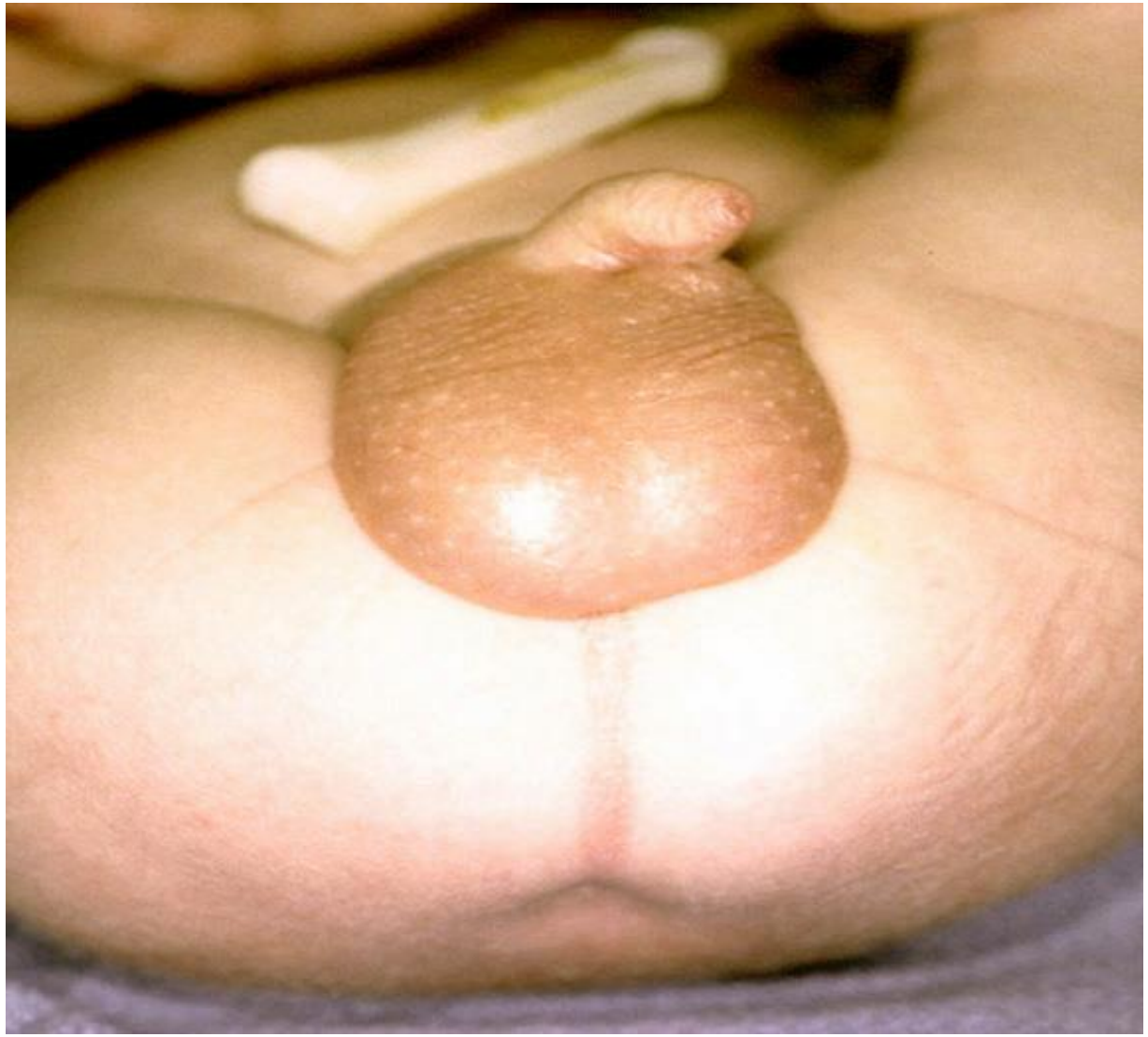
Examination of buttocks

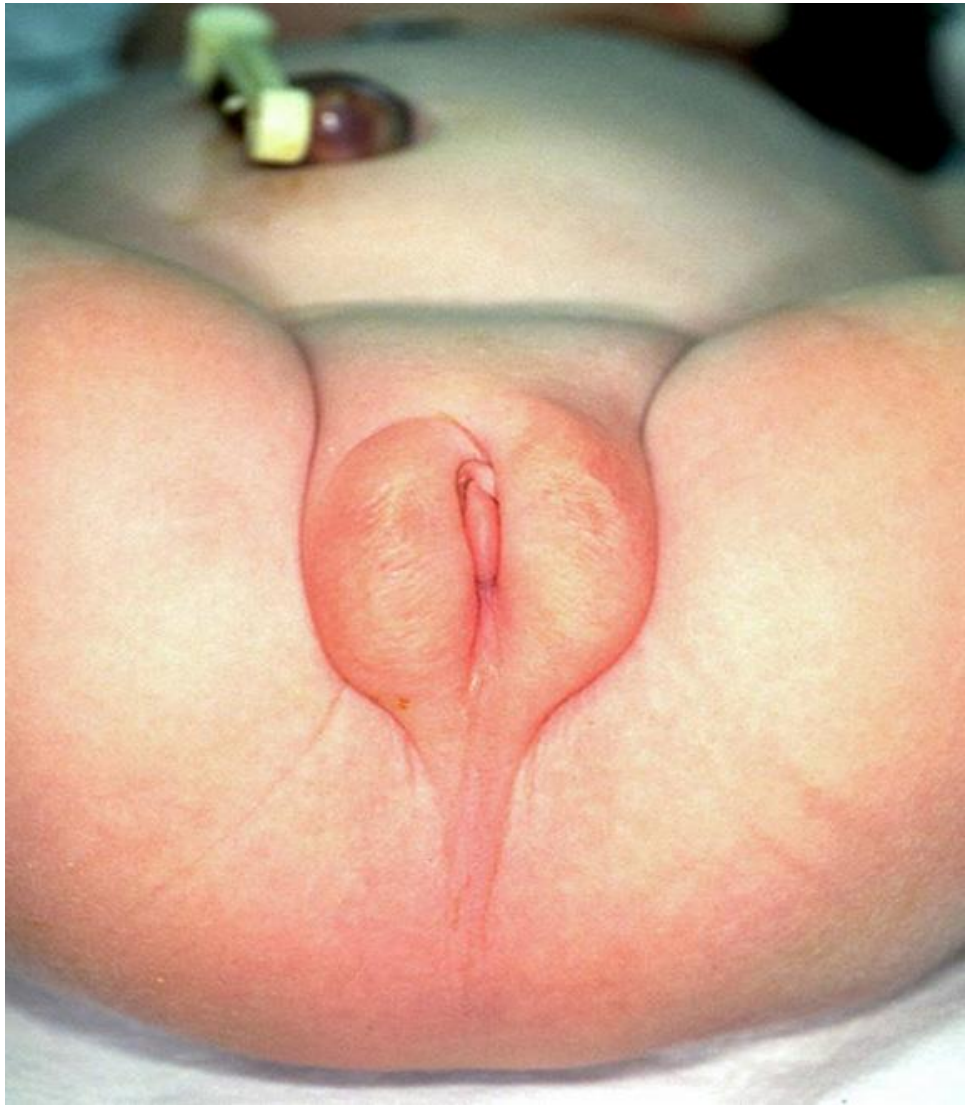
- ✓ High anomalies: sacro – coccygeal agenesis, flat buttock
  - ✓ *Flat bottom is a poor prognostic factor.*
- ✓ Low/ intermediate anomalies: pigmentation, sacral dimple
- ✓ Examine where the anus should be:
  - ✓ Bucket handle deformity: mild ARM, Rx is straight forward
  - ✓ Stenotic opening
  - ✓ Covered anus/ fistulous ano – cutaneous tract

Findings associated with a high malformation

- ✓ Flat perineum: the lack of a midline & inferior gluteal fold.
- ✓ Absence of anal dimple indicates the patient has poor muscles in the perineum.

Cont.

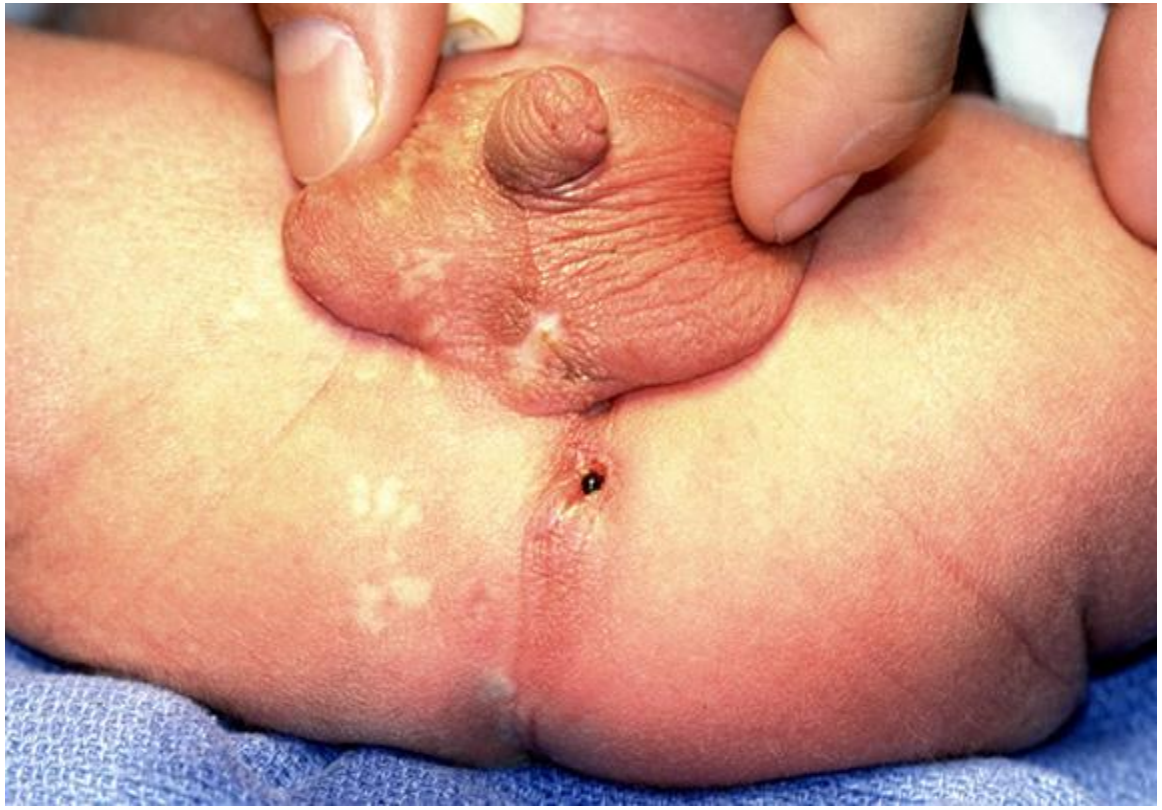


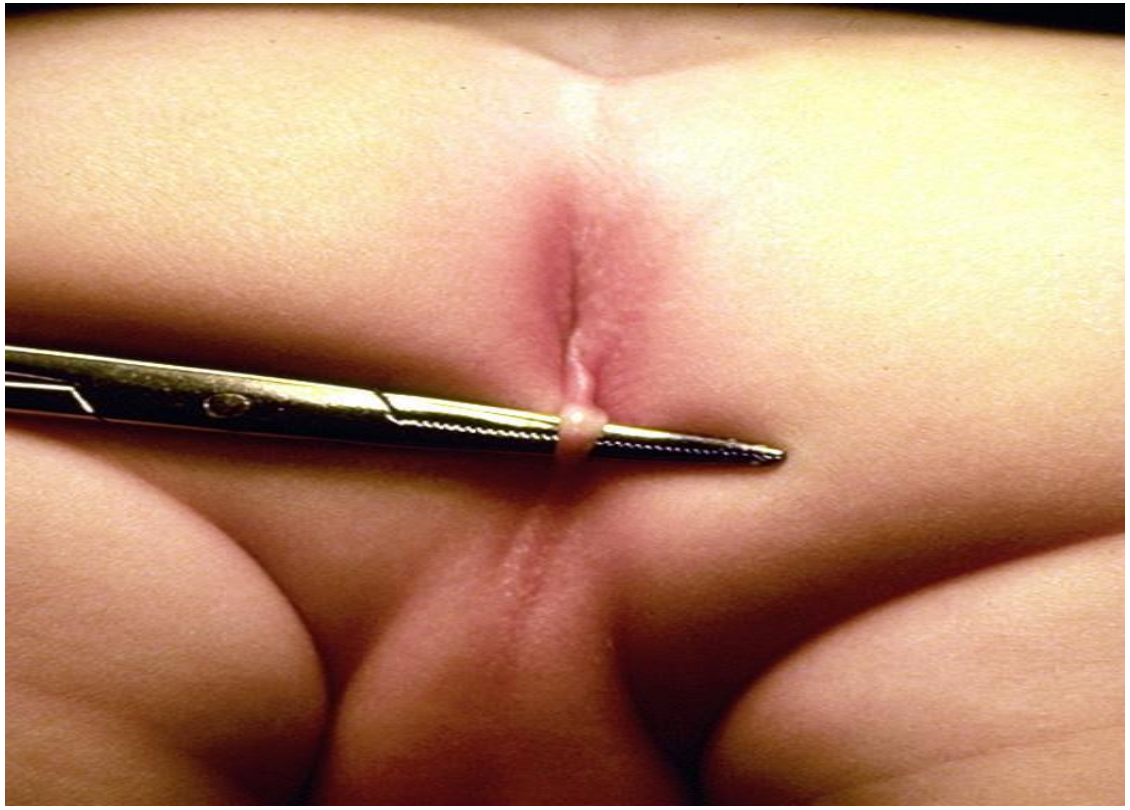


Findings associated with a low malformation

- ✓ Presence of meconium at the perineum
- ✓ Bucket – handle malformation
- ✓ Anal membrane through which meconium is visible

Cont.





Investigations

- ✓ To determine the malformation
  - ✓ Invertograms (not done anymore)
  - ✓ Cross – table lateral plain abdominal radiograph with a radio – opaque marker on the perineum: can give the distance from where the anus is ending & where the rectum is starting
- ✓ ***Abdominal U/S + KUB***: genitourinary system (hydronephrosis, hydrocolpos, pre – sacral mass, abdominal mass)

Cont.

- ✓ USG trans – perineal
- ✓ Spinal U/S or MRI: spinal anomalies (tethered cord, spinal lipoma)
- ✓ MCUG, IVP
- ✓ CT scan, MRI: to know where the muscles are
- ✓ X ray of lumbosacral spine: ***sacral ratios, sacral spine defects (Hemi – vertebrae, pre – sacral masses)***

Anatomy of malformation

- ✓ ***Distal pressure colostogram***: Done after colostomy to demonstrate presence of recto urinary fistula & to clarify the anatomy (reducing risk of injury to other structures; posterior urethra, seminal vesicles, vas deferens, and ectopic ureters).
- ✓ CT scan, MRI: to know where the muscles are

Cont.

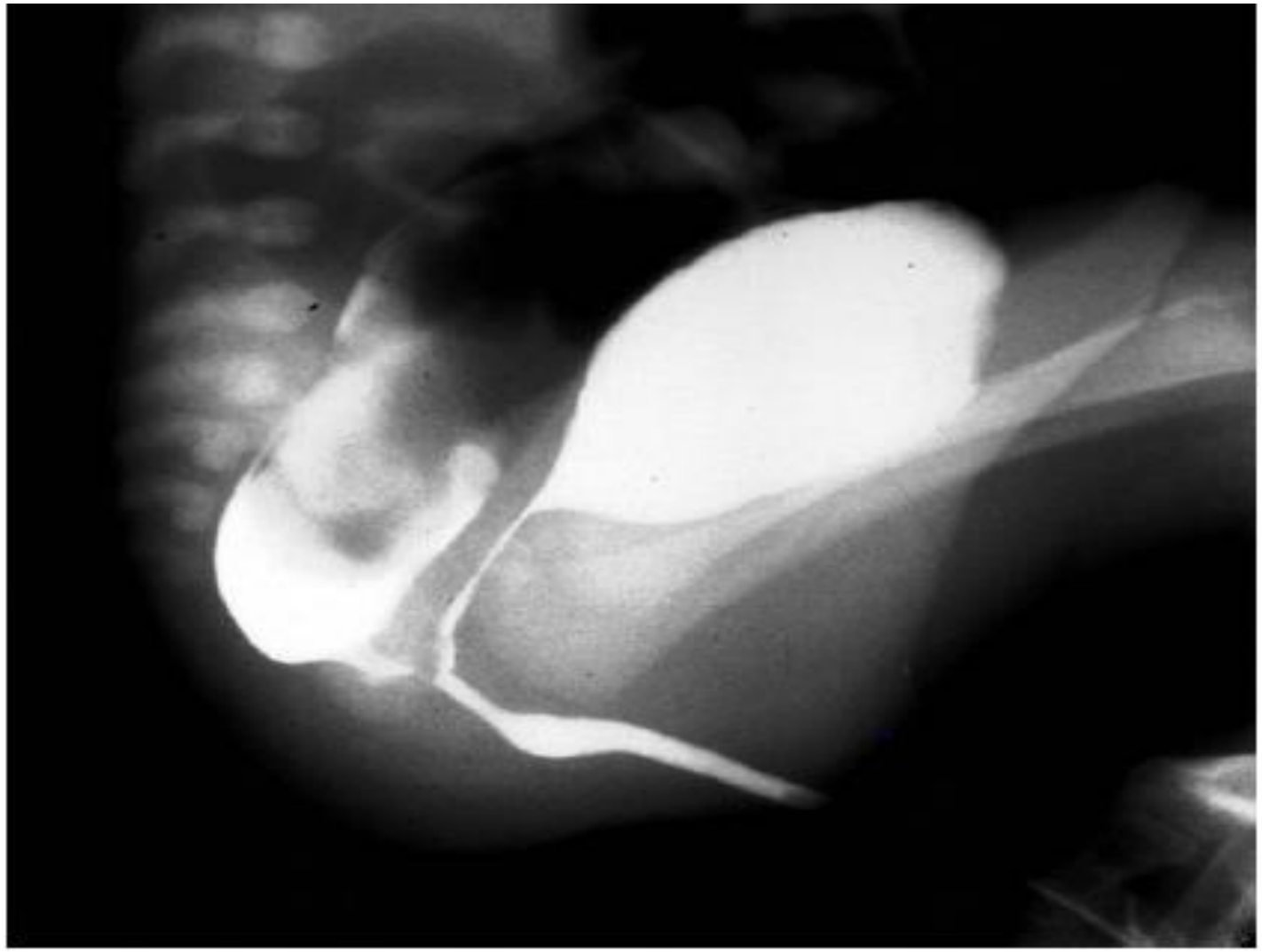


FIGURE 103-9 Distal colostogram showing a rectourethral bulbar fistula.

Approach to management

- ✓ Initial assessment steps are:
  - ✓ Determining in the newborn period whether the infant will ***require a colostomy or a primary Anoplasty procedure.***
  - ✓ Documenting any associated anomalies that may affect survival.

Cont.

- ✓ During the 1st 24 hrs. the baby should receive ***IVFs (5% Dextrose)***, ***ABs, NGT decompression*** to prevent aspiration.
- ✓ Monitor for the appearance of meconium in & around the perineum or in the urine (muconuria)
- ✓ The clinician should use these hours to R/O the presence of associated defects such as vertebral anomalies, cardiac malformations, TEF, renal anomalies & limb defects (VACTERL).
- ✓ If in a peripheral facility: refer for definitive care.

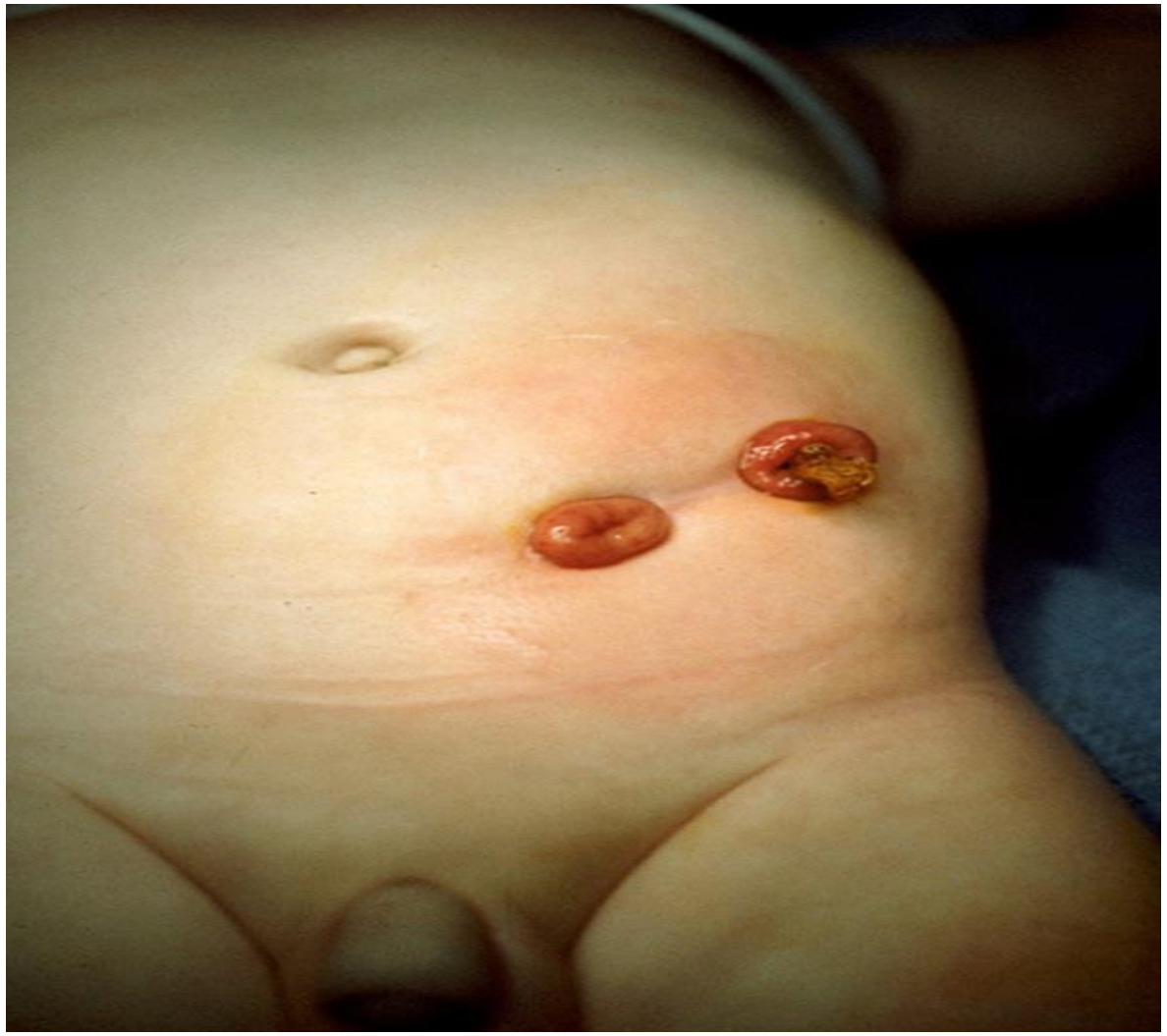
Search for other comorbidities

- ✓ R/O immediate life threatening conditions
  - ✓ Cardiac anomalies: echocardiography
  - ✓ Renal anomalies: U/S
  - ✓ Esophageal atresia: pass NGT

Neonatal colostomy

- ✓ In general, when a low lesion is present, only a perineal operation is required without a colostomy. ***Infants with a high lesion require colostomy in the newborn period***, followed by a ***pull – through procedure at approximately 2 months of age***.
- ✓ Persistent cloaca: evaluate urinary tract at time of colostomy formation to ensure normal emptying can occur and to determine whether the bladder needs drainage via Vesicostomy.
- ✓ ***If there is any doubt about the type of lesion, it is safer to perform a colostomy rather than jeopardize the infant's long term chances for continence by an injudicious perineal operation.***

Colostomy



- ✓ Neonatal colostomy is performed in children who are not amenable to primary pull through either because of malformation complexity or associated comorbidity.

Complex malformations requiring neonatal colostomy

- ✓ Males: any urinary fistula
- ✓ Females: Vestibular fistula, cloaca
- ✓ Either sex: no fistula > 1cm from perineal skin

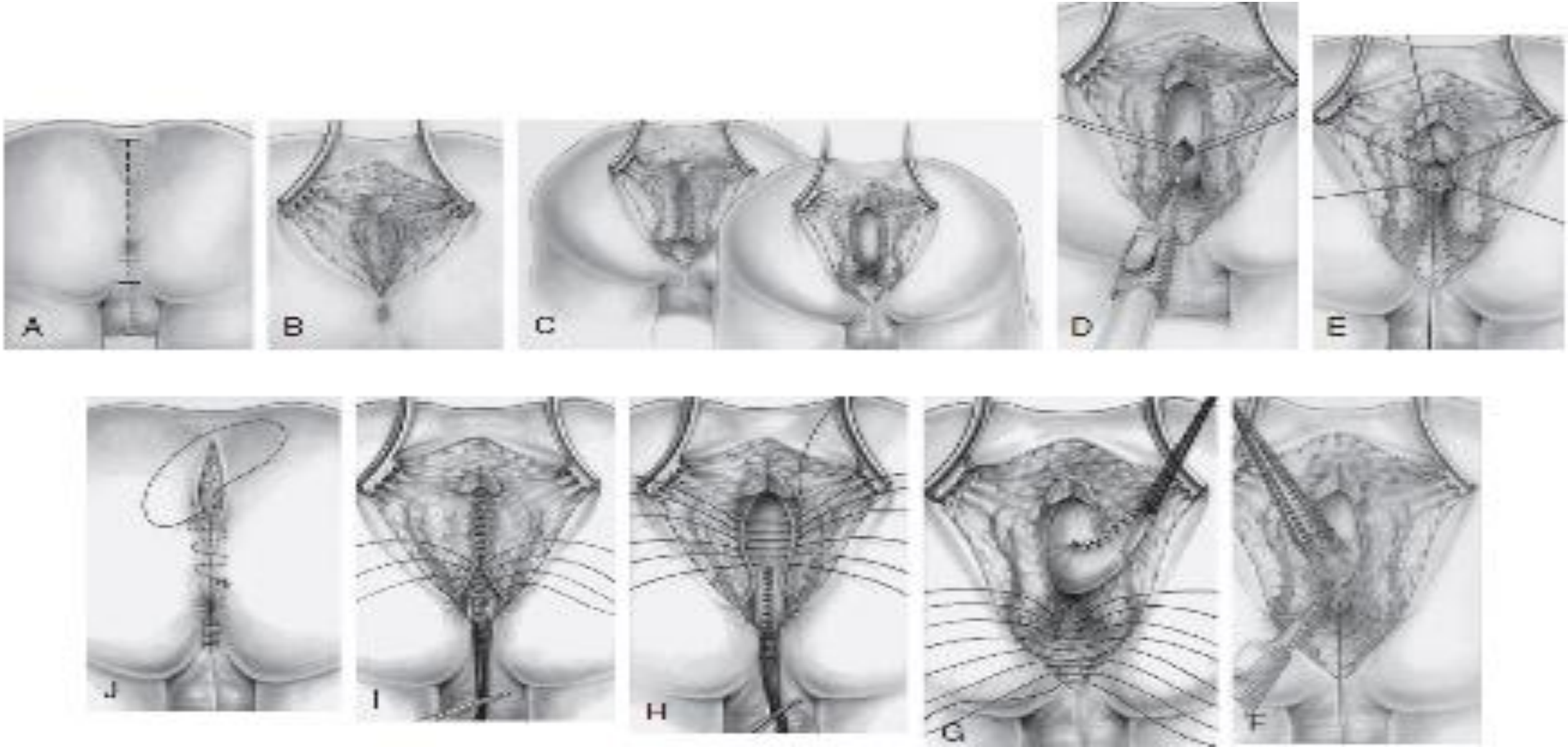
Intermediate and high anomalies

- ✓ Staged surgical procedure
  - ✓ Preliminary divided sigmoid colostomy (double barrel)
  - ✓ A pull through operation
  - ✓ Closure
- ✓ Management of patients with high lesions can be greatly facilitated using a laparoscopic – assisted approach.

Definitive pull through procedure

- ✓ Anoplasty
- ✓ Anterior Sagittal Ano – Rectoplasty (ASARP)
- ✓ ***Posterior Sagittal Ano – Rectoplasty (PSARP): most favored;*** Patient is placed in the prone jackknife position.
- ✓ PSARVP

ASARP or PSARP



Cont.

A. Planned posterior sagittal incision

B. Posterior sagittal approach with the parasagittal fibres & ischiorectal fat split in the midline.

C. Posterior rectal wall exposed.

D. Posterior rectal wall opened in the midline.

E. Posterior rectal wall opened going anteriorly until the recto – urethral fistula is identified.

Cont.

- ✓ F: Separation of the rectum from the posterior urethra with dissection above the fistula.
- ✓ G: The rectum fully mobilized and in this case tapered. Sutures are placed anteriorly to close the perineal body.
- ✓ H: The rectum pulled through and placed within the limits of the sphincter mechanism.
- ✓ I: Closure of the levator & tucking of the posterior edge of muscle complex to the posterior rectal wall.
- ✓ J: Closure of the posterior sagittal incision & completed anoplasty

Outcome

- ✓ 75% of patients were found to have voluntary bowel movements & nearly 40% were considered totally continent.
- ✓ Patients with high lesions demonstrate an increased incidence of incontinence whereas those with low lesions are more likely to be constipated.

Early complications

- ✓ Wound infection
- ✓ Wound dehiscence
- ✓ Retraction

Late complications

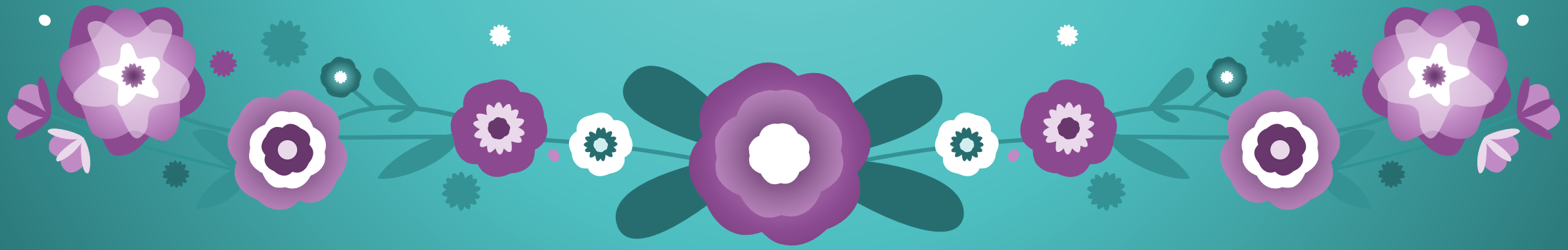
- ✓ Rectal strictures, vaginal strictures, incontinence, acquired atresias, recurrent fistulas, pelvic fibrosis
- ✓ Rectal mucosal prolapse: documented in up to 5% post – PSARP
- ✓ Constipation: the lower the malformation, the more likely. A vicious cycle ensues with mega – recto – sigmoid leading to more constipation which will lead to more mega – recto – sigmoid resulting in overflow pseudo – incontinence.

EDITED BY NAILA KAMADI

7. COMMON PEDIATRIC UROLOGICAL CONDITIONS

12TH/6/2019

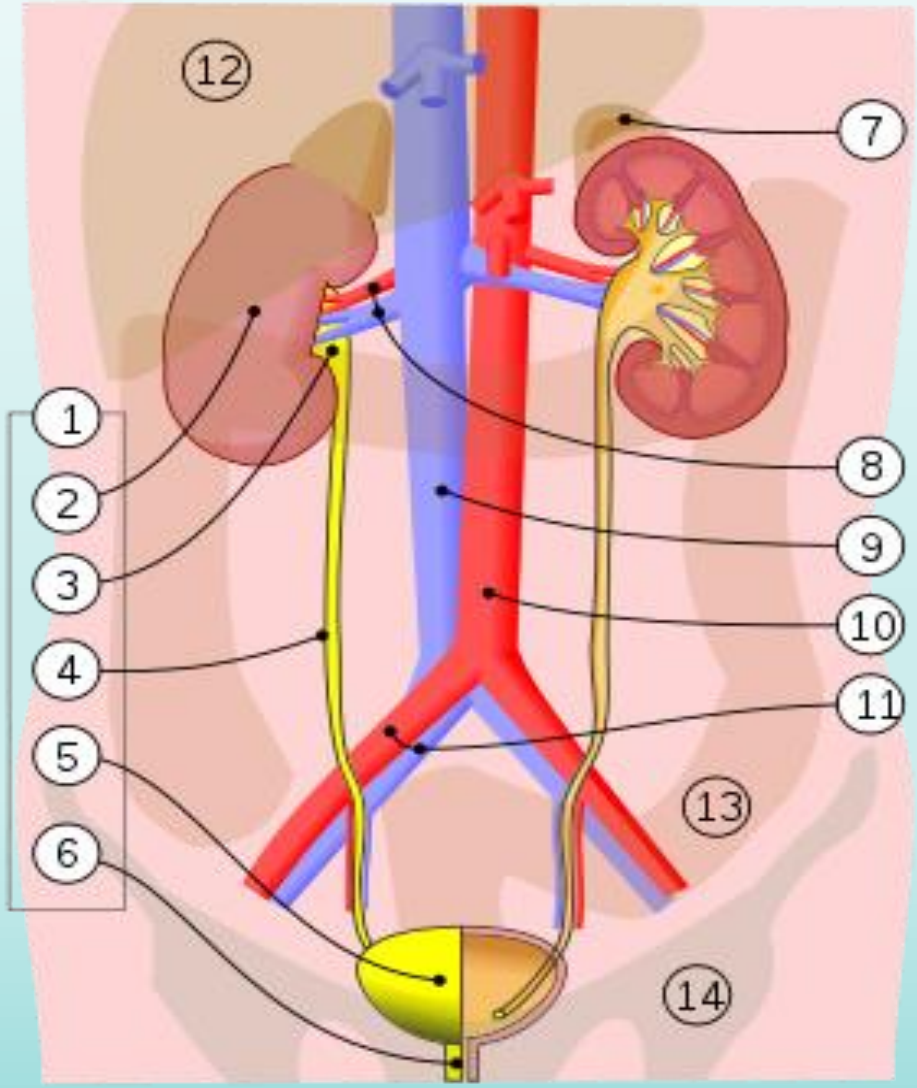
BY: DR. F. OSAWA



OBJECTIVES

- Define & describe the treatment of ***phimosis***, ***paraphimosis*** & ***balanitis.***
- Outline the investigation & management of ***a febrile pediatric UTI.***
- List the common causes of ***antenatal hydronephrosis*** & ***collecting system abnormalities***.
- Define ***cryptorchidism*** & ***hypospadias***.
- List the urologic causes of an ***abdominal mass*** in a child.

PEDIATRIC UROLOGY IS A SURGICAL SUBSPECIALTY DEALING WITH THE DISORDERS OF CHILDREN'S GENITOURINARY SYSTEMS



HISTORY

- Age
- Presenting complaint
- History
- Medications & allergies
- PMH_x: antenatal U/S

Cont.

- Elimination history:
  - Voiding frequency: overactive bladder, inflammation, very small bladder
  - Holding maneuvers
  - Incontinence (day & night)
  - Bowel movements (hard stool): the bladder neck in males is adjacent to the pouch of Douglas & a full rectum may cause urinary obstruction.
  - Fluid intake

Cont.

- Family history
  - UTI
  - Nocturnal enuresis
  - VUR: presents with urinary sepsis
  - Cystic kidney disease: associated with cystic fibrosis
  - Absence of kidney
  - Hypospadias: relatively common (~30%)
  - Cryptorchidism

PHYSICAL EXAMINATION

- Abdominal exam
  - Masses
  - Pain
  - Palpable bladder
- Back exam:
  - Dimples
  - Hairy patches

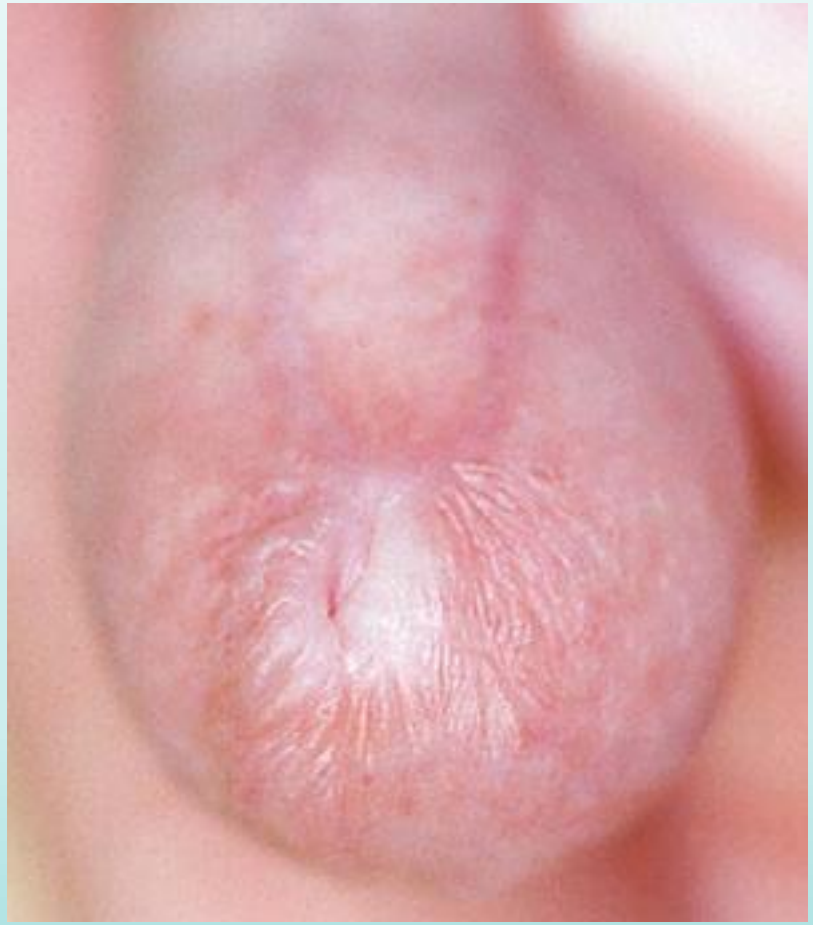
Cont.

- Genitourinary exam
  - Rash
  - Labial adhesions
  - Urethral prolapse: causes PV bleeding in female children
  - Ureterocele prolapse
  - Urethral opening (location)
  - Foreskin (phimosis/ paraphimosis vs. retractable)
  - Testicular position
  - Testicular masses
- Watch them pee!
- Post – void residual volume.

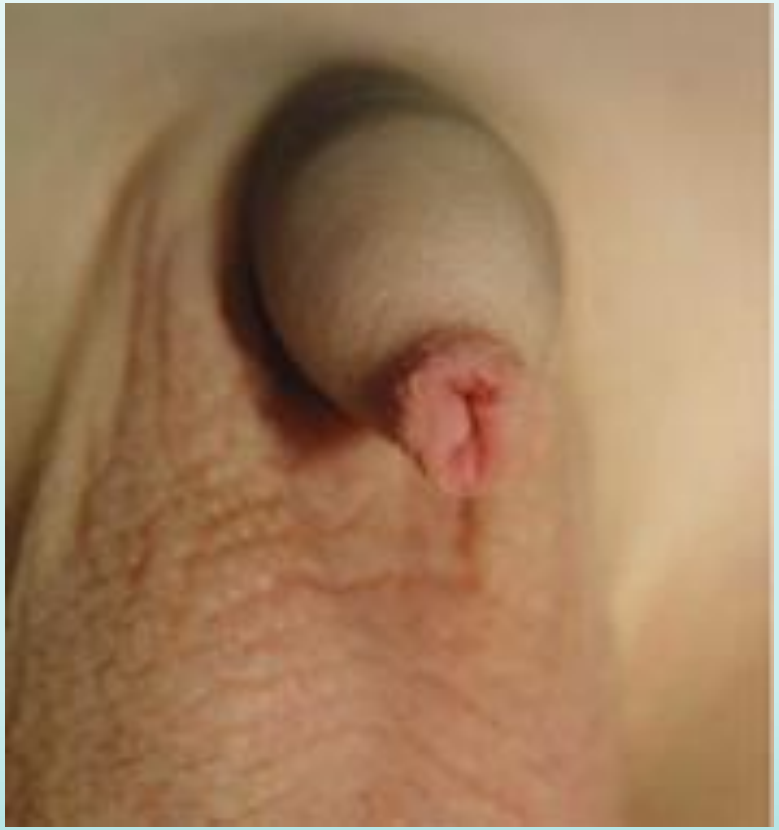
1. PHIMOSIS

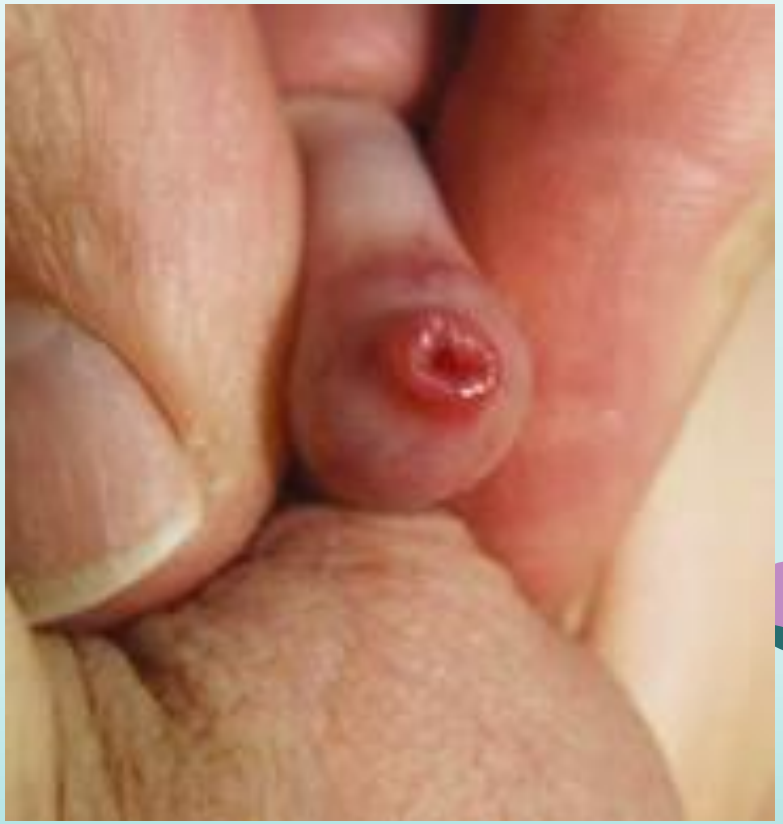
- Definition: narrowing of the prepuce
- Physiological phimosis:
  - Important to differentiate from pathologic forms
- Issues:
  - Incomplete emptying of urine
  - Urine remaining under foreskin → recurrent balanitis → ascending infection
- If asymptomatic: no treatment is offered since:
  - Forceful retraction → bleeding → scar → more adhesions

Phimosis



Physiologic phimosis: no scars, no bleeding, symmetric eversion of soft supple skin.

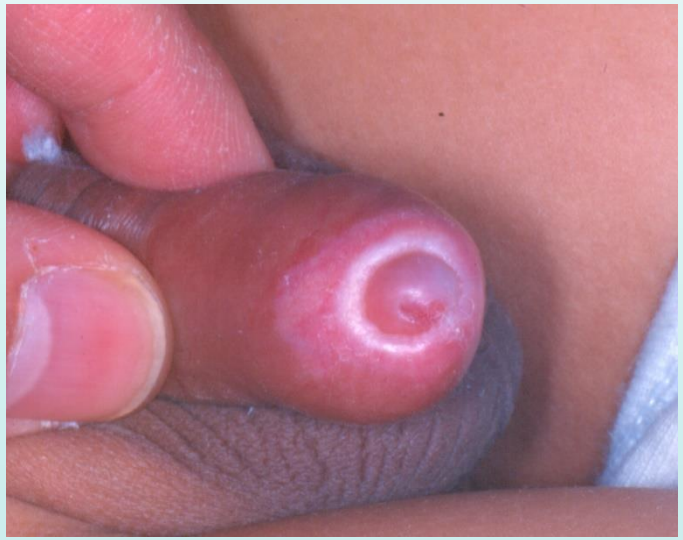


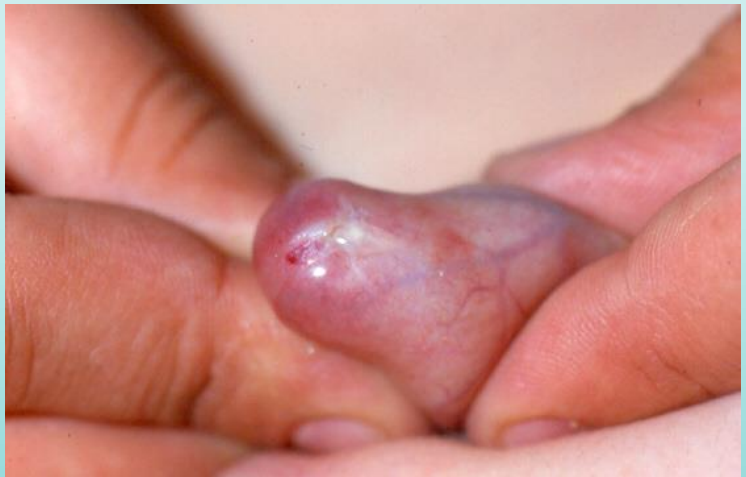


Pathological phimosis

- Distinguishing features
  - History of cracking & bleeding with retraction
  - Indurated, scarred whitened skin at tip of prepuce
  - Narrowest part is most distal
  - Painful erections
- This entity requires interventions: circumcision vs. dorsal slit.

Pathological phimosis

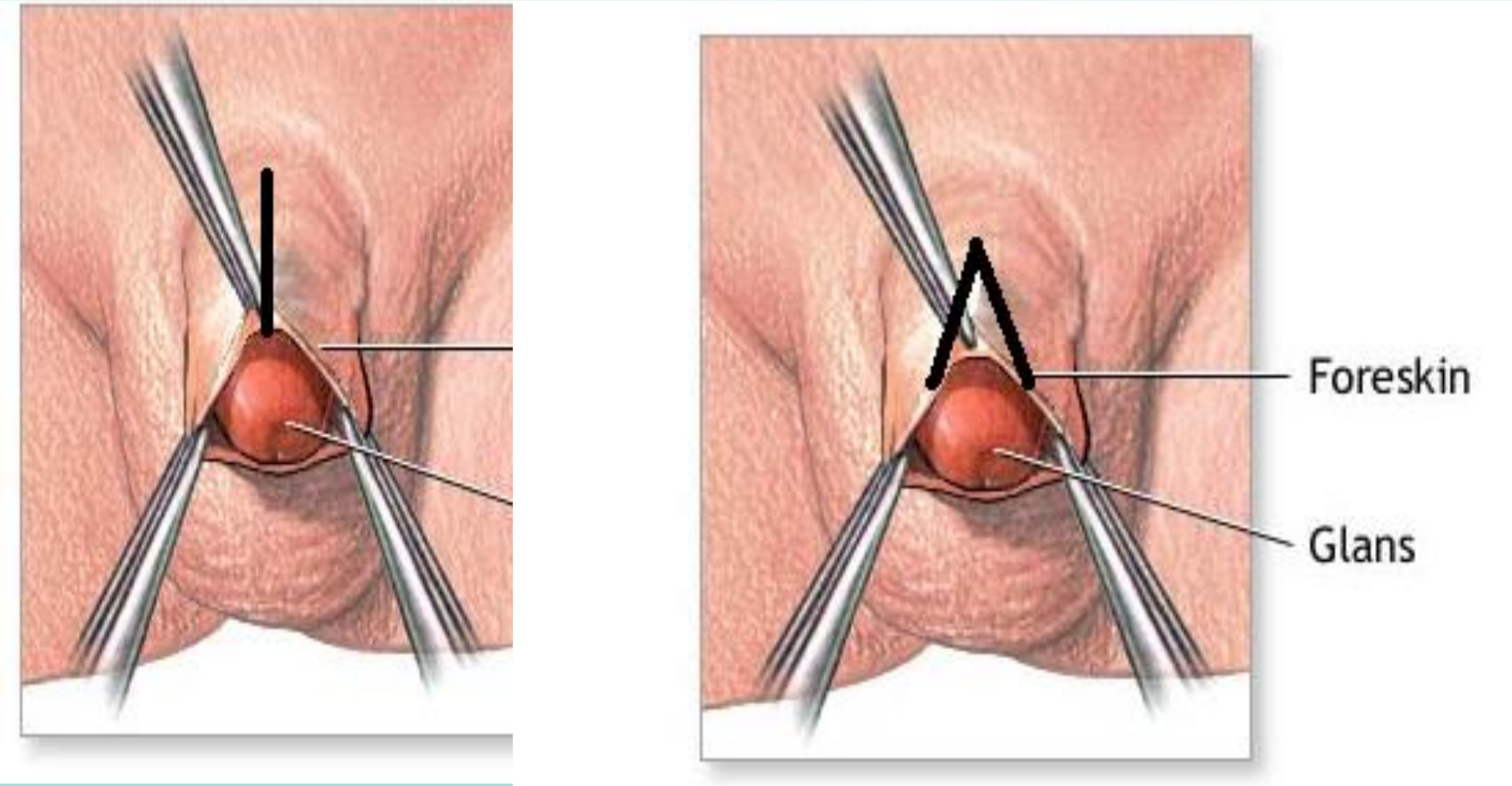




Treatment of phimosis

- Indications:
  - Symptoms
  - Pathologic phimosis
- Corticosteroid cream (short term for < 2 – 3 months)
  - Do not work
  - Not good for children
  - If used, must use strong or moderately strong steroid (betamethasone vs. clobetasol)
- Dorsal slit: incising the ‘top’ of the foreskin.

Dorsal slit



Circumcision

- There is no absolute medical indication for circumcision in the neonatal period:
  - Relative indication anomaly of urinary tract and recurrent infections.
- Potential medical advantages
  - Decrease incidence of UTIs in the first year of life
  - Prevent phimosis
  - Prevent balanoposthitis (infection of the glans penis)
  - Decrease incidence of penile cancer
  - May decrease the incidence of sexually transmitted disease

Circumcision

- ***Mainstay*** treatment.
- 2 methods
  - Use of gadgets:
    - Plastibell clamp (MC; used locally)
    - Gomco clamp
    - Mogen clamp
  - Surgical (traditional)

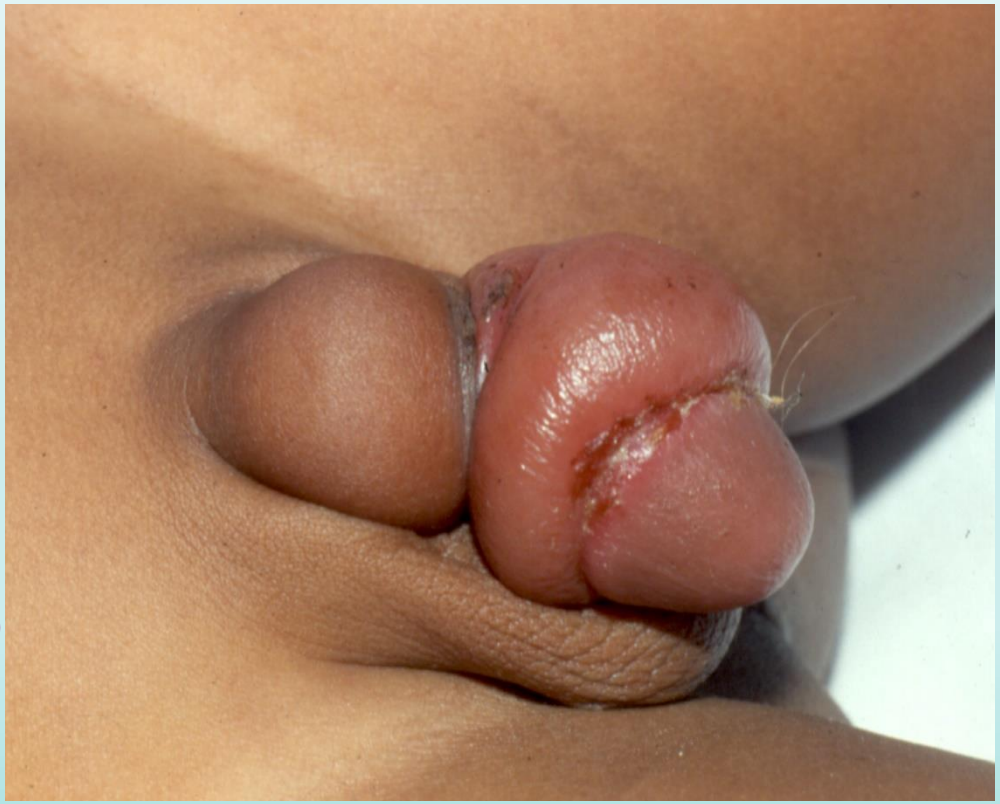
Complications (0.2 - 0.5%)

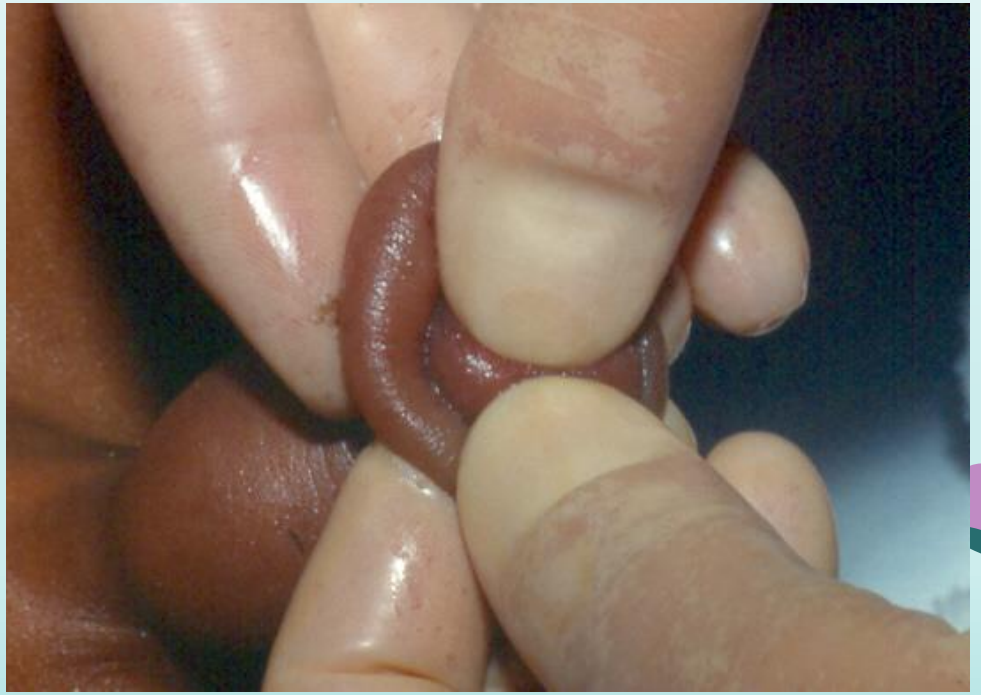
- Bleeding
- Injury to penis: amputation of glans
- Skin tissues
  - Take of too much
  - Leave on too much
  - Skin bridges
  - Inclusion cysts
  - Penile curvature
  - Urethro – cutaneous fistula
- Long term: meatal stenosis

2. PARAPHIMOSIS

- Painful constriction of glans penis by the foreskin which has been retracted at the distal penis.
- Treatment
  - Reduction under LA
    - Wrap with Coban to reduce edema
    - ? Hypertonic saline
    - Manual reduction
  - Dorsal slit
  - Circumcision

Paraphimosis

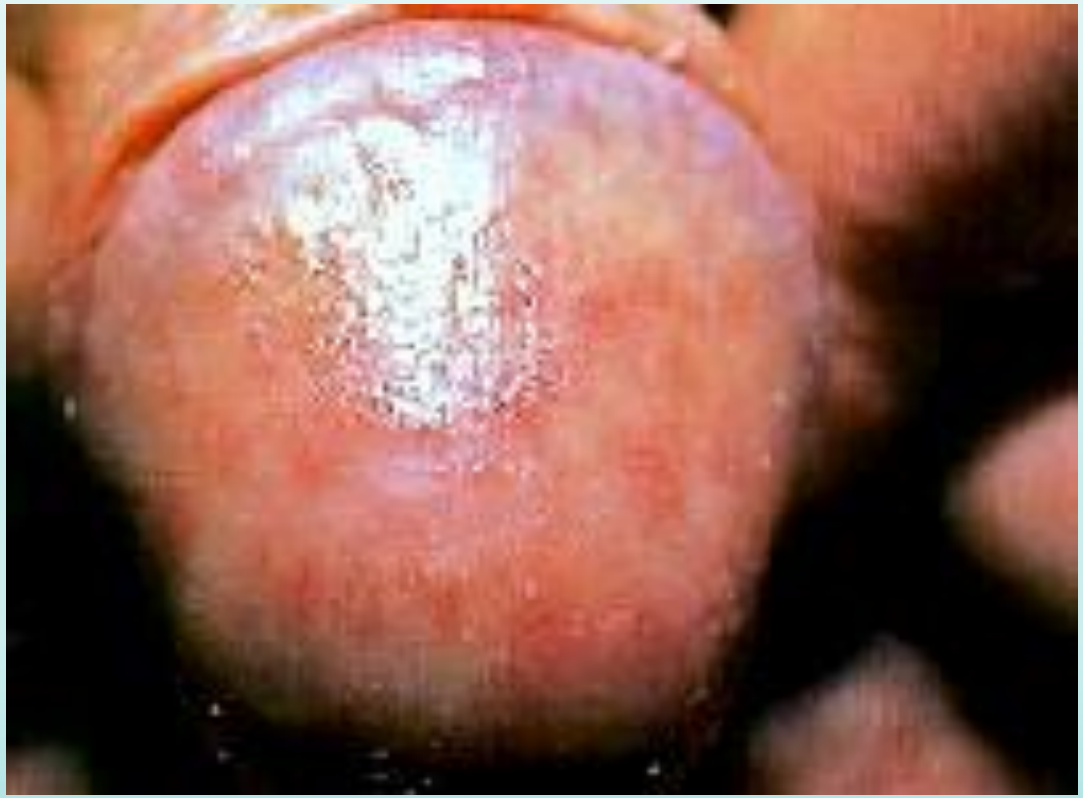




3. BALANITIS

- Consequence of phimosis
- Inflammation of underside of glans
- MCC: Chlamydia
- P/C
  - Erythema (localized)
  - Edema
  - Purulent discharge
  - Fever
  - UTI
- Recurrent balanitis is a common cause of phimosis

Balanitis



Treatment of balanitis

- Topical antibiotics (fucidin)
- Oral antibiotics for severe cases
- Warm water soaks/ baths BID for treatment
- Topical steroids
- Occasionally antifungals
- Do not retract the foreskin.

4. UTI

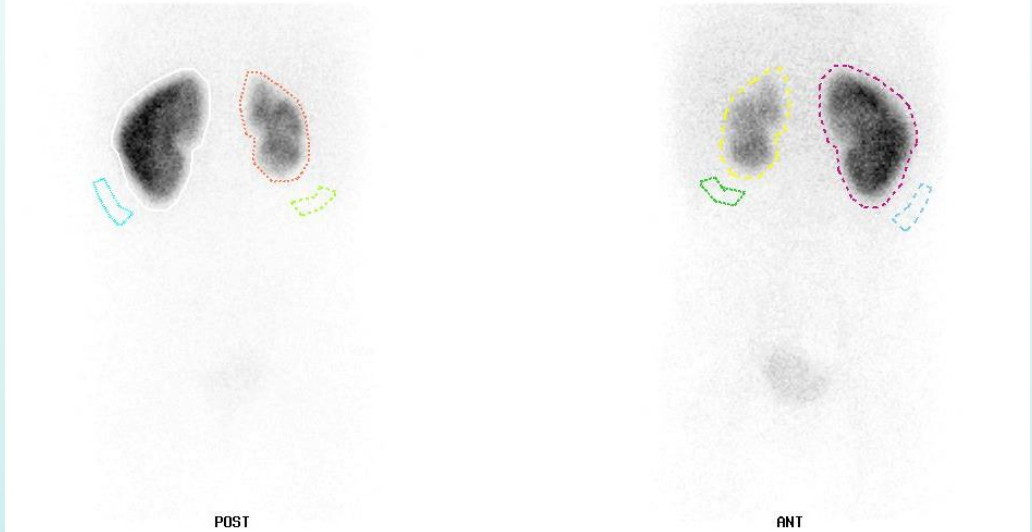
- Presentation
  - Young children (infants): febrile, vomiting, decreased appetite, lethargy
  - Older children: febrile (implies PN), dysuria, frequency, abdominal pain

Investigations

- History & PE: voiding history and/ or constipation, family history, fever
- Urine: ideally catheters specimen, bag specimens are bad (90% FP rate)
- ***Radiology examinations is recommended in all children with a febrile UTI:***
  - ***U/S***: status of bladder mucosa
  - ***VCUG:*** if US is abnormal (do U/E/Cr first as it is a contrast study)
  - ***Radionuclide studies***: look at renal parenchyma & differential glomerular function
    - ***DMSA*** scan to document a pyelonephritis
    - ***DTPA***

DMSA Scan

ghtbox 1



RESULTS

	LEFT KIDNEY	RIGHT KIDNEY
NT	127191 825	40369 997
OST	178504 595	62281 643

DIFFERENTIAL FUNCTION

OSTERIOR EAN	**74.99 %**	**25.01 %**
EOMETRIC EAN	**76.73 %**	**23.27 %**

Treatment

- Lower tract: short course antibiotics
- Upper tract (fever, back pain, nausea & vomiting)
  - 2 week course of antibiotics
  - Admission if very ill
  - Quick treatment decreases chances of scarring
  - Long term antibiotics: nitrofurantoin, Septrin; prevents renal scarring which as a direct effect on prognosis

Recurrent UTIs treatment

- Improve voiding patterns
  - Timed voiding (q2h)
  - Double voiding: seating in an ideal position to urinate and then waiting for 20 – 30 seconds to urinate again.
  - Improve emptying
    - Biofeedback
    - Alpha blocker

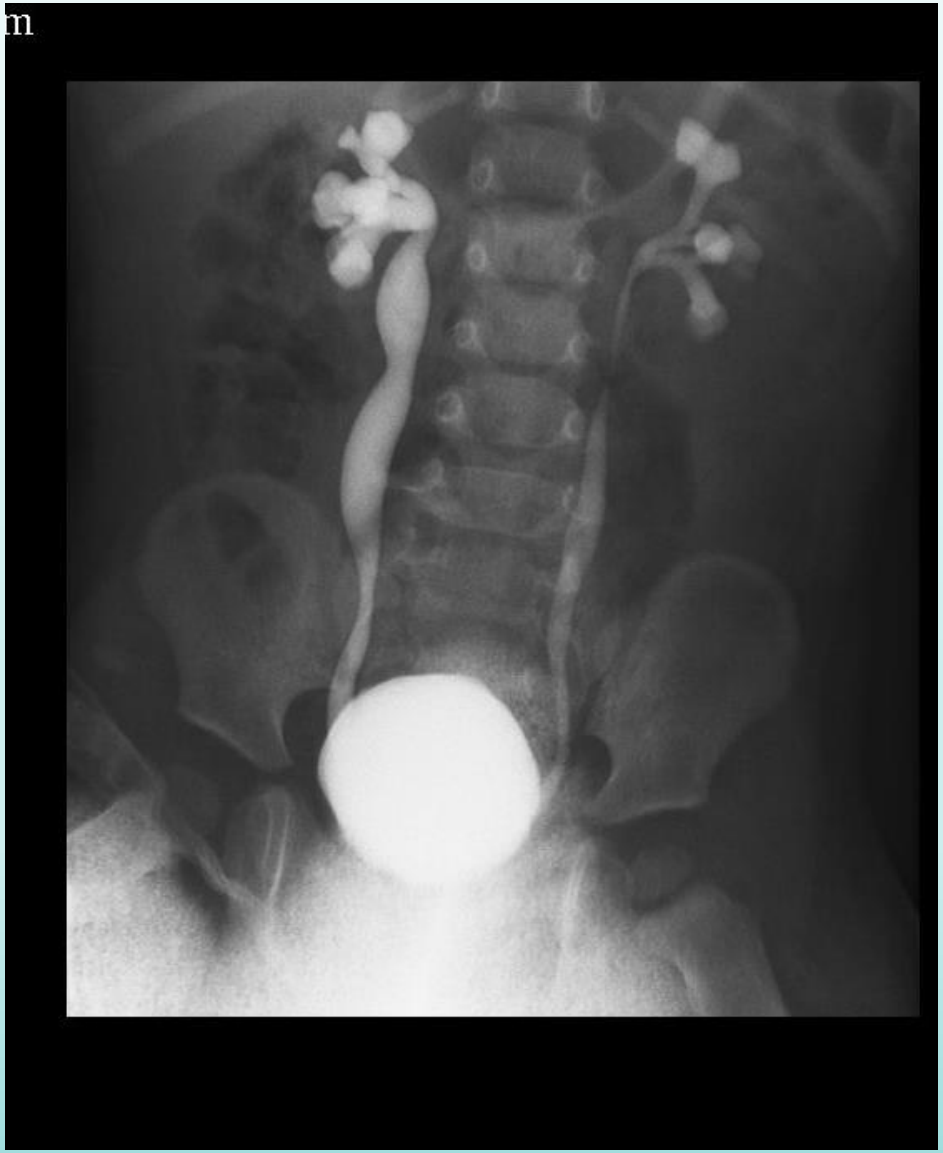
Cont.

- Increase water intake
- Stool softener
- Antibiotic prophylaxis
- Treat anatomic abnormality

4. COLLECTING SYSTEM ABNORMALITIES

- Include:
  - VUR
  - Obstruction
    - UPJ obstruction
    - UVJ obstruction
  - Duplication

VUR



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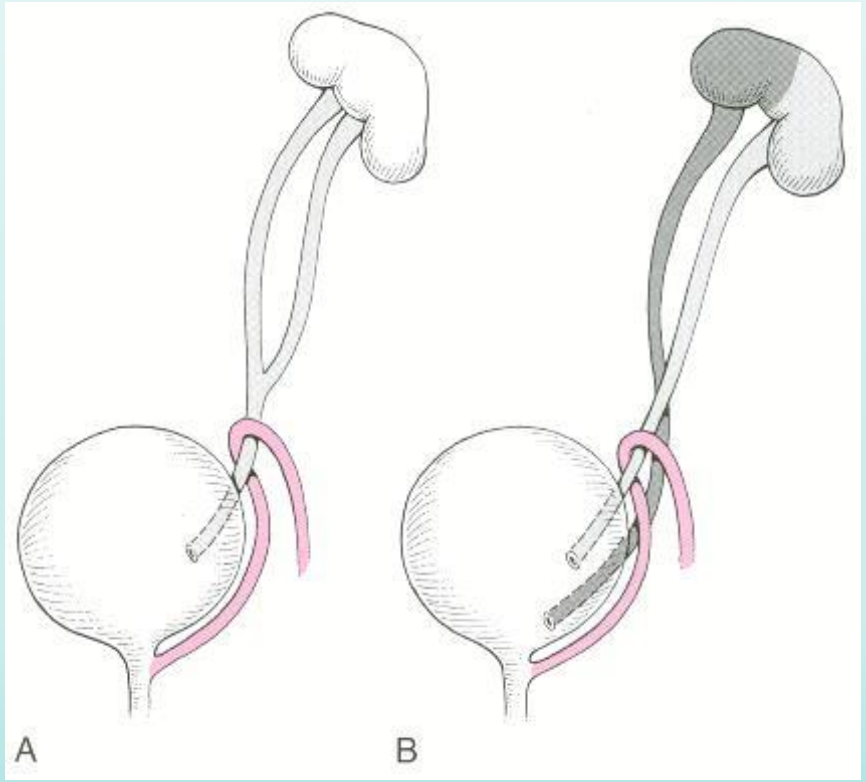
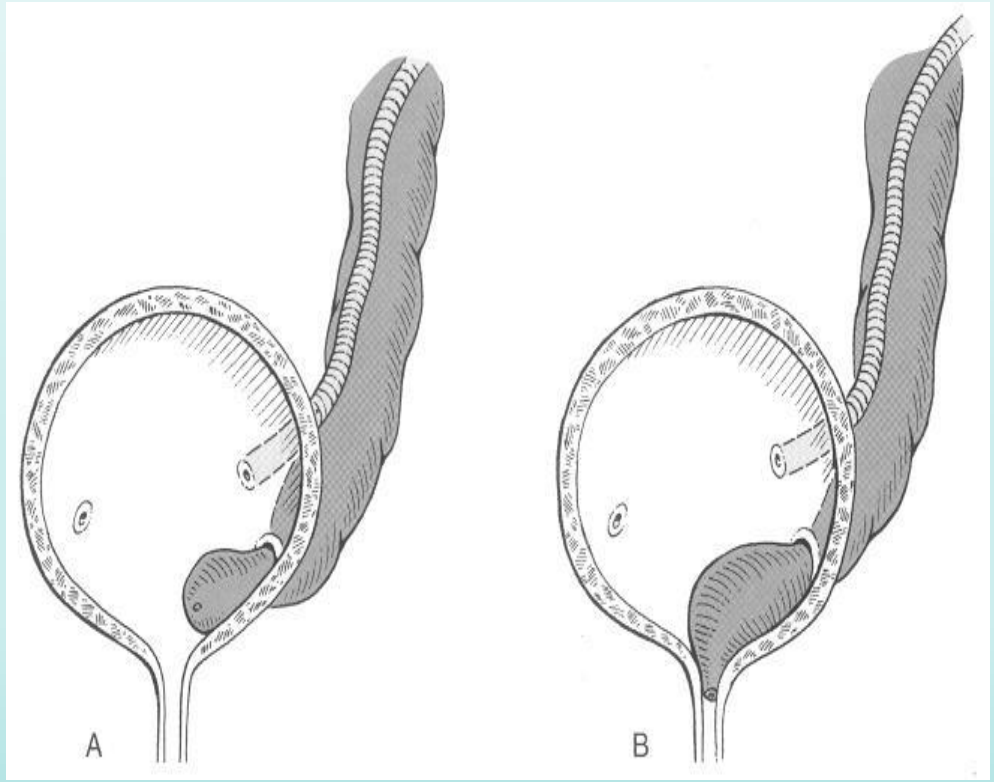
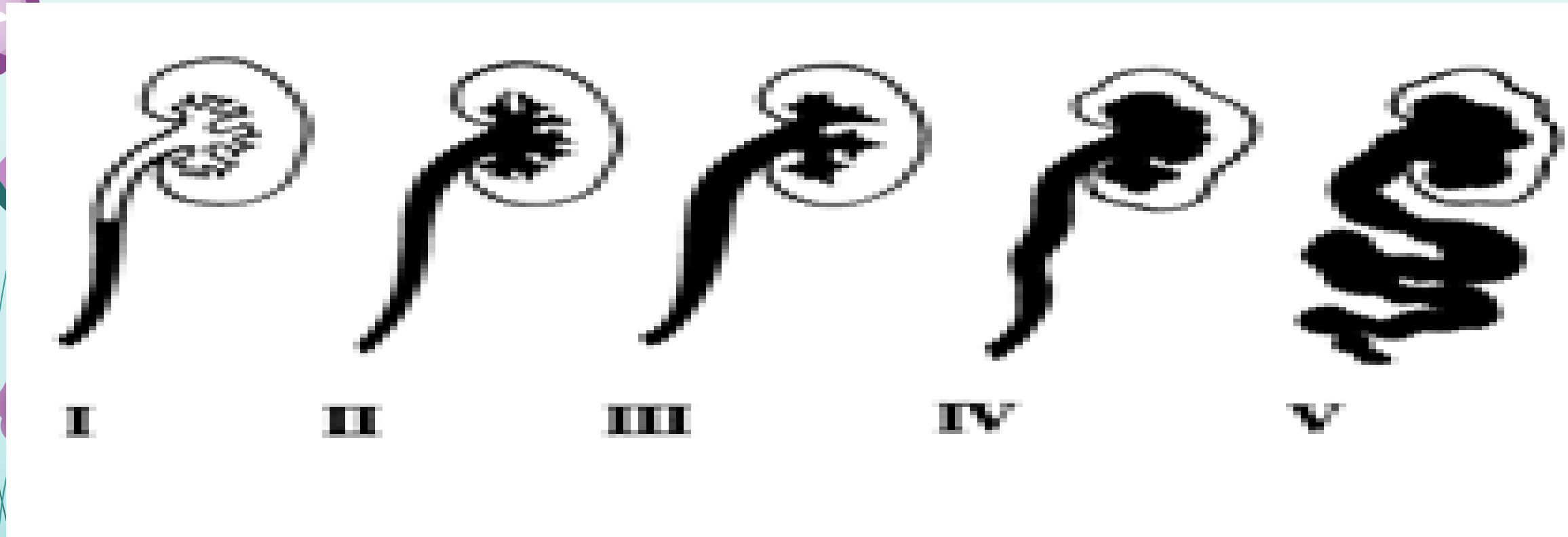




Grading of VUR



Treatment of VUR

- Ureteric reimplantation
- Cystoscopic injection of highly osmotic products (duflac)

Ureteropelvic Junction Obstruction

- Cause:
  - Congenital stricture or adynamic segment
  - Crossing vessel
- Presentation
  - Antenatal hydronephrosis
  - Intermittent severe flank pain (lasts about 6 hrs.) with nausea & vomiting
  - UTI
  - Renal calculi

Investigation & management

- Investigations
  - Ultrasound
  - Diuretic MAG3 Renal Scan
- Asymptomatic
  - Observe
  - If drop in renal function or worsening of hydro operate
- Symptomatic: pyeloplasty, ureteric re – implantation

Duplex kidney

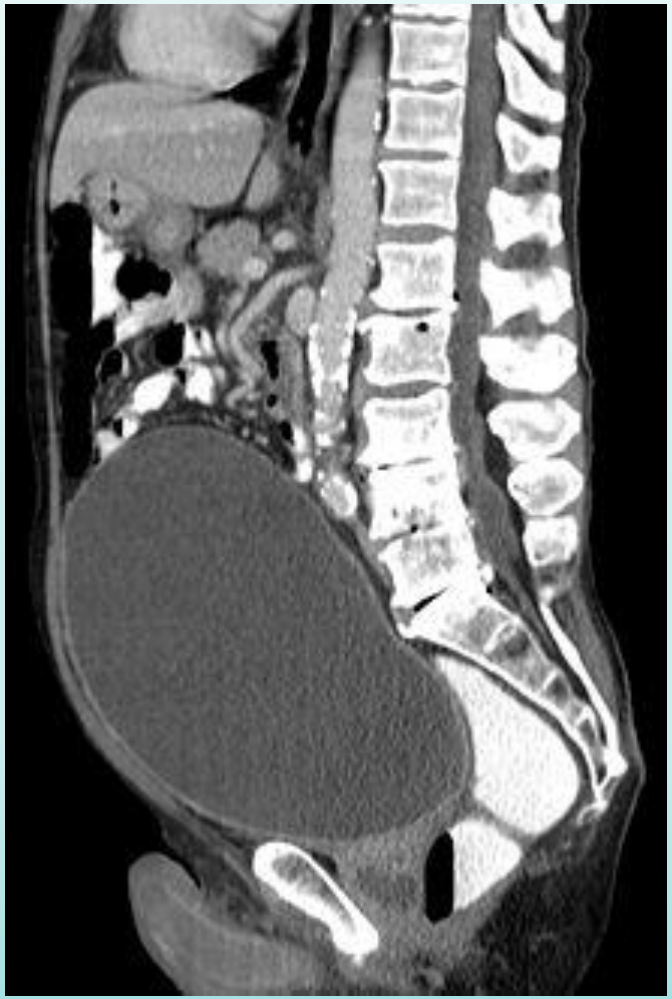
- More common in females
- Presents with incontinence

6. URINARY RETENTION (ISCHURIA)

Causes in bladder

- ***Detrusor sphincter dyssynergia***
- ***Neurogenic bladder*** (commonly pelvic splanchnic nerve damage, CES, descending cortical fibers lesion, pontine micturition or storage center lesions, demyelinating disease or Parkinson's disease)
- ***Iatrogenic*** (caused by medical treatment/ procedure): scarring of the bladder neck (commonly from removal of indwelling catheters or cystoscopy operations)
- ***Damage to the bladder***

A lack of the ability to urinate



Causes in penile urethra

- Congenital Urethral valves
- Phimosis or pinhole meatus
- Circumcision
- Obstruction in the urethra, for example a stricture (usually caused either by injury or STD), a metastasis or a precipitated pseudogout crystal in the urine
- STD lesions (gonorrhea causes numerous strictures, leading to a ‘rosary bead’ appearance, whereas chlamydia usually causes a single stricture) in the sexually active.

Other causes

- Tethered spinal cord syndrome
- Paruresis (shy bladder syndrome)
- Use of NSAIDs or drugs with anticholinergic properties
- Stones or metastases can theoretically appear anywhere along the urinary tract, but vary in frequency depending on anatomy.
  - Positional dependence
- Muscarinic antagonist: atropine, scopolamine
- Mental retardation
- Stool in bowel

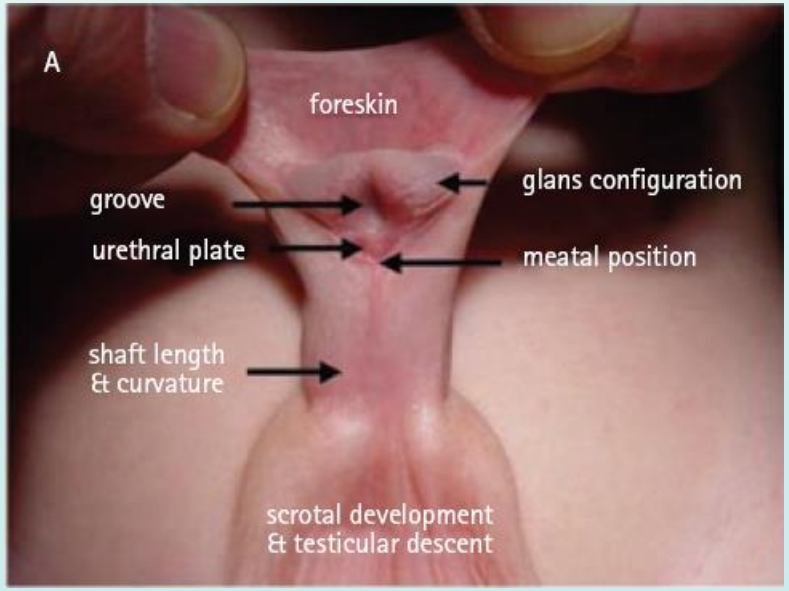
Signs & symptoms

- Poor urinary stream with intermittent flow
- Straining
- Sense of incomplete voiding
- Hesitancy
- Incontinence, nocturia, frequency
- Anuria (medical emergency)
- Pyonephrosis
- Renal failure
- Sepsis
- Hydronephrosis…

Treatment

- Sterile/ aseptic urethral catheterization
- Suprapubic cystotomy:
  - Don’t do in a neonate; do a Vesicostomy instead.
- Clean intermittent self catheterization
  - For long term obstruction & for children whose incontinence cannot be dealt with e.g. severe CP, motor neuron disease
- Treat the cause

5. HYPOSPADIAS



Epidemiology

- Incidence: 1 in 125 male births
  - Caucasian 0.3 – 0.8%
  - Other racial groups 0.05-0.4%
- Associations
  - Cryptorchidism (9.3% of patients with hypospadias)
    - Incidence of chromosomal abnormality higher with proximal hypo & UDT (22%)
  - Inguinal hernias (9%)

Risk factors for hypospadias

- Endocrine
  - Disruption in the synthetic bio pathway of androgens
  - May be a delay in the maturation of the hypothalamic-pituitary-axis
- Genetic: Familial rate 7%
- Environmental: Endocrine disrupters in the environment may be responsible for the increase in incidence
- Maternal
  - Maternal ***progestin exposure*** may increase likelihood of hypospadias. Some studies show a marked increase in hypospadias in women undergoing IVF.

Investigation

- Simple distal hypospadias: No evaluation
- Proximal hypospadias + one or bilateral impalpable testicles → Intersex evaluation
  - Electrolytes
  - Karyotype
  - 17 hydroxy – progesterone
  - Ultrasound abdomen

Treatment of hypospadias

- Referral before 6 months of age.
- Surgery usually between ***age 1 and school age***
- **Distal hypospadias**
  - Surgery mostly for cosmesis; Sometimes for urinary function
- **Proximal hypospadias**
  - Treatment for both urinary & reproductive function
  - Higher risk of complications

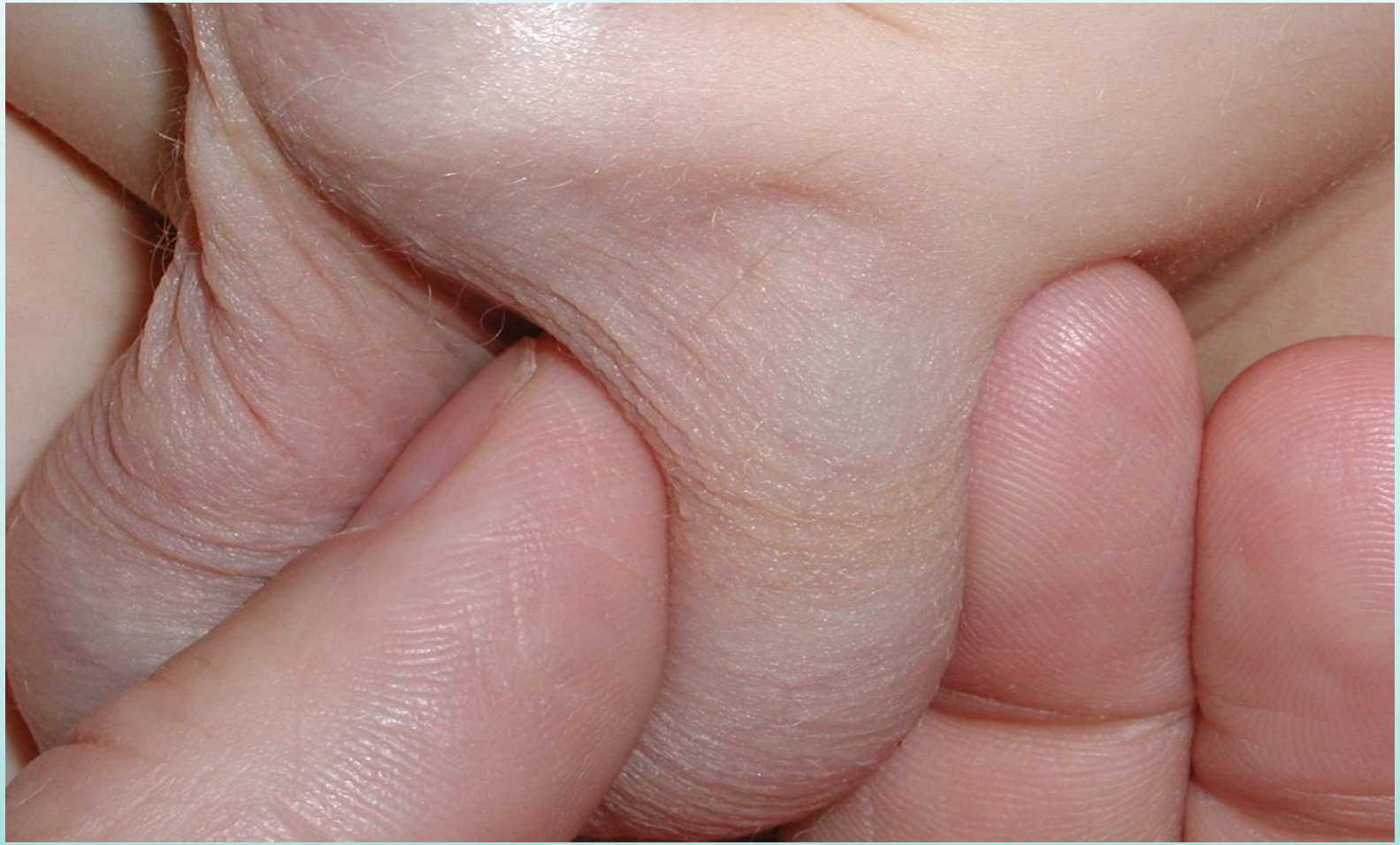
6. SCROTAL CONDITIONS: TESTICULAR PAIN

- Causes
  - Torsion of the appendix testes
  - Epididymitis
  - Testicular torsion

Torsion of appendix testes

- Symptoms/Signs
  - Pre adolescent
  - Pinpoint tenderness
  - Blue dot sign
  - Over time can cause local inflammation which looks like epididymitis on ultrasound
- Treatment: Rest, scrotal support, NSAIDS

Blue dot sign



Epididymitis

- Adolescent and older
- Gradual onset
- Tender superior portion
- Investigations:
  - U/A
  - Urine culture
  - Sexual history
  - Possible ultrasound to r/o torsion

Testicular torsion

- Symptoms:
  - Adolescent (not always)
  - Severe pain
  - Sudden onset
  - Sometimes only abdominal pain
  - Nausea & vomiting
- Examination: abnormal lie of testicle; absence of cremasteric reflex

Investigation

- ***NB: If it looks like a torsion go right to the O.R. Ideally fix in 6 hrs.!***
- Urinalysis & culture
  - If normal unlikely to be epididymitis
- Scrotal Ultrasound

Undescended Testicles (Cryptorchidism)

- The ***most common birth abnormality involving the male genitalia*** (0.8% incidence at 6 months)
- All (except premature infants) will descend in first 3 months of life:
  - If undescended at 3 months then you should refer to a pediatric urologist.
- ***Retractile testicle*** is a normally descended testicle that is pulled out of the scrotum by an overactive cremasteric reflex

Complications of UDT

- Inguinal hernia
- Risk of torsion
- Infertility: only decreased if bilateral
- Increased risk of testicular cancer
  - 4 – 10 times normal

Treatment of UDT

- Orchidopexy: placement of testicle in scrotum.
- May improve fertility
- Easier to monitor for malignancy
- Surgical correction by 6 months of age.

Hydrocele

- Communicating hydrocele
  - Persistence of a ***patent processus vaginalis***
  - Accumulation of fluid around the testicle
  - Treatment:
    - Often will close in first year of life
    - Period of observation then surgery if remains
- Non – communicating hydrocele
  - Rare in children
  - Usually a result of inflammation

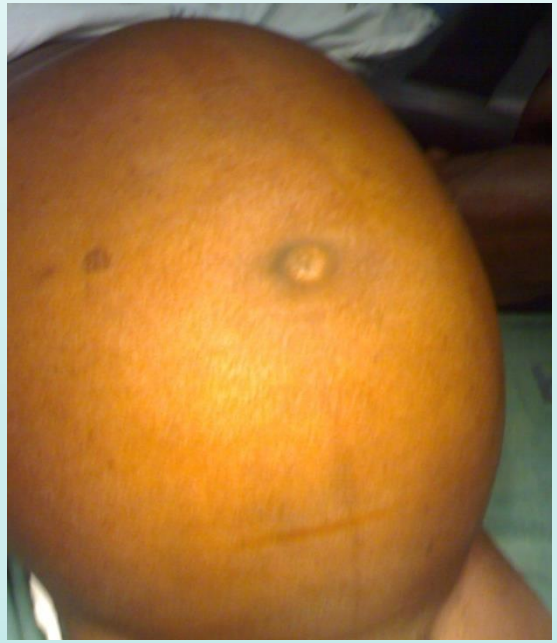
7. TUMORS

- Include:
  - Wilms' tumor
  - Rhabdomyosarcoma
  - Secondary tumors

Wilms tumor

- MC intra abdominal malignant tumor in children.
- Peak incidence: 3.5 yrs.
- P/C: large abdominal or flank mass with abdominal pain, asymptomatic hematuria & occasionally fever
- Other presentations: malaise, weight loss, anemia, left varicocele (obstructed renal vein), HTN

Wilms tumor



8. INGUINO – SCROTAL PATHOLOGY

LEVEL VI MBCHB

BY: DR. MWIKA M. P.

OUTLINE

1. Undescended Testis
2. Hernia
3. Hydrocele
4. Acute scrotum

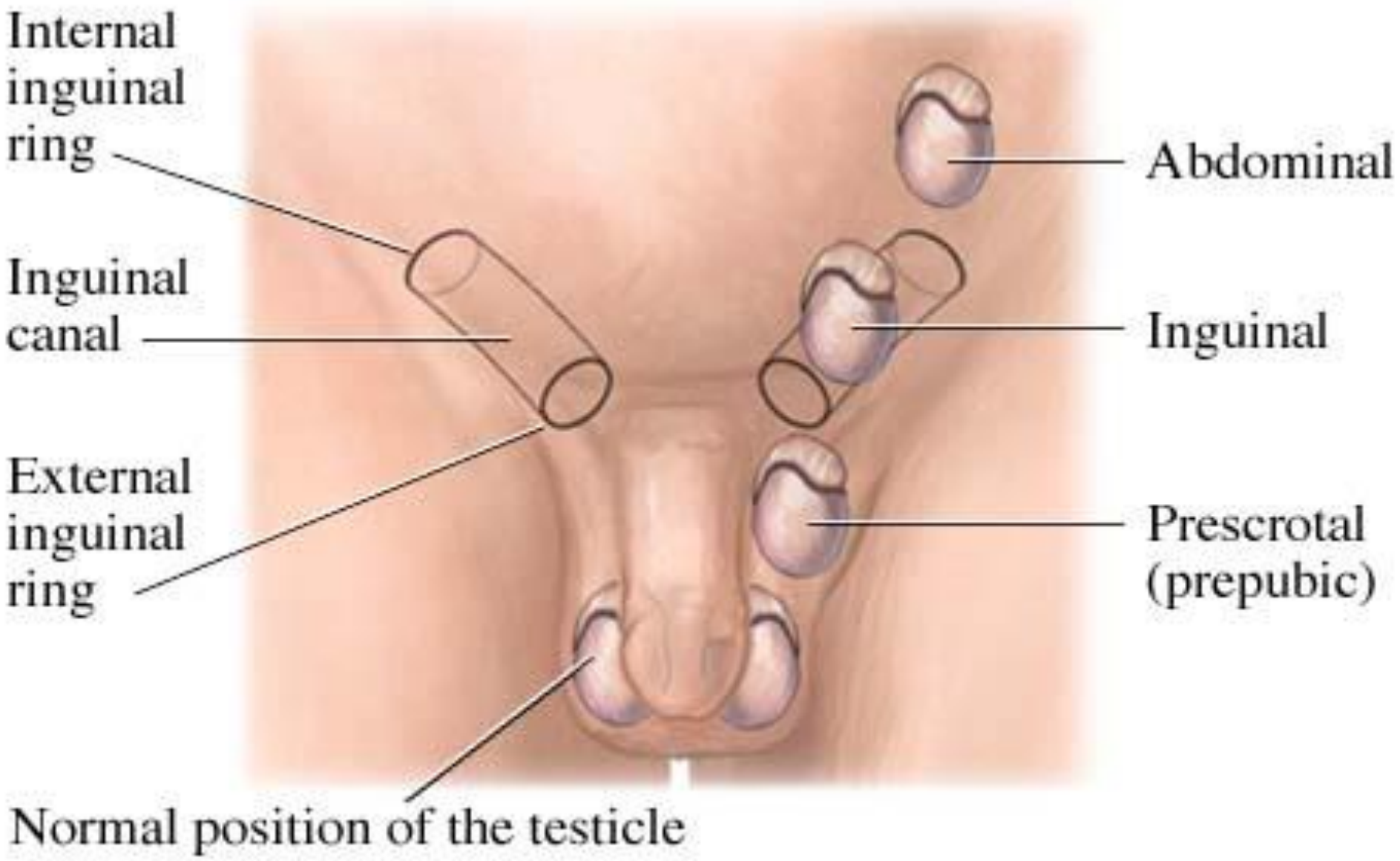
1. UNDESCENDED TESTIS (UDT)

- ✓ Definition: Testicles that ***cannot be manipulated into the scrotum***.
- ✓ Affects 4***% - 5% of full – term & 9% - 30% of premature males*** at birth.
- ✓ Incidence falls to < 1% by 3 months of age.
- ✓ Bilateral UDT occurs in 10% of children with UDT.

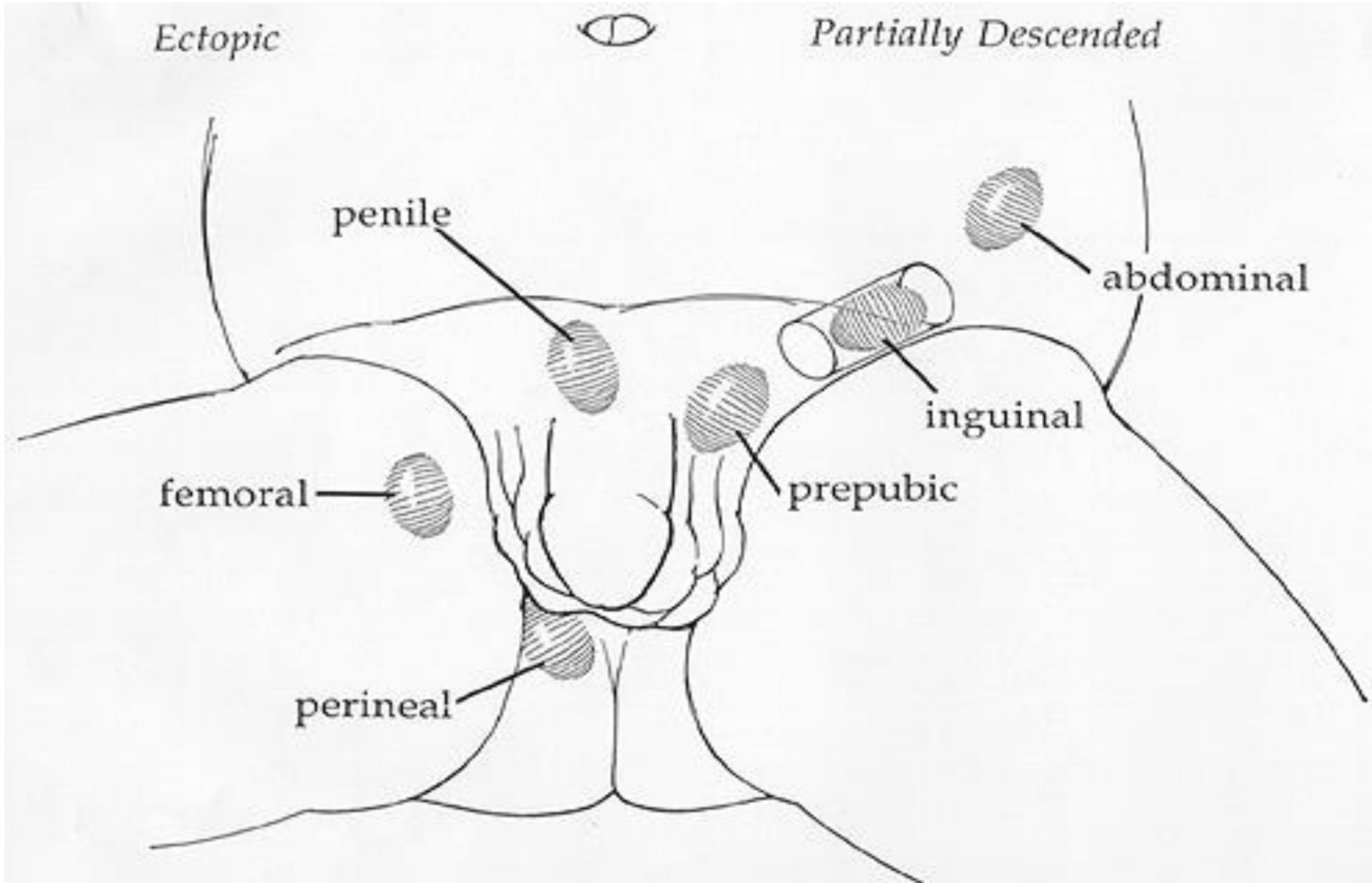
Testicular descent

- ✓ Mechanisms: not well understood
- ✓ Divided into:
  - ➢ Intra – abdominal phase (androgen independent)
  - ➢ Inguinal – scrotal phase (mechanical; hormonal dependent)

Cont.


Internal inguinal ring
Inguinal canal
External inguinal ring
Abdominal
Inguinal
Prescrotal (prepubic)
Normal position of the testicle

Anatomical classification of UDT



Cont.

- ✓ The testis can be classified in many ways, one of which is ***anatomical along the line of normal descent***.
  - ➢ Intra – abdominal, canalicular, inguinal/ pubic (prescrotal), superficial inguinal pouch
- ✓ True ectopic: pubo – penile, femoral, perineal, transverse ectopia
- ✓ Ectopic: It has been suggested that this may be the result of an ***abnormal location of the genitofemoral nerve*** with consequent abnormal migration of the gubernaculum to the wrong site.
- ✓ It can also be classified as ***palpable*** or ***non palpable***

Retractile testes

- ✓ Commonly confused with UDT.
- ✓ All retractile testes can be delivered into the scrotum.
- ✓ Whereas retractile testes will stay within the scrotum after the cremaster muscle has been overstretched, a low UDT will immediately pop back to its undescended position after it has been released.

Ascending/ acquired undescending testes

- ✓ Relatively recently recognized condition. Incidence is between 1 & 2%.
- ✓ Testes once within the scrotum but later don't reach the base of the scrotum (in later childhood/ early adolescence).
- ✓ ***Failure of the spermatic cord to elongate as the child grows***. May also be caused by either patent hernia or obliterated remnant which inhibits elongation of the adjacent vas deferens & testicular vessels.
- ✓ Risk Factor: delayed initial descent of testes
- ✓ Those with UNT at birth, followed up for possible testicular ascent.

Complications of UDT

- ✓ Infertility
- ✓ Inguinal hernia
- ✓ Torsion
- ✓ Psychological factors

Fertility

- ✓ Spermatogenesis is retarded by maldescent
- ✓ Bilateral UDT → poor fertility
- ✓ Higher UDT → more damage to seminiferous tubules
- ✓ Earlier orchiopexy (when the patient is ~6 months old) may improve chances for recovery of spermatogenesis
- ✓ Sperm counts in unilateral UDT are much lower than normal & the contralateral testis may also be defective.

Others

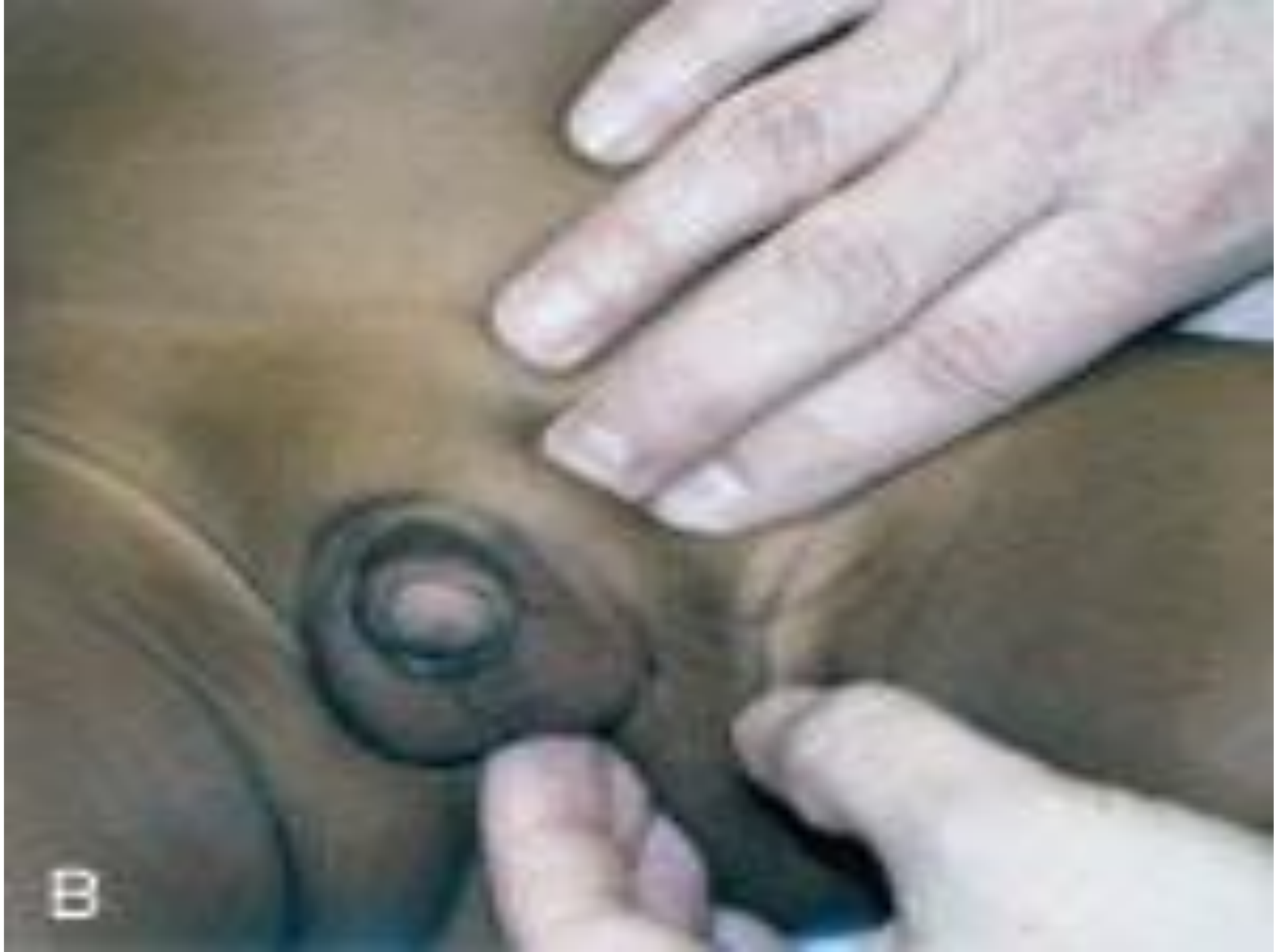
- ✓ Inguinal hernia: UDT result in a ***patent processus vaginalis*** hence a higher risk for inguinal hernia.
- ✓ Torsion: increased risk due to anatomic abnormality of a cryptorchid testis & mesentery (20% risk).
- ✓ Physiologic factors: parental anxiety about subsequent fertility.

Diagnosis

- ✓ Clinical history:
  - Has the testis **ever been palpable** in the scrotum?
  - Was the patient **born prematurely**? What was the **birth weight**?
  - Has the patient undergone **prior inguinal surgery**?
  - Family history of Cryptorchidism, Hypospadias, Infertility?

Cont.

- ✓ Physical Examination
  - Frog – leg position
  - Milk down palpating from the iliac to the scrotum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Cont.

- ✓ What are the features of the scrotum & its contents (e.g. hypoplasticity, bifidity, rugae, transposition, pigmentation)
- ✓ Is the contralateral testicle hypertrophic?
- ✓ Is the UDT located in an unusual position, such as in an ectopic site (i.e., superficial inguinal pouch or transverse scrotal, femoral, pre – penile, perineal, or contralateral hemi – scrotum)?
- ✓ Note the presence of any hypospadias or chordee. Does the patient have a normal stretched penile length?
- ✓ If the findings are equivocal, perform serial examinations.

Imaging studies

- ✓ Little role in diagnosis of UDT
- ✓ U/S
- ✓ MRI: costs, sedation
- ✓ Angiography

Management: Hormonal treatment

- ✓ Deficiency of the H-P-G axis.
- ✓ > 4 yrs. & with:
  - ➢ Bilateral UDT near the scrotal entrance
  - ➢ Retractile tests or
  - ➢ Acquired UDT
- ✓ HCG & LHRL (GnRH)
- ✓ A/E: increased scrotal rugae, pigmentation, pubic hair growth, increased penile size.

Surgical treatment

- ✓ Basic principles of orchiopexy are:
  - ➢ Localization, mobilization, isolation of processus, tension – free relocation of testis into scrotum.
- ✓ Does not reduce risk of cancer
- ✓ Recommended between 3 – 9 months
- ✓ Orchiectomy is an option for post – pubertal males and dysgenetic males.

2. INGUINAL HERNIA

- ✓ 0.8 – 4.4% of children
- ✓ 25% in premature infants
- ✓ M:F → 10:1
- ✓ More common on the right side: 60% right, 30% left ,10% bilateral.
- ✓ Increased incidence in the second twin once the first one has been diagnosed to have a hernia (***dizygotic or monozygotic?***)

Risk factors

Urogenital	Undescended testis
	Exstrophy of bladder
Increased peritoneal fluid	Ascites
	Presence of Ventriculoperitoneal shunt
	Peritoneal dialysis
Increased intra-abdominal pressure	Repair of gastroschisis/ exomphalos
	Severe ascites- liver failure, chylous etc
	Meconium peritonitis
Chronic respiratory disease	Cystic fibrosis
Connective tissue disorders	Ehlers-Danlos syndrome
	Hunter- Hurler syndrome
	Marfan syndrome
	Mucopolysaccharidosis
Miscellaneous	Developmental dysplasia of hip

Clinical presentation

- ✓ Swelling in inguinal or scrotal area.
- ✓ Classically, intermittent, when intra – abdominal pressure rises e.g. coughing, physical activity, crying.
- ✓ Occasionally, large hernia, may have a dragging or heaviness sensation.
- ✓ Significant pain signifies onset of complications: ***incarceration*** or ***strangulation*****.**

Physical Findings

- ✓ Most times hernia is suspected on basis ***of a classical history and cannot be demonstrated on examination.***
- ✓ If overt: a smooth soft mass at external ring area, cephalad and lateral to pubic tubercle.
- ✓ Enlarges when the child strains, cries or coughs.
- ✓ "***Silk Glove Sign***.": palpation of the hernia sac around the spermatic cord as it crosses the pubic tubercle

➢ The study below documented 91% sensitivity and more than 97% specificity in predicting the presence of hernia[1].

Imaging

- ✓ U/S
  - A hypoechoic structure in the inguinal canal
  - > 6mm indicates a hernia
  - 4 – 5 mm indicates a patent processus vaginalis

Contents of the hernia sac

- ✓ Commonly, small bowel and/or omentum.
- ✓ In girls, ovaries (with propensity to undergo incarceration).
  - ➢ If ovarian torsion sets in, may lead to rapid infarction. Once diagnosed to have an ovary in the sac, surgery should be performed as early as possible to prevent such an eventuality
- ✓ Sliding hernias are also known to occur in children where part of the bladder wall or fallopian tubes/ uterus have been found in the hernia defect.

Management: surgery for hernia & hydrocele

- ✓ High ligation & removal of the hernia sac
- ✓ Conventional open approach is performed via the inguinal or groin crease incision.

3. HYDROCELE

- ✓ Definition: collection of fluid between the parietal & visceral layer of the ***tunica vaginalis***.
- ✓ Causes:
  - ➢ Incomplete obliteration of the ***processus vaginalis peritonei***
  - ➢ Secondary to minor trauma
  - ➢ Testicular torsion
  - ➢ Epididymitis
  - ➢ Varicocele operation or may appear
  - ➢ Recurrence after primary repair of a communicating hydrocele

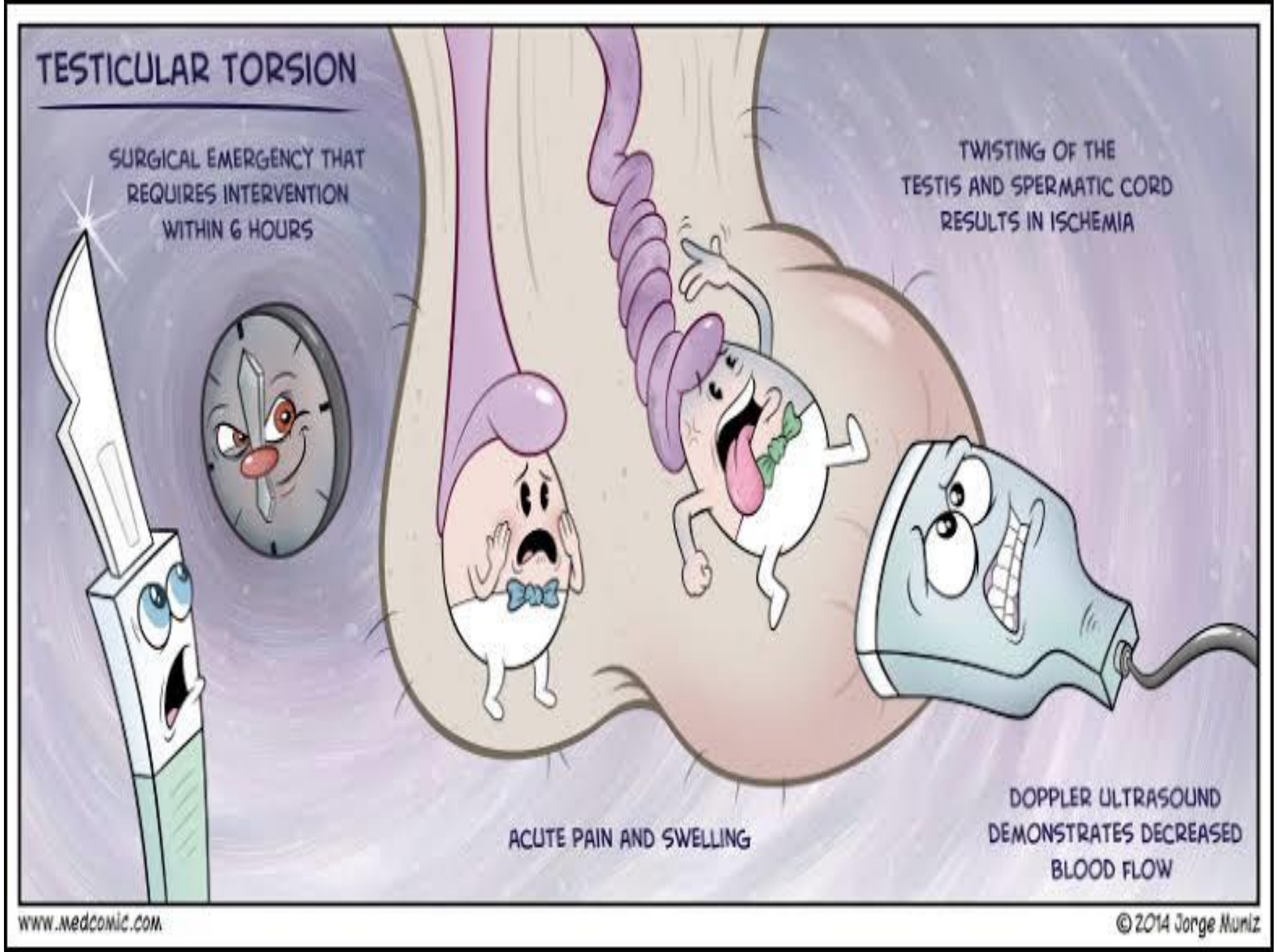
Cont.

- ✓ Diagnosis: clinical; trans – Illumination of scrotum.
- ✓ Treatment:
  - ➢ Surgical Rx of hydrocele is ***not indicated within the 1st 12 – 24 months*** because of the tendency for spontaneous resolution.
  - ➢ Early surgery is indicated if ***there is a suspicion of a concomitant inguinal hernia or underlying testicular pathology.***
  - ➢ Late onset hydrocele, non – communication, spontaneous resolution (75%) & expectant management of 6 – 9 months is recommended.

4. ACUTE SCROTUM

✓ Causes:

- Testicular torsion
- Epididymitis/ epididymo – orchitis
- Mumps orchitis
- Idiopathic scrotal edema
- Scrotal hematoma
- Incarcerated hernia
- Systemic disease: Henoch – Schönlein Purpura


TESTICULAR TORSION
SURGICAL EMERGENCY THAT REQUIRES INTERVENTION WITHIN 6 HOURS
TWISTING OF THE TESTIS AND SPERMATIC CORD RESULTS IN ISCHEMIA
ACUTE PAIN AND SWELLING
DOPPLER ULTRASOUND DEMONSTRATES DECREASED BLOOD FLOW
www.medcomic.com
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Diagnosis

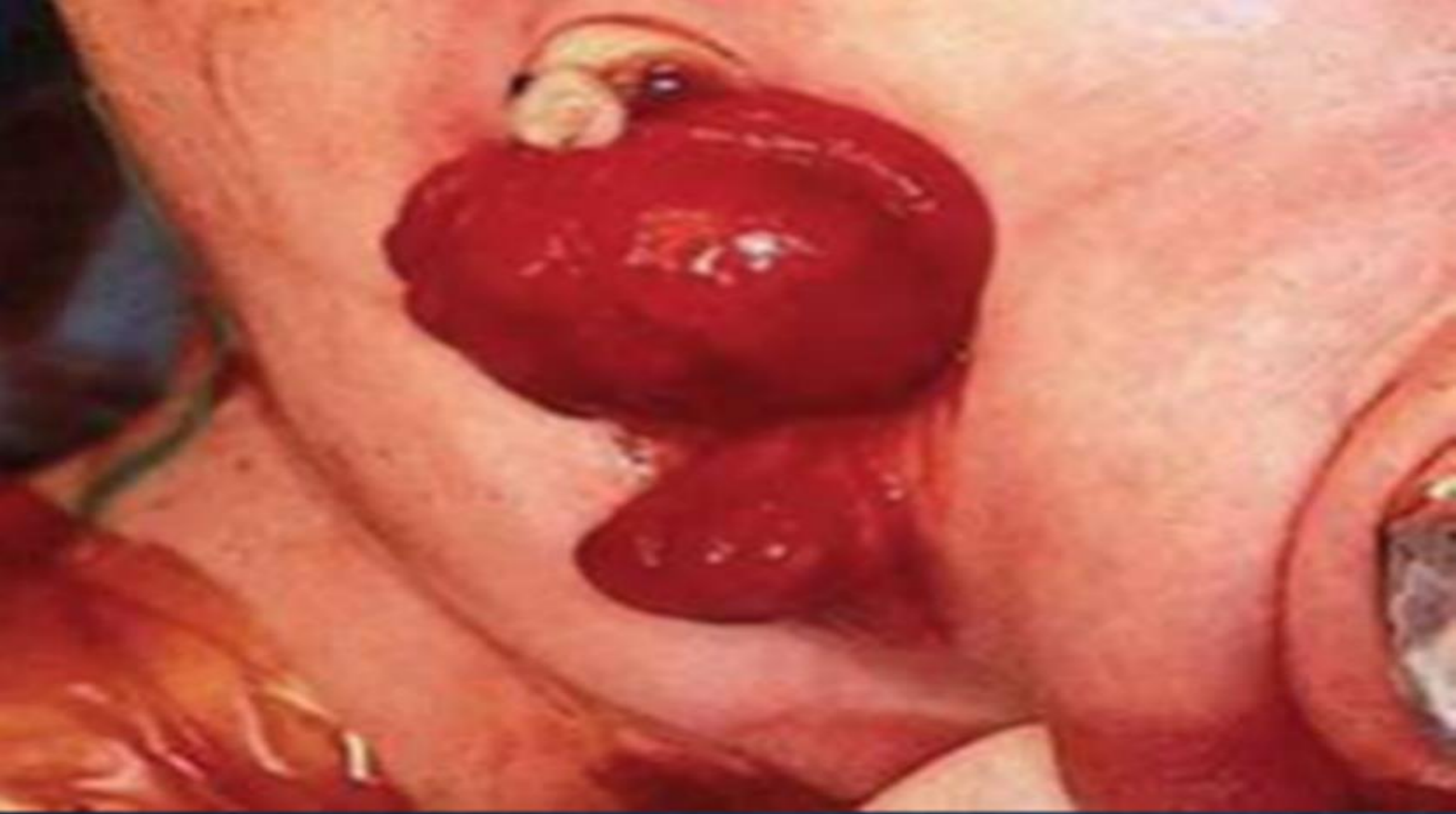
- ✓ Clinical
- ✓ Labs: TBBS, U/A
- ✓ Doppler U/S

9. BLADDER EXSTROPHY – EPISPADIA COMPLEX

BY: DR. MWIKA PETER

CONSULTANT PEDIATRIC SURGEON

EDITED BY NAILA KAMADI



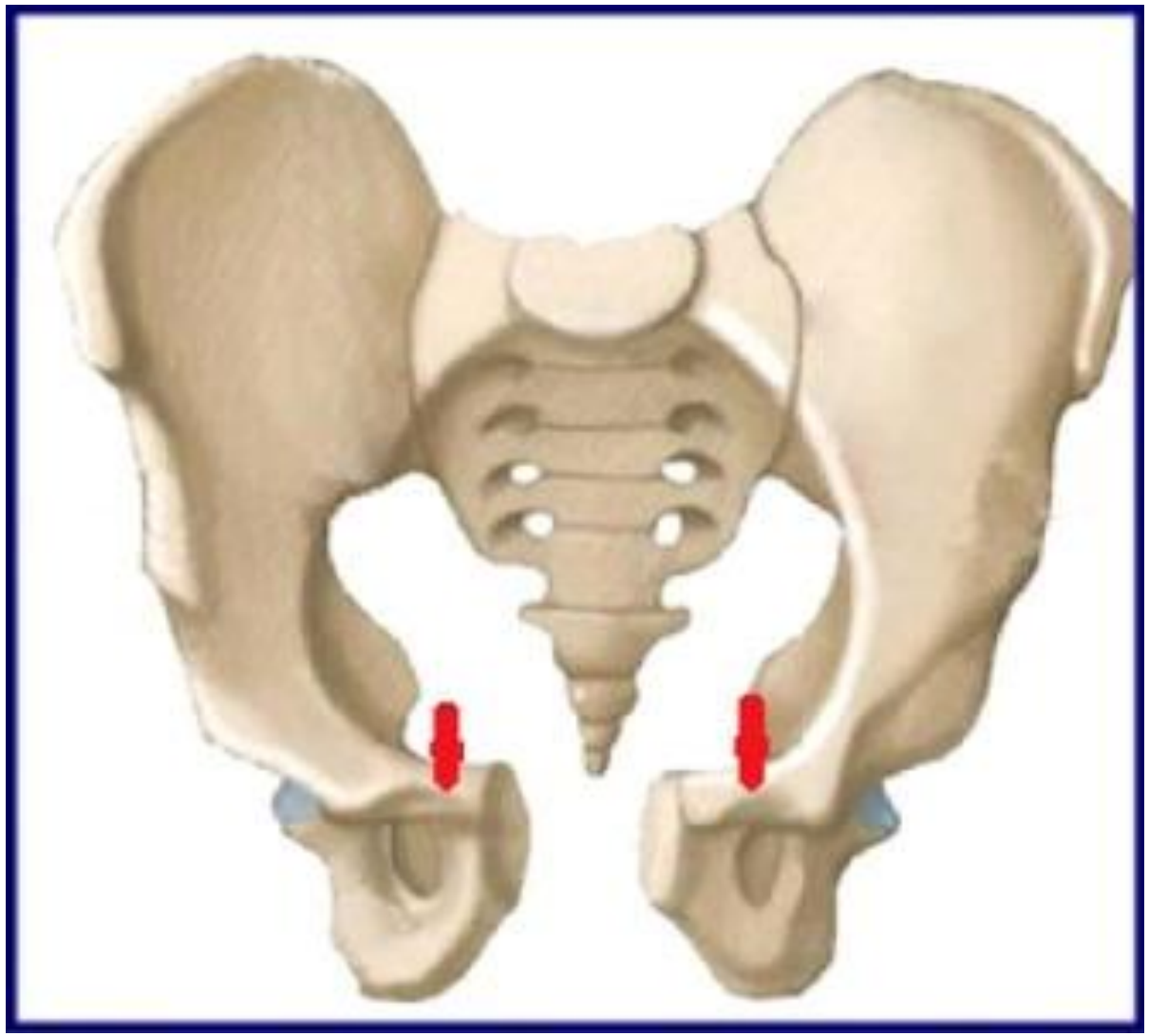
Introduction

- Definition: eversion/ prolapse of the posterior bladder wall through an anterior abdominal wall defect.
- Embryological basis:
  - The bladder & the posterior urethra are derived from the ***urogenital sinus***.
  - Failure of medial migration of mesodermal tissue anterior to the developing urogenital sinus.
  - Improper lower abdominal wall development accompanied by a pubic diastasis.

Epidemiology

- Male predominance; M:F → 2:3 – 6:1
- Incidence is 2.2 per 100,000 live births
- Risk is 500 times more in off springs of affected parents
- Increased incidence in mothers who received large doses of progesterone in the early part of 1st trimester e.g. due to ***threatened abortion***.

Pubic diastasis: external rotation of the posterior pelvis/ iliac wings

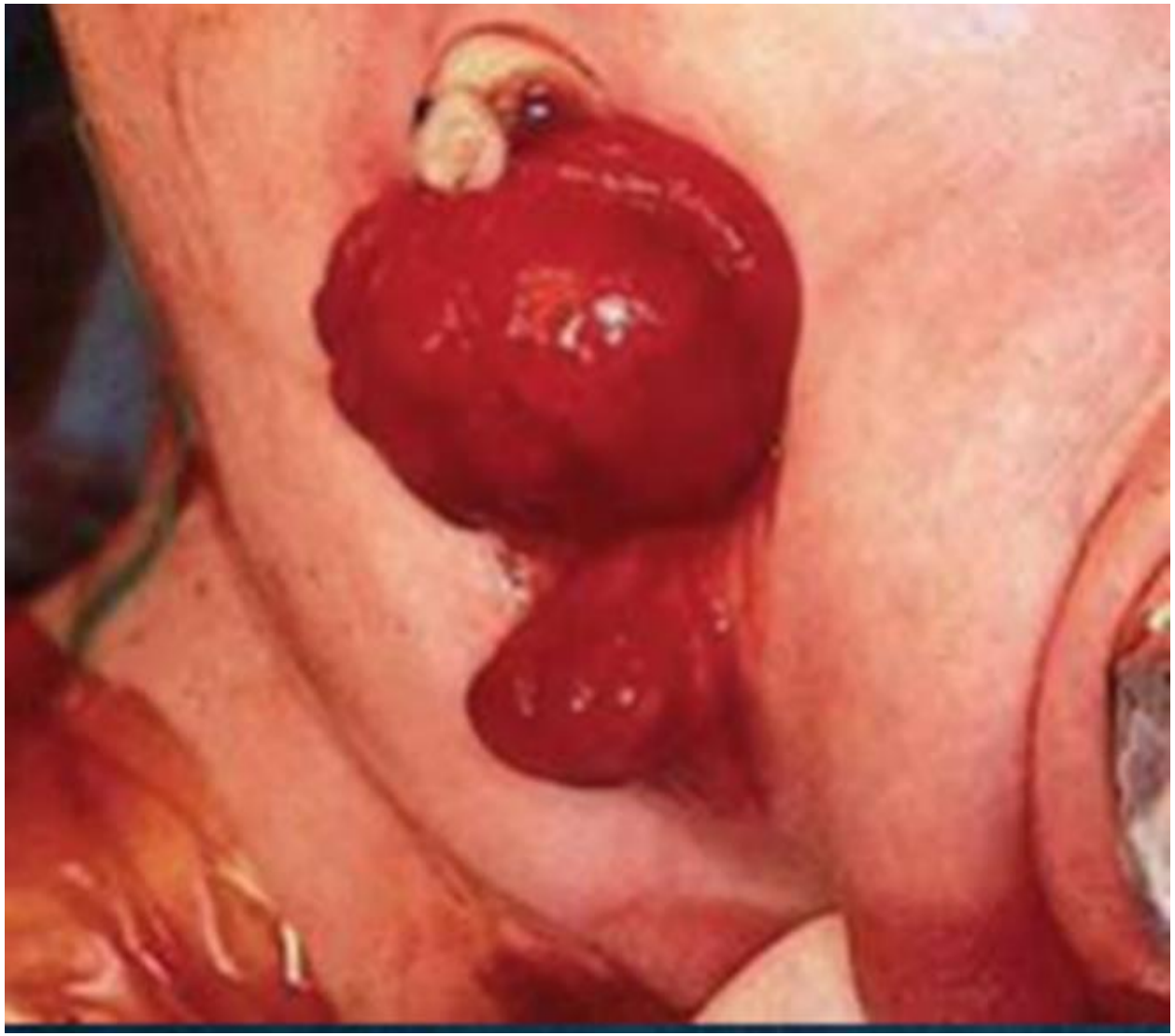


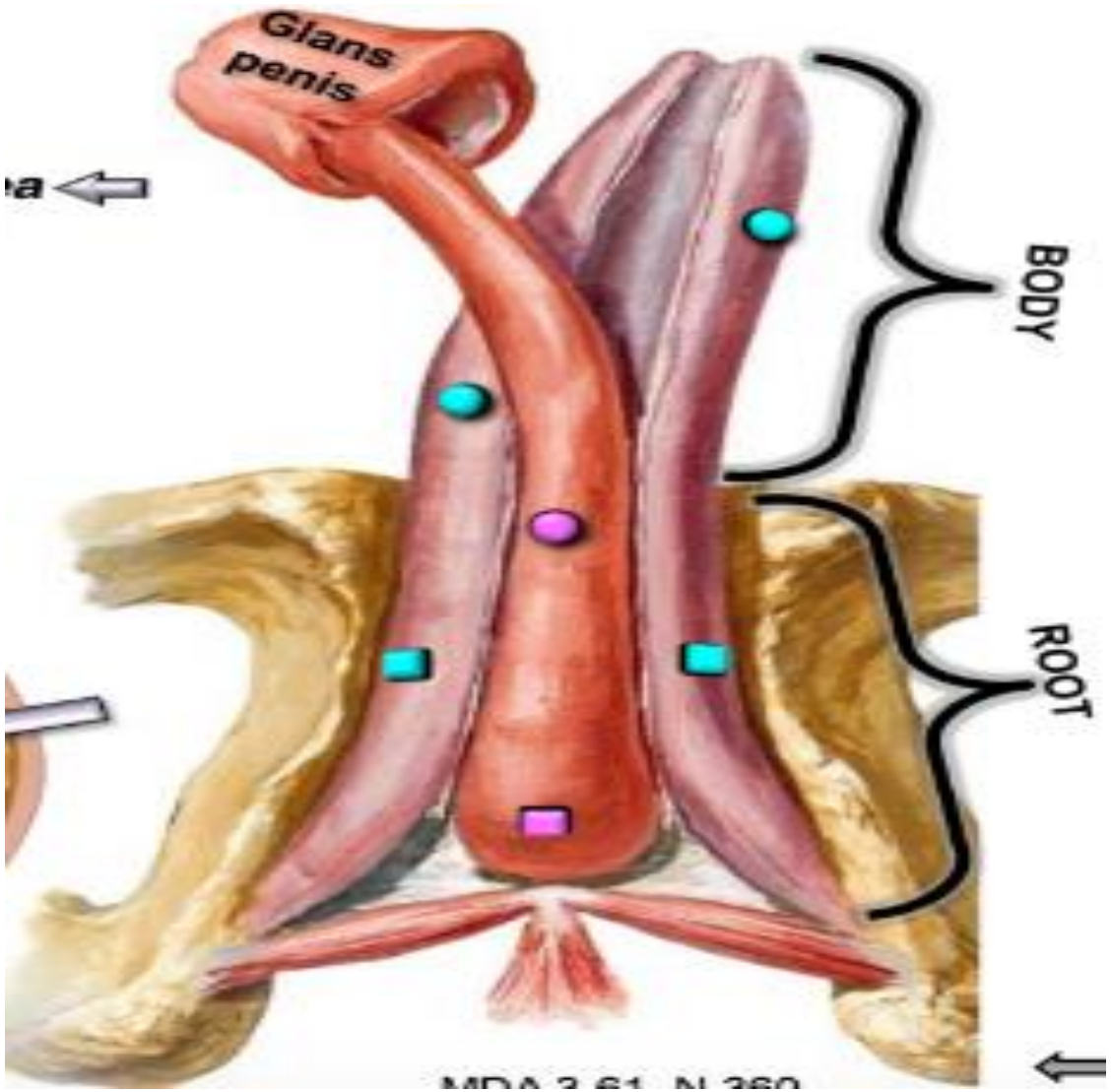
- Implications of pubic diastasis:
  - Bony deformities due to bony defects
  - Waddling gait
  - Short, pendulous penis
  - Associated ***indirect inguinal hernias*** can be seen owing to *persistent processus vaginalis*, *large superficial and deep rings*, & lack of obliquity of inguinal canal.

Male genital defects

- Corporeal bodies separated & triangular in shape.
- Long convex ventral surface and short wedge shaped dorsal surface.
- Varying length of neurovascular supply
- Prostate and accessory sex organs remain unaffected
- Testis remain normal & descend normally without the need for orchiopexy

Male

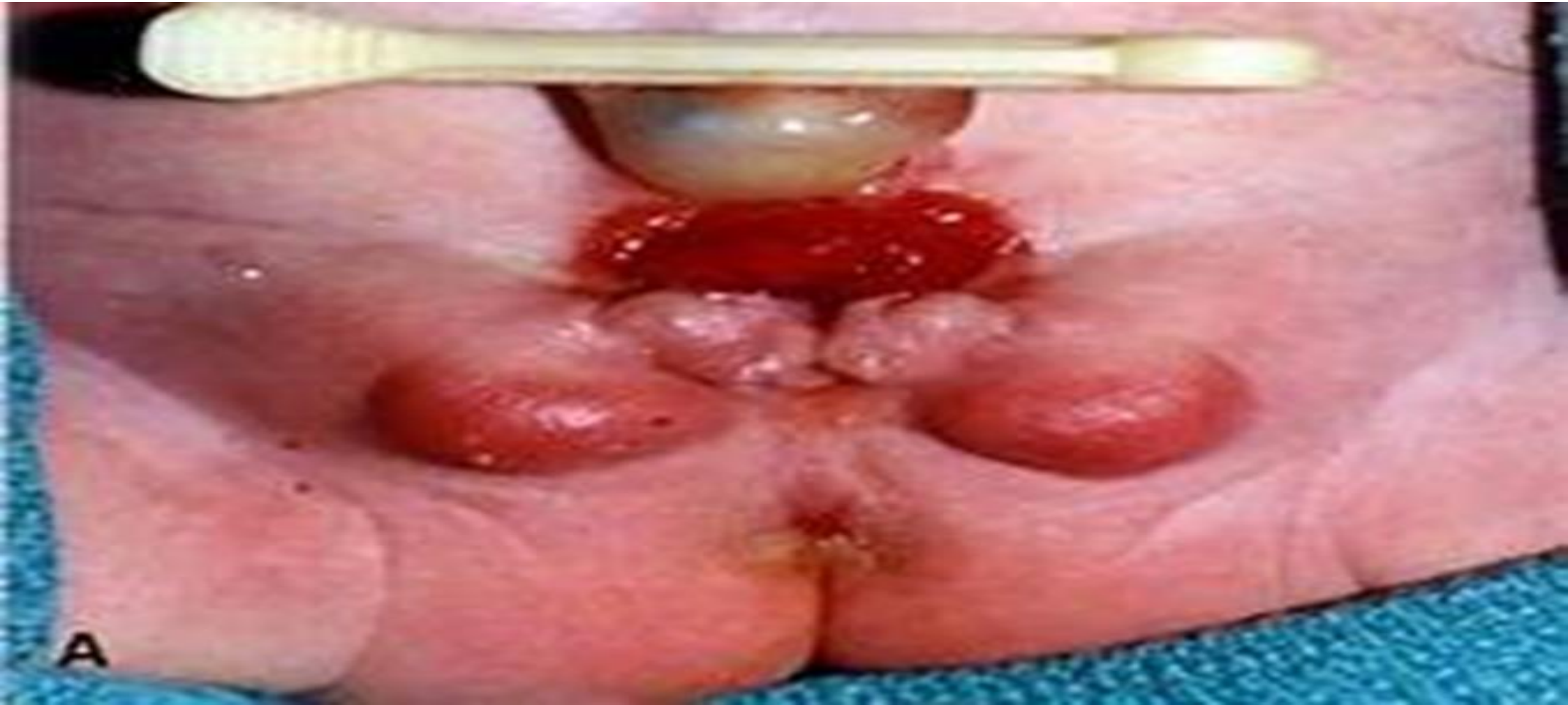




Female genital defects

- Vagina is shorter in length but the caliber is normal.
- Vaginal opening is stenotic.
- The clitoris is bifid & the labia, mons & clitoris are divergent.
- The cervix enters the vagina more superiorly, lying in the anterior vaginal wall near the introitus.

Cont.



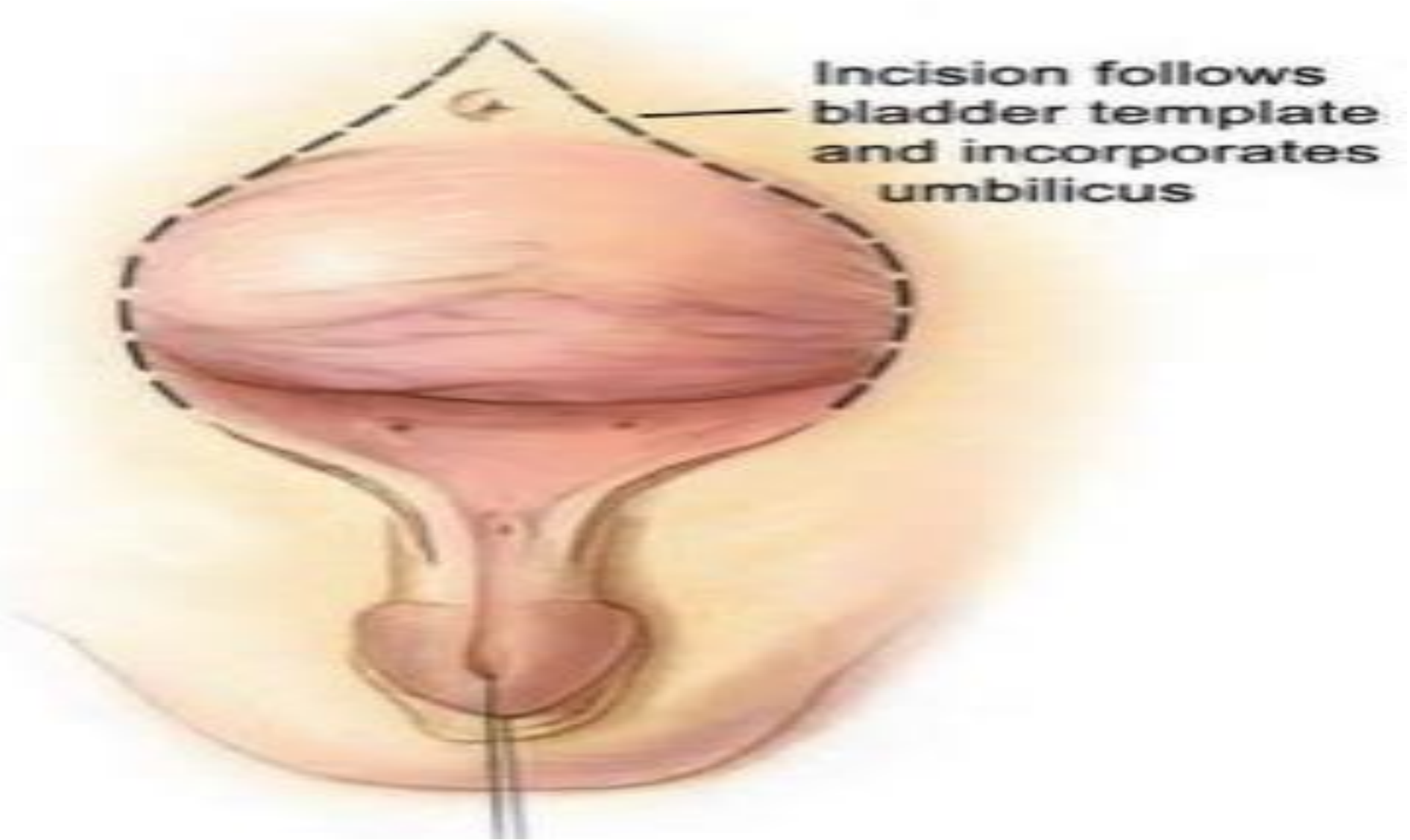
Urinary system defects

- Duplicated collecting system.
- Hypoplastic or absent kidney.
- Pelvic kidney.
- Uretero – pelvic junction obstruction.
- Multi – cystic dysplastic kidney.

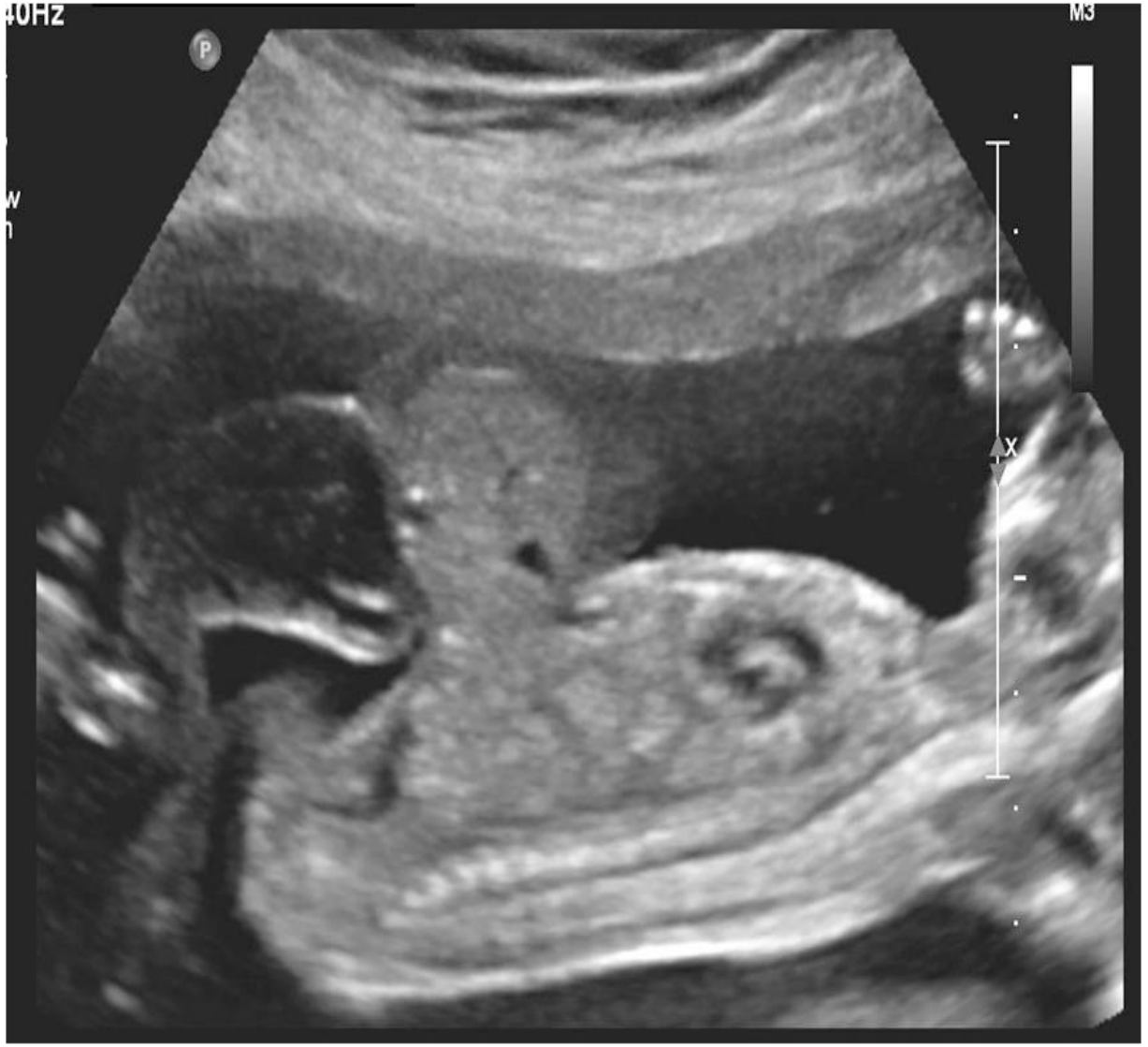
Cont.

- Reflux in exstrophy bladder ***occurs in 100% of patients***, & re – Implantation surgery is required at bladder neck reconstruction.
  - The ureters enter the bladder in an oblique fashion & traverse the bladder mucosa through a distance 5 times its diameter. This is so that with a normal Vesico – ureteric sphincter, reflux into the ureters is impossible unless the bladder raptures. With bladder exstrophy, however, the ureter does not channel through the bladder submucosa & hence there is no sphincter mechanism.

Cont.


Incision follows
bladder template
and incorporates
umbilicus

Diagnosis: anterior U/S



- Mass on the anterior abdominal wall.
- Absence of the 15 – minute bladder cycling after 14 weeks.

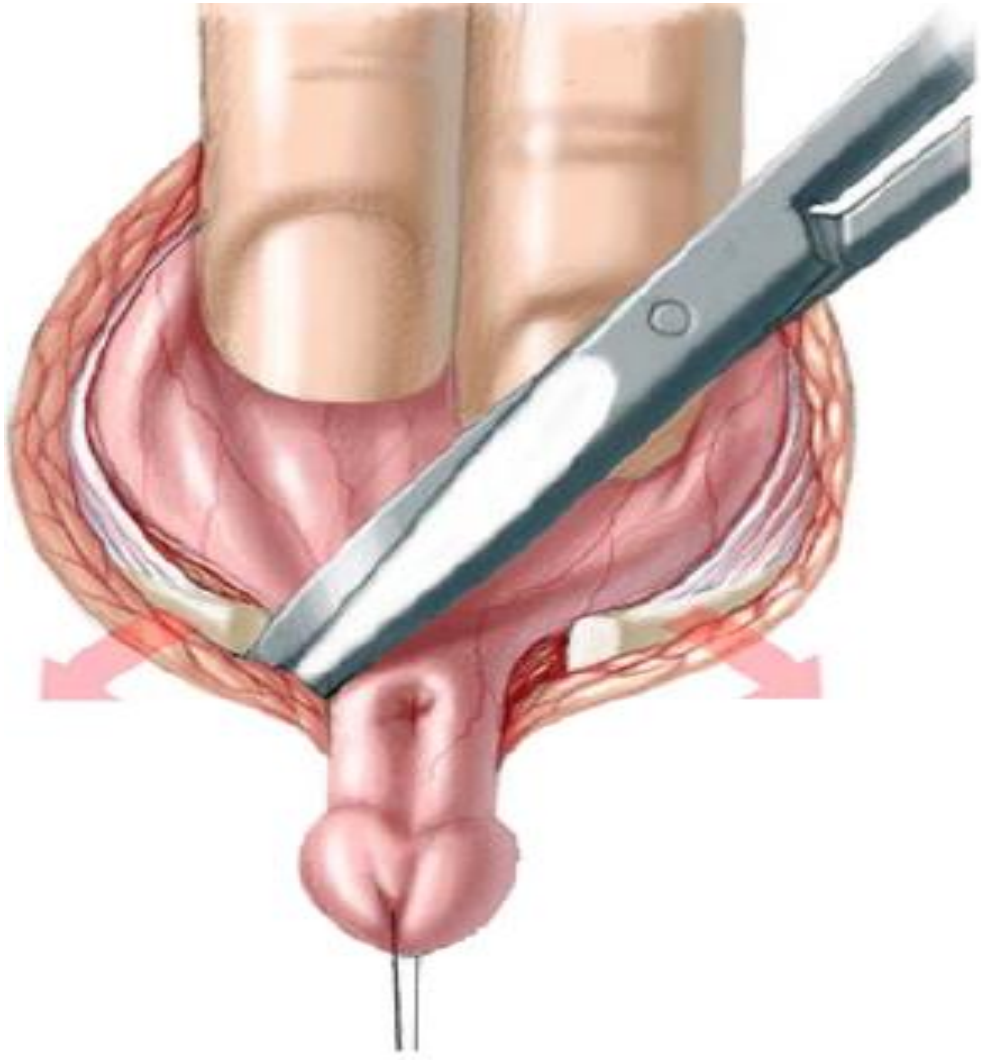
Features on U/S

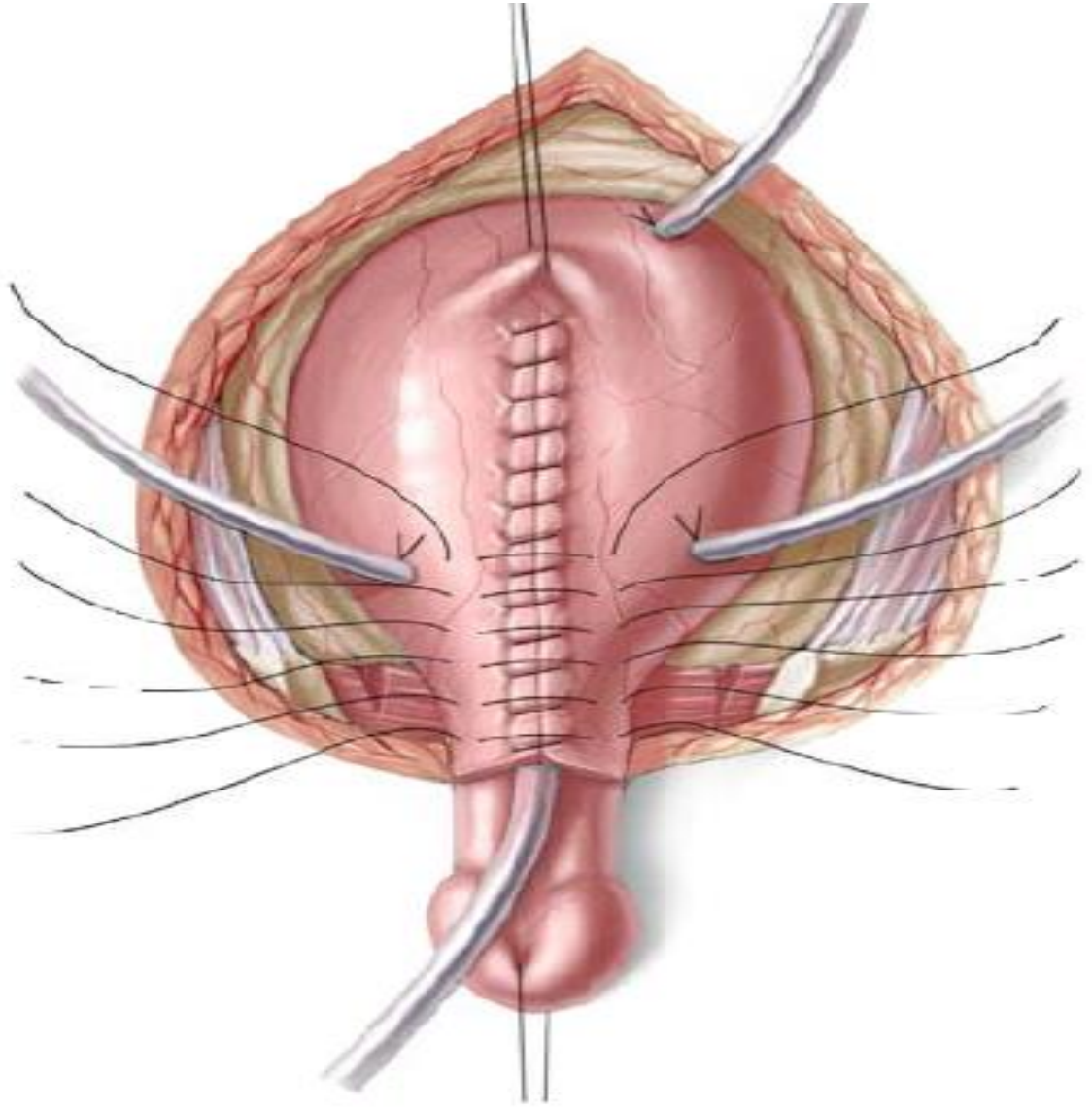
- Absence of bladder filling
- A low – set umbilicus
- Widening pubis ramus
- Diminutive genitalia
- A lower abdominal mass that increases in size as the pregnancy progresses and as the intra – abdominal viscera increase in size

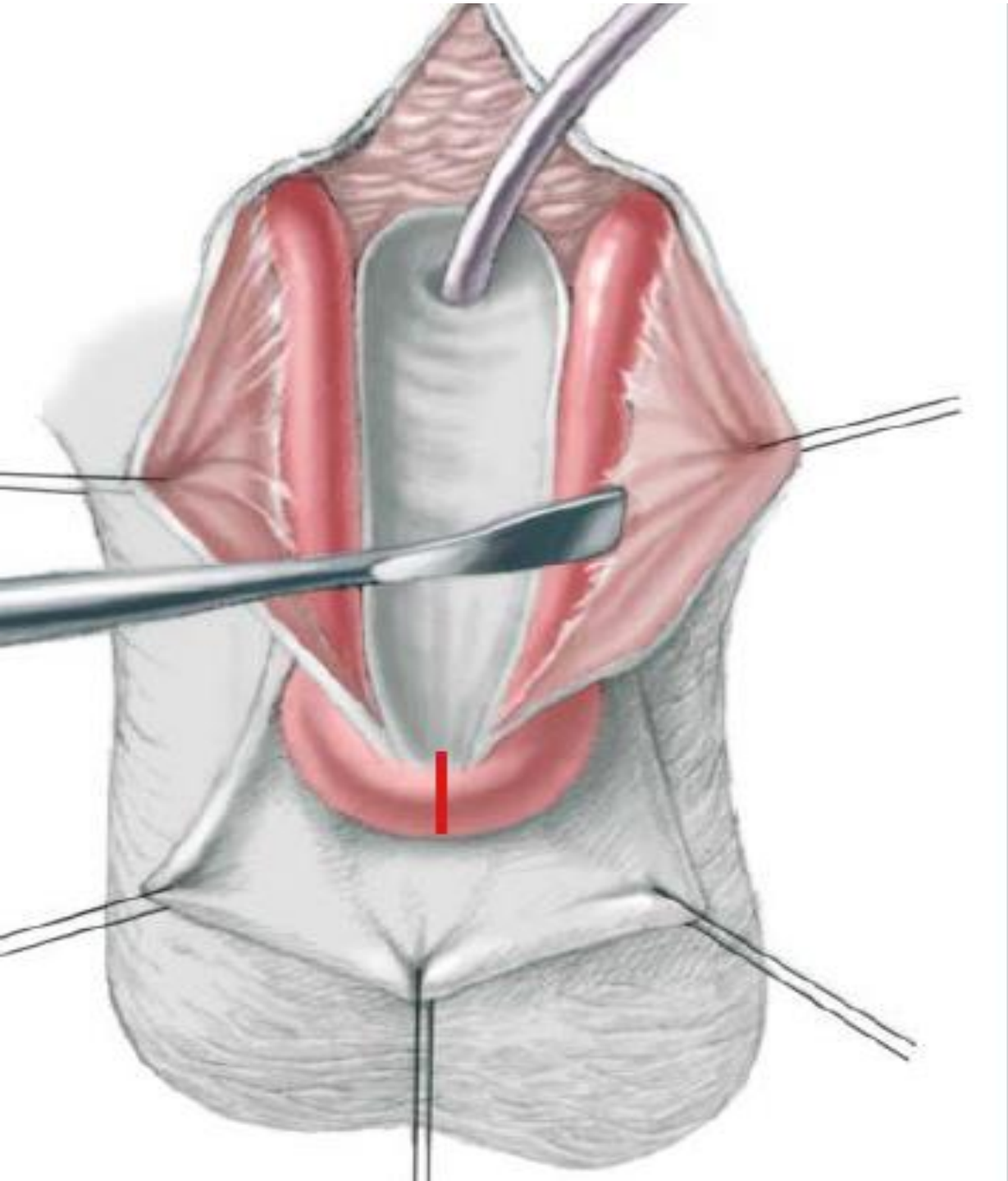
Management

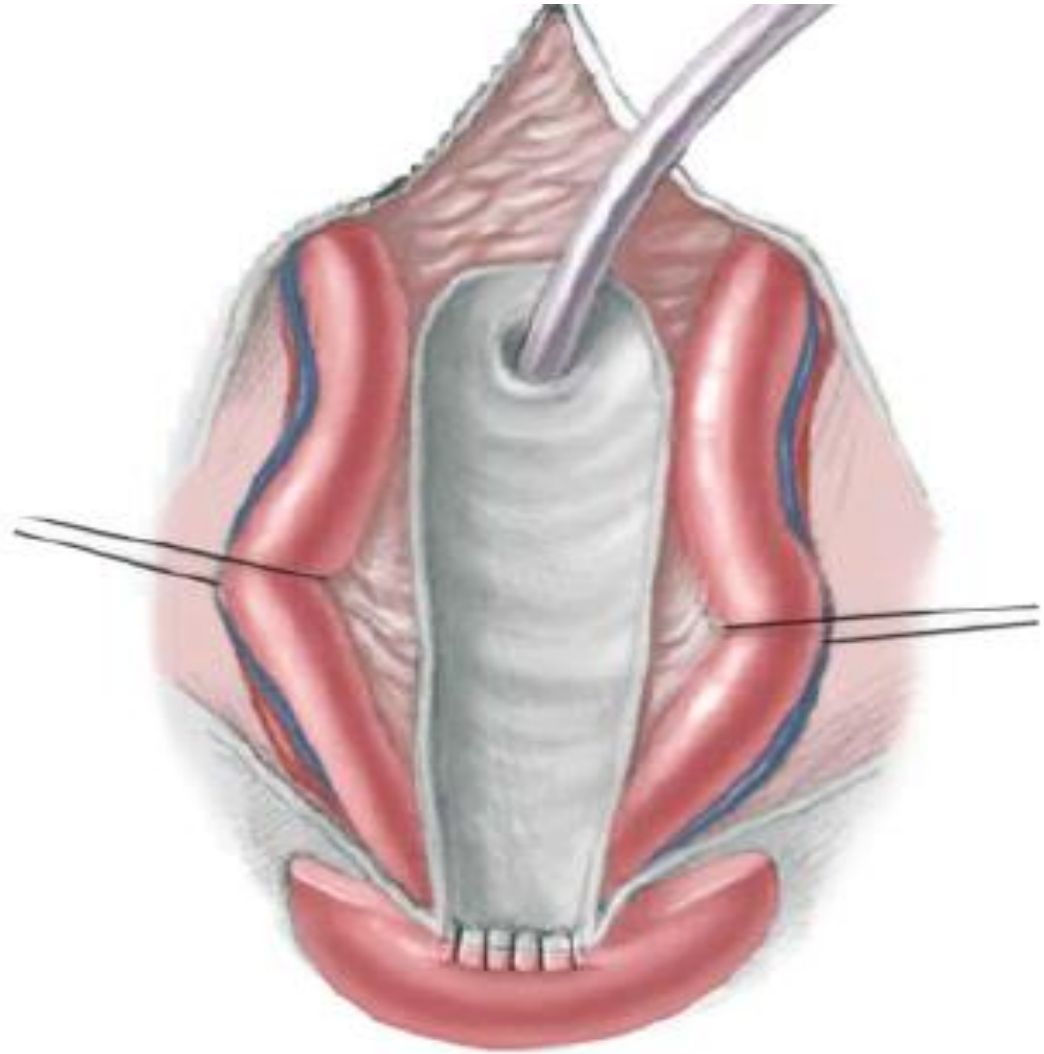
- ***Neonate***: Apply a plastic barrier for mucosal protection (*avoids formation of polyps*). Strap on the upper & lateral sides & leave the lower side open for passage of urine.
- ***Older child***: Apply jelly (*vaseline, Arimis*) to prevent polyp formation.

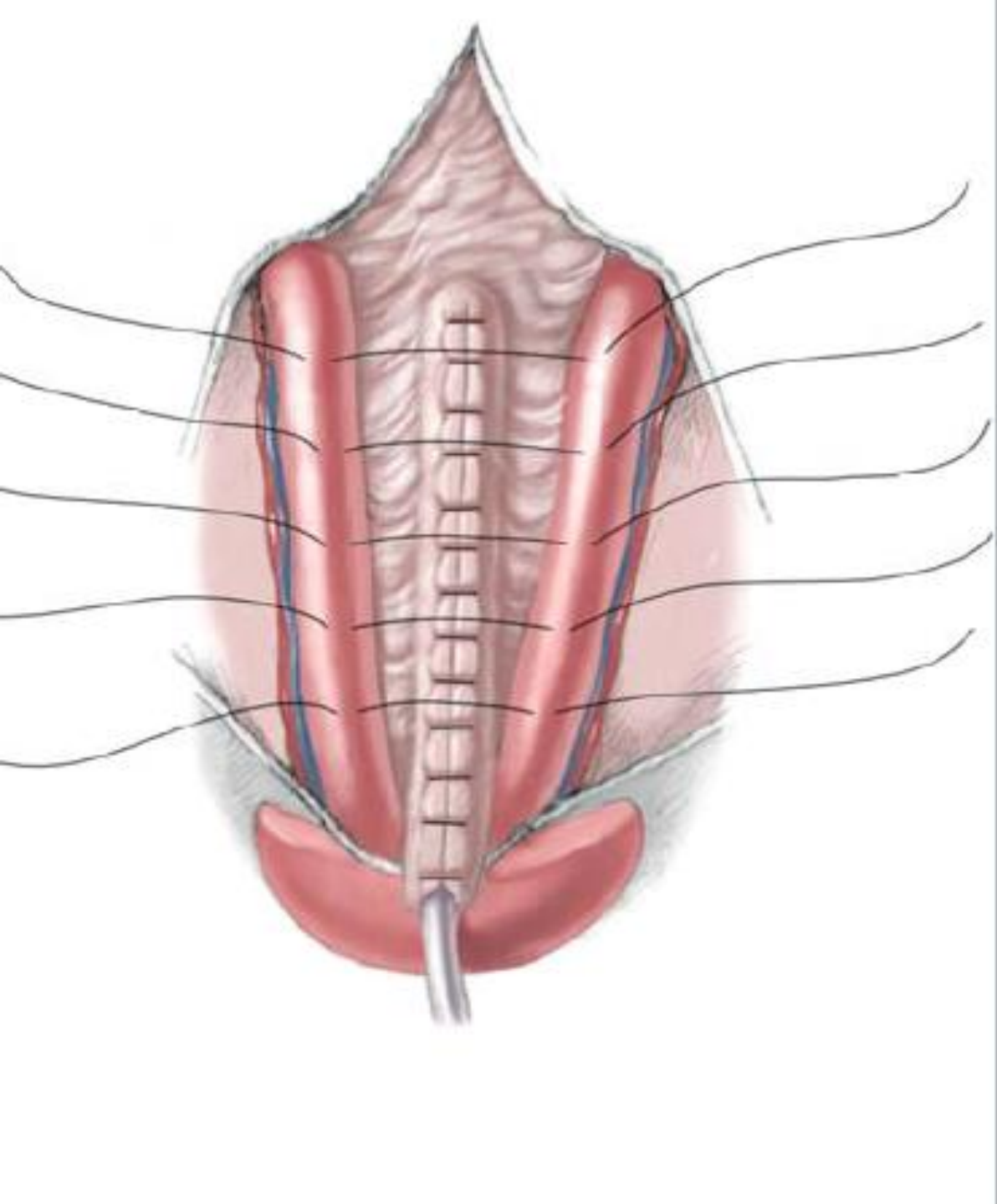
Surgery

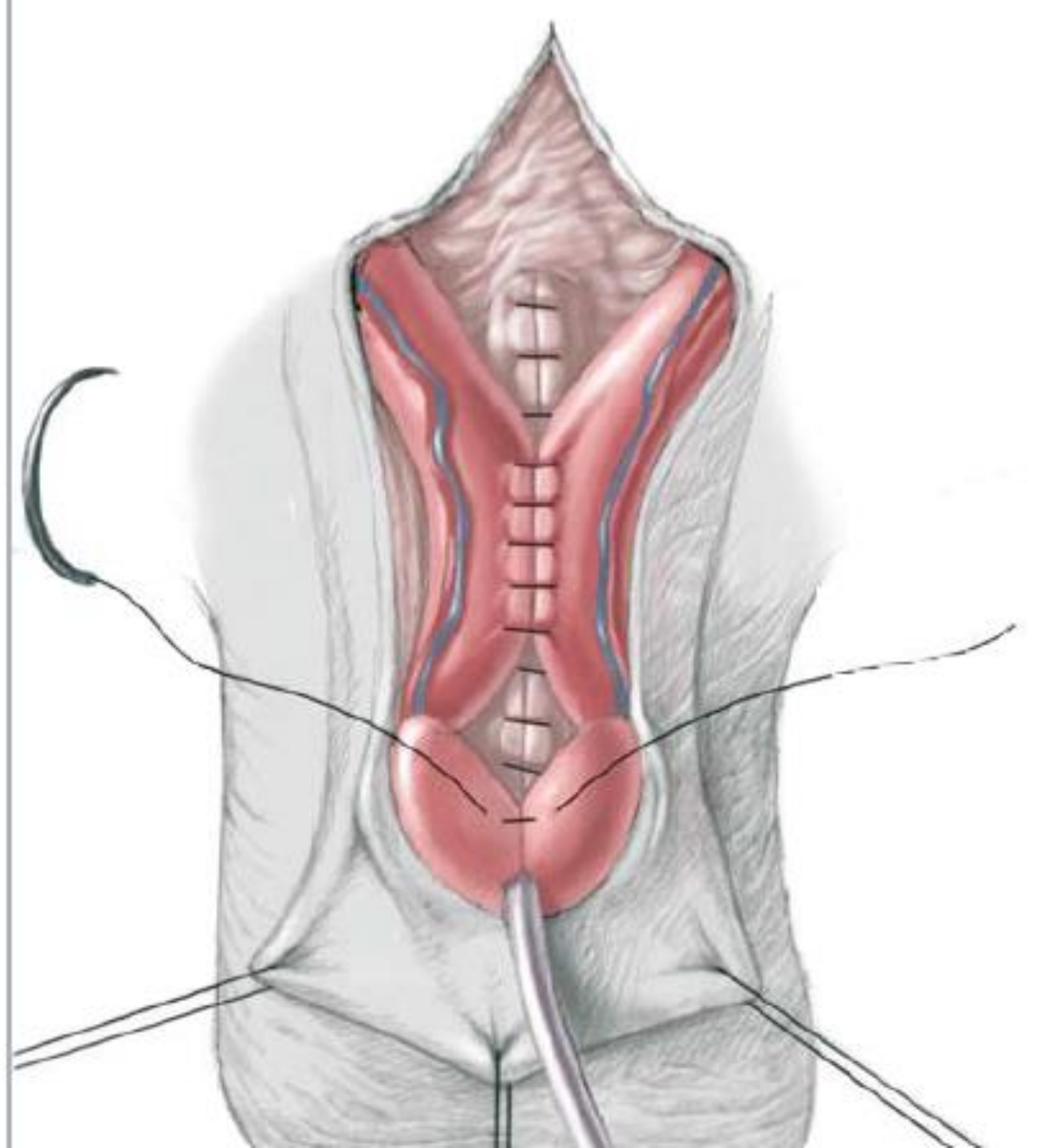










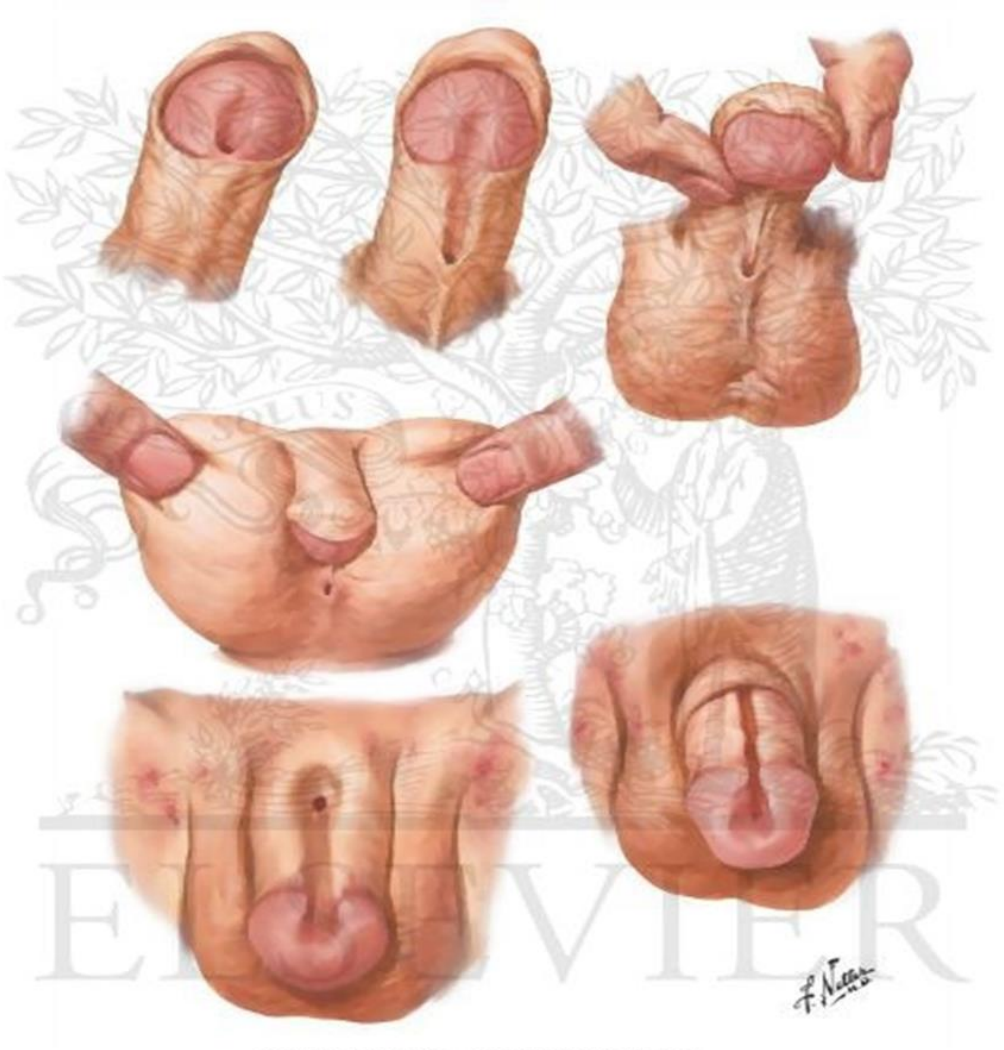



Foreskin
Urethral opening
BEFORE
AFTER

Hypospadia

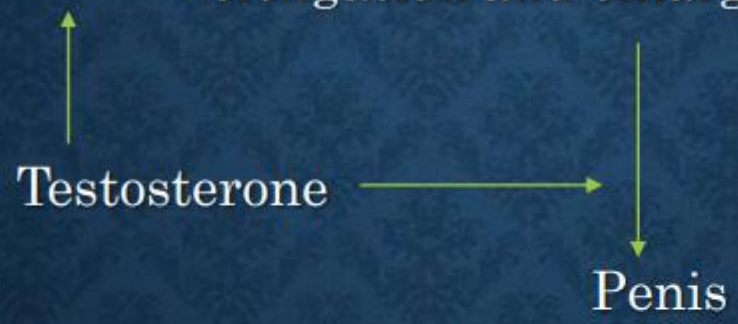
- Definition: this is an *abnormality of the anterior urethral and penile development* in which the ***urethral opening is ectopically located on the ventral aspect of the penis*** proximal to the tip of the *glans penis, which, in this condition is splayed open*.

Cont.



NORMAL DEVELOPMENT

- Genital Tubercle → elongation and enlargement + fusion of urethral folds



Embryology

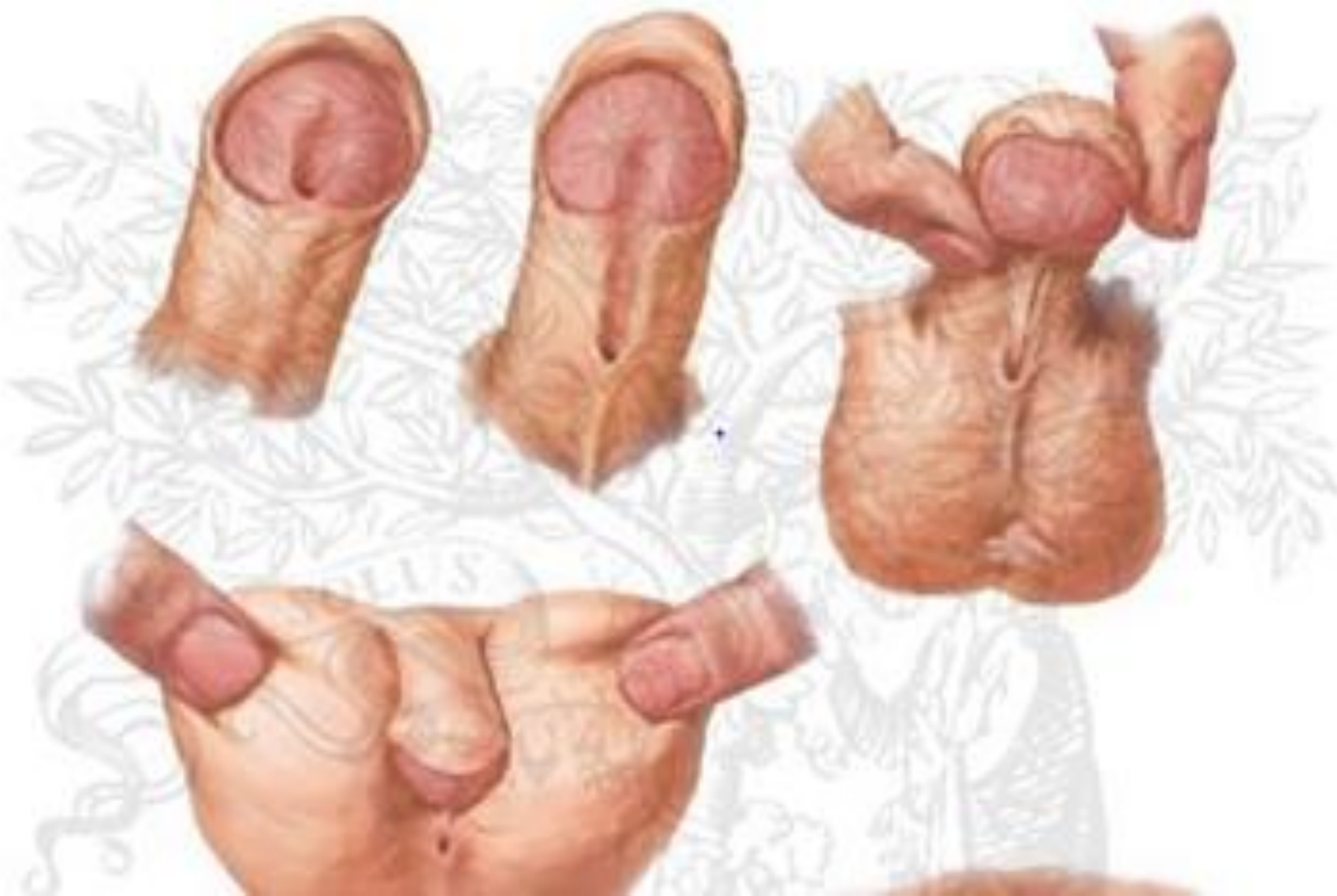
- Proliferation of mesoderm around the cloacal membrane causes the overlying ectoderm to rise up so that 3 structures are visible externally: the **phallus, urogenital folds** & **labio – scrotal swellings.**
- The phallus forms the penis; the urogenital folds form the ventral aspects of the penis (penile raphe); the labio – scrotal swellings form the scrotum.
- Hypospadia occurs when the urethral folds fail to fuse completely resulting in the external urethral orifice opening onto the ventral surface of the penis. It is generally associated with a poorly developed

Incidence

- 1:300 males
- 8% patients have father with hypospadias, 14% have male siblings with hypospadias
- 26% if father or sibling has hypospadias
- 8.5 times more risk in monozygotic twins

Associated with:

- Downward glans tilt
- Deviation of the median penile raphe:
  - Penis can twist: penile torsion
- Ventral curvature: chordee
- Midline scrotal cleft
- Scrotal encroachment onto the penile shaft: transportation

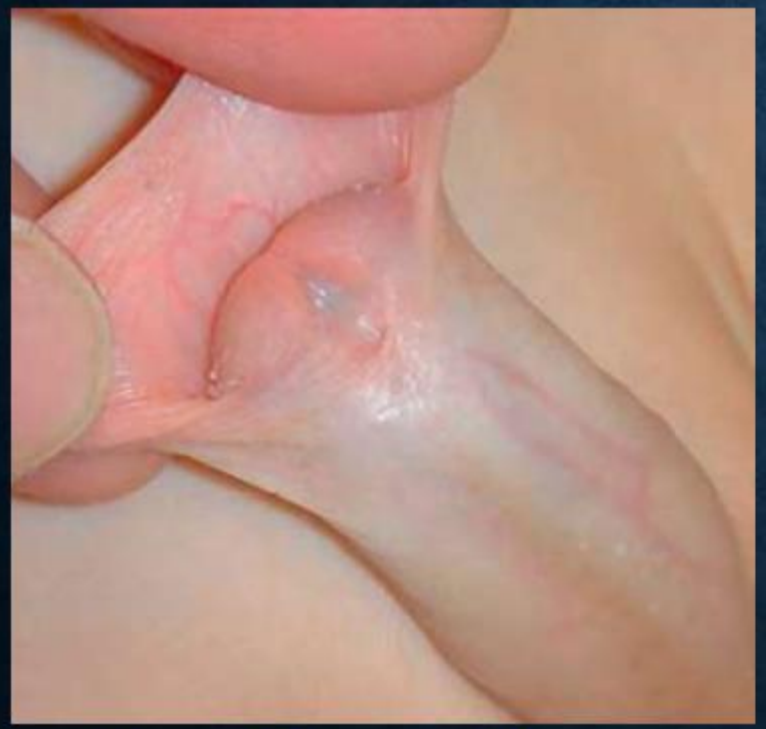


Associated anomalies

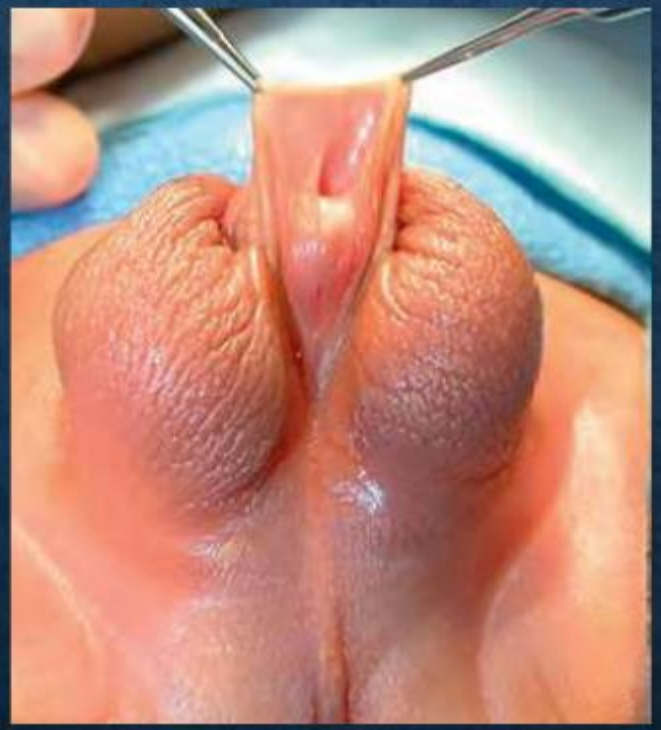
- Undescended testes: 9%
- Inguinal hernias: 9%
- Utriculus masculinus (incomplete mullerian duct regression)
- Cryptorchidism: intersexuality needs to be ruled out esp. in cases of non – palpable testis.

Anatomical types based on the opening of the meatus

- Perineal hypospadia
- Peno – scrotal hypospadia
- Proximal penile hypospadia
- Mid – penile hypospadia
- Distal penile hypospadia
- Coronal hypospadia
- Glanular hypospadia



Coronal Hypospadias



Perineal Hypospadias with
Scrotal transposition

Peno-scrotal
Hypospadias

Principles of surgical management

- Straightening the penis
- Orthotropic urine projection
- Posteriorizing the scrotum
- Penile elongation
- Any child with bilateral undescended testis & a very proximal hypospadia needs to have disorders of sexual development (intersex disorders) ruled out by being investigated further: U/S, biopsy, genitogram, karyotyping, FISH


thank you!

10. POSTERIOR URETHRAL VALVES

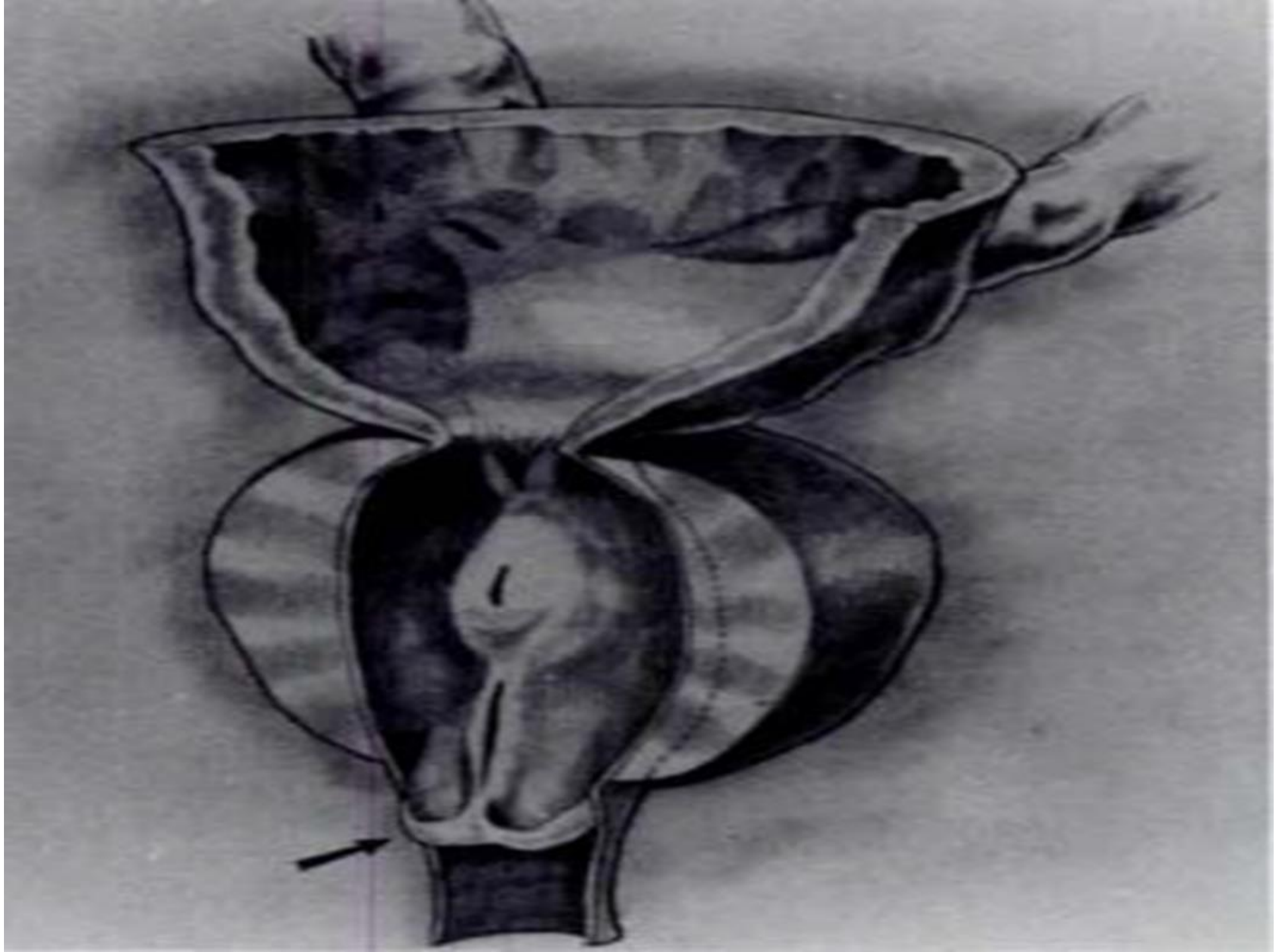
LEVEL VI

2019

BY: DR. MWIKA M. P.

Introduction

- Parts of the urethra:
  1. Prostatic urethra
  2. Membranous urethra
  3. Bulbar urethra
- PUVs are found in the ***prostatic urethra*** where they only allow retrograde flow of urine & not anterograde flow hence causing BOO.



Embryology

- The posterior urethra develops from the ***urogenital sinus (which develops from the cloaca).***
- The PUVs are a result of an aberration of the normal development of the posterior urethra.
- They are ***remnants of fused mesonephric ducts***.

Incidence

- The ***MC type of obstructive uropathy*** leading to > 50% of cases of childhood ESRD.
- 1 in every 5000 – 8000 male births.
- Causes ***10% of prenatally diagnosed hydronephrosis***.
- ***50% of such cases resolve spontaneously*** & the rest require surgical intervention.

Gunn TR, Mora JD, Pease P. Antenatal diagnosis of urinary tract abnormalities by ultrasonography after 28 weeks' gestation: incidence and outcome. Am J Obstet Gynecol 1995 Feb;172(2 Pt 1):479-86.

http://www.ncbi.nlm.nih.gov/pubmed/7856673

Pathophysiology

- Due to the presence of the PUVs primitive urinary tissues, at a critical time in organogenesis, ***mature in an abnormal environment of high intraluminal pressures and organ distention.***
- This results to **permanent maldevelopment** & **long lasting functional abnormalities** of the kidneys, ureters & bladder.
- ***These changes persist despite relief of the primary obstruction***.

Cont.

- Obstruction by PUVs → hypertrophy & hyperplasia of detrusor muscle → urine accumulation → ↑ intraluminal pressure above intramural pressure → ↓ perfusion of bladder musculature (***NB: Perfusion pressure for most viable tissues is 30 – 32***) →

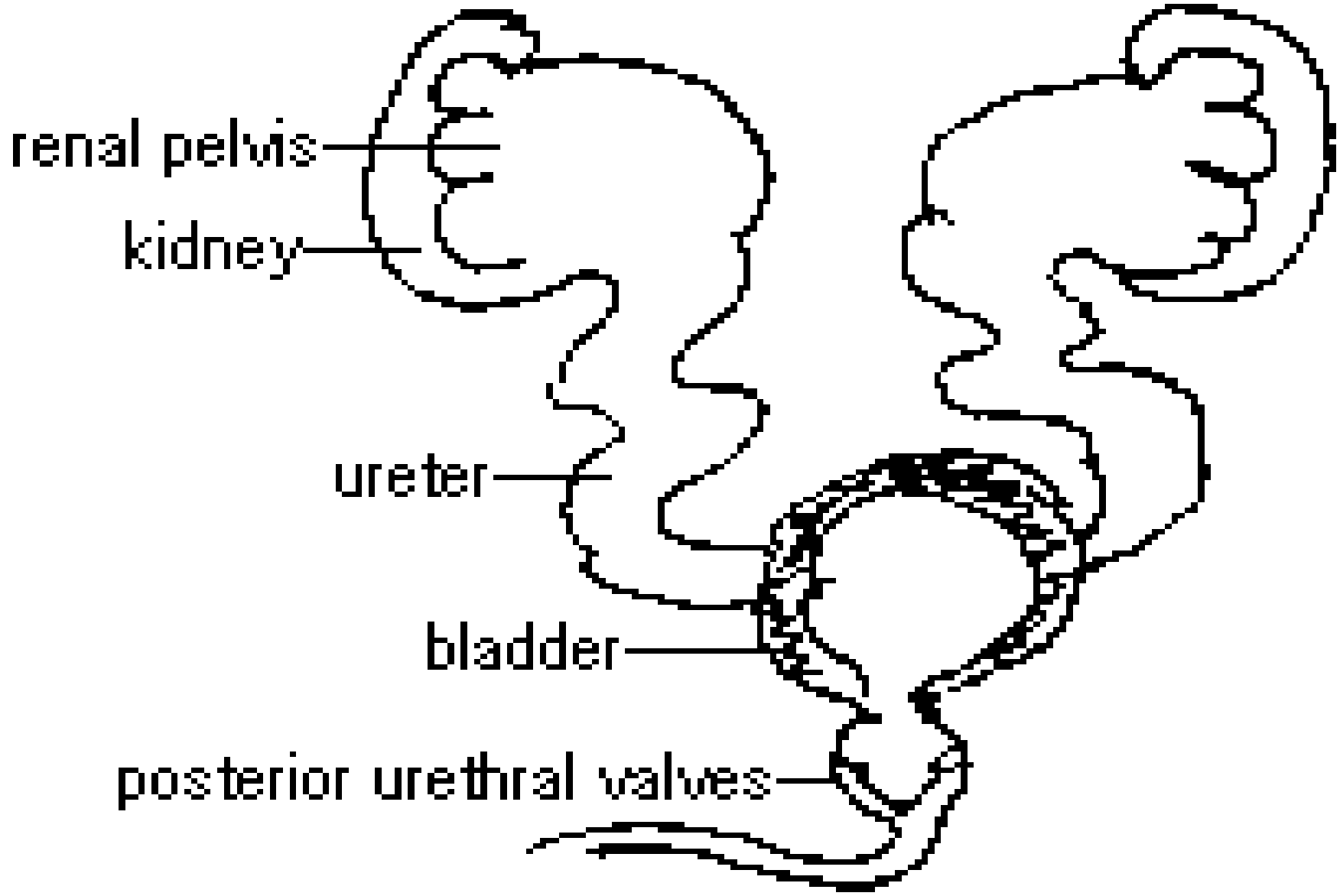
  inflammation → fibrosis in muscle →

  – → ↓ compliance

  – → Trabeculations & diverticuli

Cont.

- The same process occurs in the ureters resulting in ***hydro – ureter*** as well as in the renal pelvis resulting in ***hydronephrosis***.
- The high pressures are transmitted to the nephrons resulting in ischemia & fibrosis of the prostaglandin – producing interstitium. The higher intraluminal pressures in the proximal convoluted tubule impair filtering at the bowman's capsule. This activates the RAAS resulting in ***renal HTN***.


renal pelvis
kidney
ureter
bladder
posterior urethral valves

Functional alterations in the kidney

- Pressure changes: obstruction leads to ***rise in ureteric & intrarenal hydrostatic pressure*** which reflect back to the renal tubules → Reverse peristalsis → Dilatation of collecting system.
- Changes in tubular function → urinary obstruction leads to prostaglandins production which ***blunt cellular response to ADH leading to polyuria & electrolyte imbalance***.
- Polyuria → hypotension → poor renal perfusion → accelerated renal failure.

Mechanical damage

- Flattening & atrophy of the renal tubules which ***lose their brush border***.
- Hyalinization & sclerosis of glomeruli
- Infiltration of interstitium by inflammatory cells from recurrent infection.
- Above leads **to loss of renal parenchyma and reduced GFR.**

Bladder dysfunction

- Seen in 60 – 75% of patients.
- Voiding resistance leads to ***detrusor hypertrophy and hyperplasia***.
- High intraluminal pressures & thickened bladder wall leads to ***poor perfusion & ischemic changes within the bladder*** wall which results in ↑ collagen deposition.
- ↑ in connective tissue (collagen) ***limit bladder compliance*** during filling & results in bladder wall weakness.

3 groups of vesical dysfunction have been described

1. Detrusor hyperreflexia (29%)
2. Hypertonic & poor compliant bladder (31%)
3. Myogenic failure & overflow incontinence (40%)

Pop off mechanisms

1. ***Patent urachus***
2. Large bladder ***diverticulum***
3. ***Urinoma*** or ***ascites*** from calyceal rupture
4. ***Valves Unilateral Reflux Dysplasia (VURD)***: results in very poor or non – function of the kidney on the refluxing side (usually the left) with a relative sparing of renal function on the contralateral, non – refluxing side.

VUR

- Secondary to the BOO & co – existent bladder dysfunction. Occurs in 60% of children with PUVs.
- Unilateral in 40%. 15% of patients will demonstrate unilateral high grade VUR with non – function of the ipsilateral kidney.
- Approximately 50% of patients have VUR on the initial MCUG. (*Defoor W, et al. J Urol, 2008).*

Cont.

- Following valve ablation the severity of VUR may decrease or resolve completely in 25 – 50% of cases. This happens in 30% by 3 months & in 80% by 6 months.
- Management: Low dose prophylactic ABs

Hydronephrosis

- Virtually present in all patients.
- Bilateral in 90% of cases.
- 2^0 to urethral obstruction with subsequent high intra – vesical pressure.
- Improves gradually but significantly with relief of obstruction.

Renal Tubular Dysfunction

- High ureteral pressures affect the most distal part of nephron first. There may no significant renal failure.
- Urine concentrating abnormality → Nephrogenic DI→ hydronephrosis, vesical dysfunction dehydration
- Metabolic acidosis with hyperkalemia.
- Salt wasting due to reduced Na^+/K^+ ATPase activity

Low GFR

- Goal of management is to maximize and preserve GFR
- Raise serum creatinine 50% of the cases. Not predicative of subsequent renal function
- GFR at 1 year s/p valve resection is predictive of the final GFR. Nadir creatinine of <70 μmol predicts long term good prognosis.
- Normalizes within 6 months in some

Cont.

- Decreasing renal function may be due to renal dysplasia, hydronephrosis, UTI (vesical dysfunction), hyperfiltration - glomerulosclerosis
- Rate slow and steady from 140-440μmol/l

  at 45μmol/l/year
- Rate of deterioration very rapid after serum creatinine >440μmol/l.

Cont.

Renal dyplasia

- Single most important abnormality that determines GFR, only aspect that clinician has no impact.

Urinary ascites

- May be due to:
  - —Calyceal perforation
  - —Filtration through the urinary tract
    - Decompressive mechanism
    - Improves prognosis

Post – natal presentation

- Neonate:
  - Respiratory distress (lung hypoplasia)
  - Abdominal distension
  - Difficult voiding
- Infants:
  - FTT
  - Urosepsis
  - Poor urinary stream
  - Straining or grunting while voiding
- Older boys:
  - UTIs
  - Enuresis
  - Voiding dysfunction

Other associated anomalies

- Undescended testis
- Inguinal hernia
- Tracheal hypoplasia,
- Patent ductus arteriosus,
- Total anomalous vein drainage,
- Mitral stenosis,
- Scoliosis and lower extremity deformations,
- Imperforate anus

Investigations

- Renal U/S
- MCUG
- Radionuclide scans:
  - DPTA (diuretic renogram) → patency of the urinary tract differential GFR
  - DMSA → renal scars & differential GFR
- Urodynamic studies: bladder storage & emptying properties

Cont.

- Urinalysis: M/C/S
- U/E/Cr
- Serum bicarbonate
- Schwartz formula for estimating GFR
  - Estimated GFR (mLs/min/1.73m^2) = $\frac{\text{Height X K}}{\text{Plasma Creatinine}}$
  - K vales:
    - Adolescent boys: 62
    - Children: 48
    - Term babies: 40
    - Preterm: 29

Diagnosis

- In utero: fetal imaging
- Newborn
- Infant
- Older child

Antenatal U/S

- Distended thick – walled bladder
- Bilateral hydro – uretero – nephrosis in a male fetus.
  - Assessment of level of amniotic fluid: oligohydramnios
  - Accuracy in diagnosis: 50%

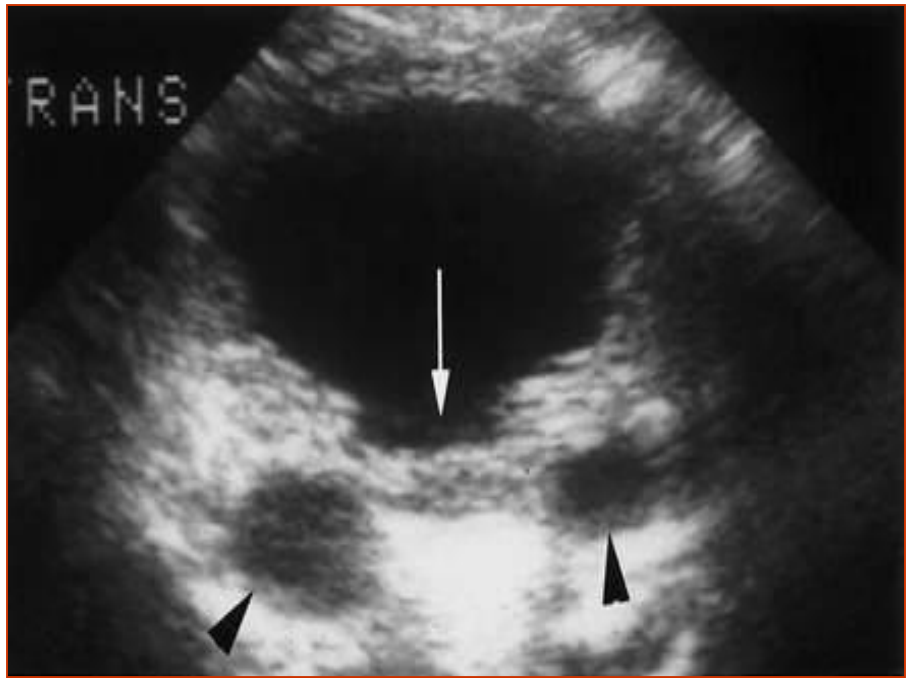
Abbott JF, Levine D, Wapner R. Posterior urethral valves: inaccuracy of prenatal diagnosis. Fetal Diagn Ther 1998; 13:179

Fetal MRI

- Supplements U/S
- Accurate in assessing ***oligohydramnios*** & ***total lung volume***.

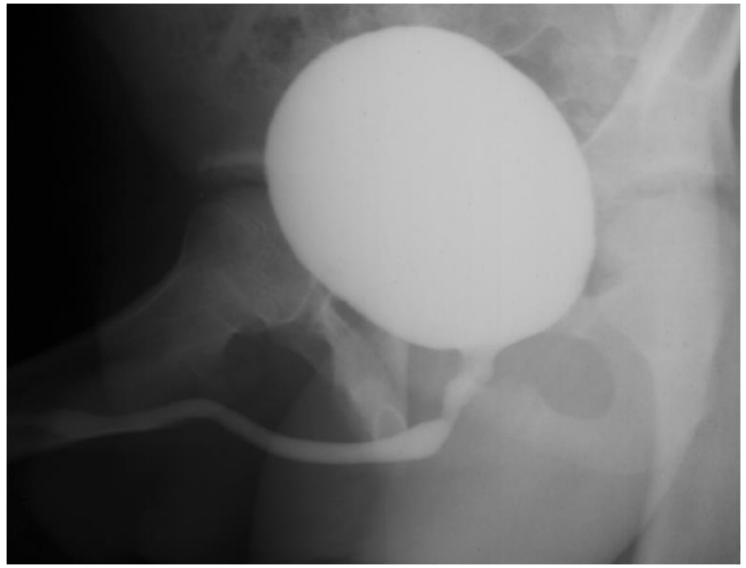
Post natal diagnosis

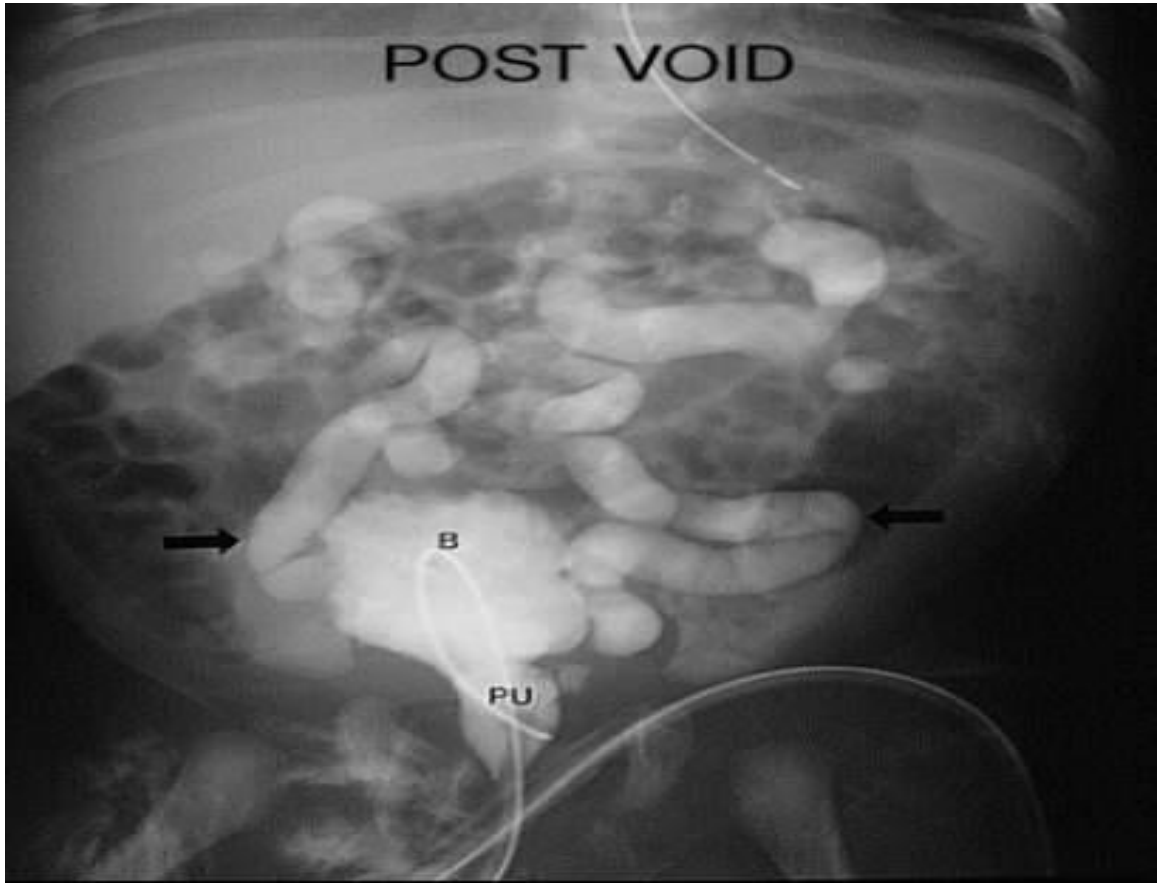
- Dilated posterior urethra: ‘keyhole sign’
- Thickened bladder wall
- Dilated upper tracts
- Assessment of renal parenchyma: cortical thinning in a newborn boy
- Urinary ascites


TRANS

MCUG

- Dilated proximal urethra
- Bladder trabeculations
- Diverticuli
- VUR
- Post – void volume (Normal: ___)





Posterior urethra is >2.5 times the diameter of the anterior urethra.

Grading VUR on MCUG

1. Reflux into a normal ureter.
2. Reflux into a normal pelvicalyceal system with no dilation.
3. Reflux into a mildly dilated ureter & pelvicalyceal system
4. Reflux into a moderately dilated ureter & pelvicalyceal system with obliteration of the sharp angle of the fornices
5. Gross dilation & tortuosity; loss of papillary impression

Radionuclide studies

- ***Dynamic radionuclide scans*** assess renal excretory function.
- Radiotracers: ***Technetium 99mTc diethylene – triamine penta – acetic acid (DTPA) or 99mTc mercapto – triglycyl – glycine (MAG-3)***
  - DTPA assesses glomerular filtration
  - MAG3 assesses tubular secretion
- Two phases involved …….

Cont.

- ***Parenchymal phase:*** Radioisotope is injected IV & renal cortical uptake is measured during the first 2 - 3 mins.
  - Split renal function is assessed quantitatively
- ***Excretory phase*** – IV Lasix administered and excretion of isotope from kidney is measured.
  - In a dilated system, if washout occurs rapidly after diuretic administration (< 15 mins), the system is not obstructed.
  - If clearance is delayed beyond 20 minutes, the pattern is consistent with obstructive uropathy

MANAGEMENT: Antenatal treatment

- Decreased UOP can result in oligohydramnios which leads to ***lung hypoplasia***
- Intrauterine attempts have been made to treat fetus with PUV;
  - Vesicoamniotic shunt: 1ST attempt 1982; has a high failure rate.
  - Fetal valve resection
- The rationale is that early decompression of the fetal renal tract will allow improved survival with:
  - Preservation of renal function
  - A reduction in the respiratory compromise
- There remains no clinical consensus about the efficacy and use of prenatal intervention.

Post – natal management

- Complex & difficult
- Depends mainly upon the ***renal status*** & ***presence of urinary infection***.
- Priority is to ***drain the bladder*** → urethral 5 FG catheter (preferable non – ballooned) or supra – pubic drainage.
  - Ballooned urethral catheter may cause bladder spasms (Rx. ***Oxybutinin***). This may involve the Vesico – ureteral junction resulting in obstruction.
  - The Ballooned urethral catheter always falls into the posterior urethra since the bladder neck is distended.

Note

- Urethral catheter causes painful bladder spasms (Rx. Oxybutynin & analgesics). The spasms may irritate the rectum resulting in constipation.
- Supra – pubic catheter: results in disuse atrophy as the bladder is not expanding and contracting. ***NEVER PUT AN ELECTIVE SUPRA – PUBIC CATHETER IN A PEDIATRIC PATIENT.***
- ***Vesicostomy*** can be an alternative to supra pubic catheterization. The anterior abdominal wall acts as a sphincter mechanism.

Cont.

- Correct ***fluid & electrolyte abnormalities.***
- ***Respiratory support*** in those with lung hypoplasia
- ***BSAs*** for those who present with Urosepsis.

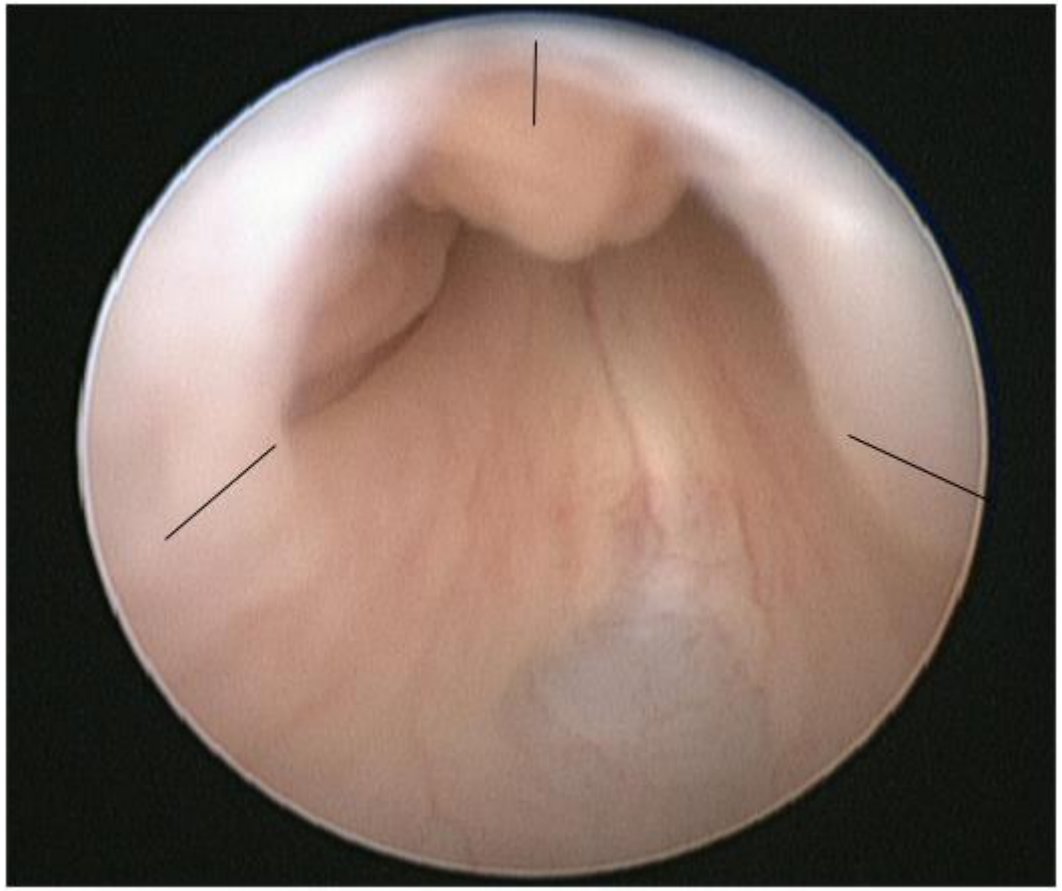
Cont.

- After 5 – 7 days, the serum creatinine determines the subsequent management:
  - If serum creatinine is < 80μmol/L:
    - Valve ablation
    - Vesicostomy for LBWT babies.

Primary valve resection

- Diagnostic cystoscopy using 0^0 6/8 Fr neonatal cystoscope.
- The configuration of the bladder neck and appearances of the bladder and ureteric orifices should also be noted.

Cont.



Cont.

- The complications of primary valve ablation include:
  - Bleeding
  - Incomplete valve resection
  - Urethral stricture
  - Inadvertent damage to the external sphincter

Cont.

- It is recommended that all boys have a ***follow – up cystoscopy at 3 months*** of the primary procedure to ensure completeness of valve ablation OR:
- A ***repeat MCU*** & proceed to cystoscopy only if the MCU suggests persisting urethral obstruction.

Early Urinary Diversion

- Recommended for patients whose:
  - ***Renal function*** remains fragile or deteriorates.
  - Suffer ***recurrent UTIs***
  - Significant deterioration in the appearance of the upper tracts
  - Bladder emptying is incomplete on serial U/S
- AIM: protect the upper tracts and minimize the risks of infection.

Who requires ureterostomy?

- In patients with persistent high serum creatinine above 160μmol/L despite trans – urethral catheterization: **high loop ureterostomy**
- **Percutaneous nephrostomy** drainage helps to identify patients who require diversion.

Poor prognosis

- 30 – 70% of patients progress to ESRD over a follow up of 11.3+/- 2.1 yrs.
- Post – operative nadir serum creatinine at 1 yr. > 70 μmol/l
- Renal dysplasia
- Bladder dysfunction
- VUR
- Onset of proteinuria

ESRD: Renal Transplant

- Occurs in 25 – 40%
- 1/3 soon after birth; 2/3 during late teenage
- Role of recurrent infection?
- Challenges: a high pressure, poorly compliant and low capacity bladder
- Solution:
  - Augmentation.
  - Should we augment before or after transplant.

Conclusion

- PUVs remains the MCC of neonatal BOO in males.
- An increasing number of cases are diagnosed antenatally but prenatal intervention does not appear to confer a benefit in the long-term outcome of renal function.
- Primary valve ablation is the recommended treatment of choice with diversion being reserved for specific individual cases.
- A significant number of boys with PUV will develop chronic kidney disease and end stage renal failure.

Trust is the feeling that a little child has when they're thrown in the air by their loving & playful father.

They know he will catch them when they fall back.

*"Pour out all your worries & stress upon him & **leave them there**, for He always tenderly cares for you – 1st Peter 5:7 (TPT)"*

#He's_A_Good_Good_Father