La-Net

SUG 403

PAEDIATRIC SURGERY

Download on the App Store

GET IT ON Google Play

Course video is available on our app

lanetonline.com

Course code: SUG 403

Course title: Paediatric Surgery

Course credit load: 2 Units

Series 1: Congenital Gastrointestinal Anomalies

- **Part 1.1: Abdominal Wall Defects**
    - Sub Part 1.1.1: Exomphalos
    - Sub Part 1.1.2: Gastroschisis
    - Sub Part 1.1.3: Management and outcomes
- **Part 1.2: Intestinal Atresia**
    - Sub Part 1.2.1: Oesophageal atresia and tracheo-oesophageal fistula
    - Sub Part 1.2.2: Duodenal and jejunoileal atresia
    - Sub Part 1.2.3: Management of intestinal atresia
- **Part 1.3: Malrotation and Midgut Volvulus**
    - Sub Part 1.3.1: Embryology and pathophysiology
    - Sub Part 1.3.2: Clinical features and diagnosis
    - Sub Part 1.3.3: Management
- **Part 1.4: Hirschsprung's Disease and Anorectal Malformations**
    - Sub Part 1.4.1: Hirschsprung's disease
    - Sub Part 1.4.2: Anorectal malformations
    - Sub Part 1.4.3: Management and complications

Introduction

Congenital gastrointestinal anomalies are structural defects present at birth that affect the alimentary tract and abdominal wall. They are among the most challenging conditions in paediatric surgical practice, requiring prompt recognition, careful resuscitation, and timely surgical correction. Many present in the neonatal period and are life-threatening if not identified and managed urgently.

This Series covers the major congenital GI anomalies relevant to Nigerian paediatric surgical practice. We begin with abdominal wall defects (exomphalos and gastroschisis), then examine intestinal atresia, including oesophageal atresia and duodenal atresia. We then address malrotation and midgut volvulus, and close with Hirschsprung's disease and anorectal malformations.

Series Learning Outcomes

Upon completing this Series, you should be able to:

- **SLO 1.1:** identify the clinical features of exomphalos and gastroschisis and describe their management,
- **SLO 1.2:** describe the clinical presentation, diagnosis, and surgical management of oesophageal atresia, duodenal atresia, and jejunoileal atresia,
- **SLO 1.3:** describe the embryological basis, clinical features, and management of malrotation and midgut volvulus, and
- **SLO 1.4:** describe the clinical features, diagnosis, and management of Hirschsprung's disease and anorectal malformations, and state the complications that may arise.

1.1 Abdominal Wall Defects

1.1.1 Exomphalos

Exomphalos (omphalocele) is a midline abdominal wall defect at the umbilical ring through which abdominal contents herniate into the base of the umbilical cord, covered by a sac of amnion and peritoneum. The sac may contain small bowel, large bowel, stomach, and liver. It results from failure of the midgut to return to the abdominal cavity during the 10th week of gestation. Exomphalos is classified as minor (sac contains only bowel; defect less than 5 cm) or major (sac contains liver; defect above 5 cm).

Associated anomalies are present in up to 70 percent of cases and include cardiac defects (most common), chromosomal abnormalities (trisomy 13, 18, and 21), Beckwith-Wiedemann syndrome (macroglossia, macrosomia, and hypoglycaemia), and neural tube defects. Antenatal

diagnosis by ultrasound is common. At birth, the sac must be kept moist and the baby resuscitated before surgery.

Fill in the blank: Exomphalos results from failure of the midgut to return to the abdominal cavity at the _____ week of gestation. Associated anomalies occur in up to _____ percent of cases, the most common being _____ defects.

1.1.2 Gastroschisis

Gastroschisis is a full-thickness defect of the abdominal wall, typically to the right of the umbilicus, through which abdominal contents herniate without any covering sac. Unlike exomphalos, the bowel is directly exposed to amniotic fluid, causing a chemical peritonitis that results in a thickened, matted, oedematous, and foreshortened bowel (the peel). The defect is usually small (2 to 4 cm). Gastroschisis is not typically associated with chromosomal anomalies, though prematurity and low birth weight are common.

Gastroschisis is a neonatal surgical emergency. The exposed bowel must be covered immediately with a warm saline-soaked gauze wrap and a plastic bag or cling film to prevent heat loss, fluid loss, and further contamination. A nasogastric tube is passed to decompress the stomach. IV fluid resuscitation is essential as fluid losses from the exposed bowel are massive.

Fill in the blank: Gastroschisis is located to the _____ of the umbilicus and has _____ covering sac. The thickened oedematous bowel in gastroschisis is caused by exposure to _____ fluid and is called the peel.

1.1.3 Management and outcomes

Surgical repair of exomphalos: small defects are closed primarily. Large defects with liver in the sac may require staged repair: the sac is preserved and used as coverage, or a silo (a synthetic mesh bag sutured to the fascial edges) is created to gradually reduce the contents over 7 to 14 days before fascial closure. Rupture of the exomphalos sac is an emergency requiring immediate surgery.

Surgical repair of gastroschisis: primary closure (reduction of all bowel into the abdomen and primary fascial closure in a single operation) is performed if the abdominal domain is adequate and the bowel is not too oedematous. If primary closure is not possible (excessive intra-abdominal pressure would compromise respiration and bowel perfusion), a silo is fashioned and the bowel reduced gradually over several days before closure. Complications include short bowel syndrome (if bowel is resected), prolonged ileus, and sepsis. Survival in centres with adequate neonatal intensive care exceeds 90 percent.

Fill in the blank: A _____ is a synthetic mesh bag used to gradually reduce abdominal contents over days before fascial closure in both exomphalos and gastroschisis. The most serious long-term complication of gastroschisis is _____ bowel syndrome if bowel is resected.

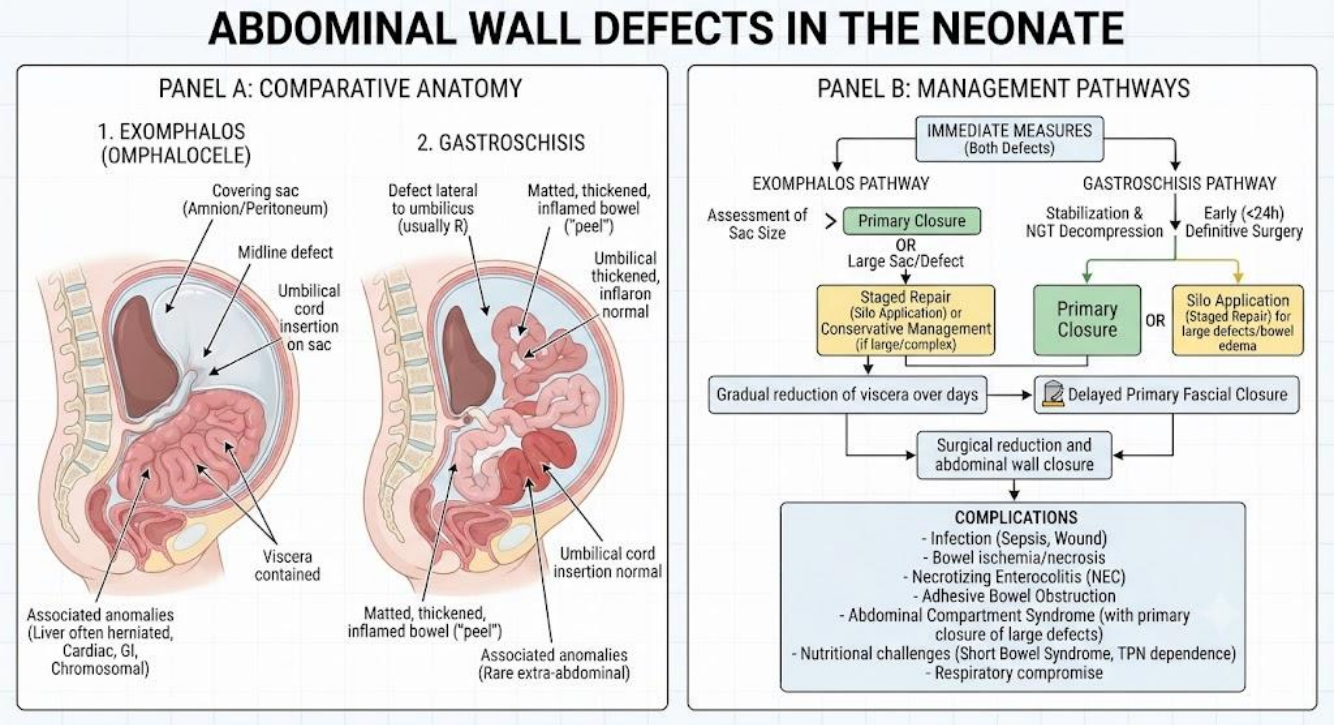

Figure 1.1: Comparison of exomphalos and gastroschisis and their management

Pilot questions 1.1

1. Distinguish exomphalos from gastroschisis in terms of location, sac, and associated anomalies. (Linked to SLO 1.1)
2. Describe the immediate management of a neonate born with gastroschisis. (Linked to SLO 1.1)
3. Describe the Beckwith-Wiedemann syndrome and state its association with exomphalos. (Linked to SLO 1.1)
4. Describe the use of a silo in the management of abdominal wall defects. (Linked to SLO 1.1)
5. State the complications of gastroschisis repair. (Linked to SLO 1.1)

1.2 Intestinal Atresia

1.2.1 Oesophageal atresia and tracheo-oesophageal fistula

Oesophageal atresia (OA) is a congenital interruption of the continuity of the oesophagus. It is classified by the Gross classification into five types. The most common type (Type C: approximately 85 percent) is OA with a distal tracheo-oesophageal fistula (TOF): the upper oesophagus ends in a blind pouch, and the lower oesophagus connects abnormally to the trachea. Type A (pure OA without a fistula: approximately 8 percent) is the next most common.

Associated anomalies are grouped in the VACTERL association (Vertebral, Anorectal, Cardiac, Tracheo-Oesophageal, Renal, and Limb anomalies).

Clinical features of OA: the neonate presents with excessive drooling and salivation (accumulation of secretions in the blind upper pouch), choking and cyanosis during feeds (aspiration), and respiratory distress. The diagnosis is suspected when a nasogastric tube cannot be passed beyond 10 to 12 cm from the mouth and is confirmed by CXR showing the tube coiled in the upper oesophageal pouch. The presence of gas in the stomach on CXR indicates a distal TOF (gas entering the stomach via the fistula).

Fill in the blank: The most common type of oesophageal atresia is Type C, which consists of OA with a _____ tracheo-oesophageal fistula. A neonate with OA is suspected when a nasogastric tube cannot be passed beyond _____ to 12 cm and the tube is seen _____ in the upper pouch on CXR.

1.2.2 Duodenal and jejunoileal atresia

Duodenal atresia is a congenital obstruction of the duodenum, most commonly at or just distal to the ampulla of Vater (post-ampullary: 85 percent). It results from failure of recanalisation of the duodenal lumen during the 8th to 10th week of gestation. There is a strong association with Down syndrome (trisomy 21: present in approximately 30 percent of cases) and with other anomalies (cardiac defects, malrotation, and annular pancreas). The classic radiographic finding is the double bubble sign (air in the stomach and the proximal duodenum with no distal gas) on plain AXR.

Jejunoileal atresia results from an intrauterine vascular accident (mesenteric ischaemia causing bowel infarction and resorption) rather than failure of recanalisation. It is classified into four types (I: mucosal web; II: fibrous cord between two blind ends; IIIa: gap with V-shaped mesenteric defect; IIIb: apple-peel or Christmas tree deformity: the most severe, with risk of short bowel syndrome; IV: multiple atresias). Unlike duodenal atresia, jejunoileal atresia is not strongly associated with chromosomal anomalies.

Fill in the blank: Duodenal atresia is associated with Down syndrome in approximately _____ percent of cases. The classic X-ray finding is the _____ bubble sign. Jejunoileal atresia results from an intrauterine _____ accident rather than failure of recanalisation.

1.2.3 Management of intestinal atresia

Management of OA: preoperative resuscitation (IV fluids, keep the baby warm, elevate the head to reduce aspiration), continuous suctioning of the upper pouch with a Replogle tube, and prone positioning if a distal TOF is present (to reduce acid reflux via the fistula into the lungs). Surgery: primary repair (division of the TOF and end-to-end oesophageal anastomosis) is the goal; if the gap between the two oesophageal ends is too long (long-gap OA), staged repair with oesophageal replacement (gastric transposition or colon interposition) is required.

Management of duodenal atresia: duodenoduodenostomy (side-to-side or diamond-shaped anastomosis bypassing the obstruction) is the standard surgical repair. Management of jejunoileal atresia: resection of the atretic segment and end-to-end anastomosis; the proximal dilated bowel may need to be tapered or plicated to improve peristalsis; bowel conservation is essential to prevent short bowel syndrome. In all intestinal atresias, a nasogastric tube is passed, IV fluids are given, and electrolyte imbalances are corrected before surgery.

Fill in the blank: The surgical repair of oesophageal atresia involves division of the TOF and _____ anastomosis. The standard repair of duodenal atresia is a _____ (bypassing the obstruction). Bowel _____ is essential in jejunoileal atresia to prevent short bowel syndrome.

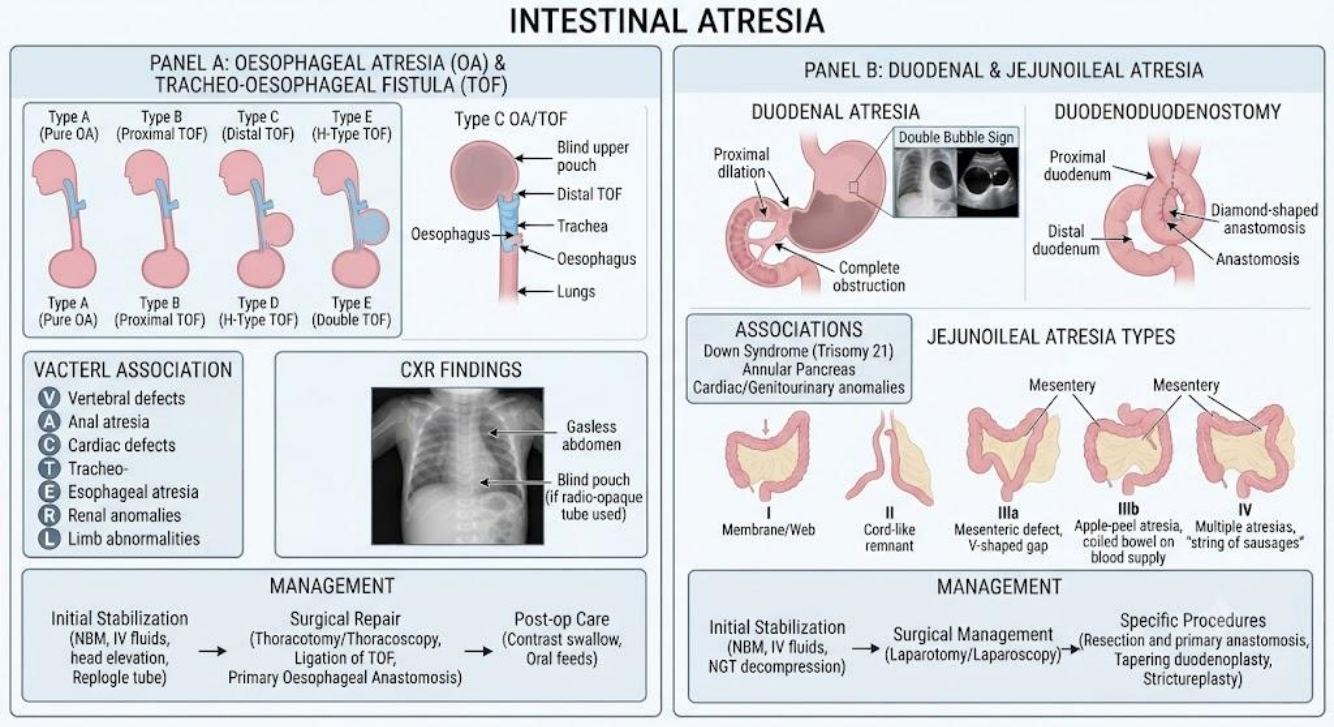

Figure 1.2: Classification and management of oesophageal atresia and intestinal atresia

Pilot questions 1.2

1. Describe the clinical features of oesophageal atresia and state how the diagnosis is confirmed. (Linked to SLO 1.2)
2. Describe the VACTERL association and explain its relevance to oesophageal atresia. (Linked to SLO 1.2)
3. Describe the double bubble sign and state the diagnosis it suggests. (Linked to SLO 1.2)
4. Distinguish duodenal atresia from jejunoileal atresia in terms of aetiology and chromosomal associations. (Linked to SLO 1.2)
5. Describe the surgical repair of duodenal atresia and jejunoileal atresia. (Linked to SLO 1.2)

1.3 Malrotation and Midgut Volvulus

1.3.1 Embryology and pathophysiology

During normal fetal development, the midgut herniates into the umbilical cord at the 6th week, rotates 270 degrees anticlockwise around the superior mesenteric artery (SMA), and returns to the abdominal cavity by the 10th week. This rotation fixes the duodeno-jejunal flexure (DJ flexure) to the left of the midline (at the ligament of Treitz) and the caecum in the right iliac fossa, creating a broad-based mesenteric attachment from the DJ flexure to the caecum that prevents volvulus.

Malrotation occurs when this rotation is incomplete or abnormal. The most common variant is non-rotation, in which the DJ flexure lies to the right of the spine and the caecum lies in the epigastrium or upper abdomen. This creates a narrow mesenteric pedicle (the midgut hangs on a narrow stalk of mesentery around the SMA) that predisposes to midgut volvulus. Ladd's bands are peritoneal bands from the caecum crossing the duodenum to the retroperitoneum, causing extrinsic duodenal obstruction.

Fill in the blank: During normal development the midgut rotates _____ degrees anticlockwise around the SMA. In malrotation, _____ bands cross the duodenum causing extrinsic obstruction, and the narrow mesenteric pedicle predisposes to _____ volvulus.

1.3.2 Clinical features and diagnosis

Malrotation presents most commonly in the first month of life (approximately 75 percent of cases in the neonatal period), but it can present at any age. The classic presentation is bilious vomiting in a neonate: bilious (green) vomiting in any neonate must be considered a surgical emergency until malrotation and midgut volvulus are excluded. Midgut volvulus causes acute intestinal obstruction with severe abdominal pain, bilious vomiting, bloody stools (a late and sinister sign indicating bowel ischaemia), and rapid deterioration to shock.

Investigations: plain AXR (may show a gasless abdomen or duodenal obstruction pattern in volvulus), upper GI contrast study (the investigation of choice for malrotation: shows the DJ flexure to the right of the spine and the corkscrew appearance of the duodenum in volvulus), and ultrasound (inversion of the superior mesenteric vein and artery: the SMA-SMV relationship is reversed in malrotation). CT scan is used in older children when the diagnosis is uncertain.

Fill in the blank: Bilious vomiting in any _____ must be treated as a surgical emergency until malrotation and midgut volvulus are excluded. The investigation of choice for malrotation is an _____ GI contrast study, which shows the DJ flexure to the _____ of the spine.

1.3.3 Management

Midgut volvulus is a surgical emergency requiring immediate laparotomy. The Ladd's procedure is the standard operation: (1) untwist the volvulus (counter-clockwise rotation), (2) divide Ladd's

bands to relieve the duodenal obstruction, (3) broaden the mesenteric base by placing the small bowel on the right and the colon on the left (returning to the non-rotated position), and (4) perform appendicectomy (as the caecum is now in the left side: an inflamed appendix may not produce right iliac fossa signs in the future). No attempt is made to restore normal rotation as this risks re-twisting.

If the bowel is ischaemic at laparotomy, the volvulus is untwisted and the bowel is wrapped in warm saline packs for 5 to 10 minutes to assess viability. Clearly non-viable bowel is resected; however, if extensive bowel is affected, a second-look laparotomy at 24 to 48 hours is planned to reassess viability and minimise resection. The risk of short bowel syndrome is significant when extensive resection is required.

Fill in the blank: The Ladd's procedure involves untwisting the volvulus, dividing _____ bands, broadening the mesenteric base, and performing _____. An appendicectomy is performed because the caecum will now lie on the _____ side.

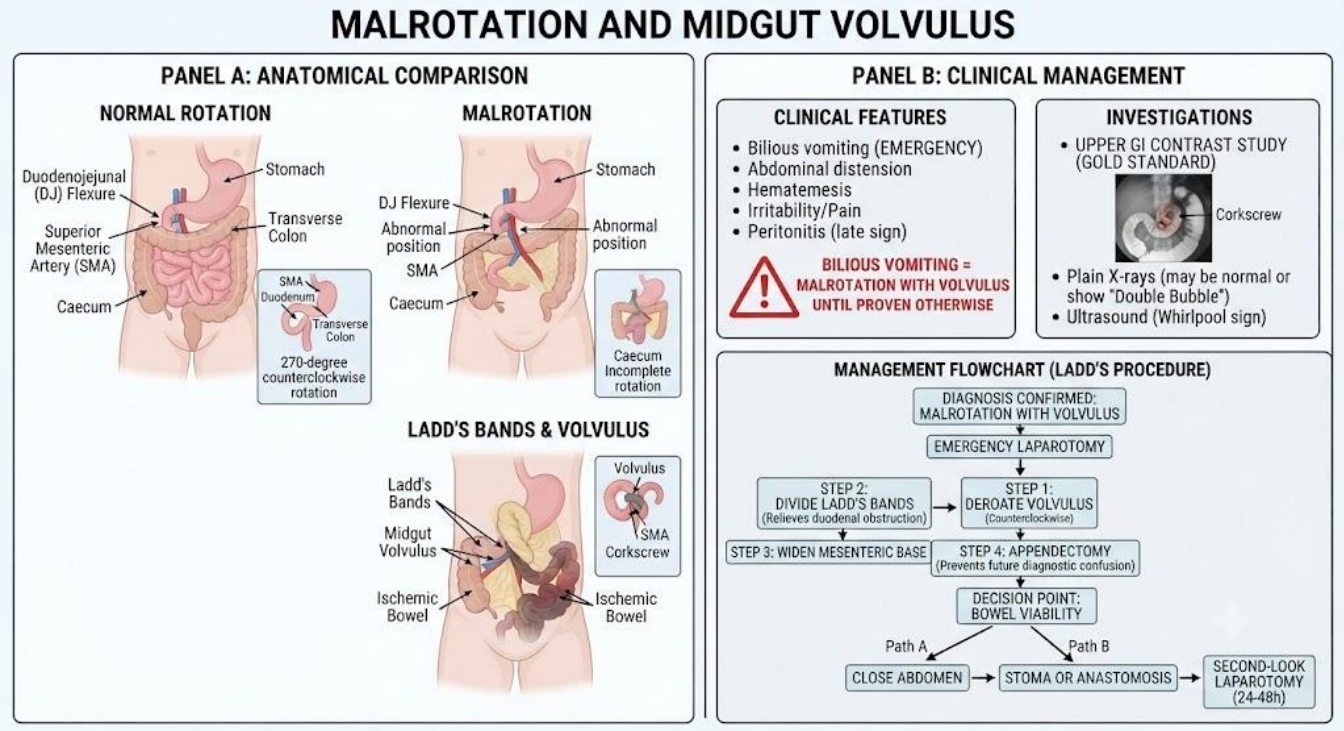

Figure 1.3: Embryology of midgut rotation, malrotation, and Ladd's procedure for midgut volvulus

Pilot questions 1.3

1. Describe the normal rotation of the midgut during fetal development. (Linked to SLO 1.3)
2. Explain why malrotation predisposes to midgut volvulus. (Linked to SLO 1.3)
3. State why bilious vomiting in a neonate is a surgical emergency. (Linked to SLO 1.3)
4. Describe the investigation of choice for malrotation and state the findings. (Linked to SLO 1.3)

5. Describe the four steps of the Ladd's procedure. (Linked to SLO 1.3)

1.4 Hirschsprung's Disease and Anorectal Malformations

1.4.1 Hirschsprung's disease

Hirschsprung's disease (HD) is a congenital absence of ganglion cells (aganglionosis) in the submucosal (Meissner's) and myenteric (Auerbach's) plexuses of the distal bowel, caused by failure of neural crest cell migration during embryogenesis. The aganglionic segment is tonically contracted and non-peristaltic, creating a functional obstruction. In 75 percent of cases the aganglionosis is limited to the rectosigmoid colon (short-segment HD); in 20 percent it extends to the sigmoid or more proximal colon; in 5 percent it involves the entire colon (total colonic aganglionosis).

Clinical features: the classic neonatal presentation is failure to pass meconium within 48 hours of birth (normally 90 percent of neonates pass meconium within 24 hours), abdominal distension, and bilious vomiting. A rectal examination or rectal washout may produce an explosive passage of gas and stool (the squirt sign). In older children and adults: chronic constipation since birth, failure to thrive, and a distended abdomen. The diagnostic investigation is a rectal suction biopsy (confirms absence of ganglion cells and the presence of hypertrophied nerve fibres with positive acetylcholinesterase staining).

Fill in the blank: Hirschsprung's disease is caused by failure of _____ crest cell migration, resulting in absence of _____ cells in the distal bowel. Failure to pass meconium within _____ hours of birth is the classic neonatal presentation.

1.4.2 Anorectal malformations

Anorectal malformations (ARM) are a spectrum of congenital defects of the anus and rectum resulting from abnormal development of the cloaca between the 4th and 8th weeks of gestation. They are classified as low (the rectum descends below the levator ani muscle: the external sphincter is well developed: good prognosis for continence), intermediate, or high (the rectum ends above the levator ani: poor sphincter development: poor prognosis for continence). In males, high lesions are associated with a rectourethral fistula (connecting the rectum to the urethra or bladder neck); in females, high lesions may have a rectovaginal or rectovestibular fistula.

The clinical diagnosis is made at birth by inspection: absence of the anal opening, passage of meconium from an abnormal site (urethra, vagina, or perineum), and perineal dimpling. Associated anomalies occur in approximately 60 percent and include the VACTERL association. Radiological assessment: invertogram (cross-table lateral X-ray with the baby inverted: identifies the position of the air-filled rectal pouch relative to the pubococcygeal line) or MRI pelvis. The Pena classification (Wingspread) is used to guide surgical planning.

Fill in the blank: In anorectal malformations, _____ lesions have a good prognosis for continence because the rectum descends below the _____ ani muscle. In males, high anorectal malformations are associated with a recto_____ fistula.

1.4.3 Management and complications

Management of Hirschsprung's disease: neonatal resuscitation with IV fluids, nasogastric tube, and rectal washouts to decompress the colon. Definitive surgery: pull-through procedure (Swenson, Duhamel, or Soave techniques) in which the aganglionic bowel is resected and normally innervated bowel is pulled through to the anus. In Nigeria and other resource-limited settings, a levelling colostomy (bringing the bowel out as a stoma at the level of normally innervated bowel) may be performed first, followed by delayed pull-through. The most feared complication is Hirschsprung-associated enterocolitis (HAEC: acute enterocolitis of the dilated proximal bowel: presents with explosive diarrhoea, fever, and abdominal distension: can be fatal).

Management of anorectal malformations: low lesions (perineal fistula in males, vestibular fistula in females) are repaired by anoplasty (a limited perineal operation) in the newborn period. High lesions require a staged approach: a diverting colostomy in the newborn period, followed by posterior sagittal anorectoplasty (PSARP, the Pena procedure) at 3 to 6 months of age, and colostomy closure 6 to 8 weeks after PSARP. Complications of ARM repair: constipation and faecal soiling (from poor sphincter function), urinary incontinence, and urinary and sexual dysfunction (from pelvic nerve injury during surgery).

Fill in the blank: The definitive surgical treatment of Hirschsprung's disease is a _____ procedure in which the aganglionic bowel is resected. The most feared complication of Hirschsprung's disease is _____ enterocolitis. High anorectal malformations are repaired by _____ sagittal anorectoplasty (PSARP).

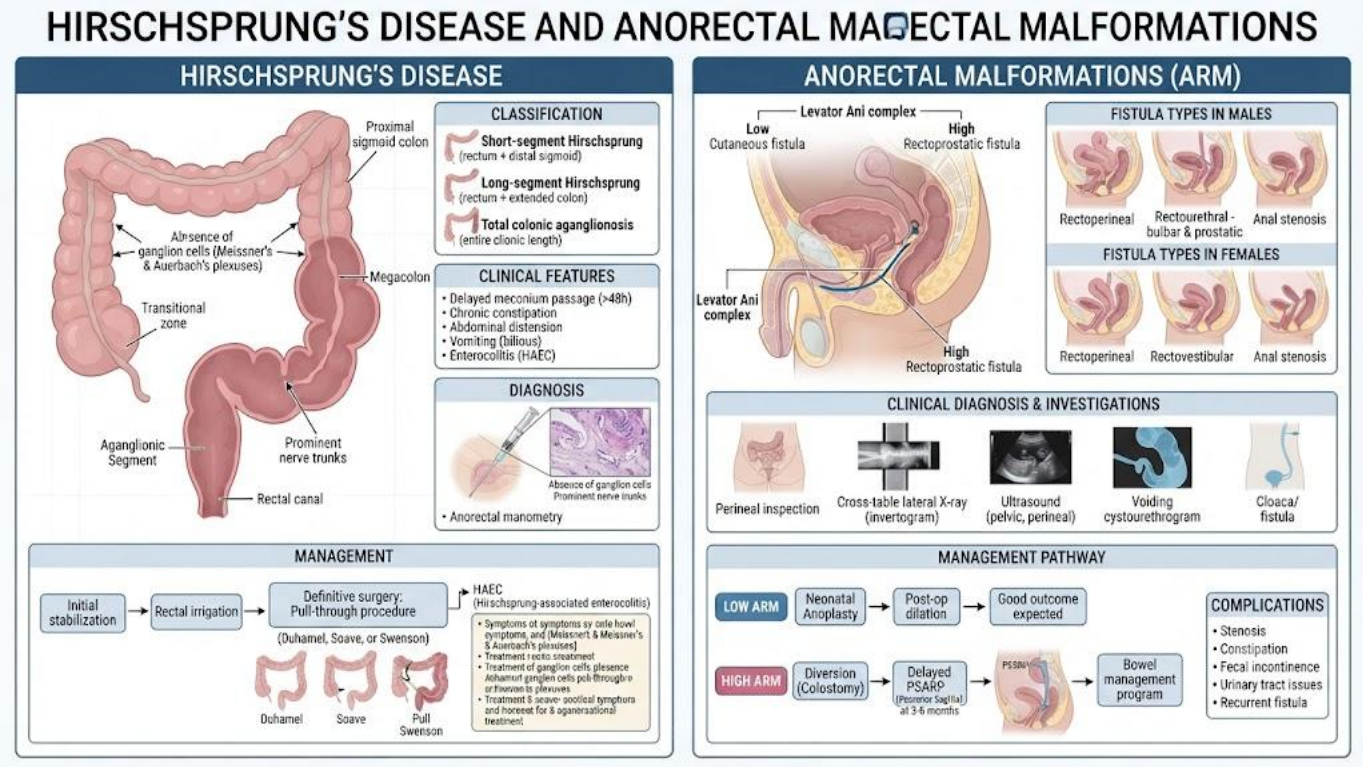

Figure 1.4: Hirschsprung's disease and anorectal malformations: pathophysiology, classification, and management

Pilot questions 1.4

1. Describe the pathophysiology of Hirschsprung's disease. (Linked to SLO 1.4)
2. Describe the clinical features of Hirschsprung's disease in a neonate. (Linked to SLO 1.4)
3. Describe the diagnostic investigation for Hirschsprung's disease and state the findings. (Linked to SLO 1.4)
4. Classify anorectal malformations and state the prognostic significance of each class. (Linked to SLO 1.4)
5. Describe Hirschsprung-associated enterocolitis and state why it is dangerous. (Linked to SLO 1.4)

Series Summary

This Series covered the major congenital gastrointestinal anomalies. We examined abdominal wall defects (exomphalos: midline with sac and associated anomalies; gastroschisis: right of umbilicus, no sac, peel bowel, neonatal emergency) and intestinal atresias (OA: bilious secretions and coiled NG tube on CXR; TOF: VACTERL association; duodenal atresia: double bubble sign and Down syndrome; jejunoileal atresia: intrauterine vascular accident and apple-peel deformity). We then explored malrotation (bilious vomiting as a surgical emergency; upper GI contrast study as the investigation of choice; Ladd's procedure) and Hirschsprung's disease and anorectal malformations (failure of neural crest migration; rectal suction biopsy; pull-through procedure; HAEC; ARM classification by levator ani level; staged PSARP for high lesions).

You should now be able to identify the clinical features of exomphalos and gastroschisis and describe their management (SLO 1.1), describe the presentation, diagnosis, and surgical management of intestinal atresias (SLO 1.2), describe the embryological basis and management of malrotation and midgut volvulus (SLO 1.3), and describe Hirschsprung's disease and anorectal malformations, including their complications (SLO 1.4).

Glossary of Terms

- **Aganglionosis:** the congenital absence of ganglion cells in the myenteric and submucosal plexuses of the bowel wall; the pathological basis of Hirschsprung's disease, resulting from failure of neural crest cell migration.
- **Apple-peel deformity (Type IIIb atresia):** the most severe form of jejunoileal atresia; the proximal jejunum ends in a blind pouch and the remaining bowel wraps around a single surviving mesenteric vessel like a spiral apple peel; associated with a high risk of short bowel syndrome.
- **Double bubble sign:** a radiographic finding on plain AXR showing two air-filled structures (the stomach and the obstructed proximal duodenum) with no gas distally; pathognomonic of duodenal atresia.
- **Gastroschisis:** a congenital full-thickness defect of the abdominal wall to the right of the umbilicus, without a covering sac, through which the bowel herniates and is directly exposed to amniotic fluid, resulting in the peel.
- **Hirschsprung-associated enterocolitis (HAEC):** the most feared complication of Hirschsprung's disease; an acute enterocolitis of the dilated proximal bowel presenting with explosive diarrhoea, fever, and abdominal distension; can be rapidly fatal.
- **Ladd's procedure:** the standard surgical treatment for malrotation and midgut volvulus: untwisting the volvulus, dividing Ladd's bands, broadening the mesenteric base, and performing appendicectomy.

- **Peel:** the thickened, oedematous, matted appearance of the bowel wall in gastroschisis caused by prolonged exposure to amniotic fluid producing a chemical peritonitis.
- **PSARP (posterior sagittal anorectoplasty):** the Pena procedure; the standard surgical repair for high anorectal malformations, performed through a posterior sagittal incision to reconstruct the anorectal canal and sphincter mechanism.
- **Silo:** a synthetic mesh bag sutured to the fascial edges of an abdominal wall defect (exomphalos or gastroschisis) to contain the herniated organs and allow gradual reduction into the abdominal cavity over days before fascial closure.
- **VACTERL association:** a non-random association of congenital anomalies: Vertebral anomalies, Anorectal malformations, Cardiac defects, Tracheo-Oesophageal fistula, Renal anomalies, and Limb defects; strongly associated with oesophageal atresia.

Concept Check Questions [Series 1]

Objective questions

1. Exomphalos results from failure of the midgut to return to the abdominal cavity at the _____ week of gestation. (Linked to SLO 1.1)
2. The most common type of oesophageal atresia (approximately 85 percent) is Type C: OA with a _____ tracheo-oesophageal fistula. (Linked to SLO 1.2)
3. The classic radiographic finding of duodenal atresia is the _____ bubble sign on plain AXR. (Linked to SLO 1.2)
4. Bilious vomiting in any neonate must be treated as a surgical _____ until malrotation and midgut volvulus are excluded. (Linked to SLO 1.3)
5. The most feared complication of Hirschsprung's disease is Hirschsprung-associated _____ (HAEC). (Linked to SLO 1.4)

Short-answer questions

6. Distinguish exomphalos from gastroschisis in terms of location, sac, and associated anomalies. (Linked to SLO 1.1)
7. Describe the VACTERL association and explain its relevance to oesophageal atresia. (Linked to SLO 1.2)
8. Explain why bilious vomiting in a neonate is a surgical emergency. (Linked to SLO 1.3)

9. Describe the diagnostic investigation for Hirschsprung's disease and state the findings. (Linked to SLO 1.4)
10. Classify anorectal malformations and state the prognostic significance of each class. (Linked to SLO 1.4)

Long-answer questions

11. Describe the clinical features, diagnosis, and management of oesophageal atresia with a distal TOF. (Linked to SLO 1.2)
12. Describe the Ladd's procedure and explain each step. (Linked to SLO 1.3)
13. Describe the pathophysiology, clinical features, and management of Hirschsprung's disease. (Linked to SLO 1.4)
14. Describe the staged management of a high anorectal malformation in a male neonate. (Linked to SLO 1.4)
15. Compare the management of exomphalos with gastroschisis, highlighting the key differences. (Linked to SLO 1.1)

Answers to Concept Check Questions [Series 1]

Objective questions

1. 10th.
2. Distal.
3. Double.
4. Emergency.
5. Enterocolitis.

Short-answer questions

6. Exomphalos: midline defect at the umbilical ring; covered by a sac of amnion and peritoneum; strongly associated with chromosomal and cardiac anomalies (up to 70 percent). Gastroschisis: defect to the right of the umbilicus; no covering sac; bowel exposed to amniotic fluid forming the peel; not associated with chromosomal anomalies but associated with prematurity and low birth weight.

7. VACTERL is a non-random association of congenital anomalies: Vertebral, Anorectal, Cardiac, Tracheo-Oesophageal, Renal, and Limb defects. It is relevant because any neonate diagnosed with OA must be screened for all other VACTERL components, as associated anomalies (particularly cardiac defects) significantly affect perioperative management and prognosis.
8. Bilious (green) vomiting in a neonate indicates that the obstruction is distal to the ampulla of Vater. Malrotation with midgut volvulus can present this way and, if unrecognised, leads to twisting of the entire midgut around the SMA, causing ischaemia and infarction of the bowel within hours. Early surgical intervention is essential to prevent loss of the entire midgut.
9. Rectal suction biopsy: a small sample of rectal mucosa and submucosa is obtained by suction and sent for histology and histochemistry. Findings: absence of ganglion cells in the submucosal plexus and hypertrophied, thickened nerve fibres. Histochemistry: positive acetylcholinesterase staining of the hypertrophied nerve fibres confirms the diagnosis.
10. Low lesions: the rectum ends below the levator ani muscle; the external sphincter is well developed; good prognosis for faecal continence; repaired by anoplasty in the newborn period. High lesions: the rectum ends above the levator ani; poor sphincter development; poor prognosis for continence; require staged repair (colostomy, then PSARP, then colostomy closure).

Long-answer questions

11. **Basic response:** Clinical features: excessive drooling and salivation (blind upper pouch fills with secretions), choking and cyanosis during feeds (aspiration), and respiratory distress. Diagnosis: a nasogastric tube cannot be passed beyond 10 to 12 cm; CXR shows the tube coiled in the upper oesophageal pouch; gas in the stomach confirms a distal TOF. Screen for VACTERL anomalies (echocardiogram, renal ultrasound, spinal X-ray). Management: Replogle tube for continuous suction of the upper pouch, IV fluids, head elevation, prone positioning. Surgery: division of the TOF and primary end-to-end oesophageal anastomosis; long-gap OA requires staged repair with oesophageal replacement.

Value-added response: The presence of a right-sided aortic arch (present in approximately 3 percent of OA cases) alters the surgical approach: a right thoracotomy is standard for OA repair, but a right-sided aortic arch in that field makes the approach hazardous and requires a left thoracotomy instead; echocardiography before surgery identifies this.

12. **Basic response:** The Ladd's procedure is the standard surgical treatment for malrotation and midgut volvulus. Step 1: untwist the volvulus by rotating the midgut counter-clockwise (reversing the direction of the twist). Step 2: divide Ladd's bands (the peritoneal bands from the caecum crossing the duodenum) to relieve the extrinsic duodenal obstruction. Step 3: broaden the mesenteric base by placing the small bowel on the right and the large bowel on the left (returning to the non-rotated position), widening the mesenteric attachment and reducing the risk of future volvulus. Step 4: perform appendicectomy (as the caecum now lies on the left side of the

abdomen: a future appendicitis would not produce the classic right iliac fossa signs and would be difficult to diagnose).

Value-added response: No attempt is made to restore normal rotation during the Ladd's procedure, because doing so would require extensive mesenteric manipulation that risks re-twisting. The non-rotated position achieved after the procedure creates the widest possible mesenteric base and is the safest configuration for the patient.

13. **Basic response:** Pathophysiology: failure of neural crest cell migration from cranial to caudal during embryogenesis results in absence of ganglion cells in the myenteric and submucosal plexuses of the distal bowel. The aganglionic segment is tonically contracted and non-peristaltic, creating a functional obstruction. Proximal bowel becomes massively dilated (megacolon). Clinical features: neonates: failure to pass meconium within 48 hours; abdominal distension; bilious vomiting; squirt sign on rectal examination. Older children: chronic constipation since birth; failure to thrive; massively distended abdomen. Management: rectal washouts to decompress; definitive pull-through procedure (Swenson, Duhamel, or Soave); in resource-limited settings: levelling colostomy then delayed pull-through. Most feared complication: HAEC (explosive diarrhoea, fever, abdominal distension: can be rapidly fatal).

Value-added response: In Nigerian practice, many children with Hirschsprung's disease present late (in infancy or childhood rather than the neonatal period) due to delayed healthcare-seeking. They may present with a massively dilated abdomen and malnutrition. Rectal washouts and nutritional rehabilitation before definitive surgery are essential to optimise the outcome.

14. **Basic response:** A high anorectal malformation in a male neonate (with a rectourethral fistula) is managed in three stages. Stage 1 (newborn period): a diverting sigmoid colostomy is fashioned to decompress the bowel and prevent faecal contamination of the urinary tract via the rectourethral fistula; the distal limb is used for a contrast study (distal colostogram) to define the anatomy of the malformation and fistula before definitive repair. Stage 2 (at 3 to 6 months of age, when the baby weighs approximately 10 kg): PSARP (posterior sagittal anorectoplasty: the Pena procedure) is performed; a posterior midline incision allows the rectum to be mobilised, the fistula to be divided and repaired, and the rectum to be placed accurately within the sphincter muscle complex. Stage 3 (6 to 8 weeks after PSARP): the colostomy is closed. Long-term complications: constipation, faecal soiling, and bladder dysfunction from pelvic nerve injury.

Value-added response: The quality of the sphincter muscle complex (assessed by electrical stimulation and MRI) is the most important determinant of long-term continence after PSARP. Patients with a well-developed sphincter mechanism, even with high lesions, can achieve social continence with bowel management programmes including bowel irrigation and dietary modification.

15. **Basic response:** Exomphalos: the sac is preserved and used as coverage in large defects; primary closure for small defects; silo for large defects; associated anomalies (cardiac and chromosomal) must be screened for and may preclude early surgery. Gastroschisis: the sac is absent and the bowel is directly exposed; immediate coverage of the bowel is essential (saline-soaked gauze and

plastic wrap); primary closure if abdominal domain allows; silo if not; surgery is more urgent as the bowel deteriorates rapidly. Key difference: exomphalos may be observed and treated semi-electively if the sac is intact; gastroschisis is always a neonatal surgical emergency. Both may require a silo for staged reduction, but the indication and urgency differ.

Value-added response: In settings where NICU care is limited (as in many Nigerian tertiary hospitals), the conservative management of exomphalos (painting the intact sac with antiseptic agents such as silver sulfadiazine or mercurochrome to allow gradual escharification and epithelialisation: the topical method) offers a viable alternative to surgery for giant exomphalos, allowing the baby to grow before eventual repair.

Answers to Pilot Questions [Series 1]

Part 1.1

1. Exomphalos: midline defect at umbilical ring; covered by a sac of amnion and peritoneum; may contain bowel and liver; associated with chromosomal and cardiac anomalies in up to 70 percent. Gastroschisis: defect to the right of the umbilicus; no covering sac; bowel directly exposed to amniotic fluid forming the peel; not associated with chromosomal anomalies.
2. Cover the exposed bowel immediately with warm saline-soaked gauze and wrap in a plastic bag or cling film to prevent heat and fluid loss. Pass a nasogastric tube to decompress the stomach. Establish IV access and give IV fluid resuscitation (the fluid losses from exposed bowel are massive). Keep the baby warm. Transfer immediately to a paediatric surgical unit.
3. Beckwith-Wiedemann syndrome is a congenital overgrowth syndrome characterised by macroglossia (large tongue), macrosomia (large body size), and neonatal hypoglycaemia (from pancreatic hyperplasia). It is associated with exomphalos (approximately 10 percent of Beckwith-Wiedemann cases have exomphalos) and with an increased risk of Wilms' tumour and hepatoblastoma.
4. A silo is a synthetic mesh bag (usually made of silastic or polyurethane) sutured to the fascial edges of the abdominal wall defect. The herniated bowel is placed inside the silo, which is then gradually reduced (squeezed down) by serial tightening over 7 to 14 days, allowing the abdominal domain to accommodate the contents progressively. Once the bowel is fully reduced, formal fascial closure is performed.
5. Short bowel syndrome (if bowel is resected for ischaemia), prolonged ileus (from the peel bowel: delayed gut motility may last weeks), catheter-related bloodstream infection from prolonged central venous access, sepsis, adhesive small bowel obstruction, and gastro-oesophageal reflux.

Part 1.2

1. Excessive drooling and salivation (upper pouch fills with secretions), choking and cyanosis during attempted feeds (aspiration), and respiratory distress. Diagnosis: a nasogastric tube cannot be passed beyond 10 to 12 cm; plain CXR shows the tube coiled in the blind upper oesophageal pouch; presence of gastric gas confirms a distal TOF.
2. VACTERL: Vertebral anomalies (hemivertebrae and sacral agenesis), Anorectal malformations, Cardiac defects (VSD most common), Tracheo-Oesophageal fistula, Renal anomalies (renal agenesis and horseshoe kidney), Limb defects (radial aplasia). Relevance: any neonate with OA must be systematically screened for all VACTERL components before surgery, as associated cardiac defects are the most common cause of death in these patients.
3. The double bubble sign is two air-filled bubbles on plain AXR: one in the stomach and one in the obstructed proximal duodenum, with no gas visible in the distal bowel. It is pathognomonic of duodenal atresia (or stenosis), indicating complete obstruction of the duodenum with no air passing distally.
4. Duodenal atresia: results from failure of recanalisation of the duodenum between the 8th and 10th weeks of gestation; strongly associated with Down syndrome (trisomy 21) in approximately 30 percent of cases. Jejunoileal atresia: results from an intrauterine vascular accident (mesenteric ischaemia with bowel infarction and resorption); not associated with chromosomal anomalies.
5. Duodenal atresia: duodenoduodenostomy (a side-to-side or diamond-shaped anastomosis between the proximal obstructed duodenum and the distal duodenum, bypassing the obstruction without resection). Jejunoileal atresia: resection of the atretic segment and end-to-end anastomosis of the two bowel ends; the proximal dilated bowel may need tapering or plication to improve peristalsis; bowel conservation is essential to avoid short bowel syndrome.

Part 1.3

1. At the 6th week of gestation, the midgut herniates into the umbilical cord and begins to rotate 270 degrees anticlockwise around the superior mesenteric artery. By the 10th week, the midgut returns to the abdominal cavity with the DJ flexure fixed to the left of the spine at the ligament of Treitz and the caecum positioned in the right iliac fossa. This creates a broad-based mesenteric attachment that prevents volvulus.
2. In malrotation, the midgut rotation is incomplete, leaving the DJ flexure to the right of the spine and the caecum in the epigastrium. The mesentery does not become broadly fixed and instead remains attached to the retroperitoneum by a narrow pedicle around the SMA. This narrow pedicle acts as an axis around which the entire midgut can twist (midgut volvulus), causing acute intestinal obstruction and bowel ischaemia.
3. Bilious (green) vomiting in a neonate indicates obstruction distal to the ampulla of Vater. Midgut volvulus is the most dangerous cause because it can lead to infarction and loss of the entire midgut within hours if untreated, resulting in short bowel syndrome or death. Any neonatal

bilious vomiting must be investigated urgently with an upper GI contrast study to exclude malrotation and volvulus.

4. The upper GI contrast study (barium or water-soluble contrast meal) is the investigation of choice. Findings in malrotation: the duodeno-jejunal flexure lies to the right of the spine (rather than to the left at the ligament of Treitz) and fails to cross the midline. In volvulus: the contrast column shows a corkscrew or spiral appearance of the duodenum and proximal jejunum as they twist around the SMA.

5. Step 1: untwist the volvulus counter-clockwise. Step 2: divide Ladd's bands (peritoneal bands from the caecum crossing the duodenum) to relieve extrinsic duodenal obstruction. Step 3: broaden the mesenteric base by placing small bowel on the right and colon on the left. Step 4: appendicectomy (caecum is now on the left: future appendicitis would not produce right iliac fossa signs).

Part 1.4

1. Neural crest cells normally migrate from the cranial neural crest to populate the enteric nervous system of the entire bowel from cranial to caudal. In Hirschsprung's disease, this migration arrests prematurely, leaving the distal bowel (most commonly the rectosigmoid) without ganglion cells. The aganglionic bowel is tonically contracted and cannot propagate peristaltic waves, creating a functional obstruction. The normal proximal bowel becomes massively distended (megacolon).

2. Failure to pass meconium within 48 hours of birth (normally 90 percent of neonates pass meconium within 24 hours), abdominal distension, and bilious vomiting. On rectal examination or after rectal washout: an explosive passage of gas and stool (the squirt sign) as the compressed proximal bowel decompresses through the relaxed aganglionic segment.

3. Rectal suction biopsy: a small sample of mucosa and submucosa is obtained from 2 to 3 cm above the dentate line using a suction biopsy instrument. Histology findings: absence of ganglion cells in the submucosal (Meissner's) plexus and hypertrophied, thickened acetylcholinesterase-positive nerve fibres. Histochemistry: positive acetylcholinesterase staining confirms the diagnosis. The biopsy must be taken above the normal hypoganglionic zone (the most distal 1 to 2 cm of the rectum is normally hypoganglionic).

4. Low lesion (below levator ani): rectum descends through the levator ani and external sphincter; good prognosis for faecal continence; treated by anoplasty in the newborn period. High lesion (above levator ani): rectum ends above the levator ani; poor sphincter muscle development; poor prognosis for continence; requires staged repair (colostomy, PSARP at 3 to 6 months, colostomy closure). Intermediate lesion: between low and high; variable prognosis.

5. Hirschsprung-associated enterocolitis (HAEC) is an acute inflammatory condition of the proximal dilated bowel in Hirschsprung's disease. It presents with explosive watery or bloody diarrhoea, fever, and rapidly progressive abdominal distension. It is dangerous because it can progress to bowel perforation, septic shock, and death within hours if not treated aggressively with IV fluids,

IV antibiotics, and immediate rectal washouts to decompress the colon. It can occur both before and after the pull-through procedure.

References and Attributions [Series 1]

Textual References

- Coran, A. G., Caldamone, A., Adzick, N. S., Krummel, T. M., Laberge, J. M. and Shamberger, R. (2012). Pediatric Surgery (7th ed.). Philadelphia: Elsevier Mosby.
- Puri, P. and Hollwarth, M. E. (2009). Pediatric Surgery: Diagnosis and Management. Berlin: Springer.
- Ameh, E. A. and Chirdan, L. B. (2011). Paediatric Surgery in the Tropics. Ibadan: Spectrum Books.
- Grosfeld, J. L., O'Neill, J. A., Fonkalsrud, E. W. and Coran, A. G. (2006). Pediatric Surgery (6th ed.). Philadelphia: Mosby Elsevier.

Figures, Plates, Tables, and Boxes

- Figure 1.1: Abdominal wall defects. AI-generated using the prompt: "Two-panel diagram: side-by-side illustrated comparison of exomphalos and gastroschisis with management flowchart showing immediate measures and diverging pathways (silo versus primary closure). Paediatric surgical diagram style." Suggested OER search string: "exomphalos omphalocele gastroschisis abdominal wall defect neonate management diagram OER."
- Figure 1.2: Intestinal atresia. AI-generated using the prompt: "Two-panel diagram: Gross classification of oesophageal atresia with Type C illustrated in detail and VACTERL box and management; duodenal atresia double bubble sign and jejunoileal atresia types with management. Paediatric surgical diagram style." Suggested OER search string: "oesophageal atresia TOF Gross classification duodenal atresia double bubble jejunoileal atresia apple-peel OER."
- Figure 1.3: Malrotation and midgut volvulus. AI-generated using the prompt: "Two-panel diagram: normal versus malrotation embryology diagrams and Ladd's bands; clinical features box, investigations box, and Ladd's procedure four-step flowchart with bowel viability assessment. Paediatric surgical diagram style." Suggested OER search string: "malrotation midgut volvulus Ladd's procedure embryology paediatric surgery OER."
- Figure 1.4: Hirschsprung's disease and anorectal malformations. AI-generated using the prompt: "Two-panel diagram: sagittal colon diagram showing aganglionic segment and megacolon with classification and management box; sagittal pelvis diagram showing low versus high ARM relative to levator ani with management flowchart. Paediatric surgical diagram style." Suggested

OER search string: "Hirschsprung disease pull-through anorectal malformations PSARP Pena procedure paediatric surgery OER."

Series 2: Groin and Scrotal Conditions

- **Part 2.1: Inguinal Hernia in Children**
    - Sub Part 2.1.1: Anatomy and pathophysiology
    - Sub Part 2.1.2: Clinical features and diagnosis
    - Sub Part 2.1.3: Management and complications
- **Part 2.2: Hydrocele**
    - Sub Part 2.2.1: Types and pathophysiology
    - Sub Part 2.2.2: Clinical features and diagnosis
    - Sub Part 2.2.3: Management
- **Part 2.3: Undescended Testis**
    - Sub Part 2.3.1: Embryology and classification
    - Sub Part 2.3.2: Clinical features and diagnosis
    - Sub Part 2.3.3: Management and complications
- **Part 2.4: Acute Scrotum**
    - Sub Part 2.4.1: Testicular torsion
    - Sub Part 2.4.2: Epididymo-orchitis and other causes
    - Sub Part 2.4.3: Management of the acute scrotum

Introduction

Groin and scrotal conditions are among the most common problems encountered in paediatric surgical practice. Many are closely related embryologically, as incomplete closure of the processus vaginalis underlies both inguinal hernia and hydrocele. Prompt diagnosis is essential because complications such as incarceration of a hernia and testicular torsion can result in irreversible organ loss if not treated urgently.

This Series covers the four major categories of groin and scrotal conditions in children: inguinal hernia, hydrocele, undescended testis, and the acute scrotum. Each section examines the embryological basis, clinical features, diagnostic approach, and management, with particular attention to the complications that arise from delayed or incorrect management.

Series Learning Outcomes

Upon completing this Series, you should be able to:

- **SLO 2.1:** describe the anatomy, clinical features, and management of inguinal hernia in children, and state the complications of incarceration,
- **SLO 2.2:** describe the types of hydrocele, their clinical features, and their management,
- **SLO 2.3:** describe the embryology, classification, clinical features, and management of undescended testis and its complications, and
- **SLO 2.4:** describe the causes of the acute scrotum, with emphasis on testicular torsion, and outline the management of each.

2.1 Inguinal Hernia in Children

2.1.1 Anatomy and pathophysiology

Inguinal hernia in children is almost always indirect (passing through the deep inguinal ring and along the inguinal canal), in contrast to adults in whom direct herniae are common. The underlying cause is a patent processus vaginalis (PPV): a peritoneal outpouching that accompanies the descending testis from the abdomen into the scrotum. Normally the PPV obliterates after testicular descent (by 1 to 2 years of age); if it remains open, abdominal contents can enter it, forming a congenital indirect inguinal hernia.

Inguinal hernia is much more common in males (approximately 9:1 male to female ratio) and in premature infants (in whom the PPV is less likely to have closed). It is more common on the right side (because the right testis descends later than the left). Bilateral herniae occur in approximately 10 to 20 percent of cases. The incidence is approximately 1 to 4 percent of term infants and up to 30 percent of premature infants.

Fill in the blank: The cause of inguinal hernia in children is a patent _____ vaginalis. The condition is more common on the _____ side because the right testis descends _____ than the left.

2.1.2 Clinical features and diagnosis

The classic presentation is an intermittent groin swelling that appears on standing, straining, or crying and disappears on lying down (reducible hernia). The swelling is smooth, non-tender, and can be felt to emerge from the external inguinal ring. The diagnosis is clinical. An irreducible (incarcerated) hernia presents as a tender, firm, non-reducible groin lump, often with vomiting

and abdominal distension from bowel obstruction. The overlying skin may be erythematous and oedematous in strangulation.

In girls, the hernia sac may contain the ovary and fallopian tube (the most common content in female inguinal herniae). An ovary incarcerated in the hernia sac can undergo torsion, making early repair essential. Transillumination is negative in a hernia (bowel is opaque) but positive in a hydrocele (fluid is translucent), helping to distinguish the two. Ultrasound is used when the clinical diagnosis is uncertain.

Fill in the blank: An inguinal hernia in a girl may contain the _____ and fallopian tube as its contents. Transillumination is _____ in a hernia (negative) but _____ in a hydrocele (positive), helping to distinguish the two.

2.1.3 Management and complications

All inguinal herniae in children should be repaired electively because the risk of incarceration is high (approximately 12 percent in the first year of life). In premature infants, repair is performed before discharge from the neonatal unit. The operation is herniotomy (high ligation of the hernial sac at the level of the deep inguinal ring, without mesh repair: the hernia in children is due to the PPV alone, not to abdominal wall weakness).

Incarcerated hernia: attempt gentle manual reduction (taxis) under sedation and analgesia. If successful, repair is planned within 24 to 48 hours (as oedema subsides). If reduction fails or strangulation is suspected, immediate surgery is required. Complications of incarceration include: bowel ischaemia and necrosis (requiring bowel resection), testicular atrophy (from compression of the testicular vessels in the inguinal canal), and ovarian infarction in girls. Recurrence after herniotomy is rare (less than 1 percent) when performed by a trained surgeon.

Fill in the blank: The surgical repair of inguinal hernia in children is called _____ (high ligation of the hernial sac), not herniorrhaphy, because the defect is due to the PPV and not abdominal wall _____. Incarceration of the hernia in the first year of life occurs in approximately _____ percent of cases.

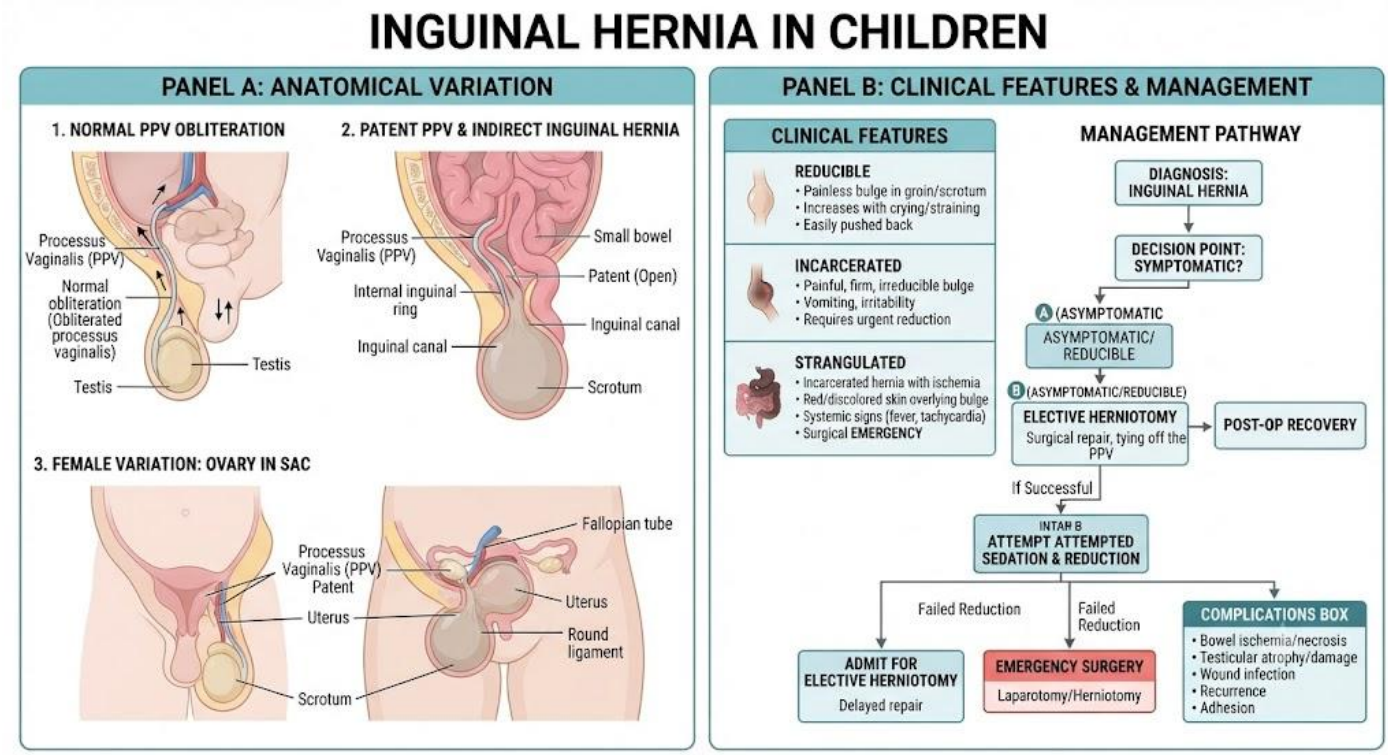

Figure 2.1: Pathophysiology, clinical features, and management of inguinal hernia in children

Pilot questions 2.1

1. Explain the embryological basis of indirect inguinal hernia in children. (Linked to SLO 2.1)
2. Describe the clinical features of a reducible and an incarcerated inguinal hernia in a child. (Linked to SLO 2.1)
3. Explain why herniotomy rather than herniorrhaphy is performed in children. (Linked to SLO 2.1)
4. Describe the management of an incarcerated inguinal hernia in a child. (Linked to SLO 2.1)
5. State the complications of a strangulated inguinal hernia in a child. (Linked to SLO 2.1)

2.2 Hydrocele

2.2.1 Types and pathophysiology

A hydrocele is a collection of fluid within the tunica vaginalis surrounding the testis. It results from an imbalance between secretion and absorption of fluid, or from a patent processus vaginalis allowing peritoneal fluid to track into the tunica vaginalis. In children, hydroceles are classified as communicating (the PPV remains open, allowing peritoneal fluid to flow freely into the tunica vaginalis: the hydrocele changes in size during the day, typically smaller in the morning and larger after activity) or non-communicating (the PPV has closed but fluid remains trapped: the hydrocele does not change in size).

A hydrocele of the cord occurs when a segment of the PPV in the inguinal canal remains patent and fluid accumulates within it, forming a cystic swelling in the groin or upper scrotum separate from the testis. An infantile hydrocele involves the entire tunica vaginalis from the internal ring to the testis. Most non-communicating hydroceles in neonates resolve spontaneously by 12 to 18 months of age as the fluid is reabsorbed. A communicating hydrocele does not resolve spontaneously and behaves like an indirect inguinal hernia in terms of surgical risk.

Fill in the blank: A _____ hydrocele has a patent PPV and changes in size during the day. Most non-communicating hydroceles in neonates resolve by _____ to 18 months of age. A hydrocele of the _____ is a cystic swelling in the inguinal canal separate from the testis.

2.2.2 Clinical features and diagnosis

A hydrocele presents as a smooth, non-tender scrotal swelling that transilluminates brilliantly (the most important clinical sign). Unlike a hernia, the upper border of a hydrocele can be clearly felt (you can get above it in the scrotum), whereas the upper border of an indirect inguinal hernia cannot be felt (it extends through the inguinal canal into the abdomen). The testis may not be separately palpable within the hydrocele.

A communicating hydrocele may be difficult to distinguish from an indirect inguinal hernia on examination alone; both share a patent PPV and both may reduce on lying. The key distinguishing feature is that a hydrocele transilluminates and contains only fluid (no bowel sounds on auscultation), whereas a hernia does not transilluminate (if bowel is the content). Ultrasound confirms the diagnosis when clinical examination is uncertain and also confirms that the testis is normal.

Fill in the blank: The most important clinical sign of a hydrocele is brilliant _____. Unlike a hernia, you can get _____ a hydrocele in the scrotum (its upper border is palpable). A communicating hydrocele differs from a hernia in that it contains only _____ and not bowel.

2.2.3 Management

Non-communicating hydroceles in neonates and infants under 12 to 18 months: observe, as the majority resolve spontaneously. Surgery is indicated if the hydrocele persists beyond 18 months of age, is large and causing discomfort, or is a hydrocele of the cord. Communicating hydroceles require surgery (at any age) because they will not resolve and carry a risk of incarceration of a hernia.

Surgical treatment: herniotomy (high ligation of the PPV at the deep inguinal ring) combined with eversion or excision of the hydrocele sac in the scrotum (Lord's procedure: plication of the sac without excision; Jaboulay's procedure: eversion of the sac behind the testis). Aspiration of a hydrocele is not recommended in children because of the high recurrence rate and the risk of missing a communicating hydrocele or a hernia. The operation is performed under general anaesthesia.

Fill in the blank: Non-communicating hydroceles are observed until _____ months; surgery is indicated if they persist beyond this age. Aspiration of a hydrocele is _____ recommended in children because of the high _____ rate.

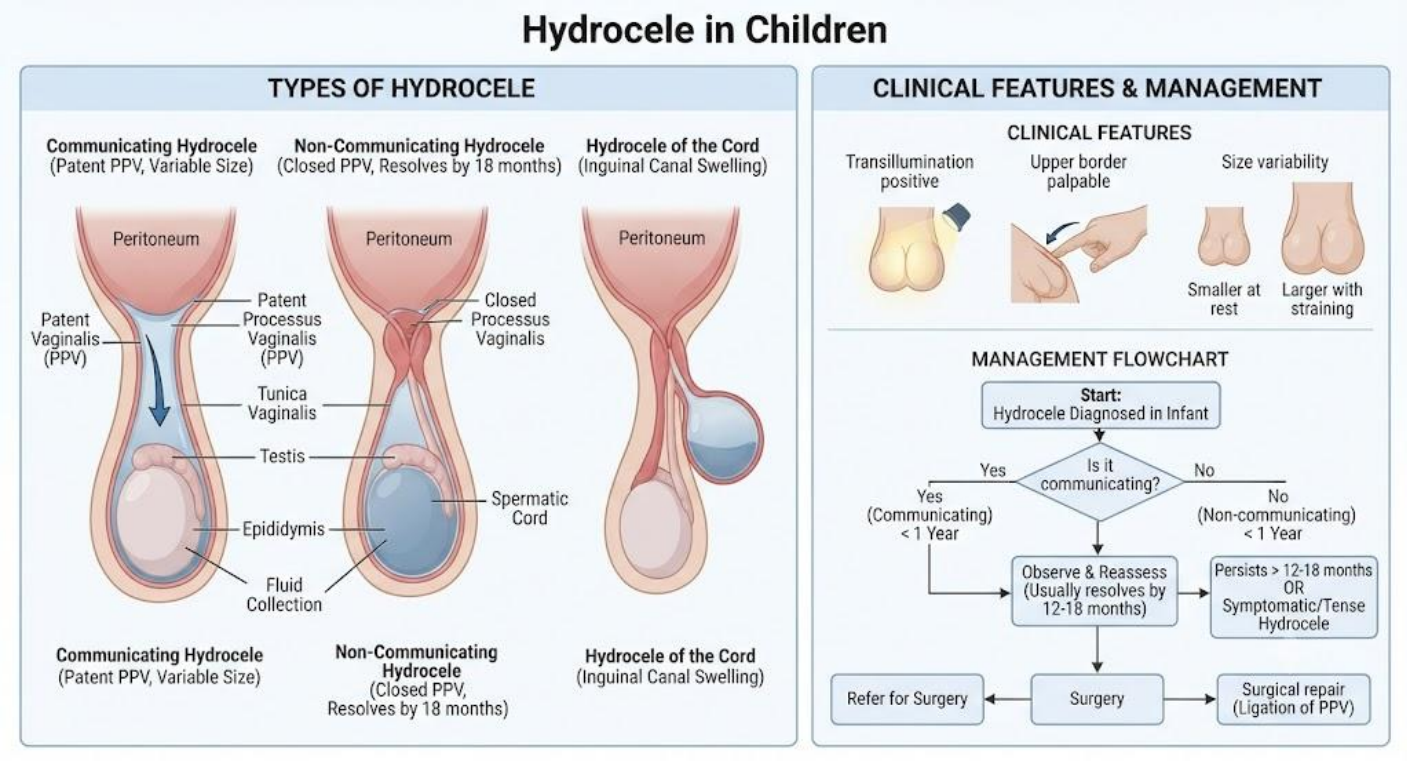

Figure 2.2: Types, clinical features, and management of hydrocele in children

Pilot questions 2.2

1. Distinguish a communicating from a non-communicating hydrocele in terms of pathophysiology and management. (Linked to SLO 2.2)
2. Describe the clinical signs that distinguish a hydrocele from an inguinal hernia. (Linked to SLO 2.2)
3. State the indications for surgical repair of a hydrocele in a child. (Linked to SLO 2.2)
4. Explain why aspiration is not recommended for hydrocele in children. (Linked to SLO 2.2)

5. Describe the surgical treatment of a communicating hydrocele. (Linked to SLO 2.2)

2.3 Undescended Testis

2.3.1 Embryology and classification

The testis develops in the retroperitoneum near the kidney and descends through the inguinal canal into the scrotum by the 32nd to 36th week of gestation, guided by the gubernaculum and influenced by androgens (testosterone and dihydrotestosterone) and the genitofemoral nerve. Descent is a two-stage process: transabdominal descent (influenced by Mullerian inhibiting substance: MIS) and inguinoscrotal descent (androgen-dependent). Failure of either stage results in an undescended testis (cryptorchidism).

Undescended testis (UDT) is classified by position: intra-abdominal (the testis remains in the abdomen: above the internal inguinal ring: 10 percent), canalicular (within the inguinal canal: 20 percent), emergent or pre-scrotal (at the external ring: 40 percent), and high scrotal (in the upper scrotum: the most common position: 30 percent). An ectopic testis is one that has left the normal path of descent and lies in an abnormal position (perineum, femoral triangle, base of penis, or superficial inguinal pouch: the most common ectopic site). A retractile testis is one that can be manually manipulated into the scrotum and remains there: it is not truly undescended and does not require surgery.

Fill in the blank: Testicular descent is guided by the _____ and is androgen-dependent. An _____ testis has left the normal path of descent and lies in an abnormal site. A _____ testis can be manipulated into the scrotum and remains there: it does not require surgery.

2.3.2 Clinical features and diagnosis

The scrotum on the affected side is underdeveloped (hypoplastic) and empty. The diagnosis is made by careful clinical examination in a warm room with warm hands: the examiner milks the testis down from the inguinal canal while the child is relaxed. A palpable testis in the inguinal canal can often be brought towards the scrotum but springs back when released (distinguishing it from a retractile testis, which stays in the scrotum). An impalpable testis (not palpable in any position: approximately 20 percent of UDT) requires further investigation.

Investigation of the impalpable testis: ultrasound (may locate an inguinal testis but is unreliable for intra-abdominal testes), MRI (better than ultrasound but not always available), and diagnostic laparoscopy (the gold standard for the impalpable testis: directly visualises whether the testis is intra-abdominal, absent, or has a blind-ending vas deferens and vessels indicating a vanishing testis). Human chorionic gonadotrophin (hCG) stimulation test (rise in testosterone after hCG injection confirms functioning testicular tissue).

Fill in the blank: The _____ testis (approximately 20 percent of UDT) is not palpable in any position and requires further investigation. The gold standard investigation for the impalpable testis is diagnostic _____.

2.3.3 Management and complications

Surgery is the definitive treatment and should be performed between 6 and 18 months of age (ideally by 12 months). Earlier surgery preserves fertility (the undescended testis undergoes progressive germ cell loss from the age of 6 to 12 months) and allows early detection of torsion and malignancy. The operation is orchidopexy: the testis is mobilised, the hernial sac (PPV) is ligated, and the testis is fixed in the scrotum in a sub-dartos pouch. Laparoscopic orchidopexy or the Fowler-Stephens procedure (dividing the testicular vessels to allow the testis to be brought down on collateral blood supply) is used for intra-abdominal testes.

Complications of undescended testis: impaired fertility (the most important complication: bilateral UDT carries a high risk of infertility if not corrected early), malignant transformation (the risk of testicular carcinoma is 3 to 8 times higher in UDT than in the general population: orchidopexy reduces but does not eliminate this risk), testicular torsion (increased risk due to abnormal fixation), psychological effects, and associated inguinal hernia (present in virtually all UDT from the patent PPV).

Fill in the blank: Orchidopexy should be performed between 6 and _____ months of age to preserve fertility. The risk of testicular carcinoma in UDT is _____ to 8 times higher than in the general population. The Fowler-_____ procedure divides the testicular vessels to bring an intra-abdominal testis into the scrotum.

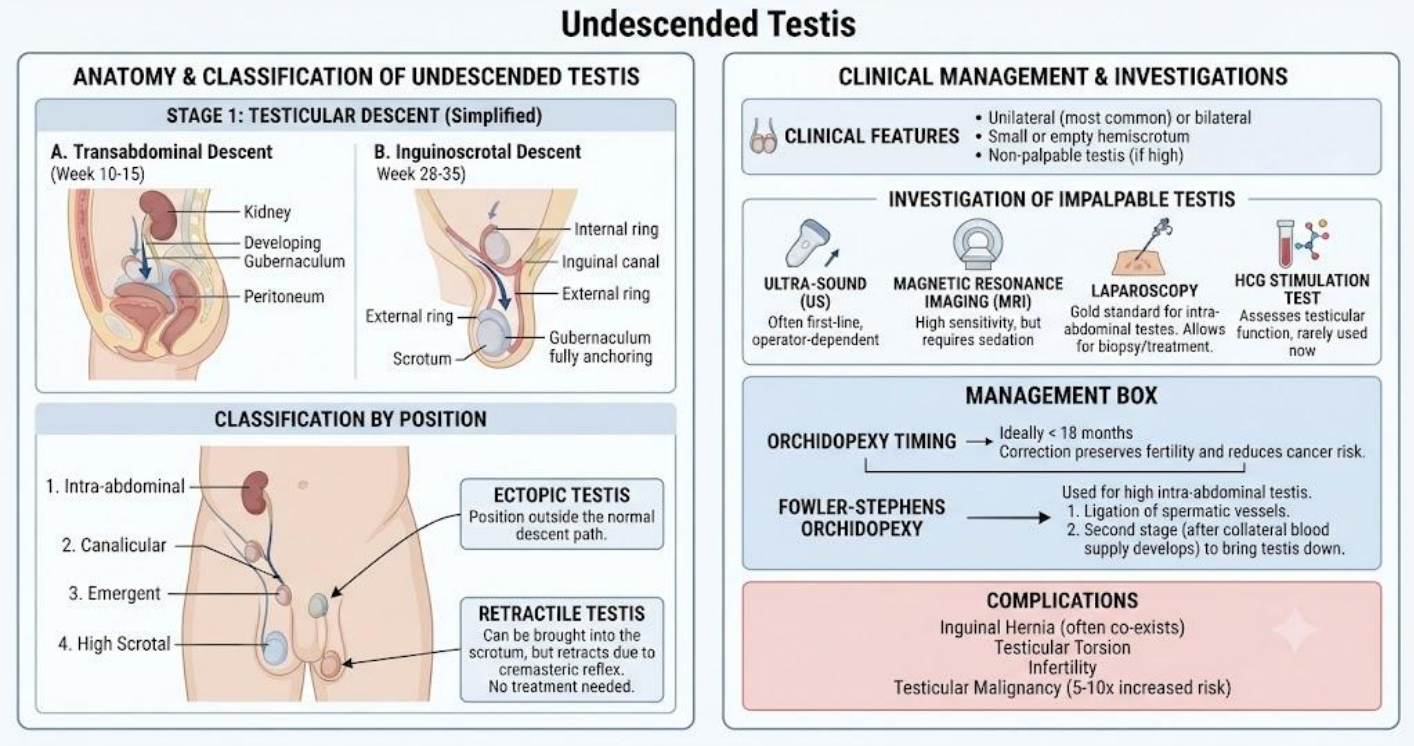

Figure 2.3: Embryology, classification, investigation, and management of undescended testis

Pilot questions 2.3

1. Describe the embryological basis of testicular descent and state why failure of descent occurs. (Linked to SLO 2.3)
2. Distinguish an undescended testis from a retractile testis and an ectopic testis. (Linked to SLO 2.3)
3. State the gold standard investigation for an impalpable testis and state what it may show. (Linked to SLO 2.3)
4. Describe orchidopexy and state the ideal timing for the operation. (Linked to SLO 2.3)
5. State the complications of an untreated undescended testis. (Linked to SLO 2.3)

2.4 Acute Scrotum

2.4.1 Testicular torsion

Testicular torsion is twisting of the spermatic cord, causing obstruction of venous outflow followed by arterial occlusion, resulting in testicular ischaemia and infarction. It is a urological emergency: the testis can be salvaged if detorsion is performed within 6 hours (salvage rate

above 90 percent); after 12 hours the salvage rate falls to approximately 50 percent; after 24 hours the testis is almost invariably infarcted. There are two types: extravaginal torsion (in neonates: the entire tunica vaginalis and its contents twist together before fixation to the scrotal wall) and intravaginal torsion (in pubertal boys: the testis twists within the tunica vaginalis due to a bell-clapper deformity: the testis hangs freely within a wide tunica vaginalis rather than being fixed posteriorly).

Clinical features of testicular torsion: sudden onset of severe unilateral scrotal pain (often waking the boy from sleep), nausea and vomiting, a tender swollen scrotal mass with the testis lying high and horizontally in the scrotum (horizontal lie: due to shortening of the cord from twisting), and absence of the cremasteric reflex (the most reliable clinical sign: the reflex is absent on the affected side). There may be a preceding history of similar episodes (intermittent torsion that spontaneously resolved).

Fill in the blank: Testicular torsion can be salvaged if detorsion is performed within _____ hours. The _____ deformity predisposes to intravaginal torsion in pubertal boys. Absence of the _____ reflex is the most reliable clinical sign of testicular torsion.

2.4.2 Epididymo-orchitis and other causes

Torsion of the hydatid of Morgagni (a small embryological remnant at the upper pole of the testis or epididymis) is the most common cause of acute scrotum in prepubertal boys. It presents with gradual onset of scrotal pain (less severe than testicular torsion), a tender nodule at the upper pole of the testis, and the blue dot sign (a blue-black nodule visible through the thin scrotal skin: pathognomonic when present). The cremasteric reflex is preserved. It resolves spontaneously and requires only analgesia.

Epididymo-orchitis (infection and inflammation of the epididymis and testis) is more common in sexually active adolescents and post-pubertal males. Causative organisms: Neisseria gonorrhoeae and Chlamydia trachomatis in sexually active adolescents; Escherichia coli and coliforms in younger boys (associated with urinary tract anomalies). Features: gradual onset of scrotal pain and swelling, fever, dysuria, and urethral discharge. The cremasteric reflex is preserved. Treatment: antibiotics guided by age and likely organism. Other causes of acute scrotum: idiopathic scrotal oedema (in prepubertal boys: bilateral scrotal erythema and swelling without tenderness: resolves spontaneously), Henoch-Schonlein purpura (HSP: scrotal involvement in approximately 2 to 8 percent of cases), and incarcerated inguinal hernia.

Fill in the blank: Torsion of the hydatid of Morgagni presents with a _____ dot sign (a blue-black nodule at the upper pole of the testis) and a _____ cremasteric reflex. Epididymo-orchitis in sexually active adolescents is most commonly caused by _____ gonorrhoeae and Chlamydia.

2.4.3 Management of the acute scrotum

Testicular torsion must be excluded in any boy presenting with an acute scrotum. The clinical diagnosis is sufficient to proceed directly to surgery: do not delay with imaging if torsion is

clinically suspected. Scrotal exploration is performed through a scrotal incision: if torsion is confirmed, the testis is detorted and assessed for viability (a viable testis: pink and pulsatile after 5 to 10 minutes of warm saline packs: is fixed in the scrotum with three-point fixation using non-absorbable sutures; a non-viable testis: black and non-pulsatile: is orchidectomised). Bilateral orchidopexy is performed (the contralateral testis is fixed at the same operation because the bell-clapper deformity is usually bilateral).

If the clinical picture is more consistent with torsion of the hydatid (blue dot sign, preserved cremasteric reflex, gradual onset, upper pole tenderness) and the diagnosis is confident, conservative management with analgesia is appropriate. If there is any doubt, surgical exploration is mandatory. Doppler ultrasound (showing absent or reduced blood flow in torsion versus increased flow in epididymo-orchitis) may be used when the diagnosis is uncertain, but must never delay surgery when torsion is suspected clinically.

Fill in the blank: In testicular torsion, bilateral _____ is performed because the bell-clapper deformity is usually _____. Doppler ultrasound _____ delay surgery when testicular torsion is clinically suspected.

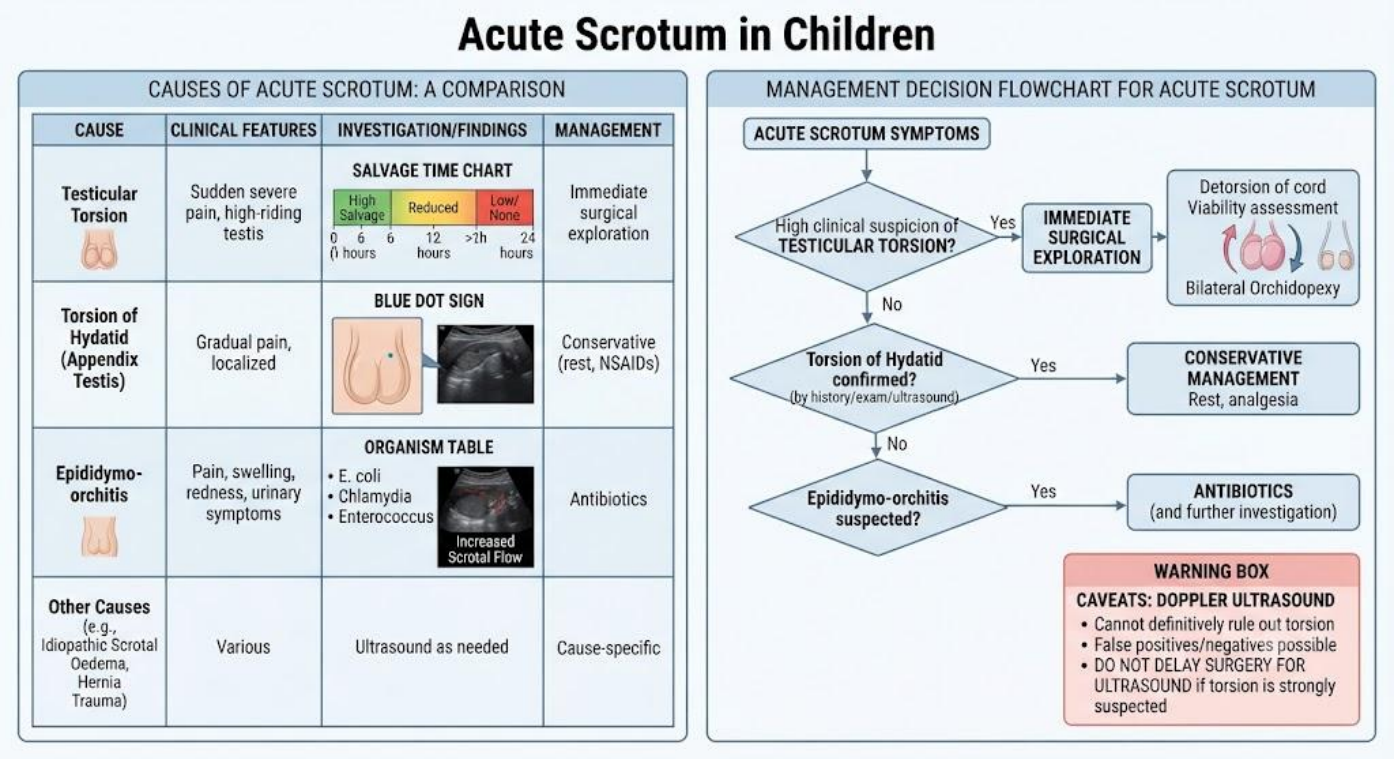

Figure 2.4: Causes and management of the acute scrotum in children

Pilot questions 2.4

1. Describe the clinical features of testicular torsion and state why it is a surgical emergency. (Linked to SLO 2.4)

2. Explain the bell-clapper deformity and state why bilateral orchidopexy is performed. (Linked to SLO 2.4)
3. Distinguish torsion of the hydatid of Morgagni from testicular torsion clinically. (Linked to SLO 2.4)
4. Describe the management of a boy presenting with a 4-hour history of acute scrotal pain. (Linked to SLO 2.4)
5. State the role of Doppler ultrasound in the acute scrotum and explain its limitation. (Linked to SLO 2.4)

Series Summary

This Series covered the major groin and scrotal conditions in children. Inguinal hernia in children is almost always indirect, caused by a patent processus vaginalis, and is treated by herniotomy; incarceration requires urgent management to prevent bowel necrosis and testicular atrophy. Hydroceles are classified as communicating or non-communicating; non-communicating hydroceles in infants may resolve by 18 months; communicating hydroceles and persistent hydroceles require herniotomy. Undescended testis should be corrected by orchidopexy between 6 and 18 months to preserve fertility; the impalpable testis is investigated by diagnostic laparoscopy; complications include infertility and malignancy.

You should now be able to describe the anatomy and management of inguinal hernia in children (SLO 2.1), describe the types and management of hydrocele (SLO 2.2), describe the embryology, classification, and management of undescended testis (SLO 2.3), and describe the causes and management of the acute scrotum with emphasis on testicular torsion (SLO 2.4). The acute scrotum is the most important condition in this Series: testicular torsion must always be excluded first, and any doubt mandates immediate scrotal exploration.

Glossary of Terms

- **Bell-clapper deformity:** an anatomical variant in which the testis is not fixed posteriorly within the tunica vaginalis but hangs freely within it like a bell clapper; predisposes to intravaginal testicular torsion.
- **Blue dot sign:** a blue-black discolouration visible through the thin scrotal skin of a prepubertal boy, located at the upper pole of the testis or epididymis; pathognomonic of torsion of the hydatid of Morgagni.

- **Communicating hydrocele:** a hydrocele in which the processus vaginalis remains patent, allowing peritoneal fluid to flow freely into the tunica vaginalis; the hydrocele varies in size during the day and does not resolve spontaneously.
- **Cremasteric reflex:** reflex contraction of the cremaster muscle causing elevation of the testis in response to stroking the medial thigh; absent on the side of testicular torsion and preserved in epididymo-orchitis and hydatid torsion.
- **Cryptorchidism:** the congenital failure of one or both testes to descend into the scrotum; synonymous with undescended testis.
- **Fowler-Stephens procedure:** a surgical technique for intra-abdominal undescended testis in which the testicular vessels are divided, allowing the testis to be brought into the scrotum based on collateral blood supply from the deferential artery.
- **Herniotomy:** high ligation of the hernial sac (patent processus vaginalis) at the level of the deep inguinal ring; the standard surgical repair for inguinal hernia and communicating hydrocele in children; does not involve mesh repair.
- **Hydatid of Morgagni:** a small embryological remnant (a vestige of the Mullerian duct) attached to the upper pole of the testis or head of the epididymis; prone to torsion in prepubertal boys, causing the acute scrotum.
- **Orchidopexy:** the surgical fixation of the testis in the scrotum; performed for undescended testis or testicular torsion; the testis is placed in a sub-dartos pouch and fixed with non-absorbable sutures.
- **Patent processus vaginalis (PPV):** failure of obliteration of the peritoneal outpouching that accompanies the descending testis; the common embryological basis of indirect inguinal hernia and communicating hydrocele in children.
- **Retractile testis:** a testis that can be manipulated into the scrotum and remains there; due to an overactive cremasteric reflex rather than true undescent; does not require surgical treatment but needs follow-up to ensure it does not become an ascending testis.

Concept Check Questions [Series 2]

Objective questions

1. Inguinal hernia in children is almost always _____ (indirect/direct), caused by a patent processus vaginalis. (Linked to SLO 2.1)

2. The most important clinical sign of a hydrocele is brilliant _____. (Linked to SLO 2.2)
3. Orchidopexy for undescended testis should be performed between 6 and _____ months of age to preserve fertility. (Linked to SLO 2.3)
4. Absence of the _____ reflex is the most reliable clinical sign of testicular torsion. (Linked to SLO 2.4)
5. The blue dot sign is pathognomonic of torsion of the _____ of Morgagni. (Linked to SLO 2.4)

Short-answer questions

6. Explain why herniotomy rather than herniorrhaphy is performed for inguinal hernia in children. (Linked to SLO 2.1)
7. Distinguish a communicating from a non-communicating hydrocele and state the management of each. (Linked to SLO 2.2)
8. State the gold standard investigation for an impalpable undescended testis and state what findings it may reveal. (Linked to SLO 2.3)
9. Describe the clinical features of testicular torsion. (Linked to SLO 2.4)
10. Explain why bilateral orchidopexy is performed when testicular torsion is found at scrotal exploration. (Linked to SLO 2.4)

Long-answer questions

11. Describe the management of an incarcerated inguinal hernia in a 3-month-old infant, including the complications of delay. (Linked to SLO 2.1)
12. Describe the embryology, classification, and management of undescended testis. (Linked to SLO 2.3)
13. Describe the causes of the acute scrotum in children and distinguish testicular torsion from its mimics. (Linked to SLO 2.4)
14. Describe the intraoperative findings and decisions during scrotal exploration for acute scrotum. (Linked to SLO 2.4)

15. Compare inguinal hernia and communicating hydrocele in terms of pathophysiology, clinical features, and management. (Linked to SLO 2.1 and 2.2)

Answers to Concept Check Questions [Series 2]

Objective questions

1. Indirect.
2. Transillumination.
3. 18.
4. Cremasteric.
5. Hydatid.

Short-answer questions

6. In children, the hernia is caused entirely by the patent processus vaginalis; there is no weakness of the abdominal wall (which is why recurrence is rare after herniotomy). Herniotomy (high ligation of the sac at the deep inguinal ring) corrects the defect without requiring mesh. Herniorrhaphy (repair of the abdominal wall with or without mesh) is used in adults where the posterior wall of the inguinal canal is weak.
7. Communicating hydrocele: the PPV remains open; peritoneal fluid flows freely into the tunica vaginalis; the hydrocele varies in size during the day (smaller in the morning); does not resolve spontaneously; requires herniotomy at any age. Non-communicating hydrocele: the PPV is closed; fluid is trapped within the tunica; the size does not change; most resolve spontaneously by 18 months; surgery only if it persists beyond 18 months or causes symptoms.
8. Diagnostic laparoscopy is the gold standard. It may reveal: (1) an intra-abdominal testis (present above the internal ring: can proceed to laparoscopic orchidopexy or plan Fowler-Stephens procedure); (2) a blind-ending vas deferens and vessels (vanishing testis: no further surgery needed); (3) a testis at or near the internal ring (can often be brought down in a single stage).
9. Sudden onset severe unilateral scrotal pain (often waking the boy from sleep), nausea and vomiting, a tender swollen scrotal mass, the testis lying high and horizontally in the

scrotum (horizontal lie from cord shortening), and absence of the cremasteric reflex on the affected side.

10. The bell-clapper deformity (the anatomical predisposition to torsion) is almost always bilateral. If only the affected testis is fixed, the contralateral testis remains at high risk of future torsion. Bilateral orchidopexy at the same operation prevents future torsion of the contralateral testis, which if it were to occur could result in bilateral testicular loss.

Long-answer questions

11. **Basic response:** Attempt gentle manual reduction (taxis) under sedation (IV morphine or intranasal midazolam) and with an ice pack applied to the groin to reduce oedema. The hernia is reduced by steady gentle pressure on the fundus of the sac. If successful, admit the infant and perform herniotomy within 24 to 48 hours (once oedema subsides). If taxis fails or strangulation is suspected (erythematous overlying skin, systemic signs of sepsis), proceed to immediate emergency herniotomy. At surgery: the sac is opened, the contents assessed (viable bowel: return to abdomen; non-viable bowel: resect and anastomose or create stoma), and the sac is ligated at the deep inguinal ring. Complications of delay: bowel ischaemia and necrosis requiring resection, testicular atrophy (compression of testicular vessels in the inguinal canal), and ovarian infarction in girls.

Value-added response: Incarceration is most common in the first year of life (approximately 12 percent incidence in this age group) and in premature infants. This is the primary reason why all inguinal herniae in children, including those in premature infants, should be repaired electively before discharge from the neonatal unit, rather than waiting until the child is older.

12. **Basic response:** Embryology: the testis develops near the kidney and descends in two stages: transabdominal (influenced by MIS from the Sertoli cells) and inguinoscrotal (androgen-dependent, guided by the gubernaculum). Descent is normally complete by 32 to 36 weeks gestation. Classification by position: intra-abdominal (10 percent), canalicular (20 percent), emergent or pre-scrotal (40 percent), high scrotal (30 percent). Ectopic testis: lies outside the normal descent path (most commonly in the superficial inguinal pouch). Retractile testis: milks into scrotum and stays: no surgery. Management: orchidopexy between 6 and 18 months (ideally by 12 months); mobilise testis, ligate PPV, fix in sub-dartos pouch with three non-absorbable sutures. Intra-abdominal testis: diagnostic laparoscopy; laparoscopic orchidopexy or Fowler-Stephens procedure. Complications of untreated UDT: infertility (most important), malignancy (3 to 8 times increased risk), torsion, and associated inguinal hernia.

Value-added response: Early orchidopexy (by 12 months) is the most important factor in preserving fertility: histological studies show progressive germ cell loss from the undescended testis from 6 months of age onwards, and this loss is significantly reduced when the testis is placed in the scrotum before 12 months.

13. **Basic response:** Causes of the acute scrotum: testicular torsion (most dangerous: sudden severe pain, high horizontal testis, absent cremasteric reflex: surgical emergency), torsion of the hydatid of Morgagni (most common in prepubertal boys: gradual onset, upper pole tenderness, blue dot sign, preserved cremasteric reflex: conservative management), epididymo-orchitis (gradual onset, fever, dysuria, preserved cremasteric reflex: antibiotics), idiopathic scrotal oedema (bilateral erythema and swelling without tenderness: resolves spontaneously), Henoch-Schonlein purpura, and incarcerated inguinal hernia. Distinguishing torsion: sudden onset, absent cremasteric reflex, and high horizontal testis strongly suggest torsion; any doubt mandates immediate scrotal exploration without waiting for imaging.

Value-added response: In pubertal boys, epididymo-orchitis and torsion can be very difficult to distinguish clinically: both present with acute scrotal pain and swelling. The rule of thumb in paediatric surgery is: if there is any clinical doubt, operate. The morbidity of an unnecessary scrotal exploration is far less than the loss of a testis from a missed torsion.

14. **Basic response:** A scrotal incision is made over the affected side. The testis and tunica vaginalis are delivered into the wound. If torsion is confirmed (twisted cord): untwist the cord manually, wrap the testis in warm saline-soaked packs, and wait 5 to 10 minutes to assess viability. A viable testis (pink, pulsatile, with return of colour after warming) is fixed in the scrotum in a sub-dartos pouch using three non-absorbable sutures (polypropylene or nylon) at three points. A non-viable testis (remains black and non-pulsatile after 10 minutes) is removed by orchidectomy (ligation of the cord at the internal ring). Regardless of the finding, the contralateral testis is then explored through a separate scrotal incision and fixed with three non-absorbable sutures (bilateral orchidopexy).

Value-added response: If hydatid torsion is found (a small infarcted nodule at the upper pole): excise the hydatid. If the testis is normal and the diagnosis was not torsion: exploration has not harmed the patient and should never be considered unnecessary: the consequences of a missed torsion always outweigh those of a negative exploration.

15. **Basic response:** Pathophysiology: both are caused by a patent processus vaginalis. In hernia, the PPV allows abdominal contents (bowel or omentum) to enter the hernial sac. In communicating hydrocele, the PPV allows peritoneal fluid to track into the tunica vaginalis. Clinical features: inguinal hernia presents as a groin swelling that does not transilluminate; upper border cannot be felt. Communicating hydrocele: scrotal swelling

that transilluminates brilliantly; upper border palpable; no bowel sounds; varies in size. Management: inguinal hernia requires herniotomy at any age (risk of incarceration). Communicating hydrocele: herniotomy at any age (will not resolve spontaneously). Non-communicating hydrocele: observe until 18 months (the majority resolve spontaneously).

Value-added response: The distinction between a communicating hydrocele and an indirect inguinal hernia can be very difficult, as both have a patent PPV. In practice, if examination under anaesthesia confirms that only fluid is entering the sac (no bowel reducible), the operation is the same: herniotomy at the deep ring with hydrocele sac management in the scrotum. Some surgeons argue that all communicating hydroceles should be treated the same as herniae.

Answers to Pilot Questions [Series 2]

Part 2.1

6. During fetal development, the processus vaginalis is a peritoneal outpouching that accompanies the descending testis from the abdomen, through the inguinal canal, into the scrotum. It normally obliterates after testicular descent, leaving the tunica vaginalis around the testis. If the PPV remains patent, abdominal contents can enter it, forming a congenital indirect inguinal hernia.
7. Reducible hernia: intermittent groin swelling appearing on straining, coughing, or crying; disappears on lying down; smooth and non-tender; emerging from the external inguinal ring; upper border cannot be felt (extends into the inguinal canal). Incarcerated hernia: a tender, firm, non-reducible groin lump; the child is distressed; vomiting and abdominal distension from bowel obstruction; overlying skin may be erythematous and oedematous in strangulation.
8. In children, the hernia is caused entirely by the patent processus vaginalis; the abdominal wall is structurally normal. Herniotomy (high ligation of the hernial sac at the deep inguinal ring) corrects the underlying defect and is all that is required. Herniorrhaphy (repair of the posterior wall of the inguinal canal with or without mesh) is performed in adults because they have an additional abdominal wall weakness; this weakness is not present in children.
9. Attempt gentle manual reduction (taxis) under adequate sedation and analgesia with the child in the Trendelenburg position. If successful, herniotomy is scheduled within 24 to 48 hours once oedema subsides. If taxis fails or strangulation is suspected (erythematous skin, systemic sepsis), proceed immediately to emergency herniotomy. At surgery, the sac contents are assessed for viability and the sac is ligated at the deep inguinal ring.

10. Bowel ischaemia and necrosis (requiring bowel resection with risk of short bowel syndrome), testicular atrophy (from compression of testicular vessels in the inguinal canal by the incarcerated hernia), ovarian infarction in girls (the ovary is particularly prone to torsion when trapped in the hernia sac), and wound infection.

Part 2.2

11. Communicating hydrocele: patent PPV allows peritoneal fluid to flow freely into the tunica vaginalis; size varies during the day (typically smaller in the morning, larger after activity or crying); does not resolve spontaneously; requires herniotomy. Non-communicating hydrocele: PPV is closed; fluid trapped within the tunica; size does not change; majority resolve spontaneously by 12 to 18 months; surgery only if persistent or symptomatic after 18 months.
12. A hydrocele transilluminates brilliantly (fluid transmits light: the scrotum glows uniformly). An inguinal hernia does not transilluminate (bowel is opaque). The upper border of a hydrocele is palpable (you can get above it in the scrotum). The upper border of an inguinal hernia cannot be felt (it extends through the inguinal canal). A hydrocele does not have bowel sounds on auscultation; a hernia containing bowel does.
13. Surgery is indicated for: (1) communicating hydrocele (at any age: will not resolve and carries risk of hernia incarceration); (2) non-communicating hydrocele persisting beyond 18 months; (3) a large hydrocele causing discomfort regardless of age; (4) hydrocele of the cord (does not resolve spontaneously).
14. Aspiration is not recommended in children because: (1) the recurrence rate is very high (the fluid re-accumulates rapidly as the underlying PPV is not corrected); (2) it may miss a communicating hydrocele or hernia; (3) it carries risks of infection and haematoma; and (4) it is unnecessary as surgery (herniotomy) is curative.
15. Herniotomy (high ligation of the PPV at the level of the deep inguinal ring) under general anaesthesia. This corrects the underlying communicating defect. In addition, the hydrocele sac in the scrotum is managed by eversion (Jaboulay's procedure: the sac is everted behind the testis so the secreting surface faces outward and fluid cannot re-accumulate) or plication (Lord's procedure: the sac is plicated without excision to reduce its volume).

Part 2.3

1. The testis develops near the kidney and descends through two stages. Stage 1 (transabdominal descent): the testis moves from the retroperitoneum to the internal inguinal ring, guided by the gubernaculum and regulated by Mullerian inhibiting substance (MIS) from Sertoli cells. Stage 2 (inguinoscrotal descent): the testis passes through the inguinal canal into the scrotum, dependent on testosterone and dihydrotestosterone stimulating the genitofemoral nerve. Failure of descent can occur at either stage: androgen deficiency or insensitivity, gubernacular abnormalities, or mechanical obstruction are common causes.

2. Undescended testis: the testis lies along the normal path of descent but has not reached the scrotum (intra-abdominal, canalicular, emergent, or high scrotal); cannot be manipulated into the scrotum; requires orchidopexy. Retractile testis: an overactive cremasteric reflex draws the testis out of the scrotum temporarily; can be milked into the scrotum with gentle pressure and remains there without being held; no surgery required but annual follow-up is needed. Ectopic testis: has descended beyond the external ring but lies in an abnormal site outside the normal descent path (superficial inguinal pouch most common); requires orchidopexy to place it in the scrotum.

3. Diagnostic laparoscopy is the gold standard for the impalpable testis. Findings may include: (1) an intra-abdominal testis (viable testis above the internal ring: proceed to laparoscopic orchidopexy or plan Fowler-Stephens procedure); (2) a blind-ending vas deferens and vessels disappearing into the internal ring (vanishing testis: no further surgery needed as no functioning testicular tissue remains); (3) testicular vessels and vas entering the internal ring (testis is in the canal or just below the ring: one-stage orchidopexy is feasible).

4. Orchidopexy: the testis is approached through an inguinal incision; the testis is mobilised, the PPV (hernial sac) is ligated at the deep ring, and a sub-dartos pouch is created in the scrotum; the testis is placed in the pouch and fixed with three non-absorbable sutures. Ideal timing: 6 to 18 months, ideally by 12 months of age. Earlier surgery preserves spermatogonial development (germ cell loss begins from 6 months) and allows long-term testicular surveillance for malignancy.

5. Infertility (the most important complication: bilateral UDT has a high risk of azoospermia if uncorrected), malignancy (3 to 8 times increased risk of testicular carcinoma: seminoma most common: orchidopexy reduces but does not eliminate this risk), testicular torsion (increased risk due to abnormal fixation of the UDT), associated inguinal hernia (patent PPV present in virtually all UDT), and psychological effects from an empty scrotum.

Part 2.4

1. Sudden onset of severe unilateral scrotal pain (often waking the boy from sleep), nausea and vomiting, a tender swollen hemiscrotum, the testis lying high and horizontally in the scrotum (horizontal lie from cord shortening by twisting), and absent cremasteric reflex on the affected side. It is a surgical emergency because the testis can be salvaged in more than 90 percent of cases if detorsion occurs within 6 hours but is almost invariably infarcted after 24 hours.
2. The bell-clapper deformity is a high insertion of the tunica vaginalis onto the spermatic cord, causing the testis to hang freely within the tunica like a bell clapper rather than being fixed posteriorly. This allows the testis to rotate freely around the cord. The deformity is almost always bilateral: even if only one testis has torted, the other is at equal risk. Bilateral orchidopexy at the same operation prevents future torsion of the contralateral testis, which, if it were to occur and result in infarction, would leave the patient with no functioning testicular tissue.
3. Hydatid torsion: gradual or moderate onset; upper pole tenderness only; blue dot sign (blue-black nodule visible through scrotal skin); cremasteric reflex preserved; patient not as systemically unwell; resolves with analgesia. Testicular torsion: sudden severe onset; entire hemiscrotum tender; no blue dot sign; cremasteric reflex absent; high horizontal testis; patient in severe distress. In any case of doubt, exploration is mandatory.
4. Assume testicular torsion until proven otherwise. Do not delay with Doppler ultrasound if the clinical suspicion is high. Establish IV access, give analgesia, and proceed immediately to scrotal exploration. Intraoperatively: assess the cord; if twisted, untwist and assess testicular viability after 5 to 10 minutes of warm saline packs. Fix a viable testis in a sub-dartos pouch; perform orchidectomy for a non-viable testis. Always perform contralateral orchidopexy at the same operation.
5. Doppler ultrasound assesses blood flow to the testis: absent or markedly reduced flow suggests torsion; increased flow suggests epididymo-orchitis or hydatid torsion. Limitation: Doppler must never delay surgical exploration when torsion is clinically suspected, because: (1) early torsion may show residual flow on Doppler; (2) the time taken to arrange and perform the scan may allow progression from a viable to an infarcted testis; (3) a false-negative result leads to dangerous delay.

References and Attributions [Series 2]

Textual References

1. Coran, A. G., Caldamone, A., Adzick, N. S., Krummel, T. M., Laberge, J. M. and Shamberger, R. (2012). Pediatric Surgery (7th ed.). Philadelphia: Elsevier Mosby.
2. Puri, P. and Hollwarth, M. E. (2009). Pediatric Surgery: Diagnosis and Management. Berlin: Springer.
3. Ameh, E. A. and Chirdan, L. B. (2011). Paediatric Surgery in the Tropics. Ibadan: Spectrum Books.
4. Hutson, J. M., O'Brien, M., Woodward, A. A. and Beasley, S. W. (2008). Jones' Clinical Paediatric Surgery (6th ed.). Oxford: Blackwell Publishing.

Figures, Plates, Tables, and Boxes

5. Figure 2.1: Inguinal hernia in children. AI-generated using the prompt: "Two-panel diagram: three illustrations showing normal PPV obliteration, patent PPV with indirect inguinal hernia, and female variation with ovary in sac; management flowchart with elective herniotomy and incarcerated hernia management. Paediatric surgical diagram style." Suggested OER search string: "inguinal hernia children processus vaginalis herniotomy incarceration paediatric surgery OER."
6. Figure 2.2: Hydrocele in children. AI-generated using the prompt: "Two-panel diagram: three illustrated diagrams showing communicating hydrocele, non-communicating hydrocele, and hydrocele of the cord; clinical features box and management flowchart with surgical treatment box. Paediatric surgical diagram style." Suggested OER search string: "hydrocele children communicating non-communicating processus vaginalis herniotomy management OER."
7. Figure 2.3: Undescended testis. AI-generated using the prompt: "Two-panel diagram: testicular descent two-stage diagram and classification diagram showing positions (intra-abdominal, canalicular, emergent, high scrotal) with ectopic and retractile boxes; investigation box (ultrasound, MRI, laparoscopy, hCG) and management box with complications. Paediatric surgical diagram style." Suggested OER search string: "undescended testis cryptorchidism orchidopexy Fowler-Stephens impalpable testis paediatric surgery OER."
8. Figure 2.4: Acute scrotum. AI-generated using the prompt: "Two-panel diagram: four-row comparison table of causes (testicular torsion with salvage time chart, hydatid torsion with blue dot sign, epididymo-orchitis, other causes); management decision flowchart (torsion: immediate exploration and bilateral orchidopexy; hydatid: conservative;

Doppler warning box). Paediatric surgical diagram style." Suggested OER search string: "testicular torsion acute scrotum children orchidopexy hydatid Morgagni epididymo-orchitis OER."

Series 3: Hepatobiliary and Jaundice Disorders

- **Part 3.1: Approach to Jaundice in Children**
    - Sub Part 3.1.1: Physiology and classification of jaundice
    - Sub Part 3.1.2: Neonatal jaundice: physiological and pathological
    - Sub Part 3.1.3: Investigation of prolonged neonatal jaundice
- **Part 3.2: Biliary Atresia**
    - Sub Part 3.2.1: Pathology and classification
    - Sub Part 3.2.2: Clinical features and diagnosis
    - Sub Part 3.2.3: Management and outcomes
- **Part 3.3: Choledochal Cyst**
    - Sub Part 3.3.1: Classification and pathophysiology
    - Sub Part 3.3.2: Clinical features and diagnosis
    - Sub Part 3.3.3: Management and complications
- **Part 3.4: Congenital Infantile Hypertrophic Pyloric Stenosis**
    - Sub Part 3.4.1: Pathology and epidemiology
    - Sub Part 3.4.2: Clinical features and diagnosis
    - Sub Part 3.4.3: Management

Introduction

Hepatobiliary disorders and jaundice in children present unique diagnostic and management challenges compared with adults. Neonatal jaundice is extremely common and must be carefully evaluated to distinguish physiological from pathological causes. Biliary atresia and choledochal

cyst, if undiagnosed, lead to progressive hepatic fibrosis and biliary cirrhosis, making early recognition and surgical intervention essential.

This Series covers the major hepatobiliary and jaundice disorders encountered in paediatric surgical practice. We begin with the clinical approach to jaundice in children, then address biliary atresia and choledochal cyst in detail. We close with congenital infantile hypertrophic pyloric stenosis (IHPS), which, while not a biliary condition, is included in this Series because its presenting feature of persistent vomiting can be confused with biliary obstruction and because it is one of the most common surgical conditions of infancy.

Series Learning Outcomes

Upon completing this Series, you should be able to:

- **SLO 3.1:** classify jaundice in children and distinguish physiological from pathological neonatal jaundice, and describe the investigation of prolonged neonatal jaundice,
- **SLO 3.2:** describe the pathology, clinical features, diagnosis, and surgical management of biliary atresia,
- **SLO 3.3:** describe the classification, clinical features, and management of choledochal cyst and its complications, and
- **SLO 3.4:** describe the pathology, clinical features, diagnosis, and surgical management of infantile hypertrophic pyloric stenosis.

3.1 Approach to Jaundice in Children

3.1.1 Physiology and classification of jaundice

Jaundice (icterus) is yellow discolouration of the skin, sclerae, and mucous membranes caused by elevated serum bilirubin (above 35 to 40 micromol/L in neonates and above 17 micromol/L in older children and adults). Bilirubin is produced by the breakdown of haem from red blood cells, transported bound to albumin to the liver, conjugated by hepatic UDP-glucuronosyltransferase (UGT1A1), and excreted in bile as conjugated (direct) bilirubin. Failure at any step leads to jaundice.

Jaundice is classified by mechanism: pre-hepatic (excess bilirubin production: haemolysis: unconjugated hyperbilirubinaemia), hepatic (failure of conjugation or intrahepatic excretion: liver parenchymal disease: mixed or conjugated hyperbilirubinaemia), and post-hepatic or cholestatic (obstruction of bile flow from the liver to the duodenum: conjugated

hyperbilirubinaemia with pale stools and dark urine). In children, the post-hepatic group is of greatest surgical interest, as biliary atresia and choledochal cyst are the principal surgical causes.

Fill in the blank: Jaundice is visible when serum bilirubin exceeds _____ to 40 micromol/L in neonates. Post-hepatic jaundice is characterised by conjugated hyperbilirubinaemia with _____ stools and _____ urine.

3.1.2 Neonatal jaundice: physiological and pathological

Physiological jaundice occurs in approximately 60 percent of term neonates and 80 percent of preterm neonates, appearing after 24 hours of life, peaking at 72 to 96 hours (day 3 to 4), and resolving by day 10 to 14 in term infants (longer in preterm infants). It results from increased red cell breakdown (fetal haemoglobin replacement), immature hepatic UGT1A1 activity, and increased enterohepatic circulation. It is unconjugated (indirect) hyperbilirubinaemia. Treatment: phototherapy (converts unconjugated bilirubin to water-soluble isomers excreted in bile and urine without conjugation); exchange transfusion for severe cases.

Pathological jaundice is suggested by: jaundice appearing within 24 hours of birth (haemolytic disease: Rh or ABO incompatibility), jaundice persisting beyond 14 days in a term infant (prolonged neonatal jaundice: requires investigation), conjugated jaundice at any age (direct bilirubin above 20 percent of total: always pathological: suggests biliary obstruction or hepatocellular disease), pale or acholic stools (biliary obstruction), dark urine, hepatosplenomegaly, or a serum bilirubin above the treatment threshold for gestational age.

Fill in the blank: Physiological jaundice appears after _____ hours of life and resolves by day _____ to 14 in term infants. Jaundice persisting beyond _____ days in a term infant requires investigation as it may be pathological.

3.1.3 Investigation of prolonged neonatal jaundice

The key investigation in prolonged neonatal jaundice is the split bilirubin (conjugated versus unconjugated fractions): a conjugated bilirubin above 20 percent of total bilirubin is always abnormal and mandates urgent investigation to exclude biliary atresia (in which early surgery: before 8 weeks of age: is essential to achieve the best outcome). Other investigations: full blood count and blood film (haemolysis), Coombs test (immune-mediated haemolysis), thyroid function tests (hypothyroidism: a cause of prolonged unconjugated jaundice), liver function tests, urine reducing substances (galactosaemia), TORCH screen (congenital infection), ultrasound abdomen (gallbladder morphology: absent or small gallbladder in biliary atresia), and hepatobiliary scintigraphy (HIDA scan).

The HIDA (hepatobiliary iminodiacetic acid) scan assesses bile flow: in biliary atresia, the isotope is taken up by hepatocytes but not excreted into the duodenum (no bowel activity); in neonatal hepatitis, excretion is delayed but eventually occurs. Liver biopsy (percutaneous or operative) is the most definitive investigation: in biliary atresia it shows bile duct proliferation and cholestasis; in neonatal hepatitis it shows giant cell transformation. Intraoperative cholangiogram is performed at the time of the Kasai procedure to confirm the diagnosis.

Fill in the blank: A conjugated bilirubin above _____ percent of total bilirubin in a neonate is always pathological. On the HIDA scan, biliary atresia shows isotope uptake by hepatocytes but no _____ into the duodenum. Liver biopsy in biliary atresia shows bile duct _____ and cholestasis.

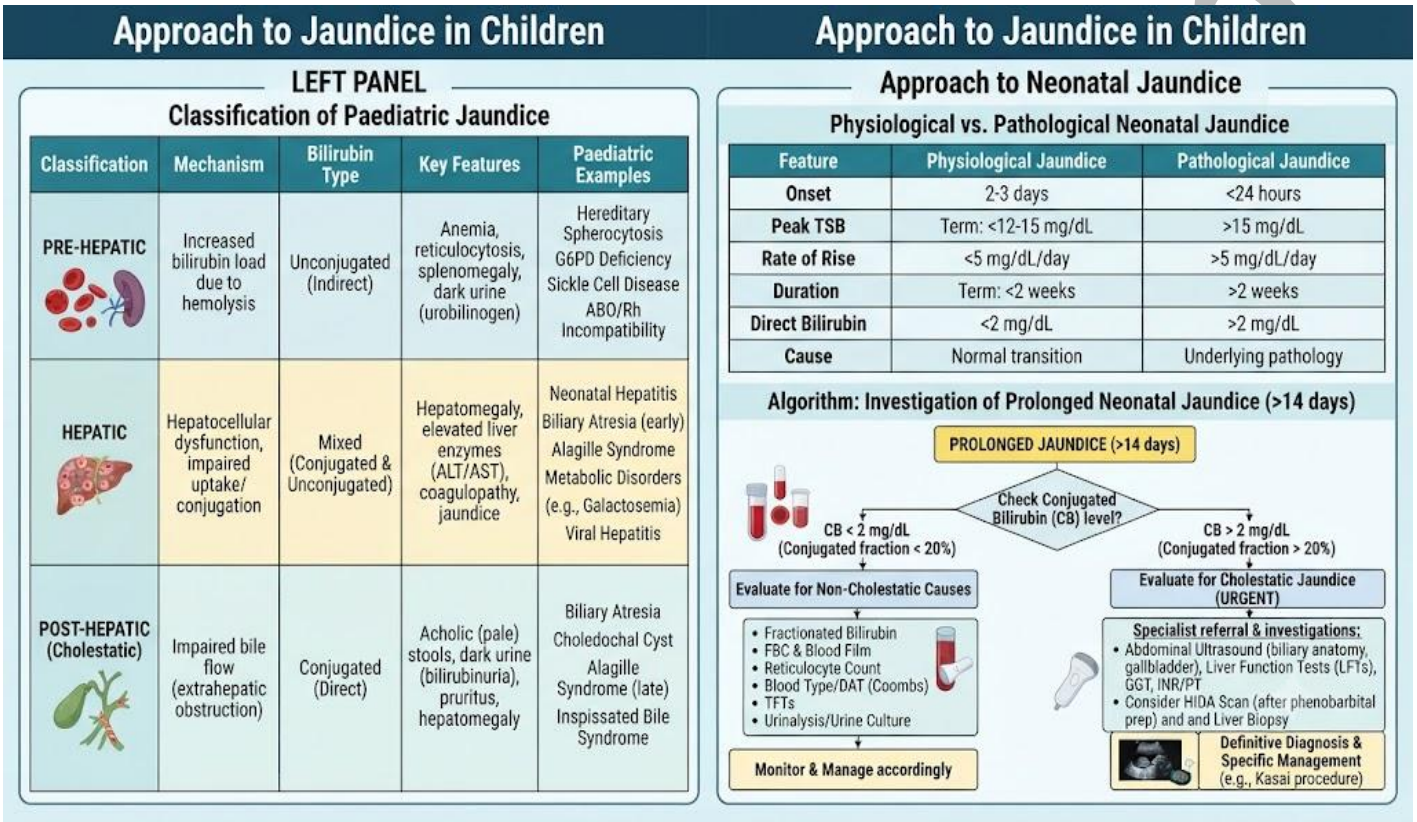

Figure 3.1: Classification of jaundice, physiological versus pathological neonatal jaundice, and investigation algorithm

Pilot questions 3.1

1. Classify jaundice by mechanism and give a paediatric example of each class. (Linked to SLO 3.1)
2. Distinguish physiological from pathological neonatal jaundice. (Linked to SLO 3.1)
3. State the key investigation in prolonged neonatal jaundice and explain its significance. (Linked to SLO 3.1)
4. Describe the HIDA scan findings in biliary atresia versus neonatal hepatitis. (Linked to SLO 3.1)
5. State the liver biopsy findings in biliary atresia and neonatal hepatitis. (Linked to SLO 3.1)

3.2 Biliary Atresia

3.2.1 Pathology and classification

Biliary atresia (BA) is a progressive fibro-inflammatory obliteration of the extrahepatic bile ducts, presenting in the neonatal period and leading to biliary cirrhosis and death if untreated by 2 years of age. It is the most common cause of prolonged neonatal jaundice requiring surgery and the most common indication for liver transplantation in children. The aetiology is not fully understood but is thought to involve perinatal viral infection (rotavirus, reovirus, or cytomegalovirus) triggering an immune-mediated inflammatory response in a genetically susceptible individual.

The Kasai classification describes three types based on the level of atresia: Type I (common bile duct atresia: the most proximal portion of the biliary tree is patent: the rarest type), Type II (common hepatic duct atresia), and Type III (involvement of the right and left hepatic ducts at the porta hepatis: the most common type: approximately 90 percent: the entire extrahepatic biliary tree is obliterated). Associated anomalies occur in approximately 20 percent and include polysplenia (the biliary atresia splenic malformation syndrome: BASM), situs inversus, and intestinal malrotation.

Fill in the blank: Biliary atresia leads to _____ cirrhosis and death if untreated by _____ years of age. The most common Kasai type is Type _____, in which the entire extrahepatic biliary tree is obliterated at the porta hepatis.

3.2.2 Clinical features and diagnosis

The typical presentation is a term neonate who appears well initially, with persistent jaundice beyond 14 days of life, progressive deepening of the jaundice, and the development of pale (acholic) stools and dark urine by 4 to 6 weeks. The stools are acholic because bile is not reaching the duodenum. The urine is dark from excreted conjugated bilirubin. Hepatomegaly develops and progresses to splenomegaly as portal hypertension develops from biliary cirrhosis.

Diagnosis: conjugated bilirubin above 20 percent of total (always investigate), ultrasound abdomen (absent or small gallbladder and absent common bile duct; the triangular cord sign: a triangular or tubular echogenic density at the porta hepatis from fibrous tissue replacing the bile ducts), HIDA scan (isotope not excreted into the duodenum), and liver biopsy (bile duct proliferation, cholestasis, periportal fibrosis). The definitive diagnosis is made at operative cholangiogram (no dye passes from the biliary tree into the duodenum). Early diagnosis is critical: the Kasai procedure performed before 8 weeks of age gives the best outcomes.

Fill in the blank: The triangular _____ sign on ultrasound (an echogenic density at the porta hepatis) is characteristic of biliary atresia. The definitive diagnosis of biliary atresia is confirmed by operative _____, which shows no dye passing into the duodenum. The Kasai procedure gives the best outcomes when performed before _____ weeks of age.

3.2.3 Management and outcomes

The Kasai portoenterostomy (hepatic portoenterostomy) is the surgical treatment of biliary atresia. The obliterated extrahepatic bile ducts are dissected and excised; a Roux-en-Y limb of jejunum is anastomosed to the transected porta hepatis (the fibrous tissue at the hilum of the liver), allowing bile from the microscopic bile ductules within the liver parenchyma to drain into the jejunum. Success depends on the age at surgery: Kasai performed before 60 days (8 to 9 weeks) of age achieves bile flow in approximately 65 to 80 percent of cases.

Even after a successful Kasai, progressive hepatic fibrosis continues in many patients and approximately 50 percent require liver transplantation by 10 years of age. Complications: cholangitis (ascending bacterial infection of the biliary system: most common postoperative complication: presents with fever, jaundice, and acholic stools: treated with IV antibiotics and prophylactic trimethoprim), portal hypertension and varices, and biliary cirrhosis. In Nigeria, many children with biliary atresia present after 8 weeks of age (due to delayed diagnosis), at which point the prognosis from the Kasai procedure is poor.

Fill in the blank: The Kasai portoenterostomy anastomoses a _____ limb of jejunum to the porta hepatis. The most common complication after the Kasai procedure is ascending _____ (infection of the biliary system). Approximately _____ percent of patients eventually require liver transplantation.

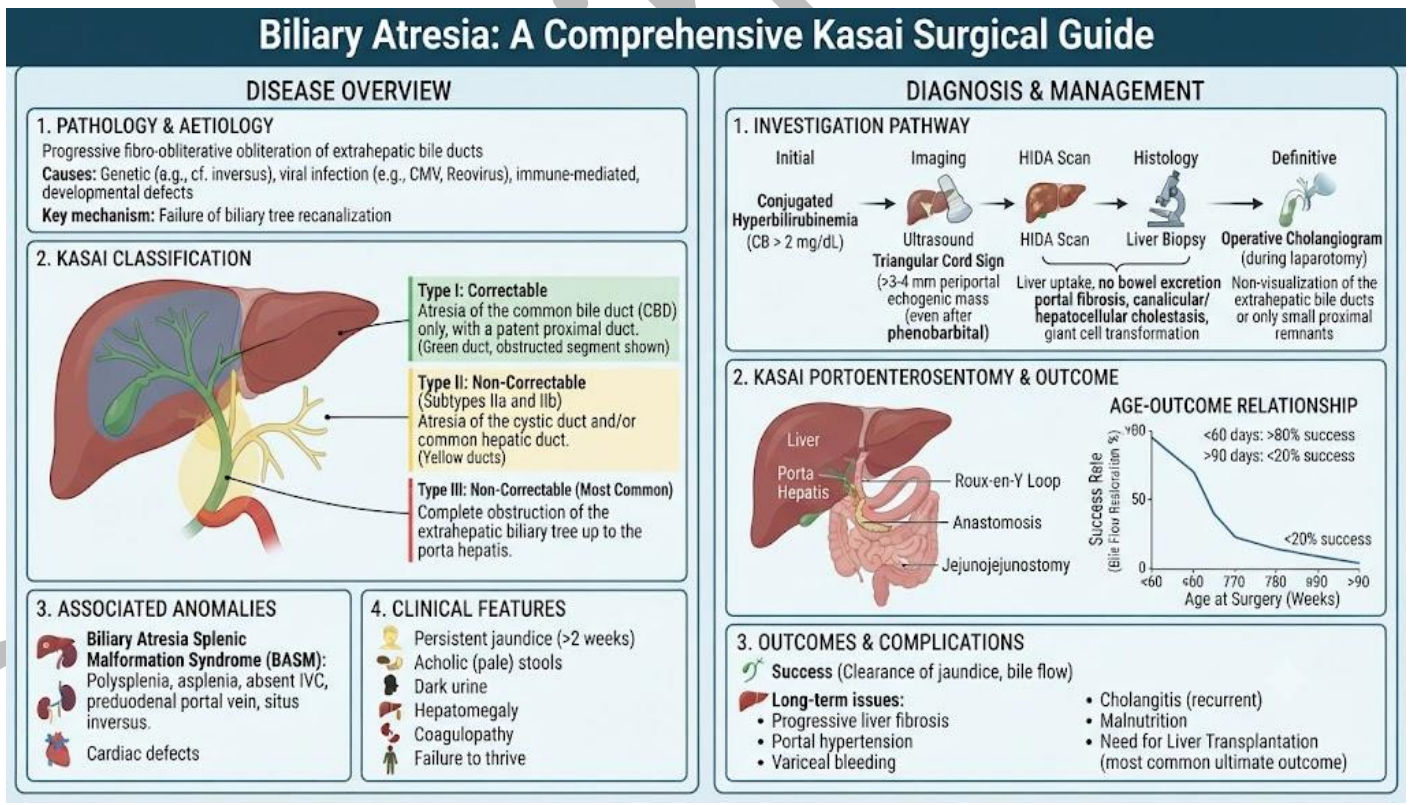

Figure 3.2: Pathology, classification, diagnosis, and management of biliary atresia

Pilot questions 3.2

1. Describe the pathology of biliary atresia and state its natural history if untreated. (Linked to SLO 3.2)
2. Describe the clinical features of biliary atresia in a neonate. (Linked to SLO 3.2)
3. Describe the triangular cord sign and state the investigation on which it is seen. (Linked to SLO 3.2)
4. Describe the Kasai portoenterostomy and explain why timing is critical. (Linked to SLO 3.2)
5. State the complications after the Kasai procedure and describe the management of cholangitis. (Linked to SLO 3.2)

3.3 Choledochal Cyst

3.3.1 Classification and pathophysiology

A choledochal cyst is a congenital cystic dilatation of the biliary tree. The Todani classification (a modification of the original Alonso-Lej classification) describes five types: Type I (cystic or fusiform dilatation of the common bile duct: the most common type: approximately 80 to 90 percent), Type II (diverticulum of the CBD), Type III (choledochocoele: cystic dilatation of the intraduodenal portion of the CBD), Type IV (multiple cysts involving both intra- and extrahepatic ducts), and Type V (Caroli disease: intrahepatic duct dilatation only: associated with congenital hepatic fibrosis).

The pathophysiology is thought to involve an anomalous pancreaticobiliary junction (APBJ): the pancreatic duct and common bile duct join outside the duodenal wall, creating a long common channel that allows pancreatic enzymes to reflux into the bile duct, causing inflammation, weakening of the duct wall, and progressive cystic dilatation. Untreated choledochal cysts carry a risk of malignant transformation (cholangiocarcinoma: risk increases with age and size) of approximately 2.5 to 28 percent over a lifetime.

Fill in the blank: The most common type of choledochal cyst is Type _____, which is a _____ or fusiform dilatation of the common bile duct. The anomalous _____ junction allows pancreatic enzymes to reflux into the bile duct, causing progressive cystic dilatation.

3.3.2 Clinical features and diagnosis

The classic triad of choledochal cyst is: abdominal pain (right hypochondrial or epigastric), jaundice (obstructive), and a right upper quadrant abdominal mass. However, the full triad is present in only approximately 20 percent of patients; most children present with only one or two features. Neonates may present with obstructive jaundice mimicking biliary atresia. Older

children and adults typically present with recurrent abdominal pain, intermittent jaundice, or complications (cholangitis, pancreatitis, or rupture).

Investigations: ultrasound abdomen (the first-line investigation: shows a cystic structure in the right upper quadrant related to the bile duct; can identify the type in most cases), MRCP (magnetic resonance cholangiopancreatography: the best non-invasive investigation for delineating the anatomy of the cyst, the biliary tree, and the APBJ), HIDA scan (assesses biliary drainage), and CT abdomen (for larger cysts and preoperative planning). ERCP is generally avoided in children (risk of pancreatitis and inadequate delineation of intrahepatic ducts).

Fill in the blank: The classic triad of choledochal cyst is abdominal pain, _____, and a right upper quadrant _____. The best non-invasive investigation for delineating the biliary anatomy in choledochal cyst is _____ (MRCP).

3.3.3 Management and complications

The treatment of all choledochal cysts (Types I, II, and IV) is complete surgical excision of the cyst with Roux-en-Y hepaticojejunostomy (anastomosis of the cut end of the hepatic duct to a defunctioned jejunal limb), to prevent malignant transformation. Simple drainage (cyst-enterostomy) is no longer recommended because it leaves the cyst wall in place and does not eliminate the risk of cholangiocarcinoma. Type III (choledochocoele) is treated by endoscopic sphincterotomy. Type V (Caroli disease) is managed medically (ursodeoxycholic acid, cholangitis treatment) with liver transplantation for advanced disease.

Complications of choledochal cyst: cholangitis (fever, jaundice, and right upper quadrant pain: treated with IV antibiotics), acute pancreatitis (from enzyme reflux via the APBJ), rupture (rare: causing bile peritonitis: emergency surgery), portal hypertension (from biliary cirrhosis in longstanding cases), and cholangiocarcinoma (the most feared long-term complication: risk increases with age: complete cyst excision is mandatory to eliminate this risk). Laparoscopic excision with Roux-en-Y hepaticojejunostomy is now the preferred approach in experienced centres.

Fill in the blank: The definitive treatment of Type I choledochal cyst is complete surgical _____ with Roux-en-Y _____. Simple drainage is no longer recommended because it leaves the _____ wall in place and does not eliminate the risk of cholangiocarcinoma.

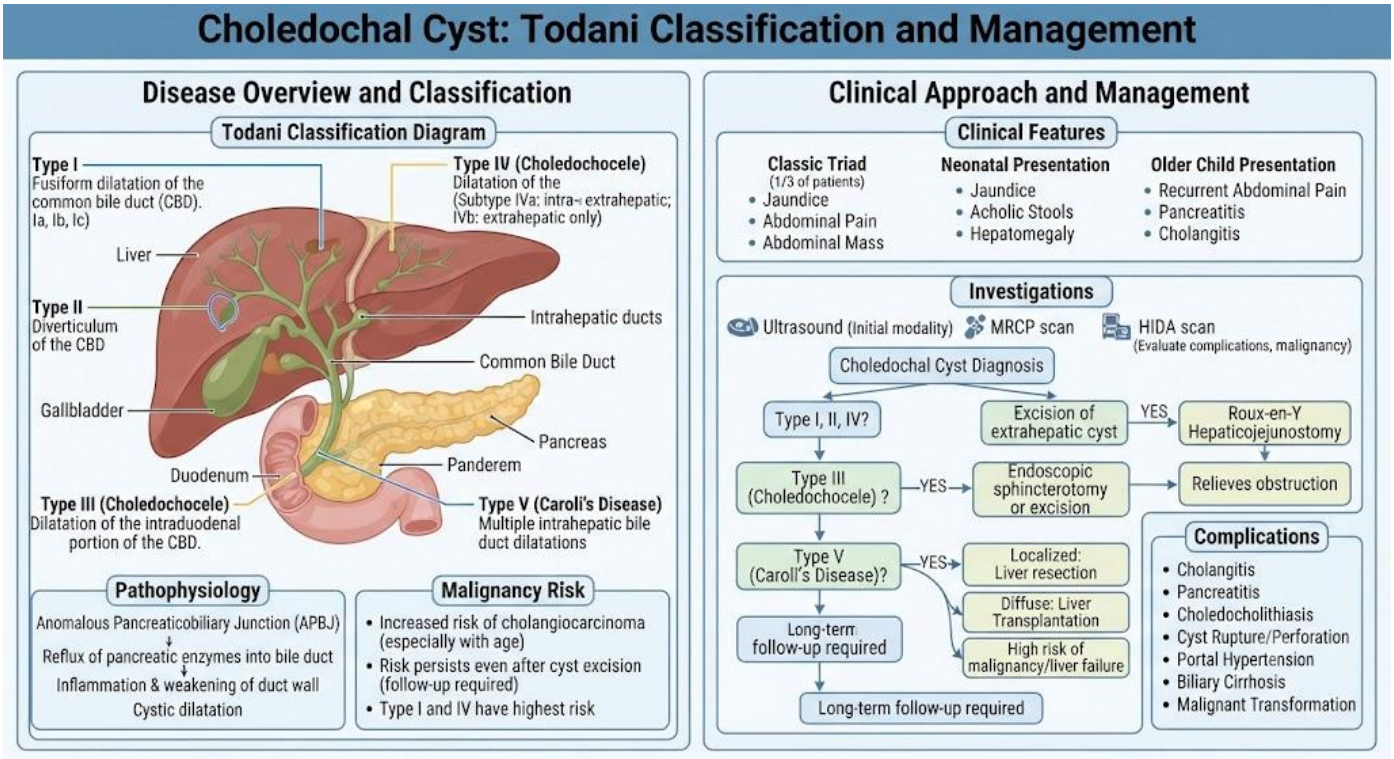

Figure 3.3: Classification, pathophysiology, clinical features, and management of choledochal cyst

Pilot questions 3.3

1. Describe the Todani classification of choledochal cyst and state the most common type. (Linked to SLO 3.3)
2. Explain the role of the anomalous pancreaticobiliary junction in the pathophysiology of choledochal cyst. (Linked to SLO 3.3)
3. Describe the classic clinical triad of choledochal cyst and state how common it is. (Linked to SLO 3.3)
4. Describe the definitive surgical treatment of Type I choledochal cyst and explain why simple drainage is no longer recommended. (Linked to SLO 3.3)
5. State the complications of choledochal cyst and identify the most feared long-term complication. (Linked to SLO 3.3)

3.4 Congenital Infantile Hypertrophic Pyloric Stenosis

3.4.1 Pathology and epidemiology

Infantile hypertrophic pyloric stenosis (IHPS) is a congenital hypertrophy and hyperplasia of the circular muscle of the pylorus, causing progressive gastric outlet obstruction. The aetiology is multifactorial: genetic predisposition (the risk is 10 to 20 percent in male offspring of an affected mother), environmental factors, and possibly early erythromycin exposure (erythromycin use in the first 2 weeks of life is associated with a significantly increased risk of IHPS). The pyloric muscle hypertrophies progressively, narrowing and elongating the pyloric channel until it is virtually obstructed.

IHPS is the most common surgical cause of vomiting in infancy. The incidence is approximately 2 to 4 per 1000 live births. It is much more common in males (male to female ratio approximately 4:1) and in firstborn children. It typically presents between 2 and 8 weeks of age (rarely before 2 weeks or after 12 weeks). There is a higher incidence in Caucasian populations, and in Nigerian practice it is less commonly encountered than in high-income countries, but cases are not rare in tertiary paediatric surgical centres.

Fill in the blank: IHPS is caused by hypertrophy of the _____ muscle of the pylorus. The condition is _____ to 4 times more common in males and is most common in _____ children. It typically presents between 2 and _____ weeks of age.

3.4.2 Clinical features and diagnosis

The classic presentation is a hungry, alert infant who develops projectile non-bilious vomiting (the vomiting is forceful and may travel across the room; it is non-bilious because the obstruction is proximal to the ampulla of Vater and bile cannot enter the stomach). Vomiting occurs shortly after every feed and the infant is hungry immediately after vomiting. Progressive weight loss and dehydration develop. The classic metabolic derangement is a hypochloraemic, hypokalaemic metabolic alkalosis (from loss of hydrochloric acid and potassium in the vomit: the kidneys retain hydrogen ions and excrete potassium to maintain electrochemical neutrality, worsening the alkalosis).

Clinical signs: visible gastric peristalsis (a wave passing from left to right across the epigastrium shortly after a feed, produced by the hypertrophied stomach attempting to force food through the obstructed pylorus), a palpable pyloric tumour (the olive: a firm, mobile, olive-shaped mass in the right upper quadrant felt during or after a test feed, between the liver edge and the right lateral border of the rectus abdominis: the pathognomonic sign). Ultrasound: a pyloric muscle thickness above 4 mm and a pyloric channel length above 14 to 16 mm confirm the diagnosis. An upper GI contrast study (barium meal) shows the string sign (a thin thread of contrast passing through the elongated narrow pyloric channel).

Fill in the blank: The classic metabolic abnormality in IHPS is a hypochloraemic, hypokalaemic metabolic _____. The palpable pyloric tumour is described as the _____ (olive-shaped

mass in the right upper quadrant). On ultrasound, a pyloric muscle thickness above _____ mm confirms the diagnosis.

3.4.3 Management

IHPS is a medical emergency before it becomes a surgical emergency: the metabolic alkalosis must be corrected before surgery. Resuscitation: IV fluids (0.45 percent saline with 5 percent dextrose and 20 to 40 mmol/L potassium chloride: replaces the ongoing chloride and potassium losses), nasogastric tube for gastric decompression. Surgery is performed once the serum electrolytes are normalised (target: serum chloride above 100 mmol/L, serum sodium above 135 mmol/L, serum potassium above 3.5 mmol/L, and urinary pH below 6 indicating resolution of paradoxical aciduria).

The surgical treatment is Ramstedt's pyloromyotomy: a longitudinal incision through the hypertrophied pyloric muscle down to the submucosa (without cutting the mucosa), allowing the mucosa to bulge through and the pyloric channel to open. It can be performed as an open operation (supra-umbilical or right upper quadrant incision) or laparoscopically (the preferred approach in experienced centres). Complications: mucosal perforation (recognised and repaired intraoperatively: the pyloromyotomy is rotated and re-done), incomplete pyloromyotomy (persistent vomiting: requires revision), and wound infection. The results of Ramstedt's pyloromyotomy are excellent: feeding usually resumes within 6 to 12 hours.

Fill in the blank: In IHPS, surgery must be preceded by correction of the _____ alkalosis. The surgical treatment is Ramstedt's _____, which splits the hypertrophied pyloric muscle without cutting the _____. Feeding resumes within _____ to 12 hours after successful pyloromyotomy.

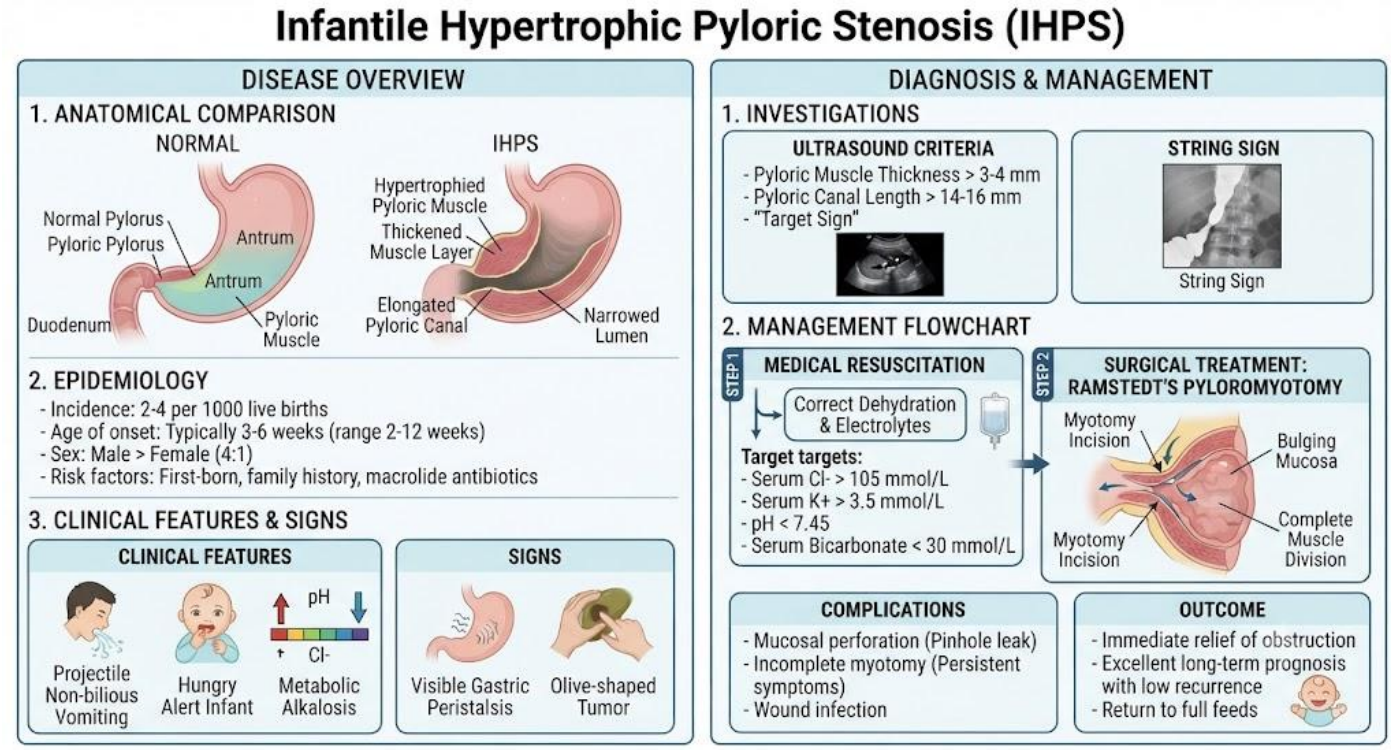

Figure 3.4: Pathology, clinical features, investigation, and management of infantile hypertrophic pyloric stenosis

Pilot questions 3.4

1. Describe the clinical presentation of infantile hypertrophic pyloric stenosis. (Linked to SLO 3.4)
2. Explain the metabolic alkalosis in IHPS and state the electrolyte targets before surgery. (Linked to SLO 3.4)
3. Describe the ultrasound findings that confirm the diagnosis of IHPS. (Linked to SLO 3.4)
4. Describe Ramstedt's pyloromyotomy and state why the mucosa must not be cut. (Linked to SLO 3.4)
5. State the complications of Ramstedt's pyloromyotomy. (Linked to SLO 3.4)

Series Summary

This Series covered the major hepatobiliary and jaundice disorders in children. We classified jaundice by mechanism (pre-hepatic, hepatic, post-hepatic), distinguished physiological from pathological neonatal jaundice, and described the investigation algorithm for prolonged neonatal

jaundice (split bilirubin, ultrasound, HIDA scan, liver biopsy). We described biliary atresia in detail: progressive fibro-inflammatory ductal obliteration, the triangular cord sign, the Kasai portoenterostomy, and the complications of cholangitis and biliary cirrhosis. We then covered choledochal cyst: Todani classification, the role of APBJ, MRCP, and complete cyst excision with hepaticojejunostomy. We closed with IHPS: projectile non-bilious vomiting, the olive tumour, hypochloraemic hypokalaemic metabolic alkalosis, and Ramstedt's pyloromyotomy.

You should now be able to classify jaundice and investigate prolonged neonatal jaundice (SLO 3.1), describe the pathology, diagnosis, and management of biliary atresia (SLO 3.2), describe the classification and management of choledochal cyst (SLO 3.3), and describe the presentation and surgical management of infantile hypertrophic pyloric stenosis (SLO 3.4).

Glossary of Terms

- **Acholic stools:** pale, clay-coloured stools resulting from obstruction of bile flow into the duodenum; a key clinical sign of biliary atresia and other causes of post-hepatic jaundice in children.
- **Anomalous pancreaticobiliary junction (APBJ):** a congenital variant in which the pancreatic duct and common bile duct join outside the duodenal wall, creating a long common channel that allows pancreatic enzyme reflux into the biliary tree; implicated in the pathophysiology of choledochal cyst and biliary malignancy.
- **Biliary atresia:** a progressive fibro-inflammatory obliteration of the extrahepatic bile ducts presenting in the neonatal period; treated by Kasai portoenterostomy; leads to biliary cirrhosis and death if untreated by 2 years.
- **Choledochal cyst:** a congenital cystic dilatation of the biliary tree classified by the Todani system into five types; associated with risk of cholangiocarcinoma; treated by complete surgical excision and Roux-en-Y hepaticojejunostomy.
- **HIDA scan:** hepatobiliary iminodiacetic acid scintigraphy; a nuclear medicine scan assessing bile flow; in biliary atresia the isotope is taken up by hepatocytes but not excreted into the duodenum; used to distinguish biliary atresia from neonatal hepatitis.
- **Kasai portoenterostomy:** the surgical treatment of biliary atresia in which the obliterated extrahepatic ducts are excised and a Roux-en-Y jejunal limb is anastomosed to the porta hepatis to restore bile drainage; outcomes are best when performed before 60 days of age.
- **Olive tumour:** the palpable pyloric tumour in infantile hypertrophic pyloric stenosis: a firm, mobile, olive-shaped mass felt in the right upper quadrant during or after a test feed; pathognomonic when present.

- **Physiological jaundice:** unconjugated hyperbilirubinaemia appearing after 24 hours of life in neonates; caused by increased red cell breakdown, immature conjugating enzyme activity, and enterohepatic circulation; resolves by day 10 to 14 in term infants.
- **Ramstedt's pyloromyotomy:** the standard surgical treatment of infantile hypertrophic pyloric stenosis: a longitudinal incision through the hypertrophied pyloric muscle to the submucosa without entering the mucosa, allowing the mucosa to bulge through and the pyloric channel to open.
- **String sign:** a radiographic finding on upper GI contrast study in IHPS: a thin thread of contrast passing through the elongated, narrowed pyloric channel; indicative of pyloric stenosis.
- **Todani classification:** the standard classification of choledochal cysts into five types (I to V) based on the location and pattern of biliary dilatation; Type I (CBD dilatation) is the most common; Type V (Caroli disease) involves intrahepatic ducts only.
- **Triangular cord sign:** an echogenic triangular or tubular density at the porta hepatis on abdominal ultrasound, representing fibrous tissue replacing the obliterated extrahepatic bile ducts; characteristic of biliary atresia.

Concept Check Questions [Series 3]

Objective questions

1. Post-hepatic jaundice is characterised by conjugated hyperbilirubinaemia with _____ stools and dark urine. (Linked to SLO 3.1)
2. Physiological jaundice appears after _____ hours of life and resolves by day 10 to 14 in term infants. (Linked to SLO 3.1)
3. The triangular cord sign on abdominal ultrasound is characteristic of _____ atresia. (Linked to SLO 3.2)
4. The most common Todani type of choledochal cyst is Type _____, a cystic or fusiform dilatation of the common bile duct. (Linked to SLO 3.3)
5. The classic metabolic abnormality in infantile hypertrophic pyloric stenosis is a hypochloraemic, hypokalaemic metabolic _____. (Linked to SLO 3.4)

Short-answer questions

6. Distinguish physiological from pathological neonatal jaundice. (Linked to SLO 3.1)
7. Describe the clinical features of biliary atresia in a neonate. (Linked to SLO 3.2)
8. Describe the Kasai portoenterostomy and explain why timing is critical. (Linked to SLO 3.2)
9. Describe the classic clinical presentation of infantile hypertrophic pyloric stenosis. (Linked to SLO 3.4)
10. Explain why surgery for IHPS must be preceded by medical resuscitation. (Linked to SLO 3.4)

Long-answer questions

11. Describe the investigation algorithm for a neonate with prolonged jaundice beyond 14 days. (Linked to SLO 3.1)
12. Describe the pathology, diagnosis, and surgical management of biliary atresia. (Linked to SLO 3.2)

13. Describe the classification, pathophysiology, and management of choledochal cyst. (Linked to SLO 3.3)
14. Describe the clinical features, investigation, and management of infantile hypertrophic pyloric stenosis. (Linked to SLO 3.4)
15. Compare biliary atresia and choledochal cyst in terms of pathology, presentation, and surgical management. (Linked to SLO 3.2 and 3.3)

Answers to Concept Check Questions [Series 3]

Objective questions

1. Pale (acholic).
2. 24.
3. Biliary.
4. I.
5. Alkalosis.

Short-answer questions

6. Physiological: appears after 24 hours; unconjugated; resolves by day 10 to 14 in term infants; no pale stools or dark urine; no associated features. Pathological: appears within 24 hours or persists beyond 14 days; may be conjugated (always pathological); associated with pale stools, dark urine, hepatomegaly; requires investigation and treatment.
7. A term neonate who appears well initially; persistent jaundice beyond 14 days deepening progressively; acholic (pale) stools by 4 to 6 weeks; dark urine; progressive hepatomegaly developing into splenomegaly as biliary cirrhosis and portal hypertension develop.
8. The Kasai portoenterostomy excises the obliterated extrahepatic bile ducts and anastomoses a Roux-en-Y jejunal limb to the porta hepatis, allowing bile from microscopic bile ductules to drain into the jejunum. Timing is critical because: performed before 60 days, bile flow is achieved in 65 to 80 percent of cases; after 60 days, progressive fibrosis reduces success rates significantly, and many patients will require liver transplantation.

9. A hungry, alert infant aged 2 to 8 weeks with forceful projectile non-bilious vomiting after every feed; the infant is hungry immediately after vomiting; progressive weight loss and dehydration; visible gastric peristalsis (left to right wave across the epigastrium after a feed); palpable olive tumour in the right upper quadrant during or after a test feed.

10. IHPS causes loss of hydrochloric acid and potassium in the vomit, producing a hypochloraemic, hypokalaemic metabolic alkalosis. General anaesthesia in the presence of metabolic alkalosis risks apnoea and respiratory depression because the alkalosis blunts the respiratory drive. Electrolytes must be normalised before surgery (serum chloride above 100 mmol/L, potassium above 3.5 mmol/L, and urine pH below 6 indicating resolution of paradoxical aciduria).

Long-answer questions

11. **Basic response:** Step 1: Split bilirubin (conjugated versus unconjugated). If conjugated bilirubin above 20 percent of total: always pathological: urgent investigation. Step 2: FBC and blood film and direct Coombs test (haemolysis: Rh and ABO incompatibility and G6PD deficiency). Step 3: Thyroid function tests (hypothyroidism: a cause of prolonged unconjugated jaundice). Step 4: Urine reducing substances (galactosaemia). Step 5: TORCH screen (congenital infection). Step 6: Liver function tests (transaminases, albumin, prothrombin time). Step 7: Ultrasound abdomen (absent or small gallbladder and triangular cord sign: biliary atresia; cystic biliary structure: choledochal cyst; gallstones). Step 8: HIDA scan (biliary atresia: isotope uptake but no duodenal excretion; neonatal hepatitis: delayed excretion). Step 9: Liver biopsy (biliary atresia: bile duct proliferation and cholestasis and periportal fibrosis; neonatal hepatitis: giant cell transformation). Step 10: Operative cholangiogram at the time of Kasai procedure (definitive: no dye into duodenum confirms biliary atresia).

Value-added response: The most critical step in this algorithm is the urgent split bilirubin: any neonate with conjugated jaundice (direct bilirubin above 20 percent of total) should be referred immediately to a paediatric surgical centre for assessment. In Nigeria, awareness of this threshold among primary care physicians and general paediatricians is key to ensuring timely referral before the optimal surgical window for the Kasai procedure (before 60 days of age) is missed.

12. **Basic response:** Pathology: biliary atresia is a progressive fibro-inflammatory obliteration of the extrahepatic bile ducts. It leads to biliary cirrhosis, portal hypertension, and death if untreated by 2 years. The most common type (Kasai Type III: 90 percent) involves complete obliteration at the porta hepatis. Associated anomalies: polysplenia, situs inversus, and malrotation (BASM). Diagnosis: conjugated bilirubin above 20

percent; ultrasound (absent gallbladder, triangular cord sign); HIDA scan (no duodenal excretion); liver biopsy (bile duct proliferation, cholestasis, periportal fibrosis); operative cholangiogram confirms diagnosis. Management: Kasai portoenterostomy: excise obliterated extrahepatic ducts; anastomose Roux-en-Y jejunal limb to porta hepatis; best outcomes when performed before 60 days. Complications: cholangitis (most common: IV antibiotics and prophylactic trimethoprim), portal hypertension and varices, biliary cirrhosis, liver transplantation in approximately 50 percent by 10 years.

Value-added response: In Nigerian practice, the biggest challenge in biliary atresia is late presentation: many infants are referred after 8 to 12 weeks of age, often after prolonged treatment for presumed physiological or breast milk jaundice. At this stage, the Kasai procedure has a much lower success rate and liver transplantation is the realistic long-term treatment. Improving awareness of the split bilirubin threshold among Nigerian paediatricians is therefore the single most impactful intervention to improve outcomes.

13. **Basic response:** Classification: Todani Type I (cystic or fusiform CBD dilatation: most common: 80 to 90 percent), Type II (CBD diverticulum), Type III (choledochocoele: intraduodenal CBD), Type IV (multiple intra- and extrahepatic cysts), Type V (Caroli disease: intrahepatic cysts only). Pathophysiology: anomalous pancreaticobiliary junction (APBJ) creates a long common channel outside the duodenal wall; pancreatic enzymes reflux into the bile duct; inflammation and progressive cystic dilatation occur; untreated cysts carry a 2.5 to 28 percent lifetime risk of cholangiocarcinoma. Management: Type I, II, IV: complete excision of the cyst and Roux-en-Y hepaticojejunostomy (hepatic duct anastomosed to defunctioned jejunal limb); simple drainage is no longer recommended; laparoscopic excision preferred in experienced centres. Type III: endoscopic sphincterotomy. Type V: ursodeoxycholic acid and cholangitis treatment; liver transplantation for advanced disease. Complications: cholangitis, pancreatitis, rupture (bile peritonitis), portal hypertension, cholangiocarcinoma.

Value-added response: The timing of surgery for choledochal cyst differs from biliary atresia: while biliary atresia has a critical window before 60 days, choledochal cysts can be operated electively but should not be delayed indefinitely because the risk of cholangiocarcinoma and cholangitis increases with age. In practice, once the diagnosis is confirmed (usually by MRCP), surgery should be planned within weeks rather than months.

14. **Basic response:** Clinical features: a hungry, alert infant aged 2 to 8 weeks presenting with forceful projectile non-bilious vomiting after every feed; immediately hungry again after vomiting; progressive weight loss and dehydration. Metabolic derangement: hypochloraemic, hypokalaemic metabolic alkalosis (HCl and K lost in vomit; kidneys retain H+ and excrete K+ to maintain electrochemical neutrality). Signs: visible gastric peristalsis (left to right wave after a feed); palpable olive tumour (firm mobile mass right upper quadrant after test feed). Investigation: ultrasound (pyloric muscle thickness above

4 mm and channel length above 14 to 16 mm confirms diagnosis); upper GI contrast study (string sign) if ultrasound equivocal. Management: medical resuscitation first (IV 0.45 percent saline plus dextrose plus KCl; NG decompression; correct electrolytes to target levels); then Ramstedt's pyloromyotomy (longitudinal incision through pyloric muscle to submucosa; mucosa must not be cut); laparoscopic or open approach; feeding resumes 6 to 12 hours postoperatively.

Value-added response: IHPS is a classic example of a condition that requires meticulous medical preparation before surgery: operating on a baby with significant hypochloraemic alkalosis carries a real risk of postoperative apnoea from blunted respiratory drive. Anaesthetists in Nigerian teaching hospitals must be familiar with the electrolyte targets and the surgeon and anaesthetist should agree that criteria are met before the operation proceeds.

15. **Basic response:** Pathology: biliary atresia is fibro-inflammatory obliteration of extrahepatic bile ducts (solid fibrous remnants); choledochal cyst is congenital cystic dilatation of the biliary tree (a patent but abnormally dilated bile duct). Presentation: biliary atresia presents in the neonatal period with obstructive jaundice, acholic stools, dark urine, and a normal-sized gallbladder (absent or atretic); choledochal cyst may present in neonates mimicking biliary atresia or in older children with the classic triad (pain, jaundice, right upper quadrant mass). Investigation: both show conjugated hyperbilirubinaemia; ultrasound differentiates (absent gallbladder and triangular cord sign in biliary atresia; cystic biliary structure in choledochal cyst); MRCP is the key investigation for choledochal cyst. Surgical management: biliary atresia: Kasai portoenterostomy (anastomose jejunum to porta hepatis); choledochal cyst: complete excision of cyst with hepaticojejunostomy (anastomose jejunum to hepatic duct). Both require a Roux-en-Y jejunal limb; both are associated with long-term complications requiring liver transplantation in advanced disease.

Value-added response: A critical practical point: biliary atresia has a time-sensitive surgical window (before 60 days) while choledochal cyst does not. However, both conditions can present with identical clinical features in the neonatal period (obstructive jaundice with acholic stools), making the distinction between them urgent. The ultrasound finding of an absent gallbladder versus a cystic biliary structure is often the key differentiating investigation.

Answers to Pilot Questions [Series 3]

Part 3.1

6. Pre-hepatic: excess bilirubin production (haemolysis: unconjugated): example: Rh incompatibility and G6PD deficiency. Hepatic: failure of conjugation or intrahepatic excretion (liver parenchymal disease: mixed or conjugated): example: neonatal hepatitis

and galactosaemia. Post-hepatic or cholestatic: obstruction of bile flow (conjugated: pale stools and dark urine): example: biliary atresia and choledochal cyst.

7. Physiological: appears after 24 hours; unconjugated only; resolves by day 10 to 14 in term infants; no pale stools; no hepatomegaly; no associated pathological features. Pathological: may appear within 24 hours of birth; may be conjugated (always pathological); may persist beyond 14 days; may be associated with acholic stools, dark urine, hepatomegaly, or a serum bilirubin above the treatment threshold for gestational age.
8. The key investigation is the split bilirubin (direct versus indirect fractions). A conjugated bilirubin above 20 percent of total bilirubin in a neonate is always pathological. Its significance is that it mandates urgent investigation and referral to a paediatric surgical unit to exclude biliary atresia, because the Kasai procedure must be performed before 60 days of age to achieve optimal bile flow.
9. In biliary atresia: the isotope (HIDA) is taken up normally by hepatocytes but is not excreted into the duodenum (no bowel activity) because the extrahepatic bile ducts are obliterated. In neonatal hepatitis: the isotope is taken up by hepatocytes with delayed excretion, but eventually some activity appears in the bowel (excretion occurs, albeit delayed, because the bile ducts are patent even though hepatocyte function is impaired).
10. Biliary atresia: bile duct proliferation (increased number of bile ductules at the portal tracts), cholestasis (bile plugs within ductules and hepatocytes), periportal fibrosis (progressive scarring of the portal tracts from ongoing biliary obstruction), and inflammatory cell infiltration. Neonatal hepatitis: giant cell transformation of hepatocytes (hepatocytes fuse to form large multinucleated giant cells), lobular disarray, and intrahepatic cholestasis without significant bile duct proliferation.

Part 3.2

11. Biliary atresia is a progressive fibro-inflammatory obliteration of the extrahepatic bile ducts. The obliteration is progressive from birth and leads to biliary cirrhosis (from back-pressure of bile in the liver), portal hypertension, hepatic failure, and death within 2 years if untreated. It is the most common cause of prolonged neonatal jaundice requiring surgery and the most common indication for liver transplantation in children.
12. A term neonate who initially appears well; persistent jaundice beyond 14 days deepening progressively; acholic (pale, clay-coloured) stools from approximately 4 to 6 weeks of age (bile not reaching the duodenum); dark urine (conjugated bilirubin excreted by the

kidney); progressive hepatomegaly (from biliary back-pressure and developing cirrhosis); later: splenomegaly, ascites, and bruising from portal hypertension and coagulopathy.

13. The triangular cord sign is an echogenic triangular or tubular density at the porta hepatis on abdominal ultrasound, representing the fibrous tissue that has replaced the obliterated extrahepatic bile ducts. It is seen on abdominal ultrasound and is a characteristic finding in biliary atresia. It may be associated with an absent or small gallbladder.

14. The Kasai portoenterostomy: the obliterated extrahepatic bile ducts are dissected down to the porta hepatis and excised. A Roux-en-Y limb of jejunum is fashioned and anastomosed to the cut surface of the porta hepatis, allowing bile to drain from the microscopic bile ductules within the liver parenchyma into the jejunum. Timing is critical because: before 60 days of age, the ductules at the porta hepatis are still patent at a microscopic level and bile flow can be restored in 65 to 80 percent of cases; after 60 days, progressive fibrosis obliterates even these microscopic ductules and the success rate falls dramatically.

15. Complications: cholangitis (most common: ascending bacterial infection of the biliary system presenting with fever, jaundice, and acholic stools: IV broad-spectrum antibiotics and prophylactic long-term trimethoprim), portal hypertension and oesophageal or gastric varices (from progressive biliary cirrhosis: managed with beta-blockers and endoscopic band ligation or sclerotherapy), biliary cirrhosis and hepatic failure, and liver transplantation (required in approximately 50 percent of patients by 10 years).

Part 3.3

1. Todani classification: Type I (cystic or fusiform dilatation of the common bile duct: most common: 80 to 90 percent), Type II (diverticulum of the common bile duct), Type III (choledochocoele: cystic dilatation of the intraduodenal portion of the CBD), Type IV (multiple cysts involving both intra- and extrahepatic ducts), Type V (Caroli disease: intrahepatic duct dilatation only: associated with congenital hepatic fibrosis). The most common type is Type I.

2. The anomalous pancreaticobiliary junction (APBJ) is a congenital variant in which the pancreatic duct and common bile duct join outside the duodenal wall (rather than within the duodenal wall at the ampulla of Vater), creating a long common channel. This long common channel allows pancreatic enzymes (amylase and lipase) to reflux freely into the bile duct, as the normal sphincter mechanism of Oddi does not protect the junction. The enzyme reflux causes inflammation of the bile duct wall, progressive weakening, and cystic dilatation.

3. The classic triad is: abdominal pain (right hypochondrial or epigastric), obstructive jaundice, and a right upper quadrant abdominal mass. The full triad is present in only approximately 20 percent of patients. Most children present with one or two features: recurrent abdominal pain and intermittent jaundice are the most common presenting complaints in older children; neonates may present with obstructive jaundice mimicking biliary atresia.

4. The definitive treatment of Type I choledochal cyst is complete surgical excision of the entire cyst with Roux-en-Y hepaticojejunostomy (the hepatic duct is anastomosed to a defunctioned Roux-en-Y jejunal limb). Simple drainage (cyst-enterostomy) is no longer recommended because it leaves the cyst wall in place: the mucosal lining of the cyst wall has a high risk of malignant transformation to cholangiocarcinoma, and this risk is not eliminated by drainage. Only complete excision of the cyst wall removes the at-risk mucosa.

5. Cholangitis (fever, jaundice, and right upper quadrant pain: treated with IV antibiotics), acute pancreatitis (from pancreatic enzyme reflux via the APBJ), rupture (rare: causing bile peritonitis: emergency surgery required), portal hypertension (from biliary cirrhosis in longstanding cases), and cholangiocarcinoma (the most feared long-term complication: risk is 2.5 to 28 percent over a lifetime; increases with age and cyst size; only complete cyst excision eliminates this risk).

Part 3.4

1. A hungry, alert infant aged 2 to 8 weeks presenting with forceful projectile non-bilious vomiting after every feed; the vomit is milk only (no bile: obstruction is proximal to the ampulla of Vater); the infant is hungry immediately after vomiting; progressive weight loss and dehydration; visible gastric peristalsis (a wave passing left to right across the epigastrium shortly after a feed); a palpable olive-shaped tumour in the right upper quadrant felt during or after a test feed (the pathognomonic sign).

2. IHPS causes loss of HCl and KCl in the vomit. Loss of HCl causes metabolic alkalosis (insufficient hydrogen ions in the blood). Loss of KCl causes hypokalaemia. The kidneys respond to alkalosis and hypovolaemia by retaining sodium and hydrogen ions and excreting potassium (paradoxical aciduria: the urine is acidic despite systemic alkalosis, because the kidney is excreting H+ to retain Na+). Electrolyte targets before surgery: serum chloride above 100 mmol/L, serum sodium above 135 mmol/L, serum potassium above 3.5 mmol/L, and urine pH below 6 (confirming resolution of the alkalosis).

3. Ultrasound is the first-line investigation for IHPS. Diagnostic criteria: pyloric muscle thickness above 4 mm (normal is less than 3 mm) and pyloric channel length above 14 to

16 mm (normal is less than 14 mm). The pyloric channel appears as an elongated, echogenic, non-relaxing structure during real-time scanning. An experienced paediatric radiologist can confirm the diagnosis in the vast majority of cases with ultrasound alone, avoiding the need for a contrast study.

4. Ramstedt's pyloromyotomy: a longitudinal incision is made through the serosa and the hypertrophied circular muscle fibres of the pylorus from the pyloric vein of Mayo to the prepyloric antrum, down to the level of the submucosa, which is left intact. The muscle fibres spring apart, allowing the mucosa to bulge through the incision and the pyloric channel to open. The mucosa must not be cut because perforation of the mucosa leads to contamination of the peritoneal cavity with gastric contents and requires immediate repair and rotation of the pyloromyotomy to a different site.

5. Mucosal perforation (recognised intraoperatively by air or bile leaking: repaired primarily; the pyloromyotomy is extended to a different site), incomplete pyloromyotomy (persistent vomiting beyond 48 hours: confirmed by upper GI contrast study: requires re-operation), wound infection, and incisional hernia (rare with laparoscopic approach).

References and Attributions [Series 3]

Textual References

1. Coran, A. G., Caldamone, A., Adzick, N. S., Krummel, T. M., Laberge, J. M. and Shamberger, R. (2012). Pediatric Surgery (7th ed.). Philadelphia: Elsevier Mosby.
2. Puri, P. and Hollwarth, M. E. (2009). Pediatric Surgery: Diagnosis and Management. Berlin: Springer.
3. Ameh, E. A. and Chirdan, L. B. (2011). Paediatric Surgery in the Tropics. Ibadan: Spectrum Books.
4. Kasai, M. (1959). Surgery of biliary atresia. Japanese Journal of Surgery, 57, 114 to 116.

Figures, Plates, Tables, and Boxes

5. Figure 3.1: Approach to jaundice in children. AI-generated using the prompt: "Two-panel diagram: three-row jaundice classification table (pre-hepatic, hepatic, post-hepatic); physiological versus pathological neonatal jaundice comparison table and prolonged neonatal jaundice investigation algorithm. Paediatric surgical diagram style." Suggested OER search string: "neonatal jaundice classification investigation biliary atresia HIDA scan liver biopsy paediatric OER."
6. Figure 3.2: Biliary atresia. AI-generated using the prompt: "Two-panel diagram: pathology and Kasai classification diagram (Types I, II, III on biliary tree) and associated anomalies and clinical features; investigation pathway and Kasai portoenterostomy diagram with age-outcome relationship and complications box. Paediatric surgical diagram style." Suggested OER search string: "biliary atresia Kasai portoenterostomy triangular cord sign HIDA scan cholangitis OER."
7. Figure 3.3: Choledochal cyst. AI-generated using the prompt: "Two-panel diagram: Todani classification (Types I to V on biliary tree illustration) and APBJ pathophysiology and malignancy risk box; clinical features box and investigations box (ultrasound, MRCP) and management flowchart with complications box. Paediatric surgical diagram style." Suggested OER search string: "choledochal cyst Todani classification MRCP hepaticojejunostomy cholangiocarcinoma paediatric surgery OER."
8. Figure 3.4: Infantile hypertrophic pyloric stenosis. AI-generated using the prompt: "Two-panel diagram: longitudinal stomach-pylorus normal versus IHPS diagram and epidemiology and clinical features and signs boxes; investigation box (ultrasound criteria and string sign) and two-step management flowchart (resuscitation with electrolyte

targets then Ramstedt's pyloromyotomy with complications). Paediatric surgical diagram style." Suggested OER search string: "infantile hypertrophic pyloric stenosis Ramstedt pyloromyotomy metabolic alkalosis ultrasound OER."

Series 4: Genitourinary Malformations

- **Part 4.1: Hypospadias**
    - Sub Part 4.1.1: Anatomy, classification, and pathophysiology
    - Sub Part 4.1.2: Clinical features and associated anomalies
    - Sub Part 4.1.3: Management and complications
- **Part 4.2: Epispadias and Bladder Exstrophy**
    - Sub Part 4.2.1: Embryology and spectrum of anomalies
    - Sub Part 4.2.2: Clinical features and diagnosis
    - Sub Part 4.2.3: Management
- **Part 4.3: Posterior Urethral Valves**
    - Sub Part 4.3.1: Pathology and classification
    - Sub Part 4.3.2: Clinical features and diagnosis
    - Sub Part 4.3.3: Management and long-term outcomes
- **Part 4.4: Wilms' Tumour**
    - Sub Part 4.4.1: Pathology and epidemiology
    - Sub Part 4.4.2: Clinical features and diagnosis
    - Sub Part 4.4.3: Management and prognosis

Introduction

Genitourinary malformations are among the most common congenital anomalies, affecting approximately 3 to 4 percent of all live births. Many are identified antenatally on routine ultrasound screening; others present in the neonatal period or in infancy with urinary tract infection, poor urinary stream, or a palpable abdominal mass. Prompt and accurate diagnosis is essential to prevent permanent renal damage and to achieve optimal functional outcomes.

This Series covers the major genitourinary malformations in paediatric surgical practice: hypospadias (the most common penile anomaly), epispadias and bladder exstrophy (the most severe end of the exstrophy-epispadias complex), posterior urethral valves (the most common cause of severe obstructive uropathy in boys), and Wilms' tumour (the most common intra-

abdominal malignancy in children). Each condition is examined from pathophysiology through diagnosis to surgical management.

Series Learning Outcomes

Upon completing this Series, you should be able to:

- **SLO 4.1:** describe the anatomy, classification, and surgical management of hypospadias and state its complications,
- **SLO 4.2:** describe the exstrophy-epispadias complex, its clinical features, and the principles of its staged surgical management,
- **SLO 4.3:** describe the pathology, clinical features, diagnosis, and management of posterior urethral valves and their long-term consequences, and
- **SLO 4.4:** describe the pathology, clinical features, staging, and management of Wilms' tumour and state its prognosis.

4.1 Hypospadias

4.1.1 Anatomy, classification, and pathophysiology

Hypospadias is a congenital malformation in which the urethral meatus opens on the ventral (under) surface of the penis proximal to its normal position at the tip of the glans. Three features are consistently present: an abnormally positioned meatus, a hooded (incomplete) foreskin (the foreskin is deficient ventrally, forming a dorsal hood), and ventral curvature of the penis (chordee: from fibrous tissue tethering the ventral shaft). The meatus may open anywhere from the glans to the perineum.

Hypospadias is classified by the position of the meatus: anterior (glanular and coronal: approximately 50 percent), middle (penile shaft: approximately 30 percent), and posterior (penoscrotal, scrotal, and perineal: approximately 20 percent). The incidence is approximately 1 in 300 male births and appears to be increasing, possibly due to endocrine disruptors. The pathophysiology involves incomplete virilisation of the genital tubercle from insufficient androgens or androgen insensitivity during the critical period of urethral development (8th to 14th weeks of gestation).

Fill in the blank: In hypospadias, the three consistent features are an abnormally positioned _____, a hooded foreskin (deficient _____), and ventral curvature called _____. Hypospadias results from incomplete _____ of the genital tubercle during the 8th to 14th weeks of gestation.

4.1.2 Clinical features and associated anomalies

Hypospadias is diagnosed at birth on clinical examination. The infant has a hooded dorsal foreskin (the foreskin resembles a hood over the dorsal glans, absent ventrally), and the meatus is visible on the ventral surface of the glans, shaft, scrotum, or perineum. Chordee (ventral penile curvature) becomes more apparent on erection. The urinary stream may be deflected downward (making standing micturition difficult in more proximal types) and, in penoscrotal and perineal types, the appearance may raise concern about a disorder of sexual differentiation.

Associated anomalies: undescended testis (present in approximately 10 percent of cases of hypospadias; higher in posterior types: up to 30 percent), inguinal hernia (from patent processus vaginalis), and a utriculus (a Mullerian duct remnant in the prostatic urethra: particularly common in severe hypospadias). In proximal (perineal or penoscrotal) hypospadias with bilateral undescended testes, a disorder of sexual differentiation (DSD: formerly called intersex) must be excluded by karyotype and hormonal evaluation before any surgery.

Fill in the blank: Associated anomalies in hypospadias include undescended testis (present in approximately _____ percent) and inguinal _____. Proximal hypospadias with bilateral undescended testes requires exclusion of a disorder of _____ differentiation by karyotype.

4.1.3 Management and complications

The goals of surgery are: to correct the chordee (straighten the penis), to create a neo-urethra extending to the tip of the glans, to achieve a cosmetically acceptable appearance with a circumcised look, and to enable the patient to void standing with a straight stream. Surgery is performed between 6 and 18 months of age (ideally 6 to 12 months). The choice of operation depends on the severity: anterior hypospadias may be corrected in one stage; posterior hypospadias often requires staged repair.

Commonly performed repairs include: MAGPI (meatal advancement and glanuloplasty incorporated: for glanular hypospadias), Mathieu (flip-flap repair: for distal hypospadias), TIP (tubularised incised plate: Snodgrass repair: the most widely used technique: a longitudinal incision is made in the urethral plate, which is tubularised over a catheter), and Duckett (transverse preputial island flap: for proximal hypospadias). In staged repairs, the first stage corrects the chordee and creates a graft bed; the second stage, 6 months later, creates the neo-urethra.

Fill in the blank: The most widely used hypospadias repair technique is the TIP (_____ incised plate or Snodgrass) repair. Surgery is ideally performed between 6 and _____ months of age. The two goals of hypospadias surgery include correcting the _____ and creating a neo-urethra to the tip of the glans.

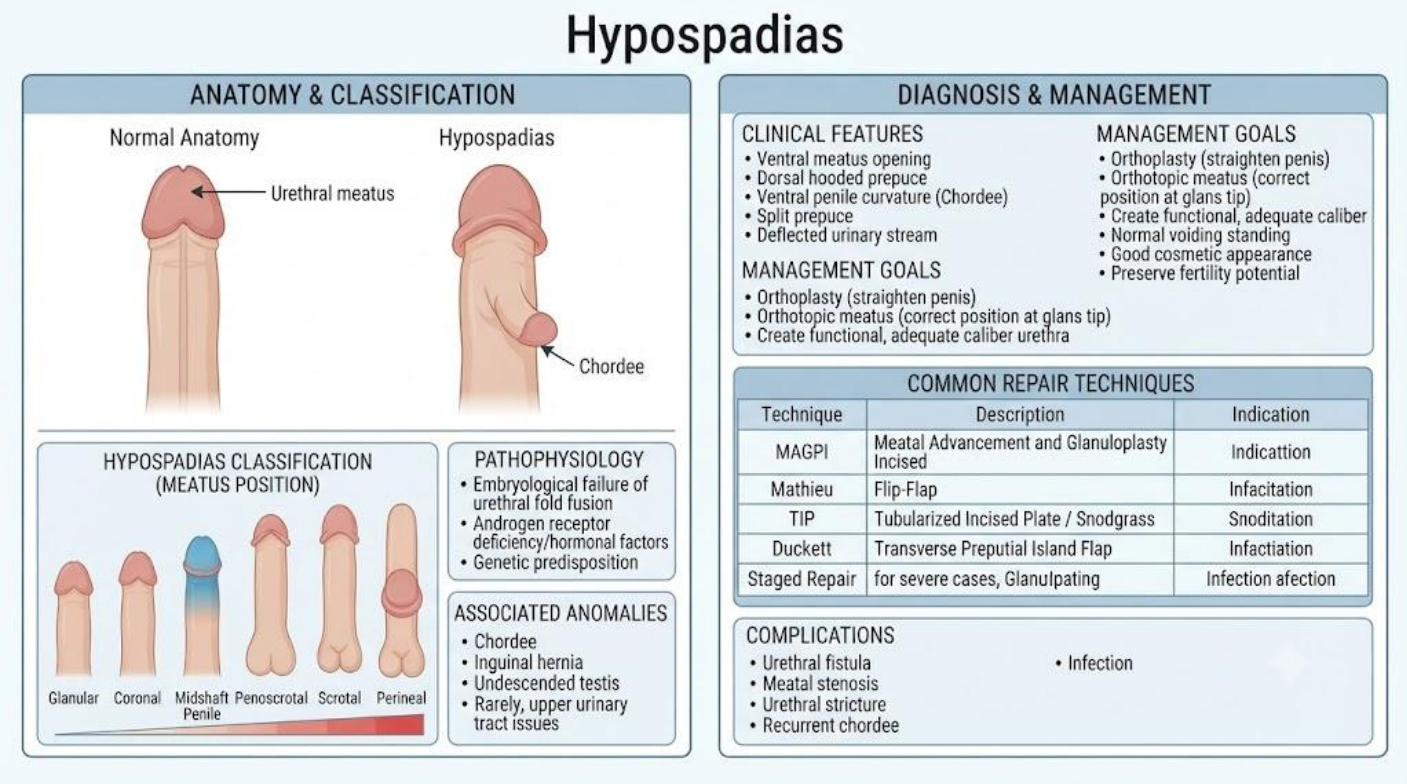

Figure 4.1: Anatomy, classification, clinical features, and management of hypospadias

Pilot questions 4.1

1. State the three consistent features of hypospadias. (Linked to SLO 4.1)
2. Classify hypospadias by meatal position and state the approximate frequency of each group. (Linked to SLO 4.1)
3. State the goals of hypospadias surgery and the ideal age for repair. (Linked to SLO 4.1)
4. Describe the TIP (Snodgrass) repair and state why it is widely used. (Linked to SLO 4.1)
5. State the most common complication of hypospadias repair. (Linked to SLO 4.1)

4.2 Epispadias and Bladder Exstrophy

4.2.1 Embryology and spectrum of anomalies

The exstrophy-epispadias complex (EEC) is a spectrum of congenital anomalies resulting from failure of mesenchymal ingrowth between the ectoderm and endoderm of the cloacal membrane during the 4th to 6th weeks of gestation. The cloacal membrane ruptures prematurely before it is reinforced by mesenchyme, resulting in exposure of the bladder and urethra. The spectrum

ranges in severity from epispadias alone (the mildest form: the urethra opens on the dorsal surface of the penis or clitoris) through classic bladder exstrophy (the most common severe form) to cloacal exstrophy (the most severe: bladder and bowel both exstrophied: associated with spinal and other anomalies).

Classic bladder exstrophy occurs in approximately 1 in 30,000 to 50,000 births (male to female ratio 2:1). The anterior bladder wall and anterior abdominal wall are absent; the posterior bladder wall is exposed and everted on the lower abdomen. The urethra is epispadiac (opens on the dorsal surface). The pubic symphysis is widely diastatic (the pubic bones do not meet in the midline: the pelvis is open anteriorly). The umbilicus is low-set. The penis in males is short and broad with dorsal chordee (upward curvature: the reverse of hypospadias chordee).

Fill in the blank: The exstrophy-epispadias complex results from failure of _____ ingrowth into the cloacal membrane, causing premature _____ of the membrane. In bladder exstrophy, the pubic _____ is widely diastatic because the pubic bones do not meet in the midline.

4.2.2 Clinical features and diagnosis

Bladder exstrophy is diagnosed at birth by inspection: the exposed, everted posterior bladder wall (the bladder plate) is visible on the lower abdomen as a red, mucosa-lined surface that continuously drips urine. The bladder plate must be protected immediately: cover it with a non-adherent dressing moistened with saline (do not use dry gauze, which damages the mucosa). The epispadiac urethra, separated pubic bones (felt on palpation), umbilicus displaced downward, and abnormal external genitalia confirm the diagnosis. Antenatal diagnosis by ultrasound is possible (absent bladder filling, abnormal genitalia, low umbilical insertion) but is often missed.

Associated anomalies in bladder exstrophy: vesicoureteric reflux (present in virtually all cases: the ureteric orifices are on the exposed bladder plate and reflux is inevitable before repair), inguinal herniae (due to wide diastasis of the pubic symphysis), undescended testes in males, and rectal prolapse (from pelvic floor weakness from the wide pubic diastasis). Epispadias alone (without exstrophy) presents in males as a dorsal urethral opening with dorsal chordee and urinary incontinence; in females as a bifid clitoris with a widened urethral opening.

Fill in the blank: In bladder exstrophy, the exposed bladder plate must be covered immediately with a _____ dressing moistened with saline. Vesicoureteric reflux is present in _____ all cases of bladder exstrophy because the ureteric orifices are on the exposed bladder plate. The pubic bones are separated causing a wide pubic _____.

4.2.3 Management

The management of bladder exstrophy is staged surgical reconstruction. The modern staged approach (the modified Cantwell-Ransley repair or the complete primary repair of exstrophy: CPRE) involves: Stage 1 (within 48 to 72 hours of birth if possible: bladder closure and abdominal wall reconstruction with iliac osteotomies to allow the pubic bones to be approximated in the midline, reducing tension on the closure), Stage 2 (at 6 to 12 months:

epispadias repair and penile reconstruction or epispadias repair in females), and Stage 3 (at 4 to 5 years: bladder neck reconstruction to achieve continence).

Long-term outcomes: with modern staged repair, most patients achieve social urinary continence (dryness for 3-hour intervals during the day), though many require bladder augmentation or continent urinary diversion for small or poorly compliant bladders. Sexual function can be preserved in most males. Cloacal exstrophy requires additional management of the exstrophied bowel, creation of colostomies, and more complex genitourinary reconstruction; the prognosis for continence and quality of life is more guarded.

Fill in the blank: Stage 1 of bladder exstrophy repair is performed within _____ to 72 hours of birth and includes bladder closure and iliac _____ to allow pubic bone approximation. Stage 3 (at 4 to 5 years) involves bladder _____ reconstruction to achieve urinary continence.

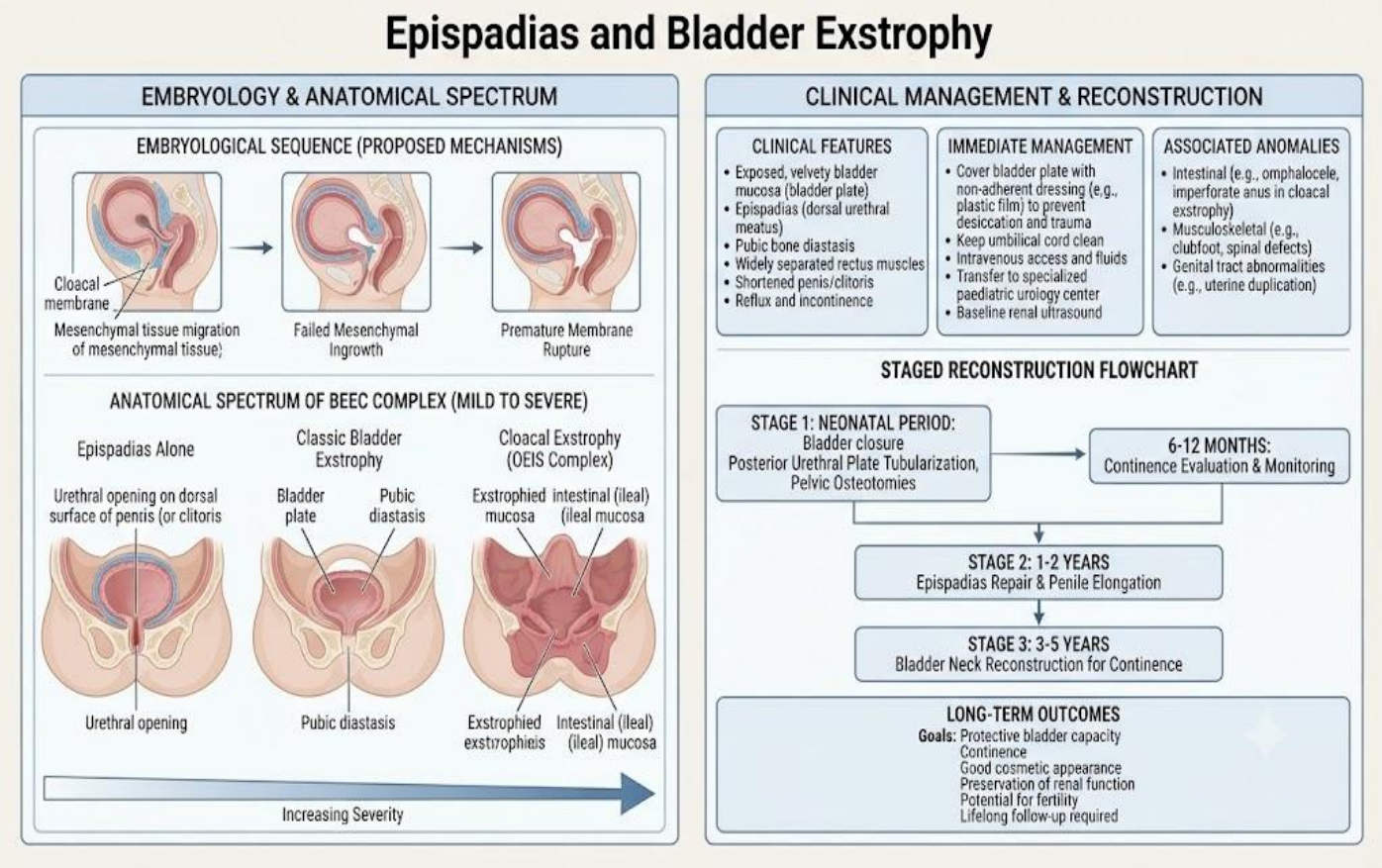

Figure 4.2: Spectrum, clinical features, and staged management of the exstrophy-epispadias complex

Pilot questions 4.2

1. Describe the embryological basis of the exstrophy-epispadias complex. (Linked to SLO 4.2)
2. Describe the clinical features of classic bladder exstrophy at birth. (Linked to SLO 4.2)
3. State the immediate management of a neonate born with bladder exstrophy. (Linked to SLO 4.2)
4. Describe the three stages of surgical reconstruction for bladder exstrophy. (Linked to SLO 4.2)
5. Explain why vesicoureteric reflux is present in virtually all cases of bladder exstrophy. (Linked to SLO 4.2)

4.3 Posterior Urethral Valves

4.3.1 Pathology and classification

Posterior urethral valves (PUV) are congenital obstructive leaflets within the posterior (prostatic) urethra of males, causing the most common and severe form of obstructive uropathy in children. They arise from abnormal fusion of the plicae colliculi (mucosal folds) at the verumontanum (the Young classification: Type I: the most common: two leaflets fusing anteriorly at the verumontanum causing a sail-like obstruction; Type III: a ring or membrane with a central opening at the verumontanum). The obstruction causes progressive bilateral hydroureteronephrosis, vesicoureteric reflux, bladder dysfunction (hypertrophied, poorly compliant, trabeculated bladder), and renal dysplasia.

The severity of renal damage depends on the degree of obstruction and the gestational age at which it began: very early obstruction (first trimester) causes severe renal dysplasia and oligohydramnios (reduced amniotic fluid from poor fetal urine output: the amniotic fluid is largely composed of fetal urine after 16 weeks). Oligohydramnios leads to pulmonary hypoplasia (the lungs require amniotic fluid for normal development: the most immediately life-threatening consequence of severe PUV), Potter sequence (oligohydramnios facies: flattened nose, low-set ears, limb deformities), and developmental limb deformities.

Fill in the blank: Posterior urethral valves cause the most common severe _____ uropathy in males. Type _____ PUV (the most common) consists of two leaflets fusing anteriorly at the verumontanum. Severe PUV causes oligohydramnios which leads to pulmonary _____ (the most immediately life-threatening complication).

4.3.2 Clinical features and diagnosis

The clinical presentation depends on the severity. Antenatal: bilateral hydronephrosis with a distended, thick-walled bladder on routine ultrasound; oligohydramnios in severe cases. Neonatal: respiratory distress (from pulmonary hypoplasia), a palpable distended bladder, bilateral flank masses (from hydroureteronephrosis), poor urinary stream or dribbling, and urosepsis. In older boys: recurrent urinary tract infections, poor urinary stream, straining to void, incontinence, and failure to thrive. A palpable bladder in a male neonate should always raise the suspicion of PUV.

Investigations: renal and bladder ultrasound (bilateral hydronephrosis and hydroureters, distended thick-walled trabeculated bladder, dilated posterior urethra: the keyhole sign), micturating cystourethrogram (MCUG: the definitive investigation: shows dilated posterior urethra with a filling defect at the valve level, trabeculated bladder, and vesicoureteric reflux), serum creatinine and electrolytes (renal function), and urine culture (urosepsis). The MCUG is performed once the infant is stabilised and urosepsis (if present) is treated.

Fill in the blank: The keyhole sign on renal ultrasound (dilated posterior urethra with distended bladder) is characteristic of _____. The definitive investigation for PUV is the micturating _____ (MCUG), which shows dilated posterior urethra and a filling defect at the _____ level.

4.3.3 Management and long-term outcomes

Initial management: urethral catheterisation (to drain the obstructed bladder and relieve back-pressure on the kidneys: a 6 to 8 French catheter is passed), IV antibiotics (for urosepsis), correction of electrolyte imbalances (hyperkalaemia and acidosis from renal impairment), and monitoring of renal function. Definitive treatment: cystoscopic valve ablation (transurethral resection or diathermy of the valve leaflets under direct vision using a paediatric cystoscope: the treatment of choice in term neonates and older boys). In premature or small neonates where the urethra is too small for a cystoscope, a temporary vesicostomy (bladder opening to the skin) is fashioned to divert urine until the child is large enough for cystoscopic ablation.

Long-term outcomes: even after successful valve ablation, renal function may continue to decline because of pre-existing renal dysplasia and ongoing bladder dysfunction (the valve bladder: a poorly compliant, high-pressure bladder that continues to damage the upper tracts even after the valve is ablated). Approximately 30 percent of boys with PUV develop end-stage renal disease by the age of 10 to 15 years and require dialysis or renal transplantation. Long-term follow-up with regular renal function tests, blood pressure monitoring, and urological assessment is essential for all patients.

Fill in the blank: Definitive treatment of PUV is cystoscopic _____ ablation (resection or diathermy of valve leaflets under direct vision). In small neonates where the urethra is too small for a cystoscope, a _____ is performed to divert urine. Approximately _____ percent of boys with PUV develop end-stage renal disease by 10 to 15 years.

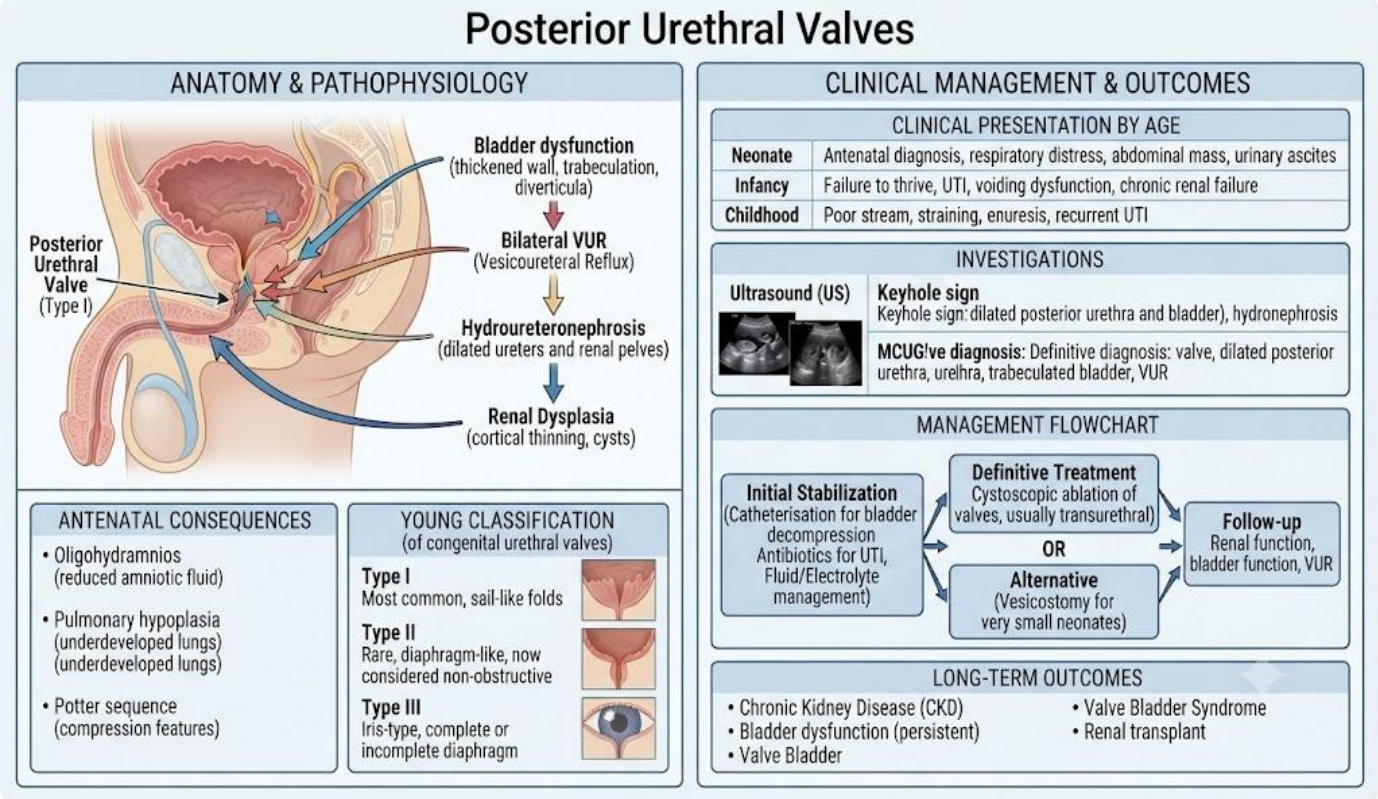

Figure 4.3: Pathology, clinical features, investigation, and management of posterior urethral valves

Pilot questions 4.3

1. Describe the Type I posterior urethral valve and the sequelae it causes. (Linked to SLO 4.3)
2. Explain how PUV causes pulmonary hypoplasia. (Linked to SLO 4.3)
3. Describe the keyhole sign and state the investigation on which it is seen. (Linked to SLO 4.3)
4. Describe the definitive treatment of PUV and state when a vesicostomy is used instead. (Linked to SLO 4.3)
5. Describe the long-term renal consequences of PUV. (Linked to SLO 4.3)

4.4 Wilms' Tumour

4.4.1 Pathology and epidemiology

Wilms' tumour (nephroblastoma) is the most common intra-abdominal malignancy in children, arising from the metanephric blastema (the embryonic precursor of the kidney). It typically occurs in children between 1 and 5 years of age, with a peak incidence at 3 to 4 years. The incidence is approximately 1 in 10,000 children. Wilms' tumour is bilateral in approximately 5 to 10 percent of cases. Histologically, it is characterised by a triphasic pattern of blastema, epithelium (tubules), and stroma. Anaplasia (unfavourable histology) is present in approximately 10 percent of cases and is the most important adverse prognostic factor.

Genetics: most cases are sporadic but approximately 10 to 15 percent are associated with specific genetic syndromes. WAGR syndrome (Wilms' tumour, Aniridia, Genitourinary anomalies, and intellectual disability: caused by deletion of chromosome 11p13 including the WT1 gene). Beckwith-Wiedemann syndrome (macroglossia, macrosomia, omphalocele, and hypoglycaemia: chromosome 11p15 abnormality). Denys-Drash syndrome (WT1 mutation: Wilms' tumour, diffuse mesangial sclerosis causing renal failure, and DSD). Children with these syndromes require regular renal ultrasound surveillance from birth.

Fill in the blank: Wilms' tumour arises from the _____ blastema and is characterised histologically by a _____ pattern (blastema, epithelium, and stroma). _____ (unfavourable histology) is the most important adverse prognostic factor. The WAGR syndrome includes Wilms' tumour, _____, genitourinary anomalies, and intellectual disability.

4.4.2 Clinical features and diagnosis

The classic presentation is a smooth, firm, non-tender abdominal mass in a well-looking child, discovered incidentally by a parent or during routine examination. The mass is typically unilateral, does not cross the midline, moves with respiration, and is dull to percussion. Associated features include haematuria (approximately 25 percent: usually microscopic), hypertension (from renin secretion by the tumour: approximately 25 percent), and anaemia. Constitutional symptoms (fever, weight loss) are uncommon and suggest advanced disease.

Investigations: ultrasound abdomen (first-line: confirms the intrarenal origin of the mass, assesses the contralateral kidney, detects renal vein or inferior vena cava thrombus), CT abdomen and chest (staging: assesses local invasion, lymph node involvement, liver metastases, and pulmonary metastases: the most common site of distant metastasis), and urine catecholamines (to exclude neuroblastoma, which may present similarly). The National Wilms' Tumour Study (NWTS) staging system: Stage I (tumour confined to kidney, completely excised), Stage II (tumour extends beyond kidney but completely excised), Stage III (residual non-haematogenous tumour in the abdomen), Stage IV (haematogenous metastases: lung or liver), Stage V (bilateral involvement).

Fill in the blank: The classic presentation of Wilms' tumour is a smooth firm _____ abdominal mass in a well-looking child. Pulmonary metastases are the most common site of distant spread and are assessed by CT of the _____. Stage _____ Wilms' tumour involves bilateral kidney involvement.

4.4.3 Management and prognosis

The management of Wilms' tumour differs between the North American (NWTS/COG) and European (SIOP) approaches. The NWTS approach: immediate nephrectomy (radical nephrectomy with regional lymph node sampling) followed by adjuvant chemotherapy (vincristine and actinomycin D for Stage I and II; add doxorubicin for Stage III; add radiotherapy for Stage III). The SIOP approach: preoperative chemotherapy (neoadjuvant) for 4 to 6 weeks to shrink the tumour before nephrectomy; this approach reduces the risk of intraoperative tumour rupture and alters the staging.

Bilateral Wilms' tumour (Stage V): treated with bilateral nephron-sparing surgery (partial nephrectomies) after preoperative chemotherapy to preserve as much renal function as possible, followed by further chemotherapy. Prognosis: the overall 4-year survival for Wilms' tumour is approximately 90 percent with modern multimodal therapy, making it one of the most curable solid tumours in children. Favourable histology: 4-year survival above 95 percent for Stage I to II. Anaplastic (unfavourable) histology Stage IV: survival approximately 55 to 60 percent.

Fill in the blank: The NWTS approach to Wilms' tumour is immediate _____ followed by adjuvant chemotherapy. Bilateral Wilms' tumour is treated with _____ nephron-sparing surgery after chemotherapy. The overall 4-year survival for Wilms' tumour is approximately _____ percent.

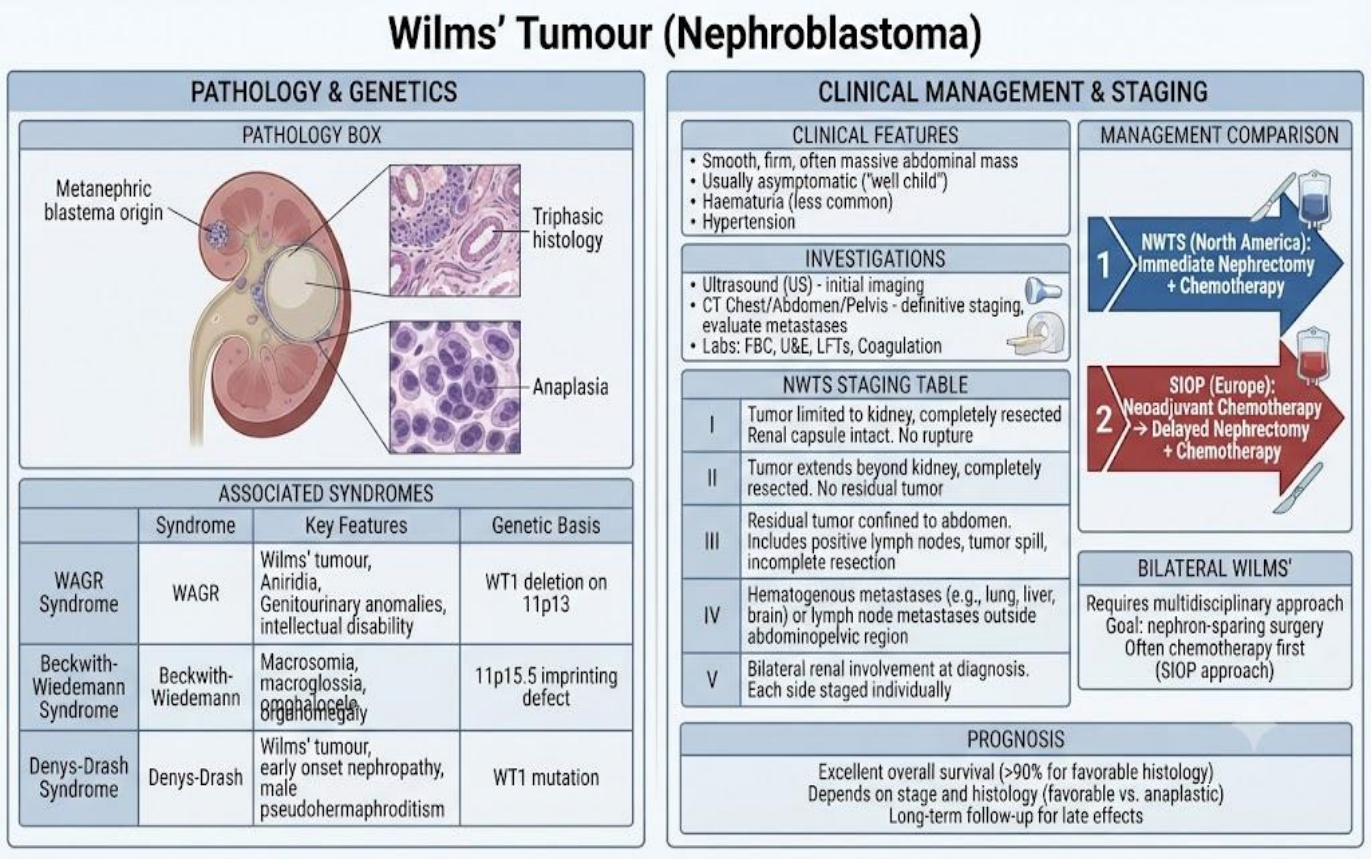

Figure 4.4: Pathology, staging, and management of Wilms' tumour

Pilot questions 4.4

1. Describe the typical clinical presentation of Wilms' tumour. (Linked to SLO 4.4)
2. Describe the NWTS staging system for Wilms' tumour. (Linked to SLO 4.4)
3. Compare the NWTS and SIOP approaches to the management of Wilms' tumour. (Linked to SLO 4.4)
4. Describe the management of bilateral Wilms' tumour. (Linked to SLO 4.4)
5. State the prognosis for Wilms' tumour and identify the most important adverse prognostic factor. (Linked to SLO 4.4)

Series Summary

This Series covered the major genitourinary malformations in paediatric surgical practice. Hypospadias presents with a ventral meatal opening, hooded foreskin, and chordee; surgery (TIP or Snodgrass repair) is performed at 6 to 12 months. The exstrophy-epispadias complex results from failed mesenchymal ingrowth; bladder exstrophy requires staged repair (bladder closure with osteotomies, epispadias repair, bladder neck reconstruction). Posterior urethral valves cause the most severe obstructive uropathy in boys; the keyhole sign on ultrasound and MCUG confirm the diagnosis; cystoscopic valve ablation is the definitive treatment, but 30 percent develop end-stage renal disease.

You should now be able to describe the anatomy, classification, and management of hypospadias (SLO 4.1), describe the exstrophy-epispadias complex and its staged management (SLO 4.2), describe the pathology, diagnosis, and management of posterior urethral valves (SLO 4.3), and describe the clinical features, staging, and management of Wilms' tumour (SLO 4.4). Wilms' tumour has one of the best prognoses among paediatric solid tumours, with an overall 4-year survival of approximately 90 percent.

Glossary of Terms

- **Anaplasia:** an adverse histological feature of Wilms' tumour characterised by nuclear enlargement (three times normal size), hyperchromatic nuclei, and multipolar mitoses; present in approximately 10 percent of cases; the most important adverse prognostic factor.

- **Chordee:** ventral curvature of the penis in hypospadias (or dorsal curvature in epispadias), caused by fibrous tethering of the ventral or dorsal shaft; must be corrected before urethral reconstruction.
- **Exstrophy-epispadias complex (EEC):** a spectrum of congenital anomalies from epispadias alone through classic bladder exstrophy to cloacal exstrophy, resulting from failure of mesenchymal ingrowth into the cloacal membrane during the 4th to 6th weeks of gestation.
- **Keyhole sign:** the ultrasound appearance of posterior urethral valves: a dilated posterior urethra with a distended thick-walled bladder resembling a keyhole in cross-section; characteristic but not pathognomonic of PUV.
- **MCUG (micturating cystourethrogram):** a fluoroscopic contrast study of the bladder and urethra performed during voiding; the definitive investigation for posterior urethral valves, showing the dilated posterior urethra, filling defect at the valve, trabeculated bladder, and vesicoureteric reflux.
- **Metanephric blastema:** the embryonic mesenchymal tissue from which the kidney develops; the cell of origin of Wilms' tumour.
- **Oligohydramnios:** reduced volume of amniotic fluid, typically below the 5th centile for gestational age; caused by reduced fetal urine output in severe posterior urethral valves; leads to pulmonary hypoplasia and Potter sequence.
- **TIP repair (tubularised incised plate):** the Snodgrass repair; the most widely used technique for hypospadias; a longitudinal incision is made in the urethral plate, which is then tubularised over a stenting catheter to create the neo-urethra.
- **Valve bladder:** the chronically dysfunctional, hypertrophied, poorly compliant, high-pressure bladder that persists after successful cystoscopic ablation of posterior urethral valves; continues to damage the upper urinary tracts and contributes to progressive renal deterioration.
- **Vesicostomy:** a surgical opening between the anterior wall of the bladder and the skin of the lower abdomen, providing temporary urinary diversion in small or premature neonates with posterior urethral valves until they are large enough for cystoscopic valve ablation.
- **WAGR syndrome:** a genetic syndrome associated with Wilms' tumour: Wilms' tumour (or predisposition), Aniridia, Genitourinary anomalies, and intellectual disability (formerly Retardation); caused by deletion of chromosome 11p13 including the WT1 tumour suppressor gene.

Concept Check Questions [Series 4]

Objective questions

1. The three consistent features of hypospadias are an abnormally positioned meatus, a hooded foreskin, and ventral curvature called _____. (Linked to SLO 4.1)
2. In bladder exstrophy, the pubic _____ is widely diastatic because the pubic bones do not meet in the midline. (Linked to SLO 4.2)
3. The keyhole sign on renal ultrasound (dilated posterior urethra with distended bladder) is characteristic of posterior urethral _____. (Linked to SLO 4.3)
4. Wilms' tumour arises from the _____ blastema and is the most common intra-abdominal malignancy in children. (Linked to SLO 4.4)
5. The overall 4-year survival for Wilms' tumour with modern treatment is approximately _____ percent. (Linked to SLO 4.4)

Short-answer questions

6. State the goals of hypospadias surgery and the ideal timing. (Linked to SLO 4.1)
7. Describe the immediate management of a neonate born with bladder exstrophy. (Linked to SLO 4.2)
8. Describe the definitive treatment of posterior urethral valves and state when a vesicostomy is used. (Linked to SLO 4.3)
9. Describe the clinical features of Wilms' tumour. (Linked to SLO 4.4)
10. Compare the NWTS and SIOP approaches to Wilms' tumour management. (Linked to SLO 4.4)

Long-answer questions

11. Describe the classification and surgical management of hypospadias. (Linked to SLO 4.1)
12. Describe the staged surgical reconstruction for classic bladder exstrophy. (Linked to SLO 4.2)
13. Describe the sequelae of posterior urethral valves and the long-term renal consequences. (Linked to SLO 4.3)

14. Describe the NWTS staging system and management of Wilms' tumour by stage. (Linked to SLO 4.4)
15. Compare posterior urethral valves and Wilms' tumour in terms of presentation and investigations. (Linked to SLO 4.3 and 4.4)

Answers to Concept Check Questions [Series 4]

Objective questions

1. Chordee.
2. Symphysis.
3. Valves (PUV).
4. Metanephric.
5. 90.

Short-answer questions

6. Goals: correct the chordee (straighten the penis), create a neo-urethra to the tip of the glans, achieve a cosmetically acceptable circumcised appearance, and enable standing micturition with a straight urinary stream. Timing: 6 to 18 months of age, ideally 6 to 12 months.
7. Cover the exposed bladder plate immediately with a non-adherent dressing moistened with saline (do not use dry gauze: damages the delicate bladder mucosa). Ensure warmth and IV access. Pass a nasogastric tube. Transfer urgently to a paediatric surgical centre for staged reconstruction. Screen for associated anomalies.
8. Definitive treatment: cystoscopic valve ablation (transurethral resection or diathermy of the valve leaflets under direct vision with a paediatric cystoscope). Vesicostomy: used in premature or very small neonates where the urethra is too small to accommodate the paediatric cystoscope; provides temporary urinary diversion until the child is large enough for cystoscopic ablation.
9. A smooth, firm, non-tender abdominal mass in a well-looking child aged 1 to 5 years, discovered incidentally. The mass is unilateral, does not cross the midline, moves with

respiration, and is dull to percussion. Associated features: haematuria (25 percent), hypertension (25 percent from tumour renin secretion), and anaemia.

10. NWTS (North American) approach: immediate radical nephrectomy with lymph node sampling, followed by adjuvant chemotherapy (vincristine plus actinomycin D for Stage I and II; add doxorubicin for Stage III; add radiotherapy for Stage III). SIOP (European) approach: preoperative neoadjuvant chemotherapy for 4 to 6 weeks to shrink the tumour before nephrectomy, reducing the risk of intraoperative tumour rupture and potentially downstaging the disease.

Long-answer questions

11. **Basic response:** Classification by meatal position: anterior (glanular and coronal: approximately 50 percent), middle (penile shaft: approximately 30 percent), posterior (penoscrotal, scrotal, and perineal: approximately 20 percent). Surgical management: goals are to correct chordee, create a neo-urethra to the glans tip, achieve a cosmetically acceptable appearance, and enable standing micturition; performed at 6 to 12 months. Common techniques: MAGPI (glanular hypospadias: meatal advancement and glanuloplasty), Mathieu flip-flap (distal hypospadias), TIP or Snodgrass (most widely used: longitudinal incision in the urethral plate tubularised over a catheter), Duckett transverse preputial island flap (proximal hypospadias). Staged repair for posterior hypospadias: Stage 1 corrects chordee and creates a graft bed; Stage 2 at 6 months creates the neo-urethra. Most common complication: urethrocutaneous fistula.

Value-added response: The TIP (Snodgrass) repair is the most widely used technique because it is applicable to most hypospadias types (distal and middle), consistently produces a slit-like meatus at the glans tip rather than a round meatus (better cosmesis), has acceptable complication rates even in inexperienced hands, and preserves the vascular urethral plate (avoiding the need for preputial tissue grafts in most cases).

12. **Basic response:** Stage 1 (within 48 to 72 hours of birth): primary bladder closure. The bladder plate is dissected free, the bladder is closed in layers, and the abdominal wall is reconstructed. Bilateral iliac osteotomies (cuts through the iliac bones) are performed to allow the pubic bones to be approximated in the midline, reducing tension on the bladder closure and improving pelvic floor support. The baby is placed in Bryant's traction or a hip spica after surgery to maintain pubic approximation. Stage 2 (at 6 to 12 months): epispadias repair and penile or clitoral reconstruction. In males: the epispadiac urethra is reconstructed and the dorsal chordee is corrected to achieve a cosmetically acceptable penis. In females: clitoral reconstruction and urethral lengthening. Stage 3 (at 4 to 5 years, when the child is developmentally ready for toilet training): bladder neck

reconstruction (Young-Dees-Leadbetter procedure) to create a competent bladder outlet and achieve urinary continence.

Value-added response: The osteotomies in Stage 1 are critical to the success of the closure: without them, the pubic bones cannot be approximated and the tension on the midline closure is excessive, leading to wound breakdown and bladder prolapse. In resource-limited Nigerian centres where osteotomy expertise is limited, outcomes of bladder exstrophy repair are correspondingly poorer, highlighting the need for centralisation of exstrophy care.

13. **Basic response:** Sequelae cascade: PUV obstructs bladder outflow, causing: (1) bladder hypertrophy and trabeculation (the detrusor muscle hypertrophies to overcome the obstruction), (2) bilateral vesicoureteric reflux (from ureteric orifice distortion by the hypertrophied bladder), (3) bilateral hydroureteronephrosis (from raised intravesical pressure transmitted to the upper tracts), (4) renal dysplasia (from the combination of obstructive uropathy and VUR during fetal development: the kidneys develop abnormally), and (5) renal failure. Antenatal: severe PUV causes oligohydramnios (reduced fetal urine output: amniotic fluid predominantly fetal urine after 16 weeks) leading to pulmonary hypoplasia (most immediately life-threatening) and Potter sequence. Long-term renal consequences: even after successful valve ablation, the valve bladder (poorly compliant, high-pressure, dysfunctional bladder) persists and continues to damage the upper urinary tracts. Approximately 30 percent of boys develop end-stage renal disease by 10 to 15 years, requiring dialysis or renal transplantation. Regular monitoring of renal function (eGFR, serum creatinine), blood pressure, and bladder function is essential lifelong.

Value-added response: The nadir creatinine (the lowest serum creatinine achieved at approximately 1 year of age after valve ablation and bladder drainage) is the single best predictor of long-term renal outcome: a nadir creatinine above 80 micromol/L is associated with a significantly higher risk of end-stage renal disease. This measurement guides the intensity of follow-up and early referral for renal transplant planning.

14. **Basic response:** NWTS staging: Stage I (tumour confined to kidney, completely excised with intact capsule, no tumour rupture, lymph nodes negative: chemotherapy with vincristine plus actinomycin D for 18 weeks), Stage II (tumour extends beyond kidney: penetrates renal capsule or involves renal sinus vessels: completely excised: chemotherapy as Stage I), Stage III (residual non-haematogenous tumour remaining in the abdomen: positive lymph nodes, positive surgical margin, tumour spill, peritoneal implants: chemotherapy with vincristine plus actinomycin D plus doxorubicin plus flank or abdominal radiotherapy), Stage IV (haematogenous metastases: most commonly pulmonary: whole-lung irradiation plus intensive chemotherapy), Stage V (bilateral: preoperative chemotherapy then bilateral nephron-sparing surgery aiming to preserve

maximum renal function; avoid nephrectomy of both kidneys; subsequent chemotherapy based on histology).

Value-added response: Lymph node sampling at nephrectomy is mandatory in the NWTS approach: failure to sample lymph nodes upstages the tumour by definition to Stage III (treatment then includes doxorubicin and radiotherapy). This is a critical intraoperative step that must not be omitted, even if the nodes appear macroscopically normal.

15. **Basic response:** PUV presentation: typically in neonates or older boys; palpable distended bladder and bilateral flank masses (from hydroureteronephrosis); poor urinary stream or dribbling; in neonates: respiratory distress (pulmonary hypoplasia); antenatal: bilateral hydronephrosis and distended bladder on ultrasound. Investigation: ultrasound (keyhole sign: dilated posterior urethra and distended bladder; bilateral hydronephrosis); MCUG (definitive: dilated posterior urethra with valve filling defect; trabeculated bladder; VUR). Wilms' tumour presentation: typically 1 to 5 years; smooth firm non-tender unilateral abdominal mass in a well-looking child; haematuria and hypertension in 25 percent each. Investigation: ultrasound (intrarenal origin; does not cross midline; assesses IVC thrombus; contralateral kidney); CT abdomen and chest (staging: lymph nodes, liver, pulmonary metastases); urine catecholamines (exclude neuroblastoma). Key distinction: PUV presents with bladder and bilateral upper tract abnormalities on ultrasound; Wilms' tumour presents as a unilateral intrarenal mass in a well child with no lower urinary tract abnormality.

Value-added response: Neuroblastoma is the most important differential diagnosis for Wilms' tumour: both present as an abdominal mass in a young child. Key distinguishing features: Wilms' tumour is intrarenal (distorts the pelvicalyceal system on imaging), smooth, does not cross the midline, and the child is well; neuroblastoma is extrarenal (displaces the kidney rather than arising from it), frequently crosses the midline (wraps around the aorta and IVC), and the child may be unwell with bone pain, weight loss, or opsoclonus-myoclonus syndrome.

Answers to Pilot Questions [Series 4]

Part 4.1

6. An abnormally positioned urethral meatus (on the ventral surface of the penis proximal to its normal position at the glans tip), a hooded (incomplete) foreskin (the foreskin is deficient ventrally and forms a dorsal hood over the glans), and chordee (ventral curvature of the penis from fibrous tethering of the ventral shaft).

7. Anterior (glanular and coronal): approximately 50 percent; the most common group; generally amenable to single-stage repair. Middle (penile shaft): approximately 30

percent; may require more complex repair. Posterior (penoscrotal, scrotal, and perineal): approximately 20 percent; the most severe; often requires staged repair; associated with higher rates of undescended testis and DSD.

8. Goals: (1) correct the chordee (straighten the penis), (2) create a neo-urethra extending to the tip of the glans, (3) achieve a cosmetically acceptable appearance (circumcised look), and (4) enable standing micturition with a straight forward urinary stream. Ideal timing: 6 to 18 months of age, ideally 6 to 12 months; at this age the penis is large enough for surgery, anaesthetic risk is low, and repair is complete before the child develops body awareness.

9. The TIP (tubularised incised plate) or Snodgrass repair: a longitudinal midline incision is made into the urethral plate (the tissue between the meatus and the glans) from the meatus to the glans tip. The incised plate is then tubularised over a stenting urethral catheter using absorbable sutures to form the neo-urethra. The glans is then wrapped around the neo-urethra (glansplasty) to create a slit-like meatus at the tip. It is widely used because it is applicable to most hypospadias types, gives a cosmetically superior slit meatus, and has good reproducibility.

10. The most common complication is a urethrocutaneous fistula: an abnormal communication between the neo-urethra and the skin surface, causing urine to leak through a pinhole opening along the shaft during voiding. Other complications include meatal stenosis, urethral stricture, residual chordee, and wound infection. Repeat surgery is required for complications in up to 10 to 20 percent of cases.

Part 4.2

11. During normal development, mesenchymal cells migrate between the ectoderm and endoderm of the cloacal membrane between the 4th and 6th weeks of gestation, reinforcing it and preventing premature rupture. In the exstrophy-epispadias complex, this mesenchymal ingrowth fails to occur, leaving the cloacal membrane unsupported. It then ruptures prematurely, exposing the underlying bladder (and bowel in cloacal exstrophy) to the surface. The severity of the anomaly depends on the timing and extent of the failure of ingrowth.

12. The exposed bladder plate (the everted posterior bladder wall) is visible as a red, moist, mucosa-lined surface on the lower abdomen, continuously dripping urine. The pubic symphysis is widely diastatic (the pubic bones are separated and felt on palpation). The umbilicus is low-set. The urethra is epispadiac (opens on the dorsal surface). In males: the penis is short and broad with dorsal chordee. In females: the clitoris is bifid and the labia are widely separated.

13. Cover the bladder plate immediately with a non-adherent dressing (such as Mepitel or paraffin gauze) moistened with warm normal saline; do not use dry gauze which adheres to and damages the bladder mucosa. Keep the baby warm. Establish IV access. Pass a nasogastric tube. Transfer urgently to a paediatric urology centre experienced in exstrophy reconstruction. Screen for associated anomalies (VUR is present in virtually all cases; check for undescended testes and inguinal herniae in males).

14. Stage 1 (within 48 to 72 hours): primary bladder closure and abdominal wall reconstruction, with bilateral iliac osteotomies to allow the widely separated pubic bones to be approximated in the midline and reduce tension on the closure; the baby is immobilised in Bryant's traction or a hip spica postoperatively. Stage 2 (at 6 to 12 months): epispadias repair (urethral reconstruction) and penile lengthening or clitoral reconstruction in females. Stage 3 (at 4 to 5 years): bladder neck reconstruction (Young-Dees-Leadbetter procedure) to create a continent bladder outlet and achieve urinary continence.

15. In bladder exstrophy, the ureteric orifices open directly onto the exposed bladder plate (not into a formed bladder with a normal trigone). Before surgical closure, the ureteric orifices are exposed and unprotected. Even after closure, the bladder is small and poorly compliant, generating high intravesical pressures during filling that force urine retrograde up the ureters. The absence of a normal anti-reflux mechanism (normally provided by the oblique intramural course of the ureter through the bladder wall) means VUR is inevitable in virtually all cases until it is corrected at subsequent ureteric reimplantation.

Part 4.3

1. Type I PUV consists of two obstructive mucosal leaflets arising from the verumontanum (an elevation on the posterior urethral wall at the level of the prostatic utricle) and fusing anteriorly to form a sail-like or crescentic obstruction across the posterior urethra. The leaflets partially obstruct urine flow from the bladder. Sequelae: bladder outflow obstruction causes bladder hypertrophy and trabeculation; raised intravesical pressure produces bilateral vesicoureteric reflux and progressive bilateral hydroureteronephrosis; renal back-pressure combined with reduced amniotic fluid (oligohydramnios) impairs fetal renal development, causing renal dysplasia; antenatally, severe cases cause oligohydramnios, pulmonary hypoplasia, and Potter sequence.

2. After approximately 16 weeks of gestation, fetal urine is the main component of amniotic fluid. Severe PUV obstructs bladder emptying, reducing fetal urine output into the amniotic sac. Oligohydramnios results. The fetal lungs require amniotic fluid to expand and develop normally: alveolar branching, surfactant production, and lung growth all

depend on repeated cycles of inhalation and exhalation of amniotic fluid (fetal breathing movements). When oligohydramnios prevents adequate fluid entry into the airways, the lungs fail to grow normally, resulting in pulmonary hypoplasia. This is the most immediately life-threatening complication of severe PUV at birth.

3. The keyhole sign is the characteristic ultrasound appearance of posterior urethral valves: a dilated posterior urethra (appearing as the shaft of the key) combined with a distended, thick-walled bladder (appearing as the circular head of the key), creating the outline of a keyhole in cross-section on transverse ultrasound imaging. It is seen on renal and bladder ultrasound and is characteristic (though not pathognomonic) of PUV.

4. Definitive treatment: cystoscopic valve ablation performed under general anaesthesia. A paediatric cystoscope is passed transurethrally; the valve leaflets are visualised directly and ablated by cold knife incision at the 5 and 7 o'clock positions (or by diathermy or laser). This relieves the obstruction at its source. Vesicostomy: used in premature neonates (typically below 34 weeks gestational age) or very small neonates where the urethra is too small (typically less than 12 to 14 French) to safely accommodate the paediatric resectoscope; a vesicostomy (opening between the bladder dome and the skin) provides temporary urinary diversion until the child has grown sufficiently for safe cystoscopic ablation (usually by 3 to 6 months of age).

5. Even after successful cystoscopic valve ablation, two factors cause continued renal deterioration. First, pre-existing renal dysplasia (from intrauterine obstructive uropathy during the critical period of nephrogenesis) is irreversible: dysplastic kidneys have a reduced nephron complement and poor reserve. Second, the valve bladder (the chronically hypertrophied, poorly compliant, high-pressure bladder that persists despite successful valve ablation) continues to transmit high pressures to the upper urinary tracts during filling and voiding, causing ongoing renal damage. Approximately 30 percent of boys develop end-stage renal disease by 10 to 15 years of age, requiring dialysis or renal transplantation. Long-term monitoring of eGFR, blood pressure, and bladder function is therefore essential.

Part 4.4

1. A smooth, firm, non-tender abdominal mass in a well-looking child aged 1 to 5 years, typically discovered incidentally by a parent while bathing the child or during routine examination. The mass is unilateral (bilateral in 5 to 10 percent), does not cross the midline, moves with respiration, and is dull to percussion. Associated features: microscopic or macroscopic haematuria (approximately 25 percent), hypertension (approximately 25 percent: from tumour renin secretion), and anaemia. Constitutional

symptoms (fever, weight loss, and anorexia) are uncommon and suggest advanced disease.

2. Stage I: tumour confined to the kidney, completely excised with intact capsule, no rupture, lymph nodes negative. Stage II: tumour penetrates the renal capsule or involves the renal sinus vessels but is completely excised with negative margins. Stage III: residual non-haematogenous tumour in the abdomen: positive regional lymph nodes, positive surgical margins, tumour spill (pre- or intraoperative), or peritoneal implants. Stage IV: haematogenous metastases, most commonly to the lungs; also liver. Stage V: synchronous bilateral Wilms' tumour.

3. NWTS (COG) approach: immediate radical nephrectomy with regional lymph node sampling is performed first, then adjuvant chemotherapy is given based on stage and histology (vincristine plus actinomycin D for Stage I and II; add doxorubicin for Stage III; add radiotherapy for Stage III abdominal disease; whole-lung irradiation and intensive chemotherapy for Stage IV). SIOP (European) approach: preoperative neoadjuvant chemotherapy for 4 to 6 weeks (vincristine plus actinomycin D) is given before nephrectomy. Advantages of SIOP: reduces tumour size and vascularity before surgery; reduces intraoperative tumour rupture risk; may allow nephron-sparing surgery in borderline cases. Disadvantage: chemotherapy may alter histology and make final staging less precise.

4. Bilateral Wilms' tumour (Stage V) is managed by bilateral nephron-sparing surgery (partial nephrectomies) aiming to excise the tumours while preserving as much normal renal parenchyma as possible to prevent end-stage renal disease. Preoperative chemotherapy (neoadjuvant: vincristine plus actinomycin D for 4 to 6 weeks) is given first to shrink both tumours and make nephron-sparing surgery more feasible. After surgery, further chemotherapy is given based on histology. Bilateral radical nephrectomies should be avoided; if one kidney must be sacrificed, the other must be preserved.

5. Prognosis: the overall 4-year survival for Wilms' tumour is approximately 90 percent with modern multimodal therapy (surgery, chemotherapy, and radiotherapy), making it one of the most curable solid tumours in children. By stage: favourable histology Stage I and II: 4-year survival above 95 percent; Stage III: approximately 90 percent; Stage IV: approximately 85 percent. Anaplasia (unfavourable histology) is the most important adverse prognostic factor: anaplastic Stage IV Wilms' tumour has a 4-year survival of approximately 55 to 60 percent. Bilateral tumours and late presentation are also associated with worse outcomes.

References and Attributions [Series 4]

Textual References

1. Coran, A. G., Caldamone, A., Adzick, N. S., Krummel, T. M., Laberge, J. M. and Shamberger, R. (2012). Pediatric Surgery (7th ed.). Philadelphia: Elsevier Mosby.
2. Puri, P. and Hollwarth, M. E. (2009). Pediatric Surgery: Diagnosis and Management. Berlin: Springer.
3. Ameh, E. A. and Chirdan, L. B. (2011). Paediatric Surgery in the Tropics. Ibadan: Spectrum Books.
4. Snodgrass, W. T. (1994). Tubularized incised plate urethroplasty for distal hypospadias. Journal of Urology, 151(2), 464 to 465.

Figures, Plates, Tables, and Boxes

5. Figure 4.1: Hypospadias. AI-generated using the prompt: "Two-panel diagram: sagittal penis illustration (normal versus hypospadias with ventral meatus and hooded foreskin and chordee), classification diagram (glanular, penile, penoscrotal, scrotal, perineal), pathophysiology and associated anomalies boxes; clinical features box, management goals box, repair techniques table (MAGPI, Mathieu, TIP/Snodgrass, Duckett, staged), and complications box. Paediatric urological diagram style." Suggested OER search string: "hypospadias classification TIP Snodgrass repair chordee paediatric urology OER."
6. Figure 4.2: Exstrophy-epispadias complex. AI-generated using the prompt: "Two-panel diagram: embryological sequence (failed mesenchymal ingrowth) and spectrum diagram (epispadias, bladder exstrophy, cloacal exstrophy); clinical features box, immediate management box, associated anomalies box, staged reconstruction flowchart (Stage 1 osteotomies, Stage 2 epispadias repair, Stage 3 bladder neck), and outcomes box. Paediatric urological diagram style." Suggested OER search string: "bladder exstrophy epispadias staged repair osteotomy bladder neck reconstruction paediatric urology OER."
7. Figure 4.3: Posterior urethral valves. AI-generated using the prompt: "Two-panel diagram: sagittal male urethra (Type I PUV) and sequelae cascade (bladder dysfunction, VUR, hydroureteronephrosis, renal dysplasia) and antenatal consequences (oligohydramnios, pulmonary hypoplasia, Potter sequence); clinical presentation by age, investigations (ultrasound keyhole sign and MCUG), management flowchart (catheterisation, antibiotics, cystoscopic ablation, vesicostomy), and long-term outcomes box. Paediatric urological diagram style." Suggested OER search string: "posterior

urethral valves MCUG keyhole sign cystoscopic ablation vesicostomy renal dysplasia paediatric OER."

8. Figure 4.4: Wilms' tumour. AI-generated using the prompt: "Two-panel diagram: pathology box (metanephric blastema, triphasic histology, anaplasia) and associated syndromes table (WAGR, Beckwith-Wiedemann, Denys-Drash); clinical features box, investigations (ultrasound, CT staging), NWTS staging table (Stages I to V), management comparison (NWTS versus SIOP), bilateral Wilms' box, and prognosis box. Paediatric oncological diagram style." Suggested OER search string: "Wilms' tumour nephroblastoma NWTS staging nephrectomy chemotherapy WAGR syndrome OER."

Series 5: Intestinal Obstruction and Other Malformations

- **Part 5.1: Intestinal Obstruction in Neonates and Infants**
    - Sub Part 5.1.1: Causes and classification
    - Sub Part 5.1.2: Clinical features and investigation
    - Sub Part 5.1.3: Management principles
- **Part 5.2: Intussusception**
    - Sub Part 5.2.1: Pathophysiology and aetiology
    - Sub Part 5.2.2: Clinical features and diagnosis
    - Sub Part 5.2.3: Management
- **Part 5.3: Meckel's Diverticulum and Intestinal Duplications**
    - Sub Part 5.3.1: Meckel's diverticulum
    - Sub Part 5.3.2: Intestinal duplications
    - Sub Part 5.3.3: Management of Meckel's diverticulum and duplications
- **Part 5.4: Necrotising Enterocolitis**
    - Sub Part 5.4.1: Pathophysiology and risk factors
    - Sub Part 5.4.2: Clinical features and staging
    - Sub Part 5.4.3: Management and outcomes

Introduction

Intestinal obstruction in children presents across a broad age range and has a wide spectrum of causes, from structural malformations in the neonate to acquired conditions such as intussusception and necrotising enterocolitis in the infant. Many of these conditions are surgical

emergencies that, if unrecognised or managed late, can result in extensive intestinal loss and lifelong nutritional consequences.

This final Series covers the major causes of intestinal obstruction and other malformations relevant to Nigerian paediatric surgical practice. We examine the general approach to intestinal obstruction in neonates and infants, then study intussusception, Meckel's diverticulum, intestinal duplications, and necrotising enterocolitis in detail. Together with the previous Series, this Series completes the core content of Paediatric Surgery.

Series Learning Outcomes

Upon completing this Series, you should be able to:

- **SLO 5.1:** classify and describe the clinical approach to intestinal obstruction in neonates and infants,
- **SLO 5.2:** describe the pathophysiology, clinical features, diagnosis, and management of intussusception,
- **SLO 5.3:** describe the pathological anatomy, clinical presentations, and management of Meckel's diverticulum and intestinal duplications, and
- **SLO 5.4:** describe the pathophysiology, staging, and management of necrotising enterocolitis and state its prognosis.

5.1 Intestinal Obstruction in Neonates and Infants

5.1.1 Causes and classification

Intestinal obstruction in neonates is classified as mechanical (a physical blockage) or functional (paralytic ileus: failure of peristalsis without a physical block). Mechanical causes are further subdivided by the level of obstruction. High obstruction (above the mid-jejunum): oesophageal atresia, duodenal atresia, annular pancreas, and jejunal atresia (causes early and profuse vomiting with minimal distension). Low obstruction (below the mid-jejunum): ileal atresia, meconium ileus, Hirschsprung's disease, and anorectal malformations (causes delayed and faeculent vomiting with marked distension and delayed passage of meconium).

Meconium ileus is obstruction of the terminal ileum by inspissated, tenacious meconium (abnormally thick from absent intestinal secretions), the earliest manifestation of cystic fibrosis (CF). It occurs in approximately 15 percent of neonates with CF. Simple meconium ileus (without perforation) presents with abdominal distension, bilious vomiting, and failure to pass meconium; the AXR shows dilated loops with a ground-glass appearance (soap-bubble sign) in

the right iliac fossa from the sticky meconium. Complicated meconium ileus involves volvulus, atresia, or perforation of the distended ileum.

Fill in the blank: High intestinal obstruction causes _____ and profuse vomiting with minimal distension. Meconium ileus is the earliest manifestation of _____ fibrosis and presents on AXR with a _____ appearance (soap-bubble sign) in the right iliac fossa.

5.1.2 Clinical features and investigation

The cardinal features of neonatal intestinal obstruction are: bilious vomiting (bile-stained green vomit: obstruction distal to the ampulla of Vater: always a surgical concern in a neonate), abdominal distension (progressive, correlating with the level of obstruction: minimal in high obstruction and maximal in low obstruction), failure to pass meconium (meconium is normally passed within 48 hours: failure suggests low obstruction: Hirschsprung's disease or anorectal malformation), and dehydration (from fluid losses in vomit).

Investigations: plain AXR (supine and erect or left lateral decubitus: identifies dilated bowel loops, air-fluid levels, and the level of obstruction; absent rectal gas in Hirschsprung's disease; soap-bubble sign in meconium ileus; double bubble in duodenal atresia), contrast enema (water-soluble contrast: identifies the level of obstruction, the transition zone in Hirschsprung's disease, the microcolon in meconium ileus), upper GI contrast study (for malrotation: the investigation of choice), and rectal suction biopsy (for Hirschsprung's disease). Serum electrolytes, blood group, and cross-match are obtained preoperatively.

Fill in the blank: Bilious vomiting in a neonate indicates obstruction _____ to the ampulla of Vater and is always a _____ concern. Meconium is normally passed within _____ hours; failure to do so suggests a low obstruction such as Hirschsprung's disease.

5.1.3 Management principles

General management principles for all neonatal intestinal obstruction: insert a nasogastric tube (decompress the stomach and prevent aspiration), establish IV access and commence fluid resuscitation (correct dehydration and electrolyte imbalances), keep the baby warm (neonates are vulnerable to hypothermia), and perform investigations to confirm the level and cause of obstruction before proceeding to surgery. Nil by mouth is mandatory once obstruction is suspected.

Specific management: simple meconium ileus is treated by Gastrografin (water-soluble contrast: the hyperosmolar solution draws fluid into the bowel lumen and softens the inspissated meconium, flushing it out) enema under fluoroscopic guidance; success rate approximately 50 percent; surgery (enterotomy and manual disimpaction or bowel resection) is required for failure of conservative management or complicated meconium ileus. All other neonatal mechanical obstructions ultimately require surgery to relieve the obstruction. Sweat chloride test is performed in all neonates with meconium ileus to confirm cystic fibrosis.

Fill in the blank: In neonatal intestinal obstruction, the first two steps are to insert a _____ tube and establish IV access. Simple meconium ileus is treated first with a _____ enema; the _____ chloride test confirms cystic fibrosis.

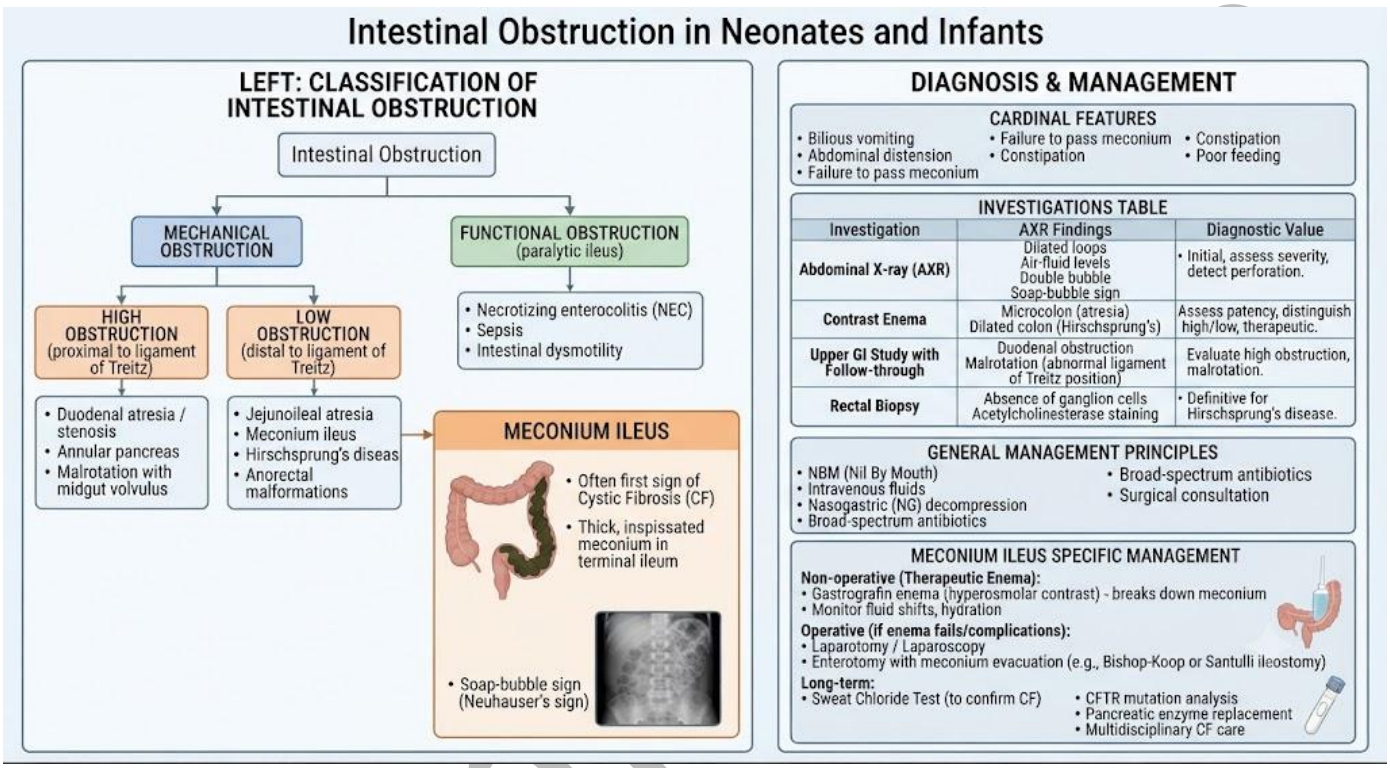

Figure 5.1: Classification, clinical features, investigation, and management of intestinal obstruction in neonates

Pilot questions 5.1

1. Classify intestinal obstruction in neonates by level and give two causes at each level. (Linked to SLO 5.1)
2. State the cardinal clinical features of neonatal intestinal obstruction. (Linked to SLO 5.1)
3. Describe the AXR findings in meconium ileus and duodenal atresia. (Linked to SLO 5.1)
4. Describe the management of simple meconium ileus. (Linked to SLO 5.1)
5. State the general management principles for neonatal intestinal obstruction. (Linked to SLO 5.1)

5.2 Intussusception

5.2.1 Pathophysiology and aetiology

Intussusception is the telescoping of a proximal segment of bowel (the intussusceptum) into the lumen of the immediately distal segment (the intussuscipiens), causing intestinal obstruction and, if unrelieved, bowel ischaemia and infarction. It is the most common cause of intestinal obstruction in infants between 3 months and 2 years of age, with a peak incidence at 5 to 10 months. The ileocaecal junction is the most common site (ileocolic intussusception: approximately 90 percent of cases).

In infants under 2 years, the majority (approximately 95 percent) are idiopathic (no anatomical lead point is found): the cause is thought to be hypertrophy of the Peyer's patches in the terminal ileum following viral infection (adenovirus is most frequently implicated), which acts as a functional lead point. In children over 2 years, a pathological lead point is found in approximately 25 percent: Meckel's diverticulum (most common), intestinal polyp, lymphoma, or a duplication cyst. Henoch-Schonlein purpura (HSP) is associated with small bowel intussusception.

Fill in the blank: Intussusception is the most common cause of intestinal obstruction in infants between _____ months and 2 years. The most common site is the _____ junction (ileocolic). In infants under 2 years, approximately _____ percent are idiopathic with no anatomical lead point.

5.2.2 Clinical features and diagnosis

The classic clinical triad is: intermittent colicky abdominal pain (the infant draws up the legs and screams with each episode: the pain is caused by peristaltic contractions pushing the intussusceptum further into the intussuscipiens; in between episodes the child may appear entirely normal or lethargic), vomiting (initially non-bilious, becoming bilious as the obstruction progresses), and red currant jelly stools (blood-stained mucus per rectum: a late and sinister sign indicating mucosal ischaemia; present in only approximately 50 percent of cases and should not be waited for before investigating).

Physical signs: a sausage-shaped mass in the right upper quadrant or epigastrium (felt in approximately 60 to 70 percent of cases on careful palpation), emptiness of the right iliac fossa (Dance's sign: the caecum has been telescoped away from the right iliac fossa), and pallor. Investigations: ultrasound abdomen (the investigation of choice: the target sign or doughnut sign: a central echogenic area of intussuscepted bowel surrounded by a hypoechoic rim of oedematous outer bowel: sensitivity above 95 percent), and plain AXR (may show a soft tissue mass in the right side of the abdomen with absent right colonic gas: Crescent sign).

Fill in the blank: The classic triad of intussusception is intermittent colicky pain, _____, and red currant jelly stools. The _____ sign (or doughnut sign) on ultrasound shows a central echogenic area surrounded by a hypoechoic rim: sensitivity above 95 percent. Dance's sign is _____ of the right iliac fossa on palpation.

5.2.3 Management

Resuscitation: IV fluid resuscitation, nasogastric tube, IV antibiotics (if peritonitis or perforation is suspected). Non-operative reduction: air enema reduction (the preferred first-line treatment: air is insufflated into the rectum under fluoroscopic or ultrasound guidance; the column of air pushes the intussusceptum back; success rate approximately 80 to 90 percent in uncomplicated cases) or hydrostatic reduction with water-soluble contrast or saline enema under ultrasound guidance. Contraindications to non-operative reduction: perforation (free air on AXR), peritonitis, and haemodynamic instability.

Operative reduction: required for failed or contraindicated enema reduction. At laparotomy (or laparoscopy), the intussusception is reduced by gentle milking of the intussusceptum backwards out of the intussuscipiens (not by pulling on the intussusceptum, which risks avulsion). If the bowel is non-viable after reduction, bowel resection and anastomosis are performed. If reduction is impossible (fixed intussusception), right hemicolectomy and ileocolic anastomosis are performed. Recurrence occurs in approximately 10 percent of cases after enema reduction; repeat enema reduction is appropriate for the first recurrence.

Fill in the blank: Air enema reduction of intussusception has a success rate of approximately _____ to 90 percent. Operative reduction involves gentle _____ of the intussusceptum backwards (not pulling). Intussusception recurs in approximately _____ percent of cases after enema reduction.

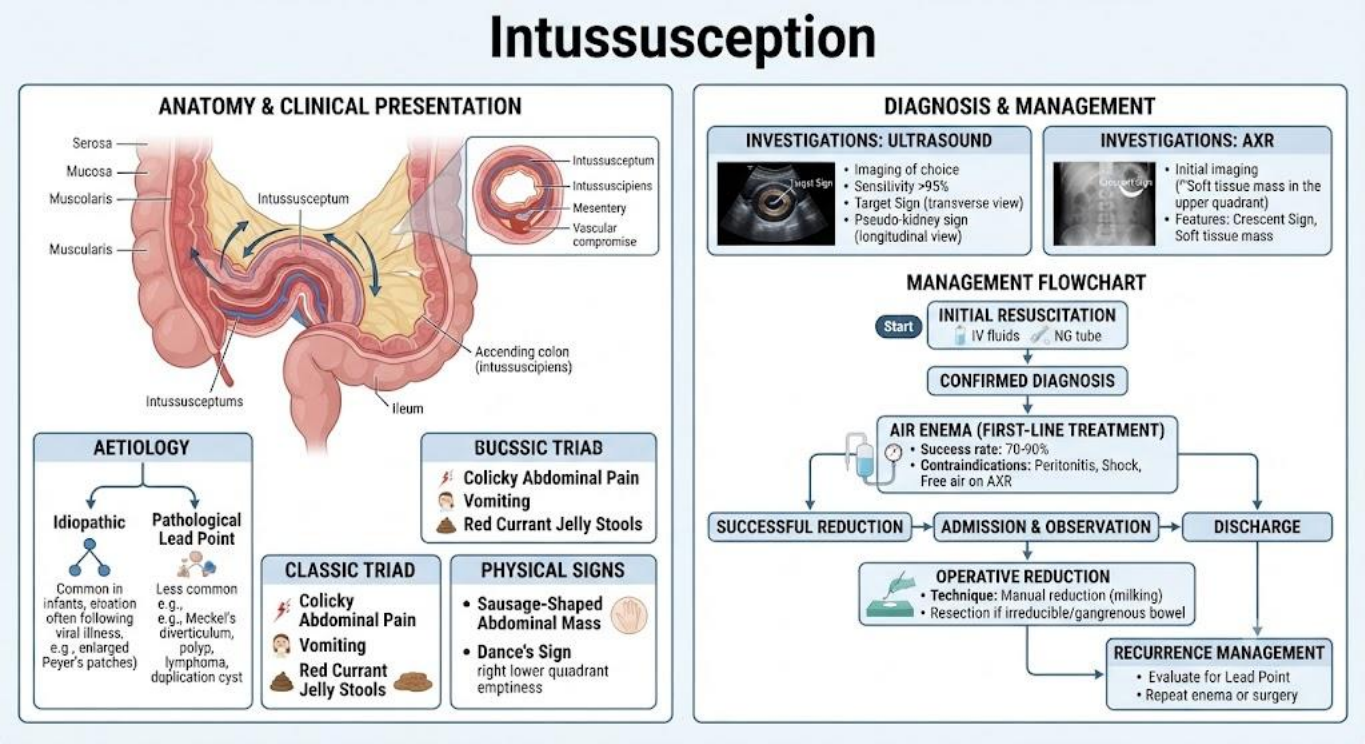

Figure 5.2: Pathophysiology, clinical features, investigation, and management of intussusception

Pilot questions 5.2

1. Describe the pathophysiology of intussusception. (Linked to SLO 5.2)
2. Describe the classic clinical triad of intussusception and state why red currant jelly stools are a late sign. (Linked to SLO 5.2)
3. Describe the target sign on ultrasound and state its sensitivity for intussusception. (Linked to SLO 5.2)
4. Describe the non-operative reduction of intussusception and state the contraindications. (Linked to SLO 5.2)
5. Describe the operative management of intussusception. (Linked to SLO 5.2)

5.3 Meckel's Diverticulum and Intestinal Duplications

5.3.1 Meckel's diverticulum

Meckel's diverticulum (MD) is the most common congenital anomaly of the gastrointestinal tract, occurring in approximately 2 percent of the population. It is a true diverticulum (containing all layers of the bowel wall), arising from the antimesenteric border of the terminal ileum, representing the remnant of the vitello-intestinal (omphalomesenteric) duct. The rule of twos: 2 percent incidence, 2 feet (approximately 60 cm) from the ileocaecal valve, 2 inches (approximately 5 cm) in length, 2 times more common in males, and 2 types of ectopic mucosa (gastric: most common and most clinically significant; pancreatic mucosa).

Ectopic gastric mucosa (present in approximately 50 percent of symptomatic MDs) secretes acid, causing peptic ulceration of the adjacent normal ileal mucosa and painless rectal bleeding (the most common presentation in children: bright red or dark red blood per rectum without pain). Other presentations: intestinal obstruction (from intussusception with the diverticulum as the lead point, volvulus around a fibrous band connecting the diverticulum to the umbilicus, or internal herniation), Meckel's diverticulitis (identical to appendicitis clinically: right lower abdominal pain and tenderness; can perforate), and umbilical fistula or sinus (if the vitello-intestinal duct remains patent to the umbilicus).

Fill in the blank: Meckel's diverticulum occurs in approximately _____ percent of the population and is the most common GI anomaly. The most common presentation in children is painless _____ rectal bleeding from ectopic _____ mucosa secreting acid. Meckel's diverticulum arises from the _____ border of the terminal ileum.

5.3.2 Intestinal duplications

Intestinal duplications are congenital cystic or tubular structures containing all layers of the bowel wall and sharing the blood supply with the adjacent native bowel. They can occur anywhere along the gastrointestinal tract from the mouth to the anus but are most common in the small bowel (ileum most frequently). They are classified as cystic (the most common type: a spherical or ovoid cystic structure closely adherent to the bowel: no communication with the bowel lumen) or tubular (a tube running parallel to the native bowel: may communicate with the bowel lumen at one or both ends).

Intestinal duplications may contain ectopic gastric mucosa (causing peptic ulceration and bleeding) or pancreatic mucosa. Clinical presentations: an asymptomatic abdominal mass (the most common: found incidentally on imaging), intestinal obstruction (from compression of the adjacent bowel), gastrointestinal bleeding (from ectopic gastric mucosa), volvulus (a tubular duplication may twist on its mesentery), and intussusception (a duplication may act as a lead point). Thoracic duplications (oesophageal or foregut duplications) may present with respiratory symptoms from mediastinal compression.

Fill in the blank: Intestinal duplications are most common in the _____ bowel (ileum most frequently). Cystic duplications are _____ with the bowel lumen, while tubular duplications may _____ with the bowel at one or both ends.

5.3.3 Management of Meckel's diverticulum and duplications

Symptomatic Meckel's diverticulum: surgical excision is mandatory for all symptomatic MDs. Options: diverticulectomy (wedge resection of the diverticulum alone: appropriate when the base is narrow and uninflamed) or segmental ileal resection and anastomosis (when the base is wide, when there is ectopic mucosa extending to the base, or when the adjacent ileum is inflamed or ischaemic). Incidental Meckel's diverticulum (found at laparotomy for another condition): excision is recommended if the diverticulum is short, has a narrow neck, or contains palpable ectopic mucosa; routine excision of all incidentally found MDs is debated.

Management of intestinal duplications: surgical excision is the treatment for all symptomatic duplications. Cystic duplications: complete excision of the cyst with its shared wall (the wall shared with the native bowel must be stripped of its mucosa rather than removed en bloc if excision would compromise the bowel blood supply); simple enucleation may damage the native bowel. Tubular duplications: segmental resection of the duplication and adjacent native bowel with end-to-end anastomosis. The Meckel's scan (technetium-99m pertechnetate scintigraphy: the isotope is taken up by ectopic gastric mucosa: sensitivity approximately 85 percent in children) is the best non-invasive investigation for identifying a bleeding Meckel's diverticulum.

Fill in the blank: For a symptomatic Meckel's diverticulum with a wide base or ectopic mucosa at the base, _____ ileal resection and anastomosis is preferred over diverticulectomy. The Meckel's scan uses _____ scintigraphy to detect ectopic _____ mucosa: sensitivity approximately 85 percent in children.

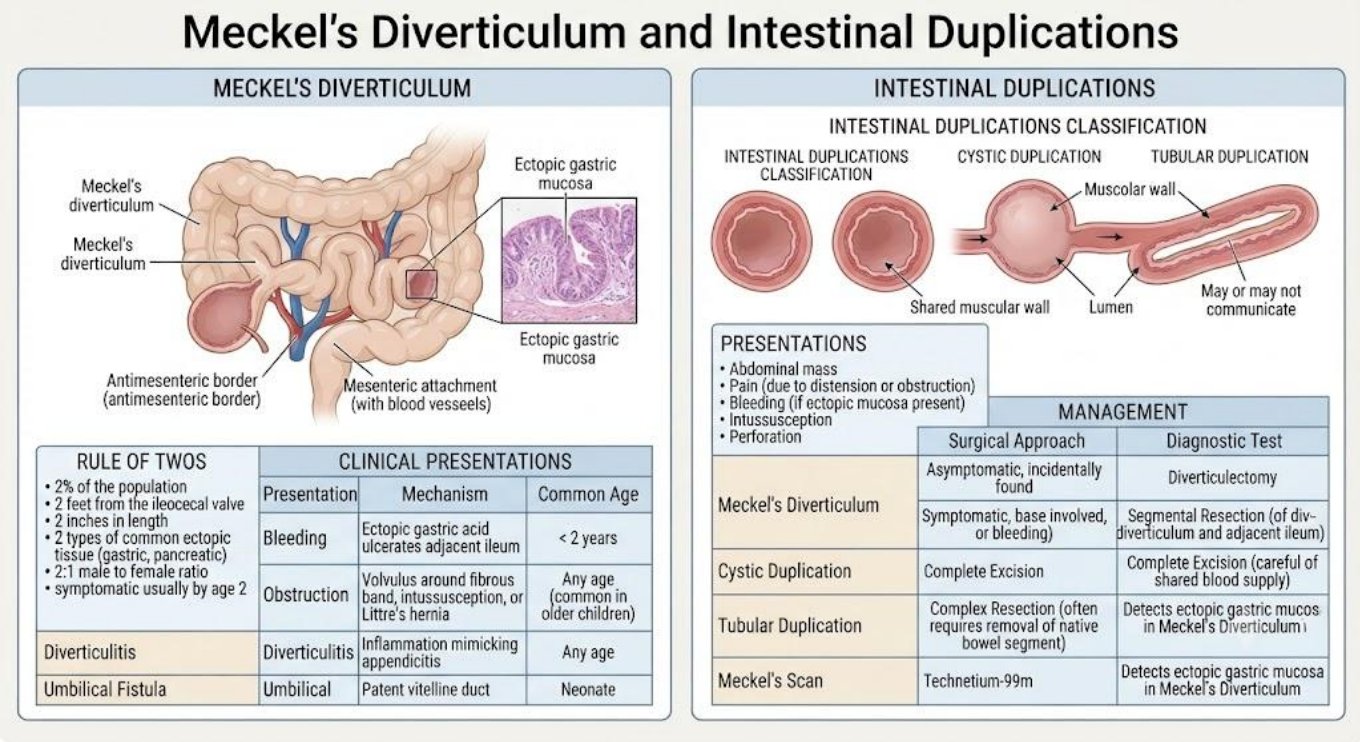

Figure 5.3: Anatomy, clinical presentations, and management of Meckel's diverticulum and intestinal duplications

Pilot questions 5.3

1. State the rule of twos for Meckel's diverticulum. (Linked to SLO 5.3)
2. Describe the most common clinical presentation of Meckel's diverticulum in children and explain the mechanism. (Linked to SLO 5.3)
3. Describe the Meckel's scan and state its diagnostic value. (Linked to SLO 5.3)
4. Distinguish cystic from tubular intestinal duplications. (Linked to SLO 5.3)
5. Describe the surgical management of a symptomatic Meckel's diverticulum. (Linked to SLO 5.3)

5.4 Necrotising Enterocolitis

5.4.1 Pathophysiology and risk factors

Necrotising enterocolitis (NEC) is an acute inflammatory necrosis of the intestinal mucosa and wall, primarily affecting premature neonates, and is the most common surgical emergency in neonatal intensive care units worldwide. The pathophysiology involves a triad of intestinal

immaturity (immature barrier function, immature immune response, and immature motility), intestinal ischaemia (from haemodynamic instability, hypoxia, or umbilical artery catheterisation), and microbial colonisation with pathogenic organisms (abnormal gut colonisation by pathogenic bacteria, enhanced by formula feeding and antibiotic use that disrupt the normal microbiome).

Risk factors: prematurity (the most important risk factor: approximately 90 percent of NEC occurs in premature infants, and the risk increases with decreasing gestational age), formula feeding (NEC is significantly less common in breast-fed infants: human breast milk is protective), hypoxia and haemodynamic instability (from respiratory distress syndrome, patent ductus arteriosus, or asphyxia), polycythaemia, and congenital heart disease (particularly left-to-right shunts that reduce gut perfusion). NEC in term neonates is less common but associated with congenital heart disease, polycythaemia, or Hirschsprung's disease.

Fill in the blank: NEC primarily affects _____ neonates and is the most common surgical emergency in NICUs. The most important risk factor is _____ (approximately 90 percent of cases occur in premature infants). Human breast milk is _____ against NEC compared with formula feeding.

5.4.2 Clinical features and staging

NEC presents in premature neonates, typically at 2 to 3 weeks of age (earlier in more premature infants), with a triad of systemic signs (temperature instability, apnoea, bradycardia, and lethargy from septicaemia), abdominal signs (abdominal distension, tenderness, erythema of the abdominal wall, and blood in stools), and radiological findings (pneumatosis intestinalis: gas in the bowel wall: the pathognomonic radiological finding; portal venous gas: a sinister sign indicating extensive disease; and pneumoperitoneum: free air indicating perforation).

The Bell staging criteria classify NEC by severity: Stage I (suspected NEC: non-specific systemic and abdominal signs without radiological confirmation: treated medically), Stage II (definite NEC: definite abdominal signs plus radiological confirmation of pneumatosis intestinalis or portal venous gas: treated medically), and Stage III (advanced NEC: signs of intestinal perforation or gangrene: pneumoperitoneum or clinical deterioration despite medical management: requires surgery). Stage IIIB has bowel necrosis without perforation (confirmed at surgery) and Stage IIIB has perforation.

Fill in the blank: The pathognomonic radiological finding of NEC is _____ intestinalis (gas in the bowel wall) on AXR. _____ venous gas is a sinister sign indicating extensive disease. Stage _____ NEC (advanced) requires surgery.

5.4.3 Management and outcomes

Medical management (Stages I and II): nil by mouth (bowel rest for 7 to 14 days), nasogastric tube decompression, IV fluids and total parenteral nutrition (TPN), broad-spectrum IV antibiotics covering Gram-negative organisms and anaerobes (gentamicin plus metronidazole, or piperacillin-tazobactam), correction of coagulopathy (fresh frozen plasma), cardiovascular

support, and close monitoring. Serial clinical and radiological assessments (AXR every 6 to 8 hours in the acute phase) are essential.

Surgical management (Stage III): indicated for pneumoperitoneum (intestinal perforation), clinical deterioration despite medical management, a fixed abdominal mass (representing necrotic loops unable to be reduced), or portal venous gas with clinical instability. The surgical options are: peritoneal drainage (a simple drain placed under local anaesthesia in the right iliac fossa: used for the very smallest and most premature infants as a temporising measure), or laparotomy and resection of the necrotic bowel (with stoma formation: the proximal end is brought out as a stoma and the distal end as a mucous fistula; primary anastomosis is rarely performed in the acute setting because of the risk of anastomotic breakdown in inflamed bowel). Stoma closure is performed when the infant has grown and recovered (usually 6 to 8 weeks later). Mortality of NEC is approximately 20 to 30 percent overall and above 50 percent when surgery is required. Long-term complications include short bowel syndrome (from extensive bowel resection) and stricture formation.

Fill in the blank: Medical management of NEC includes bowel rest (nil by mouth for _____ to 14 days), nasogastric decompression, TPN, and broad-spectrum _____. Surgical management options include peritoneal _____ (for the smallest infants) or laparotomy with resection and _____ formation.

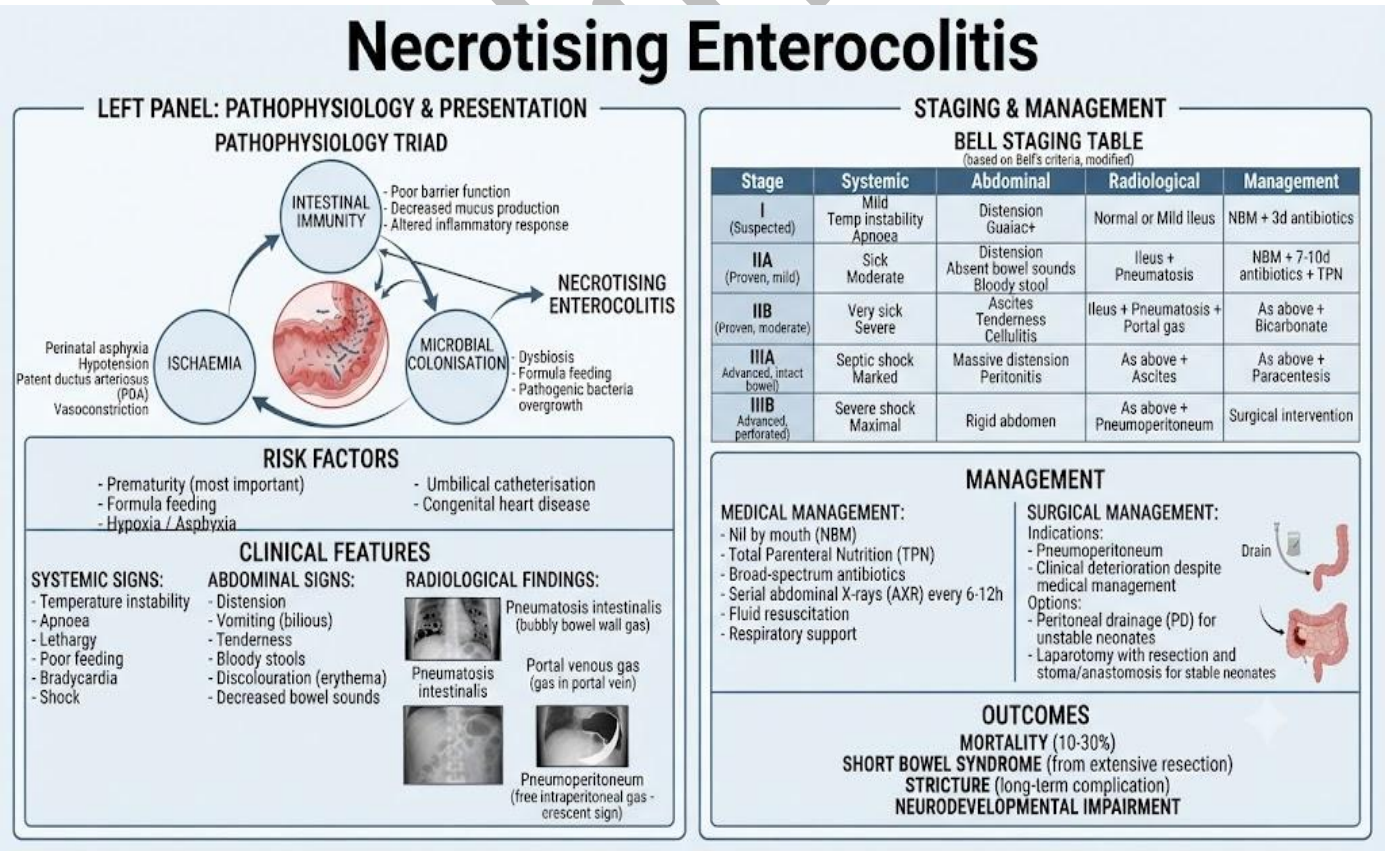

Figure 5.4: Pathophysiology, staging, and management of necrotising enterocolitis

Pilot questions 5.4

1. Describe the pathophysiology of necrotising enterocolitis. (Linked to SLO 5.4)
2. State the pathognomonic radiological finding of NEC and describe two other radiological signs. (Linked to SLO 5.4)
3. Describe the Bell staging criteria for NEC. (Linked to SLO 5.4)
4. Describe the medical management of Stage II NEC. (Linked to SLO 5.4)
5. State the surgical options for Stage III NEC and describe when each is used. (Linked to SLO 5.4)

Series Summary

This Series completed the core content of Paediatric Surgery. We examined intestinal obstruction in neonates (high versus low classification; meconium ileus as the earliest manifestation of cystic fibrosis; Gastrografin enema and sweat chloride test), intussusception (the most common intestinal obstruction in infants; ileocolic; target sign on ultrasound; air enema reduction in 80 to 90 percent; surgical milking for failures), Meckel's diverticulum (rule of twos; painless rectal bleeding from ectopic gastric mucosa; Meckel's scan; diverticulectomy or segmental resection) and intestinal duplications (cystic versus tubular; excision), and necrotising enterocolitis (premature infants; pathophysiology triad; pneumatosis intestinalis on AXR; Bell staging; medical management for Stages I and II; peritoneal drainage or laparotomy with stoma for Stage III).

You should now be able to classify and manage intestinal obstruction in neonates (SLO 5.1), describe and manage intussusception (SLO 5.2), describe and manage Meckel's diverticulum and intestinal duplications (SLO 5.3), and describe the staging and management of necrotising enterocolitis (SLO 5.4). With this Series, the full five-Series curriculum of SUG 403 Paediatric Surgery is complete.

Glossary of Terms

- **Air enema reduction:** the preferred non-operative treatment for intussusception: air is insufflated into the rectum under fluoroscopic or ultrasound guidance, pushing the intussusceptum retrograde; success rate approximately 80 to 90 percent in uncomplicated cases.

- **Bell staging:** a three-stage clinical and radiological classification of necrotising enterocolitis: Stage I (suspected), Stage II (confirmed), and Stage III (advanced with perforation or gangrene); guides the decision between medical and surgical management.
- **Dance's sign:** emptiness of the right iliac fossa on palpation in a patient with intussusception; the caecum and terminal ileum have been telescoped away from their normal position.
- **Gastrografin enema:** a therapeutic enema using hyperosmolar water-soluble contrast (diatrizoate meglumine: Gastrografin) to treat simple meconium ileus; the hyperosmolar solution draws fluid into the bowel lumen, softening and flushing out the inspissated meconium.
- **Intussusception:** the telescoping of a proximal segment of bowel (the intussusceptum) into the lumen of the distal segment (the intussuscipiens); the most common cause of intestinal obstruction in infants aged 3 months to 2 years.
- **Meckel's diverticulum:** the most common congenital anomaly of the GI tract; a true diverticulum on the antimesenteric border of the terminal ileum; remnant of the vitello-intestinal duct; follows the rule of twos.
- **Meconium ileus:** obstruction of the terminal ileum by inspissated meconium; the earliest manifestation of cystic fibrosis; presents with abdominal distension and failure to pass meconium; the soap-bubble sign on AXR is characteristic.
- **Necrotising enterocolitis (NEC):** acute inflammatory necrosis of the intestinal wall primarily affecting premature neonates; pathognomonic radiological finding is pneumatosis intestinalis (gas in the bowel wall); managed medically for Stages I and II and surgically for Stage III.
- **Pneumatosis intestinalis:** gas within the bowel wall, visible on plain AXR as a linear or bubbly lucency outlining the bowel wall; the pathognomonic radiological finding of necrotising enterocolitis.
- **Red currant jelly stools:** blood-stained mucus per rectum, appearing as red currant jelly; a late and sinister sign in intussusception indicating mucosal ischaemia; present in only approximately 50 percent of cases.
- **Target sign (doughnut sign):** the characteristic ultrasound appearance of intussusception: a central echogenic area of intussuscepted bowel surrounded by a hypoechoic rim of oedematous outer bowel; sensitivity above 95 percent for the diagnosis.
- **Vitello-intestinal duct:** the embryonic communication between the midgut and the yolk sac; normally obliterates by the 7th week of gestation; incomplete obliteration produces

Meckel's diverticulum (diverticulum), umbilical fistula (fully patent duct), or umbilical sinus (partially patent).

Concept Check Questions [Series 5]

Objective questions

1. Meconium ileus is the earliest manifestation of _____ fibrosis and shows a soap-bubble sign on AXR. (Linked to SLO 5.1)
2. Intussusception is the most common cause of intestinal obstruction in infants between 3 months and _____ years. (Linked to SLO 5.2)
3. The rule of twos for Meckel's diverticulum includes a 2 percent incidence, 2 feet from the ileocaecal valve, 2 inches in length, 2 times more common in _____, and 2 types of ectopic mucosa. (Linked to SLO 5.3)
4. The pathognomonic radiological finding of NEC is _____ intestinalis (gas in the bowel wall) on AXR. (Linked to SLO 5.4)
5. Stage _____ NEC (advanced: with pneumoperitoneum or bowel gangrene) requires surgical management. (Linked to SLO 5.4)

Short-answer questions

6. State the cardinal clinical features of neonatal intestinal obstruction. (Linked to SLO 5.1)
7. Describe the classic triad of intussusception and state why red currant jelly stools are a late sign. (Linked to SLO 5.2)
8. Describe the most common clinical presentation of Meckel's diverticulum in children. (Linked to SLO 5.3)
9. Describe the Bell staging criteria for NEC. (Linked to SLO 5.4)
10. State the surgical options for Stage III NEC and describe when each is used. (Linked to SLO 5.4)

Long-answer questions

11. Describe the classification of neonatal intestinal obstruction and the investigation of a neonate with bilious vomiting. (Linked to SLO 5.1)
12. Describe the pathophysiology, clinical features, and management of intussusception. (Linked to SLO 5.2)
13. Describe the anatomy, clinical presentations, and surgical management of Meckel's diverticulum. (Linked to SLO 5.3)
14. Describe the pathophysiology, staging, and management of necrotising enterocolitis. (Linked to SLO 5.4)
15. Compare intussusception and meconium ileus as causes of intestinal obstruction in infancy. (Linked to SLO 5.1 and 5.2)

Answers to Concept Check Questions [Series 5]

Objective questions

1. Cystic.
2. 2.
3. Males.
4. Pneumatosis.
5. III.

Short-answer questions

6. Bilious vomiting (obstruction distal to the ampulla of Vater: always a surgical concern in a neonate), abdominal distension (progressive: minimal in high obstruction and maximal in low obstruction), failure to pass meconium within 48 hours (suggests low obstruction: Hirschsprung's disease or anorectal malformation), and dehydration from fluid losses in vomit.
7. Classic triad: intermittent colicky abdominal pain (infant draws up legs and screams; appears normal or lethargic between episodes), vomiting (initially non-bilious then bilious as obstruction progresses), and red currant jelly stools (blood-stained mucus per rectum: late sign present in only 50 percent). Red currant jelly stools are a late sign

because they result from mucosal ischaemia from vascular compromise of the intussuscepted segment, which takes time to develop after the initial obstruction.

8. Painless bright red or dark red rectal bleeding in a young child, caused by ectopic gastric mucosa within the diverticulum secreting acid, which causes peptic ulceration of the adjacent normal ileal mucosa. The bleeding is painless because the ulceration is in the ileum (not in the stomach where pain receptors are sensitive to acid). It is often intermittent and may be massive.

9. Stage I (suspected NEC): non-specific systemic signs (temperature instability, apnoea, bradycardia, lethargy) and mild abdominal signs without radiological confirmation: treated medically. Stage II (definite NEC): definite abdominal signs plus radiological confirmation of pneumatosis intestinalis or portal venous gas: treated medically. Stage III (advanced NEC): Stage IIIA has signs of bowel gangrene without perforation (clinical deterioration despite medical treatment or bowel necrosis confirmed at surgery); Stage IIIB has intestinal perforation (pneumoperitoneum on AXR): both require surgery.

10. Peritoneal drainage: a simple drain placed under local anaesthesia in the right iliac fossa; used for the very smallest and most premature infants (below 1000 to 1500 g) who are too unstable for general anaesthesia and laparotomy; acts as a temporising measure. Laparotomy and bowel resection: used in larger and more stable infants; the necrotic bowel is resected; the proximal end is brought out as a stoma and the distal end as a mucous fistula; primary anastomosis is avoided in the acute setting because of the high risk of anastomotic breakdown in inflamed bowel.

Long-answer questions

11. **Basic response:** Classification: mechanical (physical block: subdivided by level: high above mid-jejunum: oesophageal atresia, duodenal atresia, annular pancreas, jejunal atresia: early profuse vomiting with minimal distension; low below mid-jejunum: ileal atresia, meconium ileus, Hirschsprung's disease, anorectal malformations: delayed faeculent vomiting with marked distension and failure to pass meconium) versus functional (paralytic ileus: sepsis, electrolyte imbalance). Investigation of bilious vomiting in a neonate: immediately insert a nasogastric tube and establish IV access. Plain AXR (supine and erect or left lateral decubitus): identifies dilated loops, air-fluid levels, level of obstruction (absent rectal gas: Hirschsprung's; soap-bubble sign: meconium ileus; double bubble: duodenal atresia). Upper GI contrast study: the investigation of choice for suspected malrotation (bilious vomiting plus gasless abdomen: surgical emergency). Contrast enema: identifies transition zone in Hirschsprung's,

microcolon in meconium ileus. Rectal suction biopsy: for Hirschsprung's disease. Serum electrolytes and blood group and cross-match.

Value-added response: In a Nigerian neonatal unit, the key practical principle is: any neonate with bilious vomiting is a surgical emergency until proven otherwise. The upper GI contrast study for malrotation must never be delayed, as midgut volvulus can progress from viable to infarcted bowel within 2 to 4 hours of onset.

12. **Basic response:** Pathophysiology: intussusception is the telescoping of a proximal bowel segment (intussusceptum: most commonly the terminal ileum) into the lumen of the distal segment (intussuscipiens: most commonly the caecum: ileocolic). The mesentery and blood vessels of the intussusceptum are compressed between the bowel walls, causing first venous congestion then arterial ischaemia and mucosal necrosis. Clinical features: classic triad of intermittent colicky pain (legs drawn up: normal between episodes), vomiting (non-bilious then bilious), and red currant jelly stools (late: mucosal ischaemia: 50 percent). Signs: sausage-shaped mass in right upper quadrant (60 to 70 percent), Dance's sign (empty right iliac fossa), pallor. Management: IV resuscitation and NG tube and antibiotics if peritonitis. First-line: air enema reduction under fluoroscopic guidance (80 to 90 percent success rate). Contraindications: pneumoperitoneum, peritonitis, haemodynamic instability. Operative: laparotomy or laparoscopy; gentle milking of intussusceptum backwards; non-viable bowel: resect and anastomose; fixed intussusception: right hemicolectomy. Recurrence: approximately 10 percent: repeat enema for first recurrence.

Value-added response: In Nigerian practice, the availability of fluoroscopic or ultrasound-guided air enema is limited to a few tertiary centres. Many children with intussusception therefore reach a surgeon only after failed or delayed diagnosis, often with an established ischaemic intussusception that requires operative reduction or resection. Improving awareness of the target sign on ultrasound among emergency physicians and general paediatricians would facilitate earlier diagnosis and higher enema reduction rates.

13. **Basic response:** Anatomy: Meckel's diverticulum is a true diverticulum (all bowel wall layers) on the antimesenteric border of the terminal ileum, approximately 60 cm from the ileocaecal valve, approximately 5 cm in length; remnant of the vitello-intestinal duct; 2 percent incidence; twice as common in males; ectopic gastric mucosa in approximately 50 percent of symptomatic cases; ectopic pancreatic mucosa less common. Clinical presentations: (1) painless rectal bleeding (most common in children: ectopic gastric mucosa secretes acid, causing peptic ulceration of adjacent ileum: bright or dark red blood per rectum without abdominal pain), (2) intestinal obstruction (intussusception with MD as lead point; volvulus around a fibrous band to the umbilicus; internal herniation around the diverticulum), (3) Meckel's diverticulitis (right lower quadrant pain identical to appendicitis: can perforate), (4) umbilical fistula or sinus (patent or partially

patent vitello-intestinal duct). Management: symptomatic MD: diverticulectomy (wedge resection) if the base is narrow and uninflamed; segmental ileal resection and anastomosis if the base is wide, ectopic mucosa extends to the base, or the adjacent ileum is inflamed. Meckel's scan (technetium-99m pertechnetate: taken up by ectopic gastric mucosa: sensitivity approximately 85 percent in children) is the best non-invasive investigation for a bleeding MD.

Value-added response: The diagnosis of Meckel's diverticulum is frequently missed or delayed because its presentations mimic other common conditions (appendicitis, lower GI bleeding, intussusception). In a child with unexplained lower GI bleeding or recurrent right iliac fossa pain who has had a normal appendix at appendicectomy, Meckel's diverticulum must be specifically excluded by Meckel's scan or diagnostic laparoscopy.

14. **Basic response:** Pathophysiology: the triad of intestinal immaturity (immature barrier function, immune response, and motility: allows bacterial translocation), intestinal ischaemia (from haemodynamic instability, hypoxia, or umbilical catheterisation: reduces mucosal perfusion), and pathological microbial colonisation (abnormal gut flora: enhanced by formula feeding and broad-spectrum antibiotic exposure: invades the ischaemic, immature mucosa and triggers an exaggerated inflammatory response). Risk factors: prematurity (most important: 90 percent of NEC), formula feeding (breast milk protective), hypoxia, polycythaemia, and congenital heart disease. Bell staging: Stage I (suspected: non-specific signs: medical management), Stage II (confirmed: pneumatosis intestinalis or portal venous gas on AXR: medical management), Stage III (advanced: pneumoperitoneum or deterioration despite medical treatment: surgical management). Medical management (Stages I and II): nil by mouth for 7 to 14 days, NG decompression, IV fluids and TPN, broad-spectrum IV antibiotics, FFP for coagulopathy, cardiovascular support, serial AXR every 6 to 8 hours. Surgical management (Stage III): peritoneal drainage (for smallest infants below 1000 to 1500 g as a temporising measure under local anaesthesia) or laparotomy with resection of necrotic bowel and proximal stoma formation. Mortality: 20 to 30 percent overall; above 50 percent when surgery required. Long-term: short bowel syndrome and intestinal stricture.

Value-added response: Prevention of NEC through exclusive human breast milk feeding is the single most effective intervention available: breast milk contains immunoglobulin A, lactoferrin, lysozyme, oligosaccharides, and growth factors that reduce NEC incidence by approximately 6 to 10 fold compared with formula feeding. In Nigerian NICUs, promoting and maintaining breast milk supply for premature infants (including donor breast milk programmes where maternal milk is unavailable) should be a priority quality improvement target.

15. **Basic response:** Intussusception: the most common intestinal obstruction in infants aged 3 months to 2 years; ileocolic in 90 percent; usually idiopathic; presents with intermittent colicky pain (infant draws up legs), vomiting, and red currant jelly stools (late in 50

percent); sausage-shaped mass in right upper quadrant; target sign on ultrasound (above 95 percent sensitive); treated by air enema reduction (80 to 90 percent success rate) or surgery. Meconium ileus: occurs in the neonatal period (first 48 hours); exclusively in neonates with cystic fibrosis (approximately 15 percent of CF); presents with abdominal distension, bilious vomiting, and failure to pass meconium; soap-bubble sign on AXR (ground-glass appearance in right iliac fossa); no abdominal mass is palpable; treated by Gastrografin enema (50 percent success) or surgery. Key differences: intussusception occurs in older infants, is idiopathic, and is not associated with systemic disease; meconium ileus occurs at birth, is always associated with cystic fibrosis, and is confirmed by sweat chloride test. Both present with intestinal obstruction but at different ages, with different imaging findings, and requiring different specific treatments.

Value-added response: In Nigerian practice, meconium ileus and the underlying diagnosis of cystic fibrosis are significantly underdiagnosed compared with high-income countries. Cystic fibrosis is often mistakenly considered a disease of Caucasian populations; however, it does occur in Africans and Nigerian children. Any neonate presenting with features of meconium ileus should have a sweat chloride test performed and be referred for CF genetic counselling.

Answers to Pilot Questions [Series 5]

Part 5.1

6. High obstruction (above mid-jejunum): early and profuse vomiting with minimal distension: causes include oesophageal atresia (Type C: OA with distal TOF) and duodenal atresia (double bubble sign and Down syndrome association). Low obstruction (below mid-jejunum): delayed faeculent vomiting with marked distension and failure to pass meconium: causes include meconium ileus (cystic fibrosis; soap-bubble sign) and Hirschsprung's disease (failure to pass meconium within 48 hours; squirt sign on rectal examination).
7. Bilious vomiting (bile-stained green vomit indicating obstruction distal to the ampulla of Vater: always a surgical concern in a neonate), abdominal distension (progressive and correlating with level of obstruction: minimal in high obstruction and maximal in low obstruction), failure to pass meconium within 48 hours (suggests low obstruction: Hirschsprung's disease or anorectal malformation), and dehydration from ongoing fluid losses in vomit.
8. Meconium ileus AXR: dilated loops of small bowel with a ground-glass or soap-bubble appearance (bubbly lucency) in the right iliac fossa, caused by gas mixing with the inspissated sticky meconium; an absence of air-fluid levels (the sticky meconium prevents layering). No air in the rectum. Duodenal atresia AXR: the double bubble sign:

two air-filled bubbles (the stomach and the obstructed proximal duodenum) with complete absence of gas in the distal bowel; the double bubble results from air swallowed by the neonate filling the stomach and the dilated proximal duodenum proximal to the atresia.

9. Simple meconium ileus is managed first with a Gastrografin (hyperosmolar water-soluble contrast) enema administered under fluoroscopic guidance. The hyperosmolar solution draws fluid from the bowel wall into the lumen, softening the inspissated meconium, which is then flushed out distally. IV fluid resuscitation is given simultaneously to compensate for the fluid shifts caused by the hyperosmolar contrast. The success rate is approximately 50 percent. If the Gastrografin enema fails or if the meconium ileus is complicated (volvulus, atresia, or perforation), surgery is required (enterotomy and manual disimpaction or bowel resection). A sweat chloride test must be performed in all neonates with meconium ileus to confirm cystic fibrosis.

10. Insert a nasogastric tube (to decompress the stomach, prevent aspiration, and allow measurement of aspirate losses). Establish IV access and commence IV fluid resuscitation (to correct dehydration and electrolyte imbalances from vomiting). Keep the baby warm (neonates are vulnerable to hypothermia from a large surface area to body weight ratio). Make the baby nil by mouth immediately. Perform investigations to confirm the level and cause of obstruction (plain AXR, contrast studies, rectal biopsy as indicated) before proceeding to surgery.

Part 5.2

11. Intussusception begins when a proximal segment of bowel (the intussusceptum, most commonly the terminal ileum) invaginates into the lumen of the immediately adjacent distal segment (the intussuscipiens, most commonly the caecum and ascending colon: ileocolic pattern). The invaginated segment carries its mesentery with it; the mesenteric vessels and lymphatics are compressed between the walls of the intussusceptum and intussuscipiens, initially causing venous congestion and oedema (which worsens the obstruction and prevents spontaneous reduction) and ultimately leading to arterial ischaemia, mucosal necrosis (producing the red currant jelly stools), full-thickness bowel necrosis, and perforation if untreated.

12. Classic triad: intermittent colicky abdominal pain (the infant draws up the legs and screams during each episode caused by a peristaltic wave pushing the intussusceptum further in; between episodes the child may appear entirely normal or pale and lethargic), vomiting (initially non-bilious, becoming bilious as obstruction progresses), and red currant jelly stools (blood-stained mucus per rectum: a late sign indicating mucosal

ischaemia and necrosis: present in only approximately 50 percent of cases). Red currant jelly stools are a late sign because they appear only after venous congestion has progressed to mucosal ischaemia: at the early stage, the mucosa is congested but still viable and no blood is shed.

13. The target sign (also called the doughnut sign or pseudo-kidney sign) is the characteristic ultrasound appearance of intussusception: in cross-section, the intussuscepted bowel appears as a central echogenic area (the intussusceptum: the bowel wall and mucosa) surrounded by a concentric hypoechoic rim (the oedematous outer layer of the intussuscipiens). In long section it resembles a pseudo-kidney. The sensitivity of ultrasound for diagnosing intussusception using the target sign is above 95 percent; ultrasound is the investigation of choice and can also guide hydrostatic reduction.

14. Non-operative reduction by air enema: the child is sedated and the air enema catheter is placed per rectum. Air is insufflated under pressure (maximum 80 to 120 mmHg) under fluoroscopic or ultrasound guidance. The column of air pushes the intussusceptum retrograde until it is fully reduced back into the terminal ileum (confirmed by free flow of air into the small bowel). Success rate: approximately 80 to 90 percent in uncomplicated cases. Contraindications: free air on AXR (pneumoperitoneum indicating perforation), peritonitis (tenderness, erythema, haemodynamic instability suggesting advanced disease), and haemodynamic instability not responding to resuscitation.

15. The child is prepared for general anaesthesia (IV fluids and NG tube and IV antibiotics if peritonitis). Laparotomy or laparoscopy is performed. The intussusception is identified. The intussusceptum is reduced by applying gentle sustained milking pressure from below (pushing the intussusceptum backwards out of the intussuscipiens): the intussusceptum must never be pulled by grasping and tugging, as this risks avulsion and bowel injury. If the bowel is viable after reduction (pink and pulsatile), it is returned to the abdomen. Non-viable bowel is resected and anastomosed. If the intussusception is fixed and irreducible, right hemicolectomy with ileocolic anastomosis is performed. A lead point (Meckel's diverticulum, polyp, or lymphoma) found at laparotomy is also resected.

Part 5.3

1. 2 percent incidence in the general population (the most common congenital GI anomaly). 2 feet (approximately 60 cm) from the ileocaecal valve. 2 inches (approximately 5 cm) in length. 2 times more common in males than females. 2 types of ectopic mucosa: gastric mucosa (most common and most clinically significant: causes peptic ulceration and bleeding) and pancreatic mucosa (less common). The rule of twos is a useful mnemonic

but it is also important to note that 2 percent of those with a Meckel's diverticulum develop symptoms in their lifetime.

2. The most common presentation in children is painless rectal bleeding. Mechanism: ectopic gastric mucosa within the diverticulum secretes hydrochloric acid and pepsin onto the adjacent normal ileal mucosa, causing a chronic peptic ulcer at the junction between the ectopic gastric mucosa and the normal ileal mucosa. The ulcer erodes blood vessels and produces rectal bleeding that is typically bright red or dark red (depending on the bleeding rate and transit time). The bleeding is painless because the ileal ulceration is in a pain-insensitive location (unlike a gastric or duodenal ulcer which causes epigastric pain from acid on pain-sensitive gastric mucosa).

3. The Meckel's scan (technetium-99m pertechnetate scintigraphy) detects ectopic gastric mucosa: the isotope (pertechnetate) is selectively taken up and secreted by the parietal cells of gastric mucosa, whether located in the stomach or ectopically in a Meckel's diverticulum. After IV injection, the isotope accumulates in ectopic gastric mucosa within 20 to 30 minutes and produces a focal area of increased uptake in the right lower abdomen (corresponding to the diverticulum) simultaneously with uptake in the normal stomach. Sensitivity: approximately 85 percent in children (lower in adults due to less ectopic gastric mucosa). It is the best non-invasive investigation for identifying the source of a bleeding Meckel's diverticulum when the diagnosis is suspected but not confirmed.

4. Cystic duplications: the most common type; a spherical or ovoid cystic structure closely adherent to the mesenteric border of the native bowel; shares the bowel wall and mesenteric blood supply with the adjacent native bowel; does not communicate with the bowel lumen; may contain ectopic gastric or pancreatic mucosa; presents as an asymptomatic abdominal mass, obstruction, or GI bleeding. Tubular duplications: a tube running parallel to the native bowel along its mesenteric border; shares the bowel wall and blood supply; may communicate with the native bowel lumen at one or both ends; may extend over a long segment of the bowel; presents similarly but also with volvulus or intussusception.

5. Symptomatic Meckel's diverticulum must always be excised. Surgical options depend on the anatomy of the diverticulum: (1) diverticulectomy (wedge resection): the diverticulum is excised with a wedge of the adjacent ileal wall and the defect is closed transversely; appropriate when the base is narrow (below 1 cm in width) and the adjacent ileum is uninflamed and there is no palpable ectopic mucosa at the base; (2) segmental ileal resection and end-to-end anastomosis: the diverticulum and a segment of adjacent ileum are resected; appropriate when the base is wide, when ectopic mucosa extends to the base of the diverticulum, when the adjacent ileum is inflamed or ischaemic (in

Meckel's diverticulitis), or when the diverticulum is the lead point of an intussusception causing ischaemia of the adjacent bowel.

Part 5.4

1. The pathophysiology of NEC involves three interacting factors. Intestinal immaturity: premature neonates have an immature intestinal epithelial barrier (increased permeability), an immature mucosal immune system (reduced immunoglobulin A and pattern recognition), and immature motility (reduced clearance of bacteria from the lumen). Intestinal ischaemia: haemodynamic instability (from respiratory distress syndrome, patent ductus arteriosus, sepsis, or asphyxia), hypoxia, and umbilical catheterisation reduce intestinal mucosal perfusion, impairing the barrier function further. Pathological microbial colonisation: formula feeding disrupts normal gut colonisation (absence of breast milk oligosaccharides and immunoglobulins that promote normal microbiome development); broad-spectrum antibiotic use eliminates normal flora; pathogenic bacteria (Klebsiella, Enterobacter, Clostridium) colonise the immature ischaemic gut, invade the mucosa, produce gas (pneumatosis intestinalis), and trigger an exaggerated systemic inflammatory response.
2. The pathognomonic radiological finding is pneumatosis intestinalis (gas within the bowel wall), visible on plain AXR as a linear or bubbly lucency outlining the bowel wall; it results from gas produced by bacteria invading the bowel wall. Two other radiological signs: (1) portal venous gas (branching lucencies within the hepatic portal venous system: visible as a tree-like branching radiolucency overlying the liver on AXR): a sinister sign indicating extensive disease and bacterial invasion of the portal venous circulation; (2) pneumoperitoneum (free air under the diaphragm or in the flanks on a left lateral decubitus AXR): indicates intestinal perforation and is an indication for emergency surgery.
3. Bell staging: Stage I (suspected NEC): non-specific systemic signs (temperature instability, apnoea, bradycardia, lethargy) and mild abdominal signs (distension, blood in stools by dipstick or microscopy) without radiological confirmation: treat medically. Stage II (definite NEC): definite abdominal signs (definite distension, tenderness, erythema of abdominal wall, absence of bowel sounds, gross blood in stools) plus radiological confirmation (pneumatosis intestinalis or portal venous gas on AXR): treat medically. Stage IIIA (advanced NEC without perforation): clinical deterioration despite medical treatment or bowel gangrene confirmed at surgery without perforation: treat surgically. Stage IIIB (advanced NEC with perforation): pneumoperitoneum on AXR indicating intestinal perforation: treat surgically.

4. Medical management of Stage II NEC: (1) nil by mouth: bowel rest for 7 to 14 days (stops feeding that provides substrate for bacterial growth and reduces bowel wall tension); (2) nasogastric tube decompression (relieves distension and prevents aspiration); (3) IV fluids and total parenteral nutrition (TPN) via a central venous line (maintains nutrition and fluid balance during bowel rest); (4) broad-spectrum IV antibiotics covering Gram-negative organisms and anaerobes (gentamicin or cefotaxime plus metronidazole, or piperacillin-tazobactam) for 7 to 14 days; (5) correct coagulopathy with fresh frozen plasma (NEC causes a consumptive coagulopathy from sepsis and intestinal necrosis); (6) cardiovascular support (ionotropes if needed for haemodynamic instability); (7) serial clinical assessment and AXR every 6 to 8 hours to detect deterioration and progression to Stage III.

5. Peritoneal drainage: a simple surgical drain (Penrose drain or similar) is placed in the right iliac fossa under local anaesthesia; the drain decompresses the peritoneal cavity, releases contaminated fluid, and may stabilise the infant. Used for the smallest and most premature infants (below 1000 to 1500 g) who are too critically ill and too small to tolerate laparotomy under general anaesthesia; it may be used as a temporising measure before definitive surgery when the infant has stabilised. Laparotomy and resection: used for larger and more stable infants; the abdomen is explored, the extent of bowel necrosis assessed, and all non-viable bowel resected; the proximal end is brought out as a stoma (ileostomy or colostomy) and the distal end as a mucous fistula; primary anastomosis is avoided in the acute setting because of the high risk of anastomotic dehiscence in inflamed, oedematous, fragile bowel; stoma closure is planned 6 to 8 weeks later when the infant has recovered.

References and Attributions [Series 5]

Textual References

1. Coran, A. G., Caldamone, A., Adzick, N. S., Krummel, T. M., Laberge, J. M. and Shamberger, R. (2012). Pediatric Surgery (7th ed.). Philadelphia: Elsevier Mosby.
2. Puri, P. and Hollwarth, M. E. (2009). Pediatric Surgery: Diagnosis and Management. Berlin: Springer.
3. Ameh, E. A. and Chirdan, L. B. (2011). Paediatric Surgery in the Tropics. Ibadan: Spectrum Books.
4. Bell, M. J., Ternberg, J. L., Feigin, R. D. et al. (1978). Neonatal necrotizing enterocolitis: therapeutic decisions based upon clinical staging. Annals of Surgery, 187(1), 1 to 7.

Figures, Plates, Tables, and Boxes

5. Figure 5.1: Intestinal obstruction in neonates. AI-generated using the prompt: "Two-panel diagram: classification diagram (mechanical high versus low versus functional) with meconium ileus box (cystic fibrosis, soap-bubble sign); cardinal features box, investigations table (AXR findings, contrast enema, upper GI study), general management principles, and meconium ileus specific management. Paediatric surgical diagram style." Suggested OER search string: "neonatal intestinal obstruction meconium ileus cystic fibrosis Gastrografin enema paediatric surgery OER."
6. Figure 5.2: Intussusception. AI-generated using the prompt: "Two-panel diagram: cross-sectional ileocolic intussusception illustration with aetiology box (idiopathic versus lead point), classic triad box, and physical signs box; investigations box (target sign on ultrasound and crescent sign on AXR) and management flowchart (resuscitation, air enema first-line, contraindications, operative milking technique, recurrence). Paediatric surgical diagram style." Suggested OER search string: "intussusception target sign air enema reduction operative management paediatric surgery OER."
7. Figure 5.3: Meckel's diverticulum and intestinal duplications. AI-generated using the prompt: "Two-panel diagram: Meckel's diverticulum anatomical diagram on antimesenteric ileum with rule of twos box and clinical presentations table (bleeding, obstruction, diverticulitis, umbilical fistula); intestinal duplications diagram (cystic versus tubular) with presentations box and management table (diverticulectomy versus segmental resection, Meckel's scan). Paediatric surgical diagram style." Suggested OER search string: "Meckel's diverticulum rule of twos Meckel's scan intestinal duplication paediatric surgery OER."

8. Figure 5.4: Necrotising enterocolitis. AI-generated using the prompt: "Two-panel diagram: NEC pathophysiology triad (immaturity, ischaemia, microbial colonisation) with risk factors box and clinical features box (systemic signs, abdominal signs, radiological findings: pneumatosis intestinalis and portal venous gas and pneumoperitoneum); Bell staging table (Stages I to III) with medical management box (nil by mouth, TPN, antibiotics, serial AXR) and surgical management box (peritoneal drainage versus laparotomy and stoma) and outcomes box. Paediatric surgical diagram style." Suggested OER search string: "necrotising enterocolitis NEC Bell staging pneumatosis intestinalis peritoneal drainage laparotomy stoma OER."