Approach to a neonate withabdominal distension

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Introduction:

- Abdominal distention is a common clinical presentation in neonates and underlying causes are multiple ranging from transient benign conditions to life threatening etiologies like necrotizing enterocolitis.
- Systematic approach including thorough history taking, physical examination, basic laboratory tests and imaging studies would narrow down the diagnostic list.
- *There is no statistical definition of abdominal distension*. Some reported definitions include: actual increase in abdominal size, measurable change in abdominal circumference, or "when the abdominal wall is ona higher plane than the xiphisternum in an infant lying on their back on a flat surface."

Differential diagnosis:

The causes of abdominal distention can range from gastrointestinal, renal, retroperitoneal to non-systemic causes like sepsis and ascites.

Intestinal obstruction:

- Intestinal obstruction occurs in approximately 1 in 2000 births.
- Classic clinical signs of neonatal intestinal obstruction are vomiting, abdominal distention, and failure to pass meconium.
- Infant's clinical presentation is dependent on the anatomic level of the obstruction

Table. 1. Obstructive causes of abdominal distention

High obstructive lesions	Low obstructive lesions	Functional Obstruction
Pyloric stenosis	Ileal atresia / stenosis	Necrotizing enterocolitis
Duodenal obstruction Atresia/ stenosis/annular pancreas/peritoneal bands	Colonic atresia or stenosis	Sepsis
Malrotation	Meconium ileus	Hypothyroidism
Malrotation with volvulus	Small left colon syndrome	Peritonitis
Proximal jejunal atresia /stenosis	Meconium plug syndrome	Electrolyte imbalance
	Hirschsprung disease	
	Anorectal malformations	

A. High obstructive lesions:

Suspect: When the neonate presents with more of vomiting and less withabdominal distention

1)***Pyloric stenosis:***

- Occurs 2 in every 1000 children usually betweenthe 3rd to 6th weeks of life although it has been reported inpreterm neonates and children over 1 year of age.
- A hungry neonate presents with increasingly projectileand non-bilious vomits after feeds.

2) ***Duodenal obstruction: atresia,stenosis, annular pancreas,peritoneal bands:***

- May present with bilious/nonbilious emesis and failure to pass stools.
- Duodenal atresia may be seen as an isolated finding or in association with trisomy 21.

3)***Malrotation:***

- Absent or incomplete bowel rotation during the embryologic process leads to abnormal bowel fixation.
- There can be true malrotation, atypical malrotation, and nonrotation.

4)***Malrotation with volvulus:***

- Bowel malrotation with volvulus is an extremely dangerous condition because it may lead to infarction of the entire midgut – a potentially fatal complication.

5)***Proximal jejunal atresia or stenosis:***

- It arises from intrauterine vascular insult, which results in necrosis and resorption, leading to segmental stenosis.
- In proximal jejunal atresia, the classicappearance is a triple bubble, with gaseousdistention of the stomach, duodenum, andproximal jejunum

B. Low obstructive lesions:

Suspect: When the neonates present with feed intolerance and abdominal distention with or without the history of delayed passage of meconium.

1)Intestinal Structural Pathologies:

1)***Ileal atresia or stenosis/ Colonic atresia or stenosis*:**

2***) Meconium ileus:***

- Meconium ileus is a functional disorder of the bowel in which the highly viscous meconiumbecomes impacted and then dehydrated, to obstruct the terminal ileum with dried meconium pellets.
- Approximately 90% of patients with meconium ileus have cystic fibrosis.

3*) Small left colon syndrome**:*

- It's a transient condition typically seen in term infants born to mothers with a historyof diabetes.
- Neonates fail to pass stool in the first 48 hours, usuallyresolves spontaneously.

4) *Meconium plug syndrome:*

- Meconium obstructs the rectum and/or lower colon.
- Increased incidence of Hirschsprung disease. More commonly seen in infantsof diabetic mothers.

5)*Hirschsprung disease:*

- Aganglionosis of distal colon/rectum leading to delayed passage of stool.
- Approximately 75% of infants with Hirschsprung disease fail to pass meconium in the first 48 hours after birth. More commonin males.

6)*Anorectal malformation:*

- It encompasses a complex group of anomalies in which the anal canal or rectum has failed to develop normally.
- In most patients, there is a fistulous connection from the bowel to theperineal skin or urogenital tract.
- Patients with imperforate anus commonly manifest multiple anomalies in other parts of the body (e.g., oesophageal atresia, cardiac defects urinary tract anomaliesand sacral agenesis).

2)Perforation:

A) *Congenital:*

- More likely to occur secondary to bowel obstructions or atresia's. Bowel perforation in utero may lead to meconium peritonitis and secondary abdominal distension.

B) *Acquired:*

1) **NEC:** Perforation occurs in approximately 30% of necrotizing enterocolitis (NEC) cases. Terminal ileum and ascending colon are the most common locations.
2) **Spontaneous intestinal perforation (SIP):**
    - occurs most commonly in extremely low birthweightinfants typically in the ileum. Risk factors include concomitant use of steroids and indomethacin and extreme prematurity.

- Presents in the first 2 weeks, often with a bluish discoloration of the abdomen and abdominal distension.

3) **Traumatic:** occur from NG/OG tube misplacementor excessive positive-pressure ventilation.

3)Necrotizing enterocolitis:

- It's primarily a disease of premature infants.
- NECstill remains the most common gastrointestinal emergency in NICU, despite a gradual decrease in incidence over the last 10 years because of improved prevention strategies.
- It is thought to be multifactorial, with immature bowel function, bowel hypoxia or ischemia, type of enteral feeding, and disruption of gut microbiota likely representing contributing factors.Overall mortality is between 10% and15%, with an increase to 30% if intestinal perforation occurs.
- Pneumatosis intestinalisis the classic sign on abdominal radiograph, and thrombocytopenia is also classically seen.

4) Sepsis:

- Sepsis occurs in approximately one-third of very low birthweight infants.
- This should be considered with or without NEC based on the entire clinical picture.
- Abdominal distention and other signs of feeding intolerance can be clinical signs of sepsis, particularly in premature patients who may not mount a fever.
- This should be considered with or without NEC based on the entire clinical picture.

5) Abdominal masses:

- Abdominal masses occur in almost 1 of every 1000 live births and have a broad differential diagnosis. The principal lesions in this category are hepatic, renal, adrenal and ovarian cyst.

6)Miscellaneous :e.g. Hypothyroidism, Dyselectrolytemia,Constipation etc.

Approach:

(A)History and physical examination:

- Detailed maternal history including prenatal sonography.
- History of polyhydramnios indicates underlyingGI obstructions.
- Oligohydramnios indicates underlying renal pathology.

Ask the following questions and perform thorough physical examination of the neonate.

1)What is the gestational age of the baby? Is baby sick?

- A well neonate with abdominal distention → most likely surgical abdomen
- A sick neonate with abdominal distention → Most likely NEC or septic ileus secondary to underlying sepsis.

2)Is the neonate on continuous positive airway pressure?

- A well baby on non invasive respiratory support → Likely CPAP belly

3) Is the baby vomiting? What colour is the emesis?

- Bilious vomiting or blood in vomitus → Likely to be pathological

4) Is baby passed meconium/stool?

- Delayed passage of meconium indicates Hirschsprung disease or meconium plug syndrome.

5)Is the baby's abdomen flat or distended? Is it soft or tender? Is there a palpable mass?

- Tender abdomen – Rule outsepsis, NEC, perforation, complicated surgical condition.
- Soft abdomen – Likely to be a benign condition or uncomplicated surgical condition.
- While abdominal distention since birth → likely to be a congenital mass (Cysts, tumours, renalmases) or ascites.

6)Is the baby septic? Are there other signs of illness: irritability, temperature instability, apnea/bradycardia episodes?

7)Does the neonate look oedematous?

- Abdominal distention could be secondary tothird spacing from ascites or other fluid.

8)Is examination of the baby's perineum normal? Is the anus patent?

- ARM presents with progressive abdominal distention.

9)What other abnormalities are present?

- Rule out syndromes like Trisomy 21 or associations like VACTERL

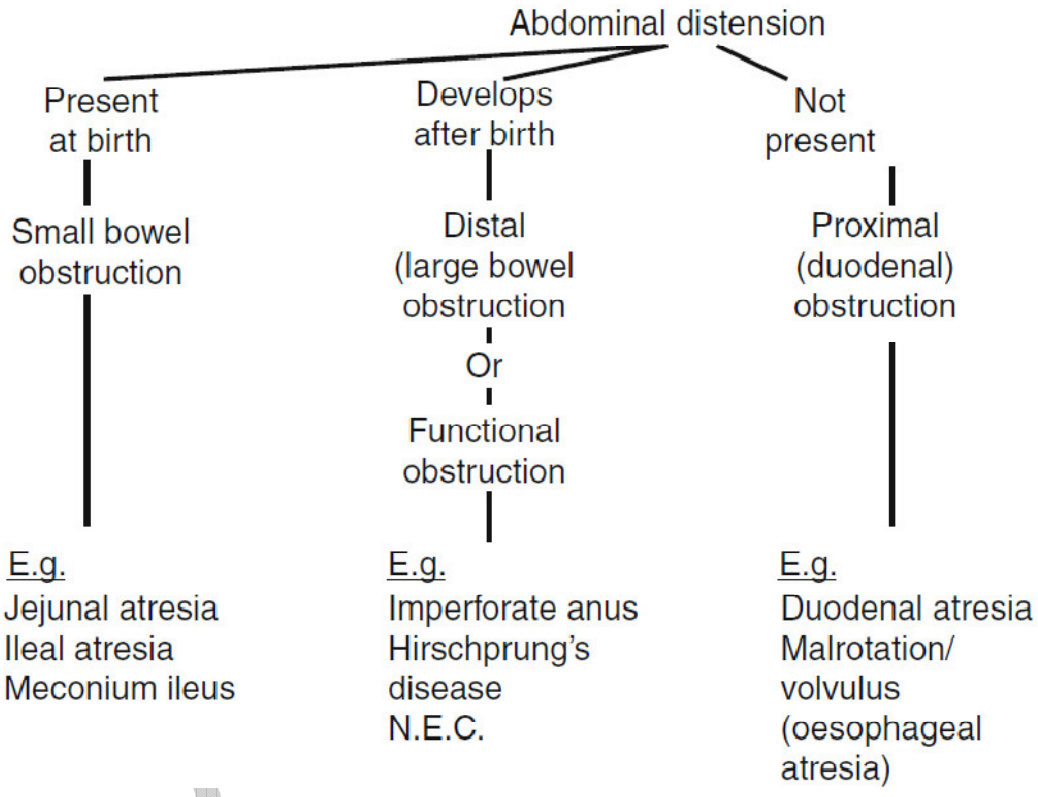


Fig :1 The relationship of abdominaldistension to underlying causes ofobstruction

(B) Investigations:

1)Air is good contrast medium.Plain X- ray abdomen is diagnostic in most of the neonates.

- Obtainhorizontal and left lateral decubitus films. Many disorders have characteristic appearances on X – ray described widely in literature for e.g., Triple bubble appearance in jejunal atresia.
- It provides some additional information such as the presence of intraperitoneal calcification in prenatal perforation and intramural gas bubbles in necrotizing enterocolitis, fixed and dilated bowel loops, rectal and colonic air.
- The number and distribution of dilated loops may allow differentiation between high and low bowel obstruction.

2)Upper GI contrast using Gastrografin

3)Contrast enema using Gastrografin

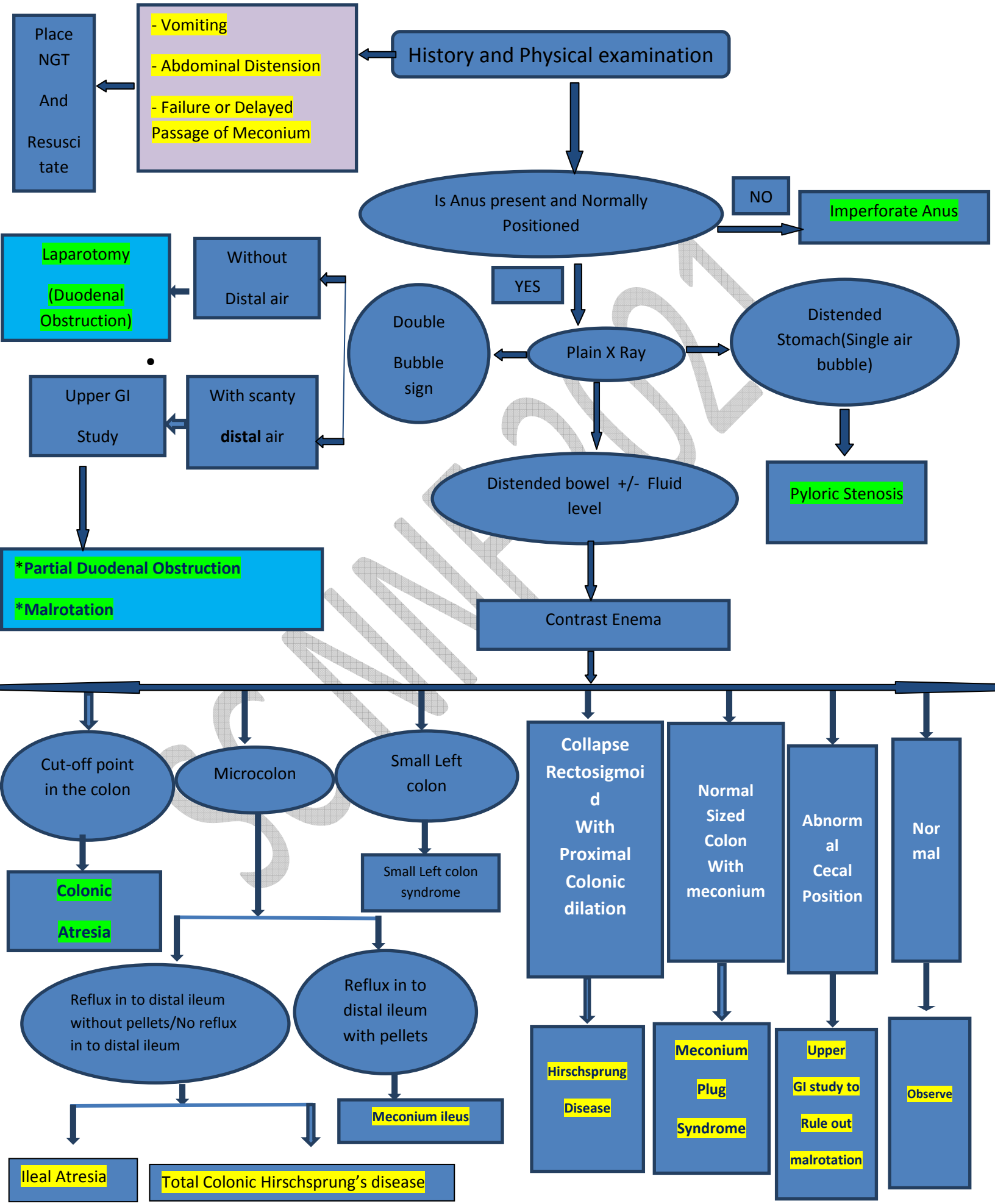
4)Abdominal ultrasound:Ultrasound can be used for diagnosing masses, ascites, or debris indicating perforation. May also be used to assess for pneumatosis intestinalis in NEC.

5)Laboratory studies:

1. Septic screen:
   - Positive screen points towards underlying sepsis.
   - Dropping platelet count along with hyponatremia and metabolic acidosis are surrogate markers of advanced stage of NEC
2. Blood cultures.
3. Liver function tests(LFTs) are helpful to evaluatehepatomegaly or ascites secondary to liverpathology. Coagulation studies may be indicated if liver function tests are abnormal.

***Summary and red flags*:**

- Congenital malformations and sepsis are the major causes of abdominal distension in newborn.
- Majority of the surgical condition are either diagnosed antenatally by ultrasound or very shortly after birth.
- Plain abdominal radiography helps diagnose most conditions.
- Bilious vomiting in neonates (in the absence of sepsis) indicates upper GI obstruction and ischaemic damage to the bowel and is a potential emergency.
- Hirschsprung disease should be suspected if a term baby fails to pass meconium in the first 48 hours.
- Obstruction occurring after the evacuation of normal meconium is likely to be (a) malrotation with volvulus, (b) intestinal atresia (c) Hirschsprung disease or (d) necrotizing enterocolitis – all of which are potentially fatal.


History and Physical examination
- Vomiting
- Abdominal Distension
- Failure or Delayed Passage of Meconium
Place NGT And Resuscitate
Is Anus present and Normally Positioned
NO
Imperforate Anus
YES
Plain X Ray
Double Bubble sign
Distended Stomach(Single air bubble)
Pyloric Stenosis
Without Distal air
Laparotomy (Duodenal Obstruction)
With scanty distal air
Upper GI Study
*Partial Duodenal Obstruction
*Malrotation
Distended bowel +/- Fluid level
Contrast Enema
Cut-off point in the colon
Microcolon
Small Left colon
Collapse Rectosigmoid With Proximal Colonic dilation
Normal Sized Colon With meconium
Abnormal Cecal Position
Normal
Colonic Atresia
Small Left colon syndrome
Reflux in to distal ileum without pellets/No reflux in to distal ileum
Reflux in to distal ileum with pellets
Hirschsprung Disease
Meconium Plug Syndrome
Upper GI study to Rule out malrotation
Observe
Meconium ileus
Ileal Atresia
Total Colonic Hirschsprung's disease

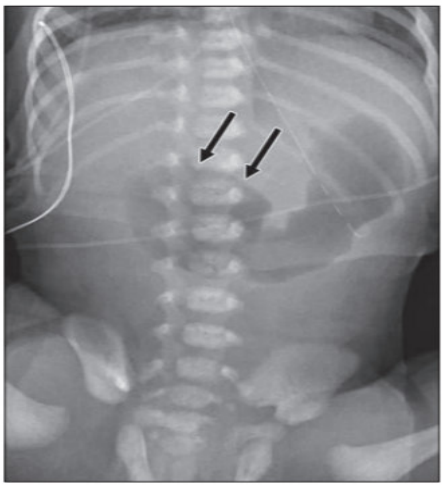


Fig 3. Duodinal atresia - X ray abdomen -showing double bubble that is pathognomonic for duodenal atresia, with pronounced dilation of gas-filled duodenal bulb (arrows) related to chronic obstruction. Gas-filled stomach is partially decompressed by nasogastric tube. No bowel gas is seen distal to level of obstruction.

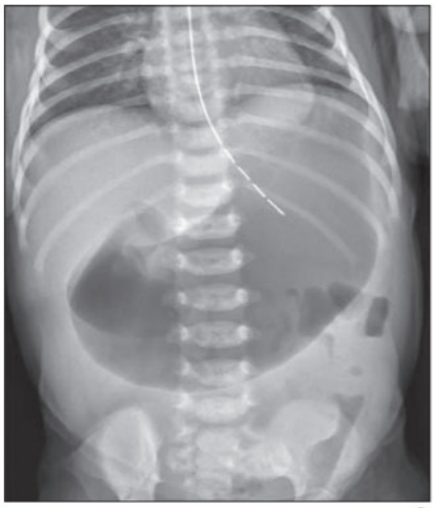


Fig 4. Malrotation and Ladd bands – X ray abdomen - showing marked gaseous distention of stomach and proximal duodenum, with small amount of bowel gas seen in distal bowel loops in left hemiabdomen.

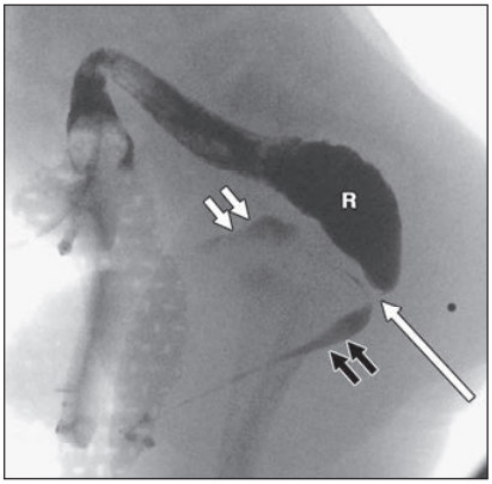


Fig 5. Imperforate anus and rectourethral fistula – Barium enema - Lateral view- shows fistulous communication (long white arrow) between rectum (R) and urethra (black arrows), with contrast material also faintly opacifying bladder (short white arrows). Note that metallic BB (black dot) was placed at expected position of anus on perineum to aid in presurgical planning.

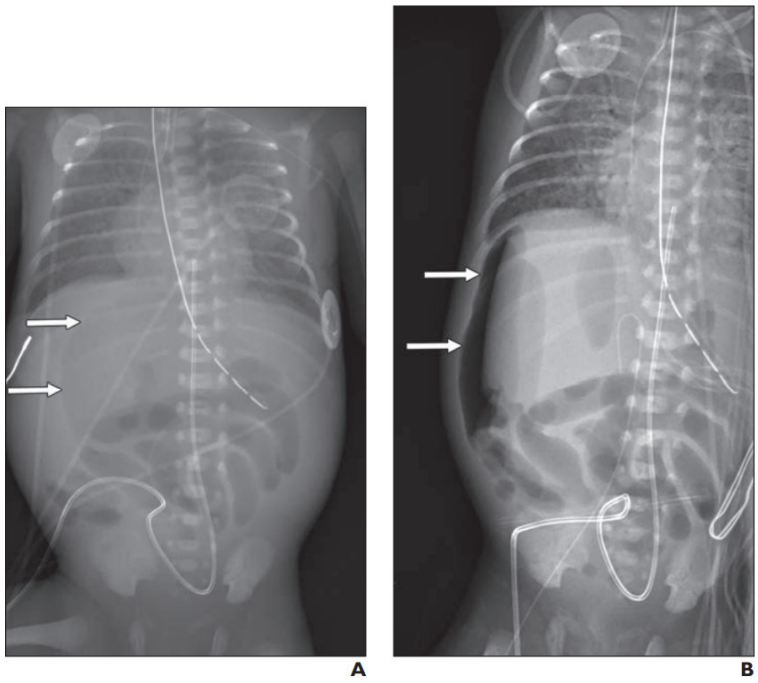


Fig 6. Necrotizing enterocolitis and pneumoperitoneum – X ray chest and abdomen - (A) shows multiple distended stacked bowel loops, with large pneumoperitoneum (arrows), which is better seen on left lateral decubitus view (arrows, B). No definite pneumatosis or portal venous gas were present.

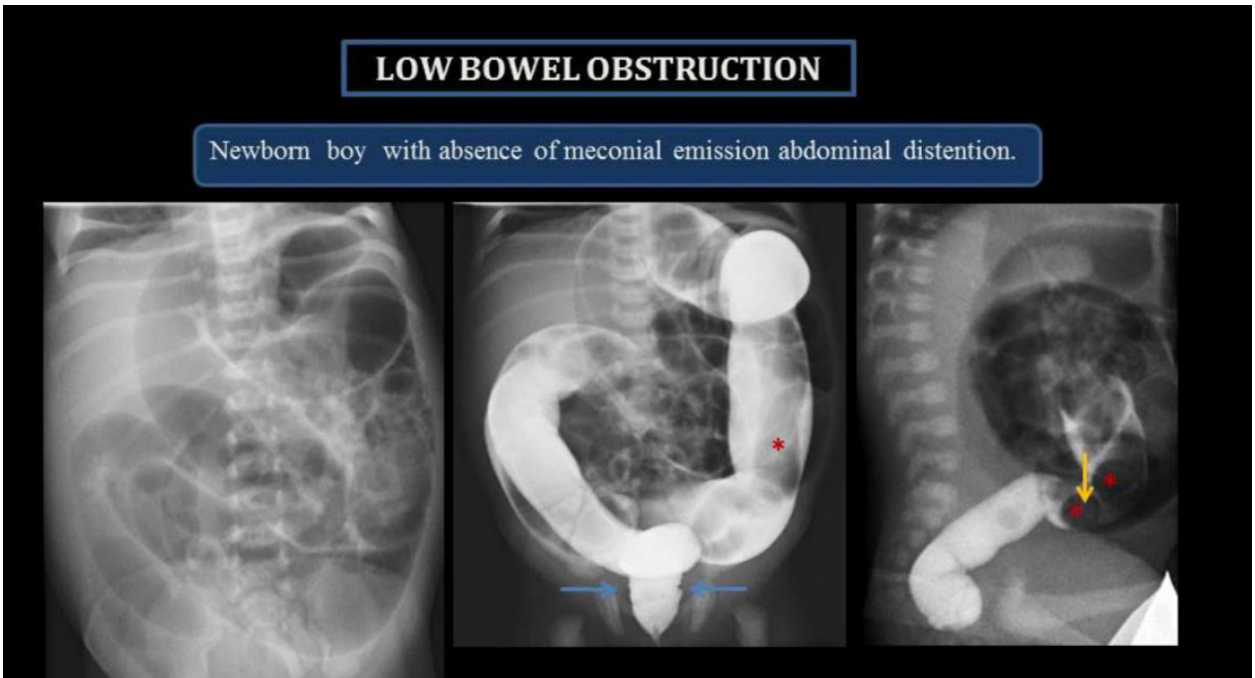


Fig. 7. Hirschsprungs Disease

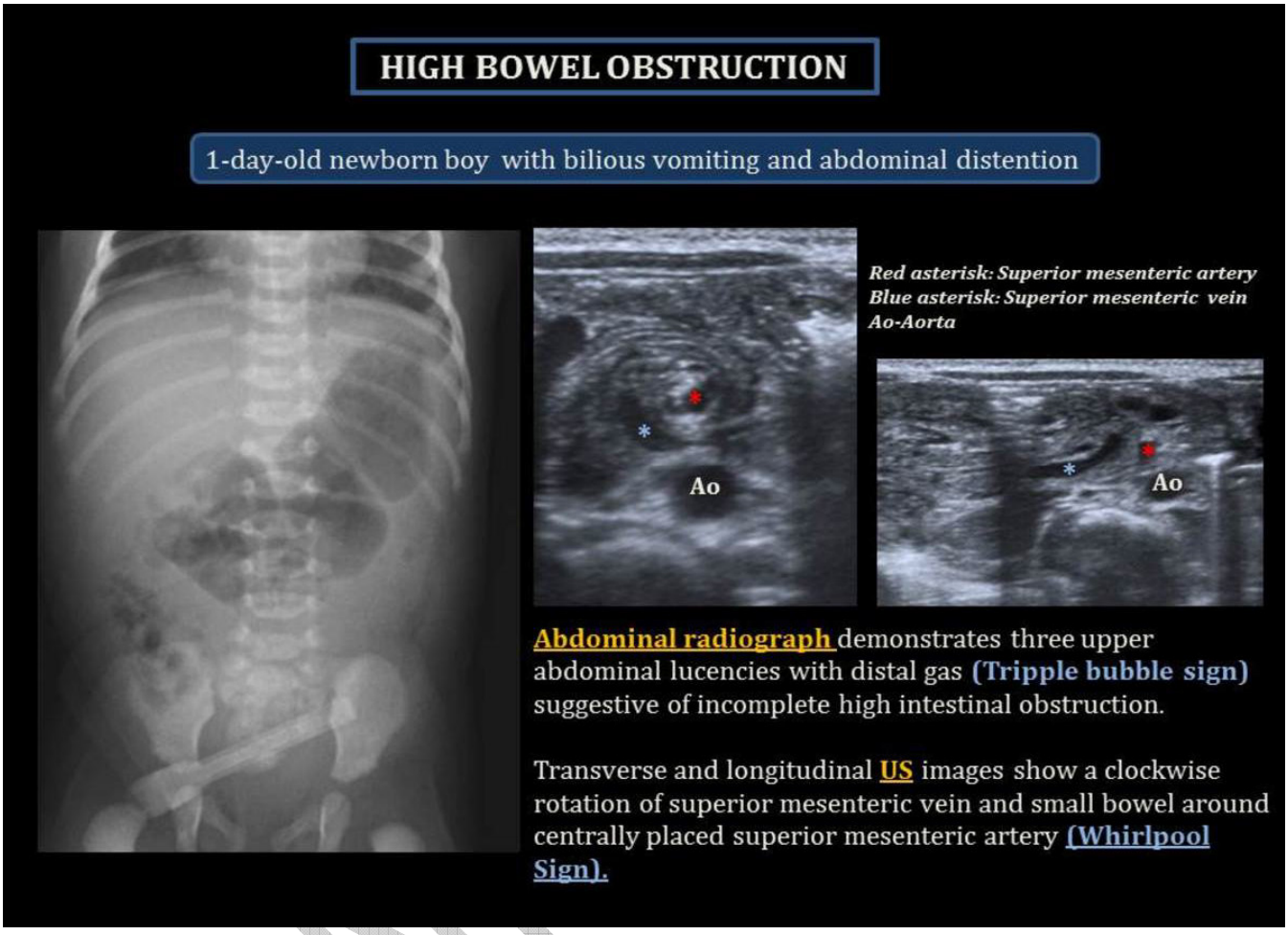


Fig 8: Intestinal malrotation with midgut volvulus

Suggested reading:

1. Gomella's Neonatology: Eighth Edition
2. American Journal of Roentgenology 2018.210:976
3. Intestinal Obstruction in the Newborn Differential Diagnosis.emedicine Feb 2020
4. J.M. Hutson, S.W. Beasley, *The Surgical Examination of Children*, 279. DOI 10.1007/978-3-642-29814-1_21, © Springer-Verlag Berlin Heidelberg 2013
5. Radiology, ComplejoHospitalario de Toledo, Virgen de la Salud - Toledo/ES