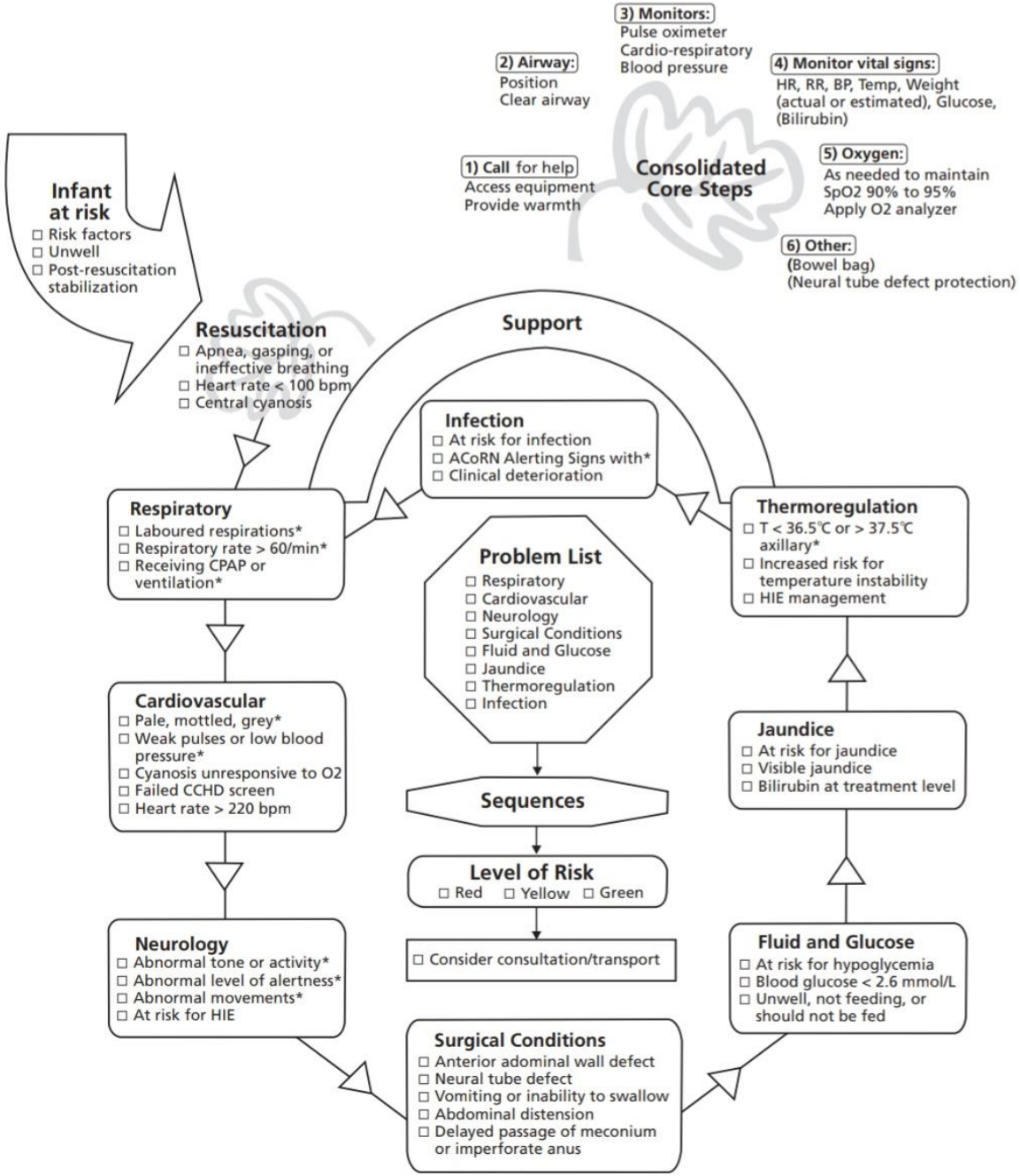
surgical condition

Dr. Dehvari
neonatologist

The ACoRN Primary Survey



The ACoRN Process (Figure 2.1) has nine key steps:

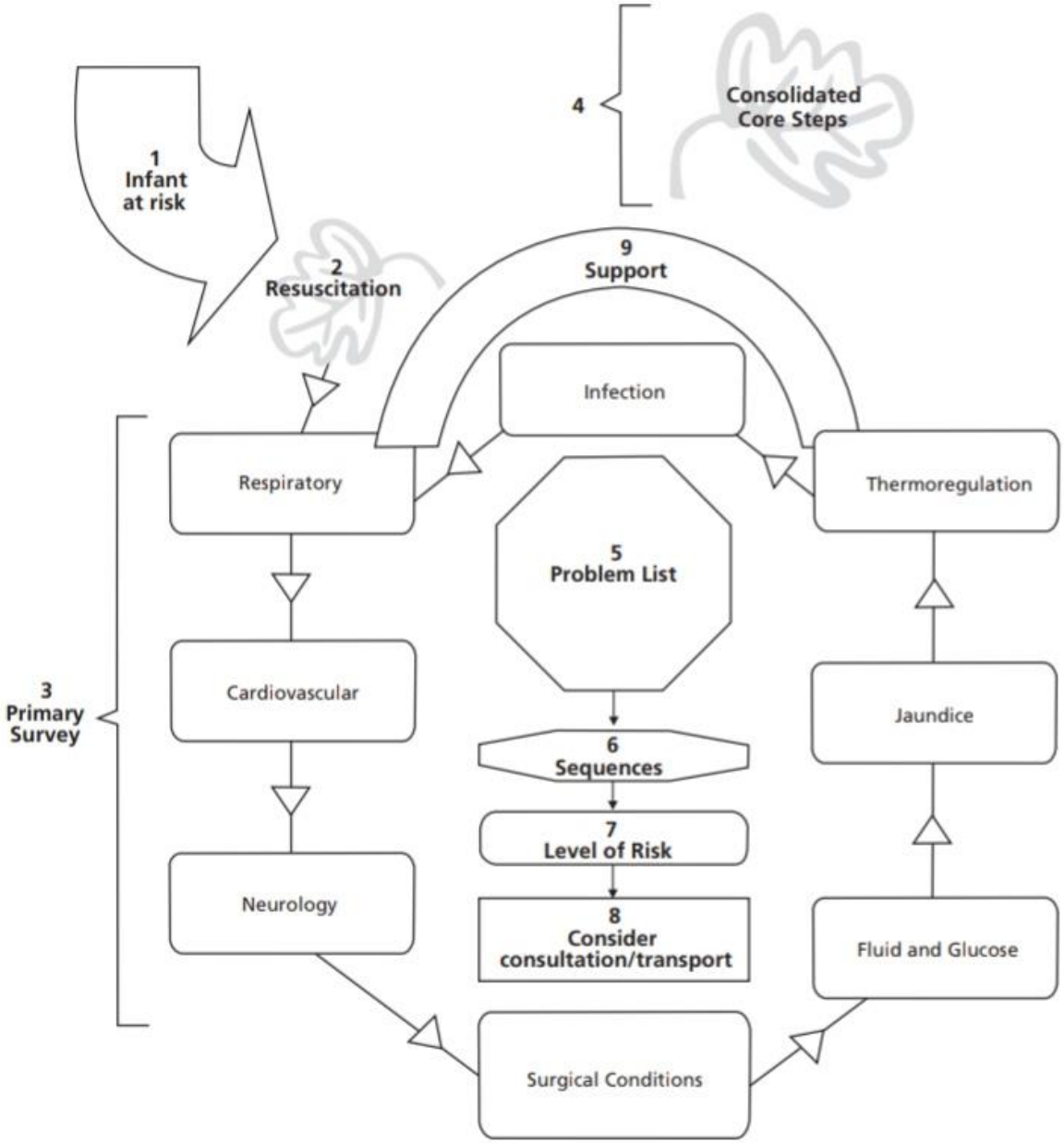


Figure 2.1. The ACoRN Process.

- Key concepts
- 1. Infants with major surgical conditions require care by a specialized medical- surgical team
- 2. Early recognition, preferably in utero, improves quality of care and outcomes.
- 3. Infants who show one or more of the ACoRN Alerting Signs for surgical conditions should
- be investigated promptly to identify or exclude pertinent associated conditions and causes.
- 4. Medical stabilization precedes surgery in all infants presenting with major surgical conditions.
- 5. The timing of surgical intervention varies based on the condition and degree of medical stabilization.
- In most cases, surgical intervention can be delayed until optimal medical stabilization
- is achieved

Prenatal diagnosis

- • Identify or exclude associated conditions and causes, such as chromosomal anomalies or genetic
- syndromes.
- • Optimize counselling for parents on potential outcomes, intervention options, and the most appropriate
- timing, location, and mode of delivery.
- • Plan special care requirements. Collaborative care planning will involve the family, the mother’s
- antenatal care provider, maternal– fetal medicine specialists, neonatologists, paediatric surgeons, and
- geneticists.
- • Arrange a family visit to the facility where the mother and infant will be cared for, and a meeting
- with members of the care team.
- • Determine need and timing for prenatal transport or family relocation.

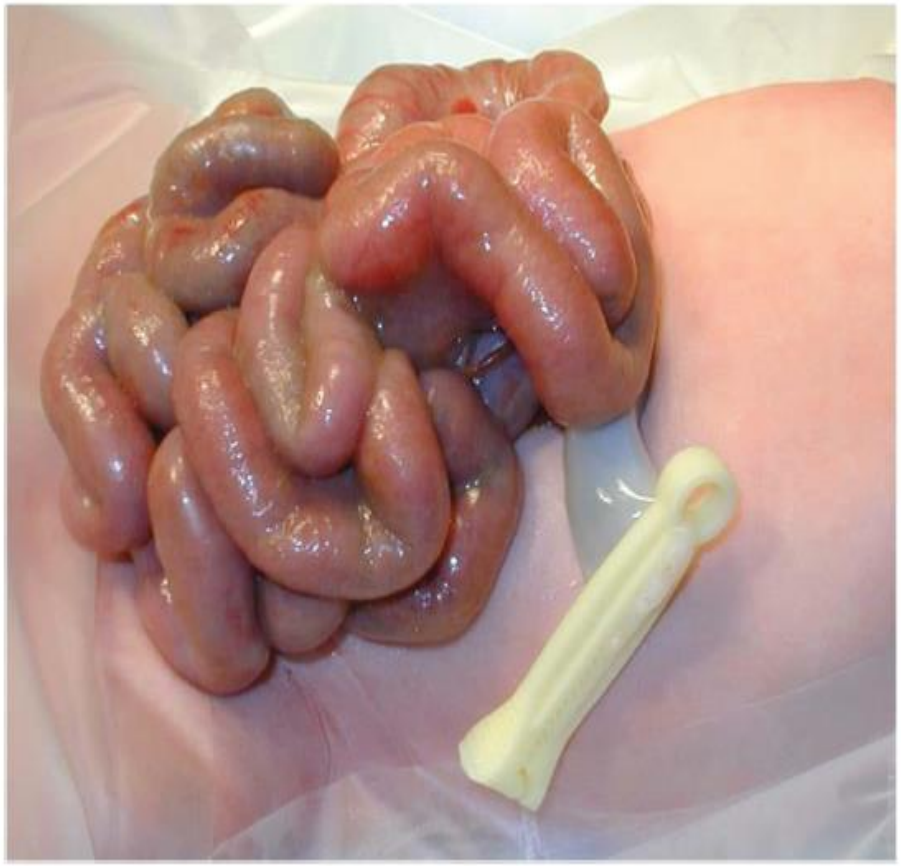
not detected during routine maternal ultrasound

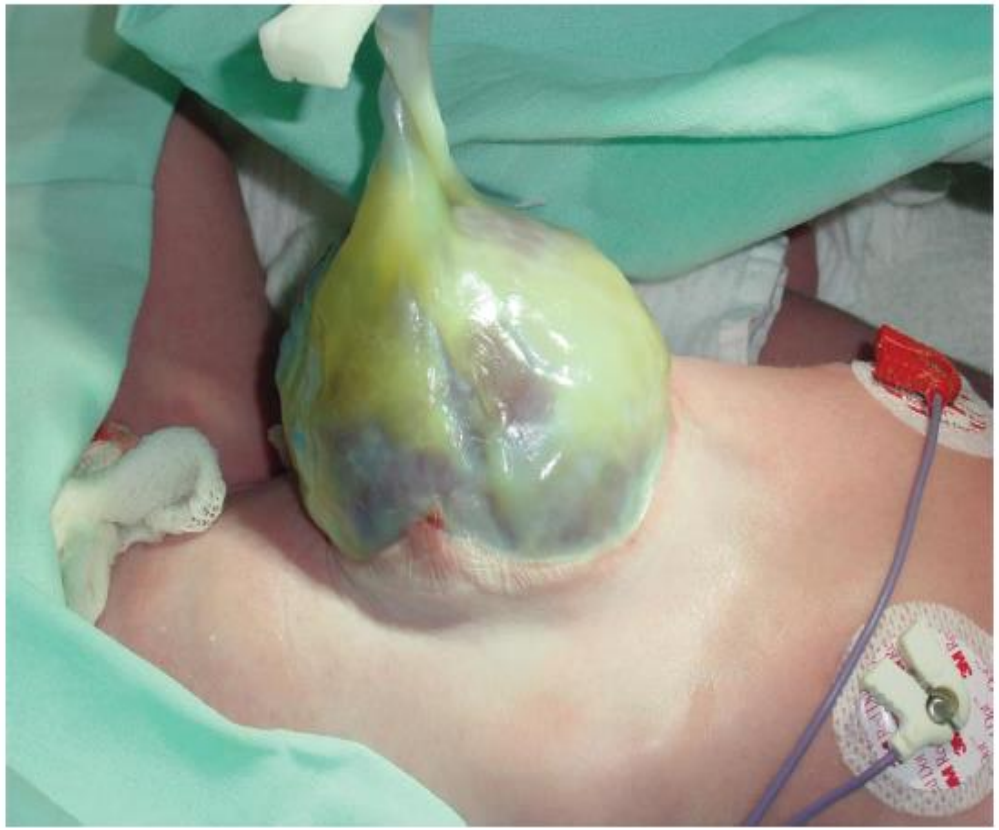
- • Prompt recognition,
- • Medical stabilization, and
- • Urgent transport to a tertiary facility

Surgical Conditions alerting signs

- Anterior adominal wall defect
- Neural tube defect
- Vomiting or inability to swallow
- Abdominal distension
- Delayed passage of meconium or
- imperforate anus

Anterior abdominal wall defect





Gastroschisis

- The small intestine and, sometimes, other organs (stomach, large bowel, spleen) protrude through a
- small opening in the abdominal wall to the right of the umbilicus (Figure 6.1). The umbilical cord is
- not involved and is anatomically normal. The exposed intestine is not covered by a peritoneal membrane
- and often appears thick, foreshortened, and matted. The intestine is usually anatomically normal,
- but all infants with gastroschisis have abnormal rotation and fixation of the intestines. Some infants may
- have other GI anomalies, including volvulus, intestinal atresia, intestinal stenosis, or intestinal perforation.
- Coexisting chromosomal anomalies are unusual in infants with gastroschisis

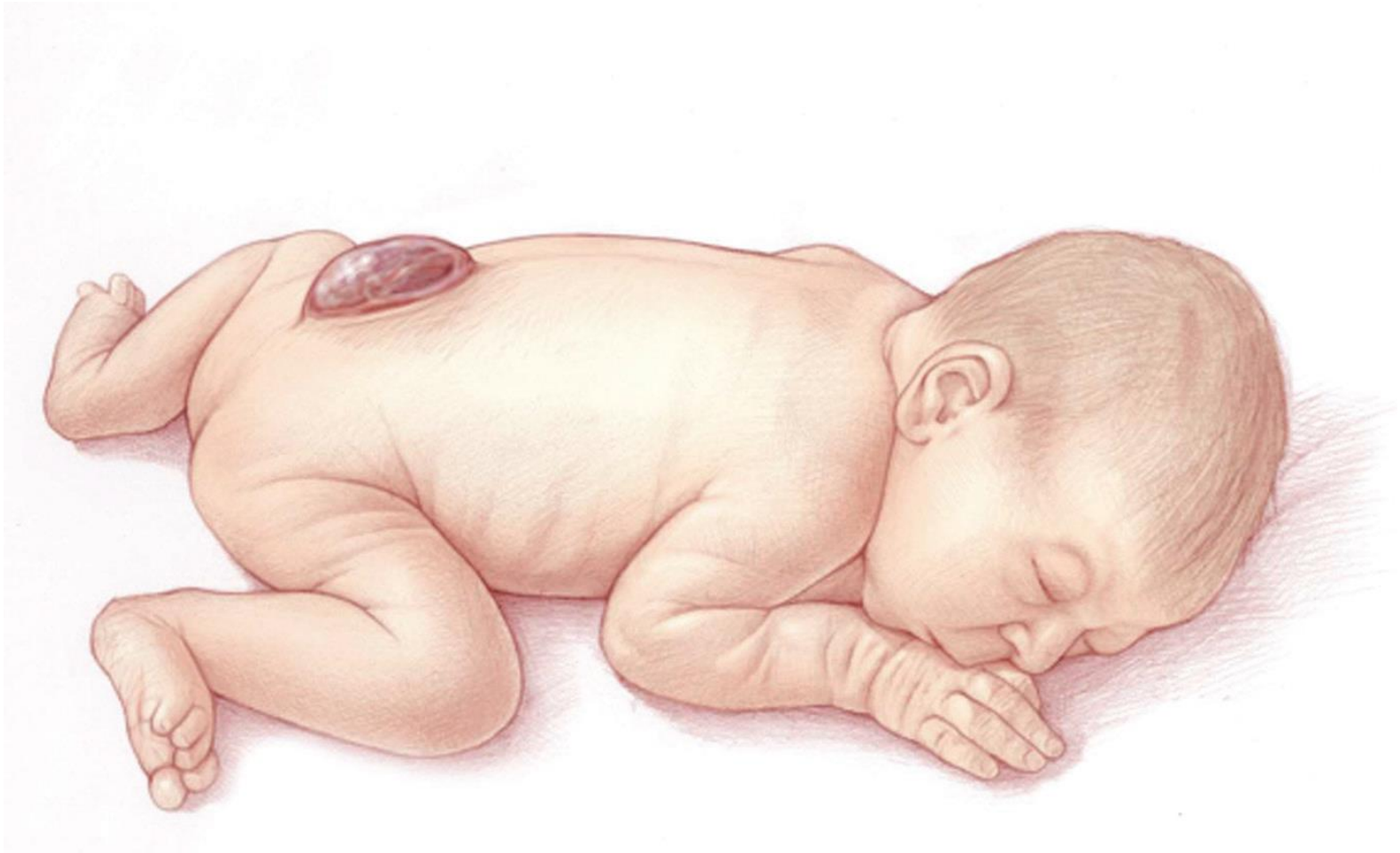
Omphalocele

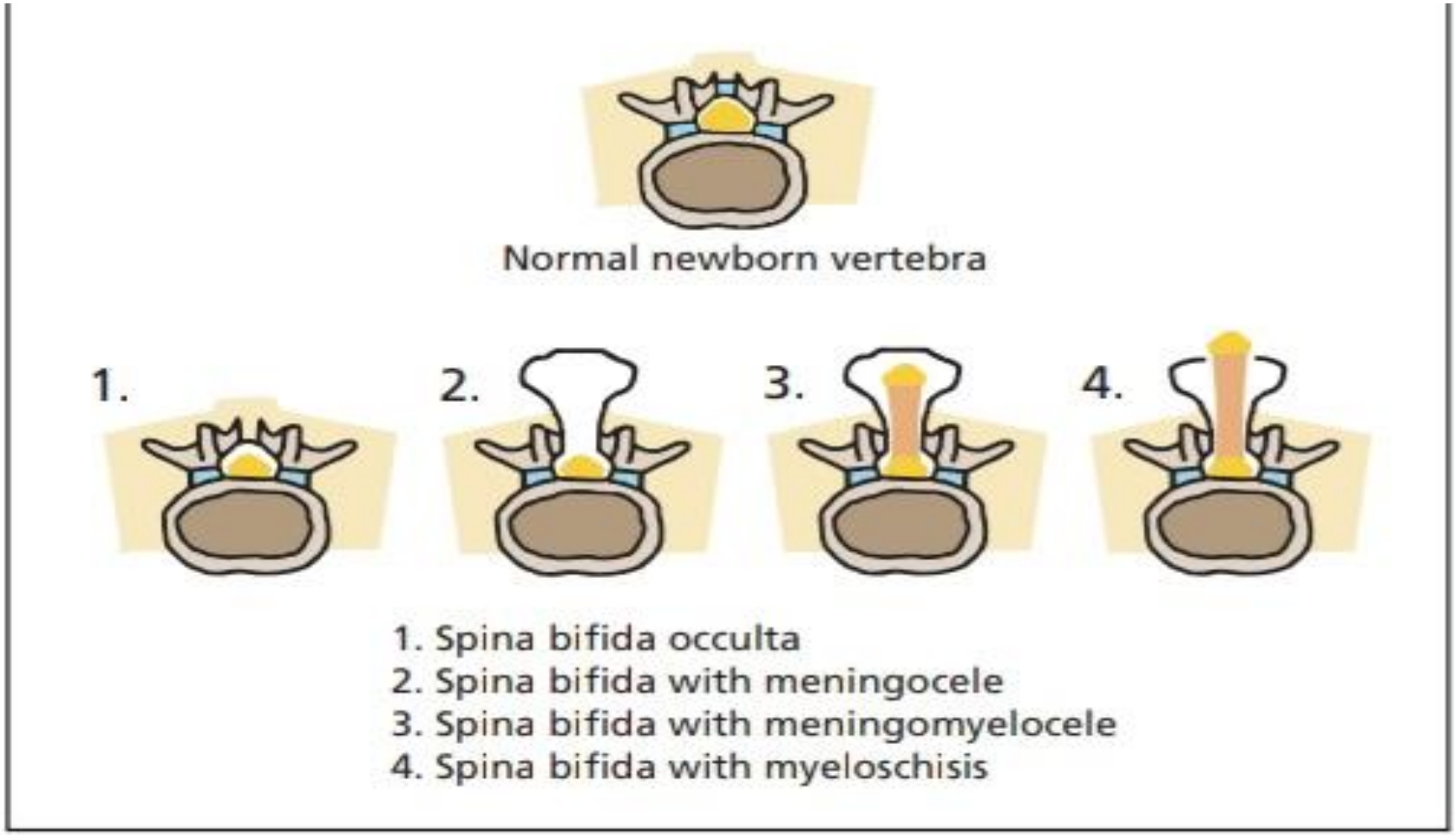
- Characteristically a midline defect, an omphalocele occurs at the insertion point of the umbilical cord,
- with a translucent sac composed of amnion, Wharton's jelly, and peritoneum covering the herniated
- abdominal contents. The umbilical cord inserts into the membrane rather than the abdominal wall. In
- most cases, only the small intestine is visible, but when the omphalocele is large (sometimes called a
- 'giant omphalocele'), the liver and other organs may be seen. A giant omphalocele (Figure 6.2) is difficult
- to repair because there may not be enough space in the abdominal cavity to replace exposed organs
- high incidence of associated conditions (30% to 40%) and chromosomal
- abnormalities in infants with omphalocele that may impact prognosis, including survival

Neural tube defect

- 1. Spina bifida occulta is the mildest form of this condition, and it is often discovered coincidentally
- during an imaging test done for unrelated reasons. Spina bifida occulta is a small separation in one
- or more of the vertebrae without spinal cord or meningeal herniation.
- 2. Meningocele is an opening in the vertebrae, through which a sac containing meninges and cerebrospinal
- fluid protrudes.

- 3. Myelomeningocele is the most common form of spina bifida. In addition to meningocele, nerve
- tissue protrudes through the vertebral opening. It is covered by a sac and may be partially or completely
- covered by skin.
- 4. Myeloschisis is an open defect of the spine that may involve exposure of the entire spinal cord. It is
- the most severe type of spina bifida





.4. Types of neural tube defect.

Vomiting or inability to swallow

- **Nonbilious vomiting can be caused by an obstruction proximal to the point where the common bile**
- duct enters the duodenum:
- • Pyloric stenosis can present as early as the first week to months after birth, with nonbilious, forceful
- vomiting, and dehydration.
- • Proximal duodenal webs or atresia will present within hours of birth.
- **Bilious vomiting in the newborn should be considered evidence of mechanical intestinal obstruction,**
- until proven otherwise. Causes include:
- • Duodenal atresia, stenosis, or web, with obstruction distal to the entry of the common bile duct into
- the duodenum; • Jejunal- ileal atresia; and
- • Malrotation with volvulus

- Volvulus is a surgical emergency. If the condition is unrecognized or surgery is delayed, necrosis of the entire intestine, shock, and death can ensue
- Bilious vomiting is not always due to a GI obstruction, but obstruction must be ruled out urgently

Inability to swallow (or handle secretions)

- **Esophageal atresia (EA)/ tracheoesophageal fistula (TEF) can present with difficulty feeding,**
- noisy and laboured breathing (especially during attempted feeds), and inability to handle oral secretions
- (excessive drooling). There may also be periods of cyanosis and apnea during attempted feeds, as the infant
- aspirates milk or oral secretions into the lungs. When a tracheoesophageal fistula is present, infants
- may experience increasing abdominal distension, especially during noninvasive respiratory support or
- mechanical ventilation

infant's lungs and also decrease risk for overdistension or perforation of the stomach. In both types, timing the esophageal anastomosis depends on the length of the gap between the proximal and distal esophagus.

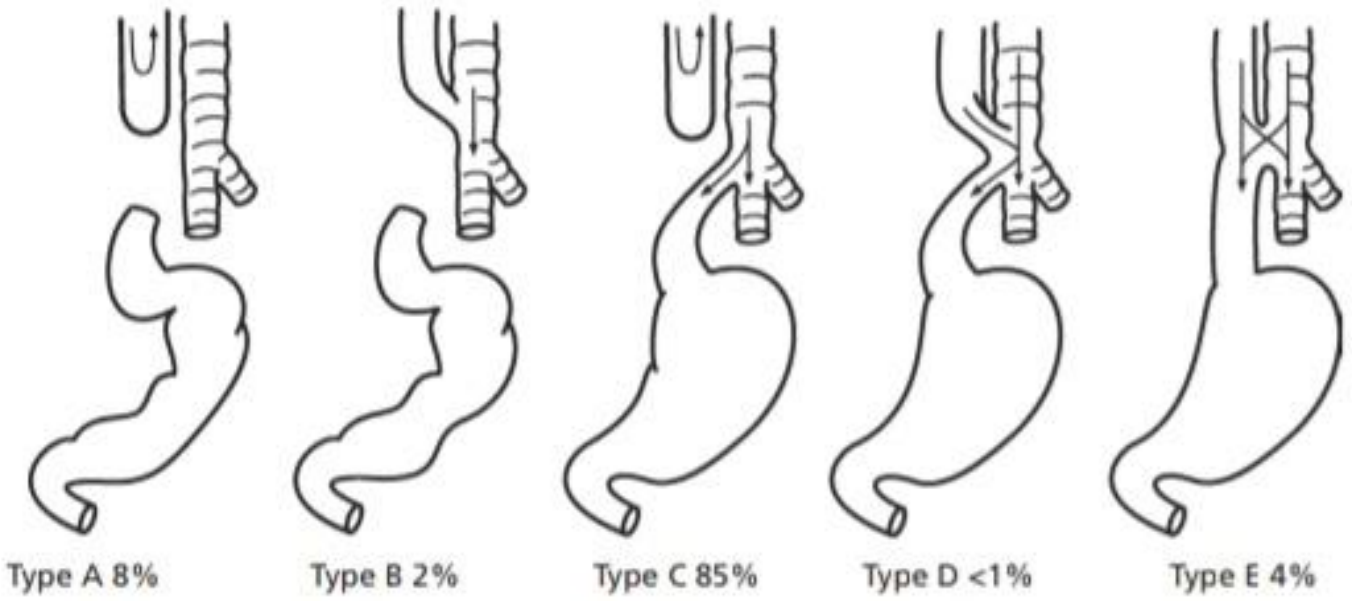


Figure 6.7. The 5 types of EA/TEF, with prevalence (%).

Abdominal distension

- When the infant's abdomen is tender as well as distended, it is important to exclude causes of
- acute abdomen, such as necrotizing enterocolitis (NEC), volvulus, or GI perforation (ruptured
- bowel).

- 1. **NEC is a condition of multifactorial pathogenesis and is more common in preterm infants. NEC**
- commonly occurs in the terminal ileum and proximal colon. Manifestations vary widely, from abdominal
- distension with fresh blood in the stool to fulminant sepsis, intestinal perforation, and necrosis
- of the entire intestinal tract. Age of onset can be days in term or late preterm infants but can
- be delayed by weeks in infants born more preterm. NEC without peritonitis or perforation will
- generally be managed medically with bowel rest, antibiotics, and close monitoring.
- 2. **Volvulus occurs when the bowel twists on itself, leading to intestinal ischemia. Infants with volvulus**
- may present with abdominal distension, bilious emesis, bloody stools, and hemodynamic instability.
- Volvulus can be secondary to malrotation or the bowel can twist on a lead point, such as in congenital
- Meckel's diverticulum or enteric duplication.
- 3. **Spontaneous gastric or intestinal perforations are focal and occur more commonly in preterm**
- infants.

Delayed passage of meconium or imperforate anus

- • Imperforate anus, with or without a perineal or genitourinary fistula,
- • Microcolon,
- • Colonic atresia,
- • Meconium ileus, or
- • Hirschsprung's disease (congenital absence of the myenteric plexus).
- Delayed passage of meconium or imperforate anus

  . An infant who has not passed meconium by 48 h (or 72 h in a preterm infant) should be identified early, preferably before the infant's abdomen becomes distended, to avoid complications such as colitis due to bacterial overgrowth.

- In all newborns, ensure that the anus is patent. A presumed anal opening may in fact be a blind pouch. There may be seepage of meconium through the urethra, vagina, or perineal fissure in infants with imperforate anus

Core Steps

- • Protect an exposed lesion with a clear cover or dressing, as appropriate (see Appendix A),
- • Place NPO

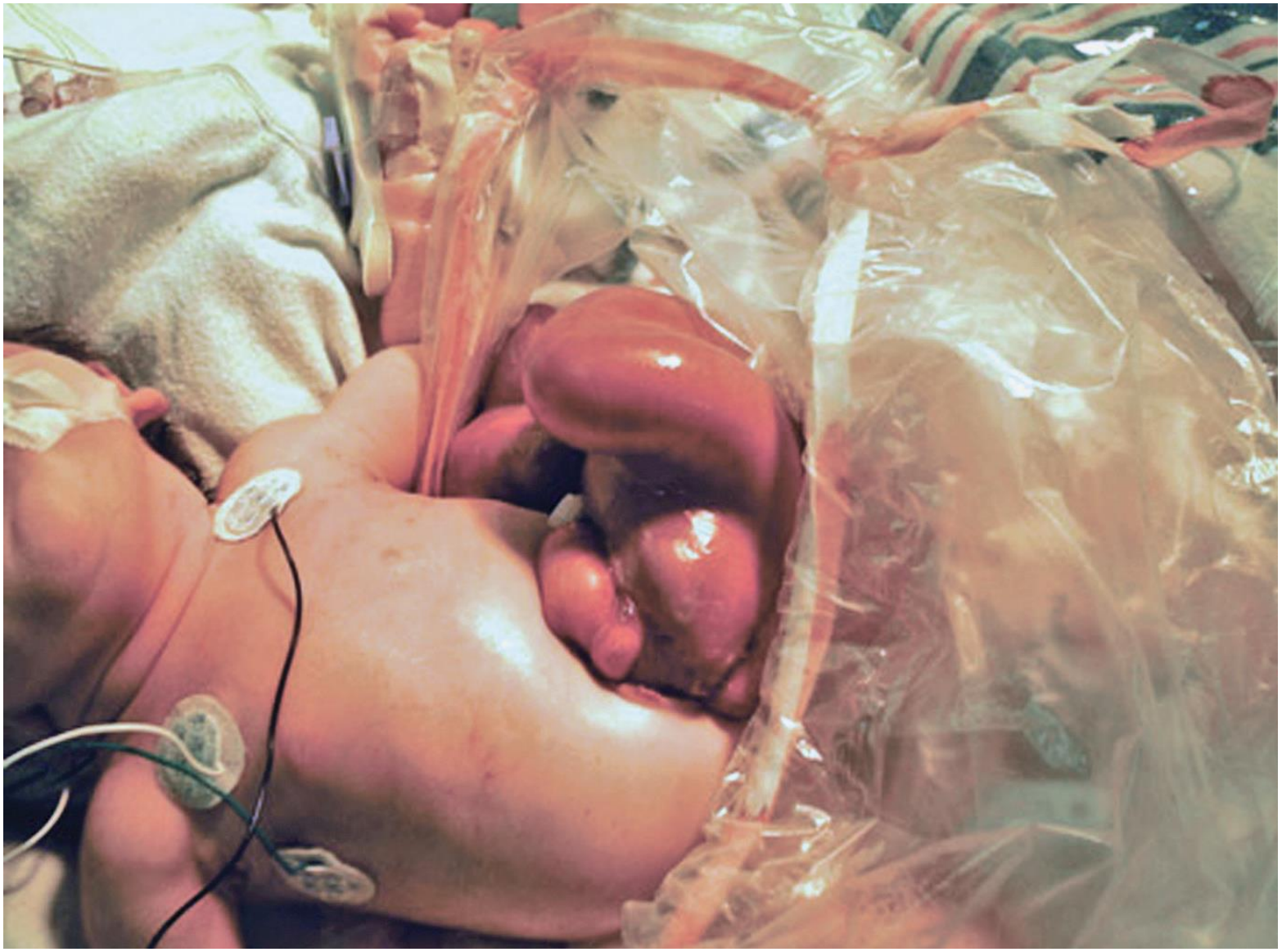
Organization of Care

- • An anterior abdominal wall defect,
- • A neural tube defect,
- • No anterior wall defect but signs of GI obstruction (i.e., vomiting, inability to swallow, abdominal
- distension, delayed passage of meconium, or an imperforate anus), *and whether a gastric tube can be*
- inserted

For the infant who has an anterior abdominal wall defect

- • Manage the infant on an overhead warmer or in an incubator, to prevent heat loss.
- • Maintain sterile technique when manipulating the bowel (wear a mask and latex- free sterile
- gloves).
- • Place the infant in a sterile bowel bag (Figure 6.5) and secure at the level of the nipples or apply a
- transparent occlusive covering to the bowel to minimize heat and fluid losses.
- • Avoid mask ventilation, continuous positive airway pressure (CPAP), and noninvasive positive pressure
- ventilation (NiPPV) to minimize gaseous distension of the intestines. Intubate early if the infant is in
- respiratory distress and needs ventilatory support.

- • Minimize handling of the exposed bowel or viscera to prevent secondary damage and infection. For
- infants with an omphalocele, take care not to tear the sac.
- • Avoid pressure on the mesentery and potential kinking of the bowel by using rolls to support the
- bowel midline in either a supine or side- lying position.
- • Decompress the GI tract by inserting a #10 Fr double-lumen Replogle ‘sump’ tube for continuous
- suction or a #8 Fr or #10 Fr single- lumen gastric tube, on low, intermittent suction



- ◦ A double- lumen Replogle tube allows continuous suction through one lumen with the other
- vented to air, to prevent the catheter from generating a vacuum against the mucosa. Start suction
- at 40 mmHg to 60 mmHg and maintain pressure in the low range, never exceeding 80 mmHg.
- ◦ If a single- lumen gastric tube is inserted to decompress the GI tract, use intermittent suction
- only. Continuous suction using this device can collapse the stomach, creating a vacuum pressure
- on the mucosa, which can cause ulceration, hemorrhage, or perforation. Use a regulator that
- has an intermittent suction setting, with preset on- and- off cycles. Start suction at 40 mmHg to
- 60 mmHg and maintain pressure in the low range, never exceeding 80 mmHg.

neural tube defect

- • Manage the infant on an overhead warmer or in an incubator to prevent heat loss.
- • Maintain sterile technique when examining or dressing the lesion (wear a mask and latex- free sterile
- gloves to minimize risk for developing latex sensitivity).
- • Ensure the defect is covered with a sterile moist protective wrap.
- • Position prone or side- lying.
- • Create or use a pre- packaged ‘donut’ dressing on skin surrounding the lesion to prevent pressure on
- the protruding sac.
- • Keep the sac intact to prevent cerebrospinal fluid leakage or damage to the nerve tissue

Unable to pass gastric tube

- • Raise the head of the infant’s bed 30° to minimize aspiration from the esophageal pouch or
- gastroesophageal reflux through a distal fistula.
- • Initiate low continuous suction using a double- lumen Replogle tube placed in the upper esophageal
- segment, and set the level of suction at 40 mmHg to 60 mmHg.
- • If a Replogle tube is not available, insert a single- lumen gastric tube to the point of resistance and
- apply low intermittent suction at 40 mmHg to 60 mmHg.
- • Avoid mask ventilation, CPAP, or NiPPV for infants with EA/TEF to minimize gaseous distension
- of a blind esophageal pouch or GI tract via a fistula. Intubate early if the infant is in respiratory distress
- and requires ventilatory support

Focused history

- • Maternal age: risk for gastroschisis increases in mothers younger than 25 years old and is highest in
- mothers under 20 years old
- • Medical, obstetrical, and family history
- • Maternal diabetes or hypothyroidism
- • Maternal medications during pregnancy, including potential teratogens
- • Other risk factors include suboptimal prenatal care, low socio- economic status, smoking during
- pregnancy, occupational or recreational exposure to solvents, or use of amphetamine- related drugs
- or cocaine during pregnancy

- • Previous fetuses or children with congenital anomalies
- • Prenatal genetic screening
- • Prenatal ultrasound findings. Most conditions described in this chapter are easily seen on ultrasounds,
- but others, such as EA/TEF, become suspected in the absence of a fluid- filled stomach or
- polyhydramnios on ultrasound

Focused history

- Because many lesions are associated with other congenital anomalies, obtaining a full antenatal record, including any fetal echocardiography results, is important for care planning and infant outcomes

Focused history

- • Gestational age
- • Birth weight: small, appropriate, or large for gestational age
- • Need for resuscitation
- • Onset of feeding and type of feeds
- • Any changes in feeding pattern
- • Difficulty feeding: coughing, choking, cyanosis, or apnea
- • If vomiting, time of onset and colour of vomitus
- • Passage of meconium at or post- delivery
- • Colour of stool and presence of blood
- • If abdominal distension, time of onset

Focused physical examination

- • The association of omphalocele with Pentalogy of Cantrell, Beckwith- Wiedemann syndrome,
- OEIS complex (omphalocele, exstrophy of the cloacae, imperforate anus, and spinal deformity) or
- with aneuploidies, such as trisomy 13 and 18; and
- • EA/ TEF or imperforate anus may be components of the VACTERL association (vertebral defects,
- anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities

Focused physical examination

- *Abdominal wall defect*
- • Check for a covering membrane and whether it is intact.
- • With gastroschisis, assess the bowel for kinking, cyanosis, or necrosis. Presence of meconium on the
- bowel surface may indicate a rupture. Positioning to avoid kinking or pressure on the bowel and
- monitoring the health of the bowel through the bowel bag, before and during transfer to a level 3
- centre, is important

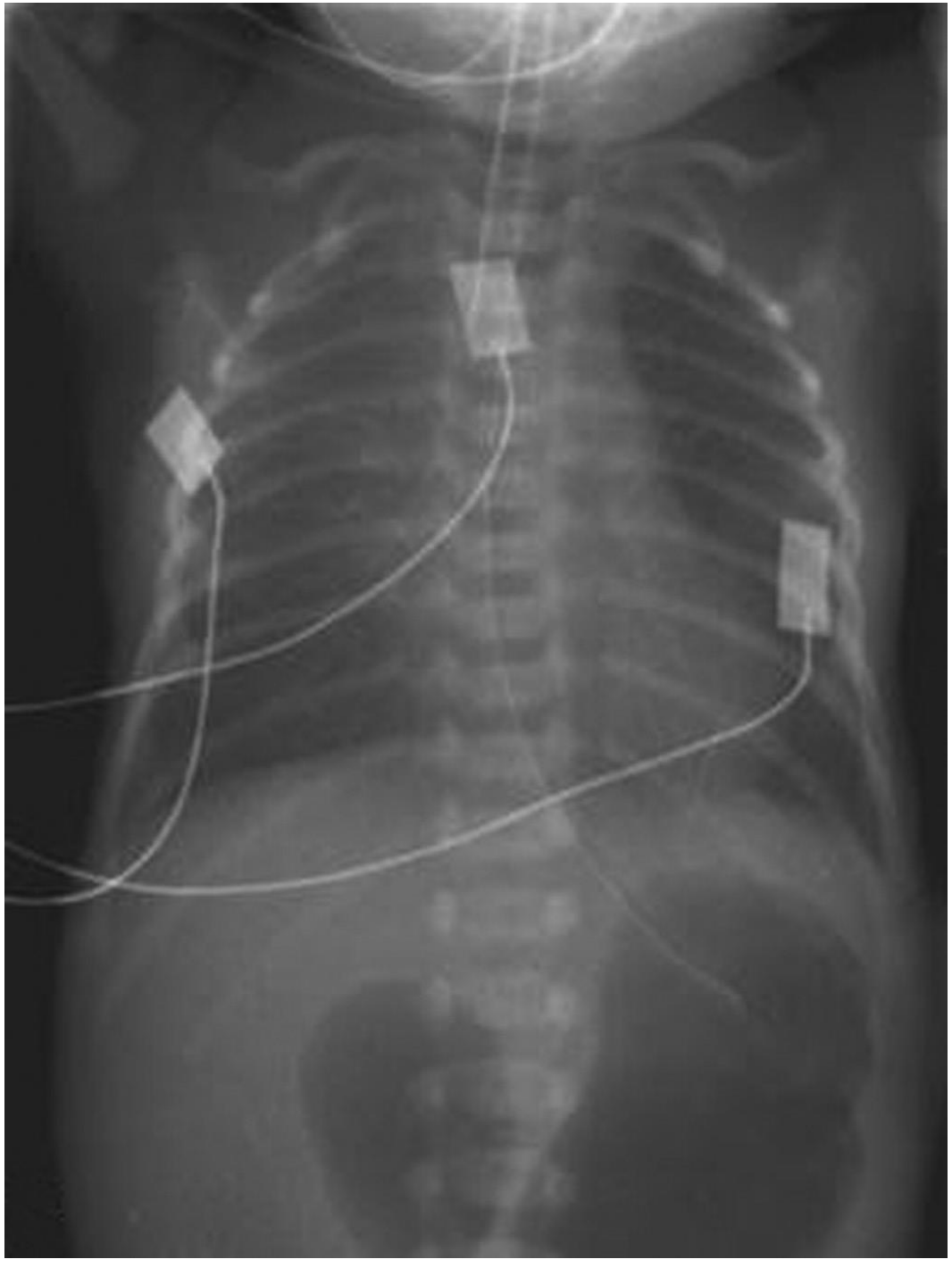
- *Neural tube defect*
- • Observe lesion for the level of spine involved and the presence of intact covering.
- • Measure head circumference and evaluate anterior fontanelle and cranial sutures. Hydrocephalus can
- complicate spina bifida cases with coincident Chiari malformation.
- • Conduct a thorough neurologic exam for signs of deficit, such as abnormal tone and movement of
- lower limbs, absence of anal sphincter tone, or dribbling of urine

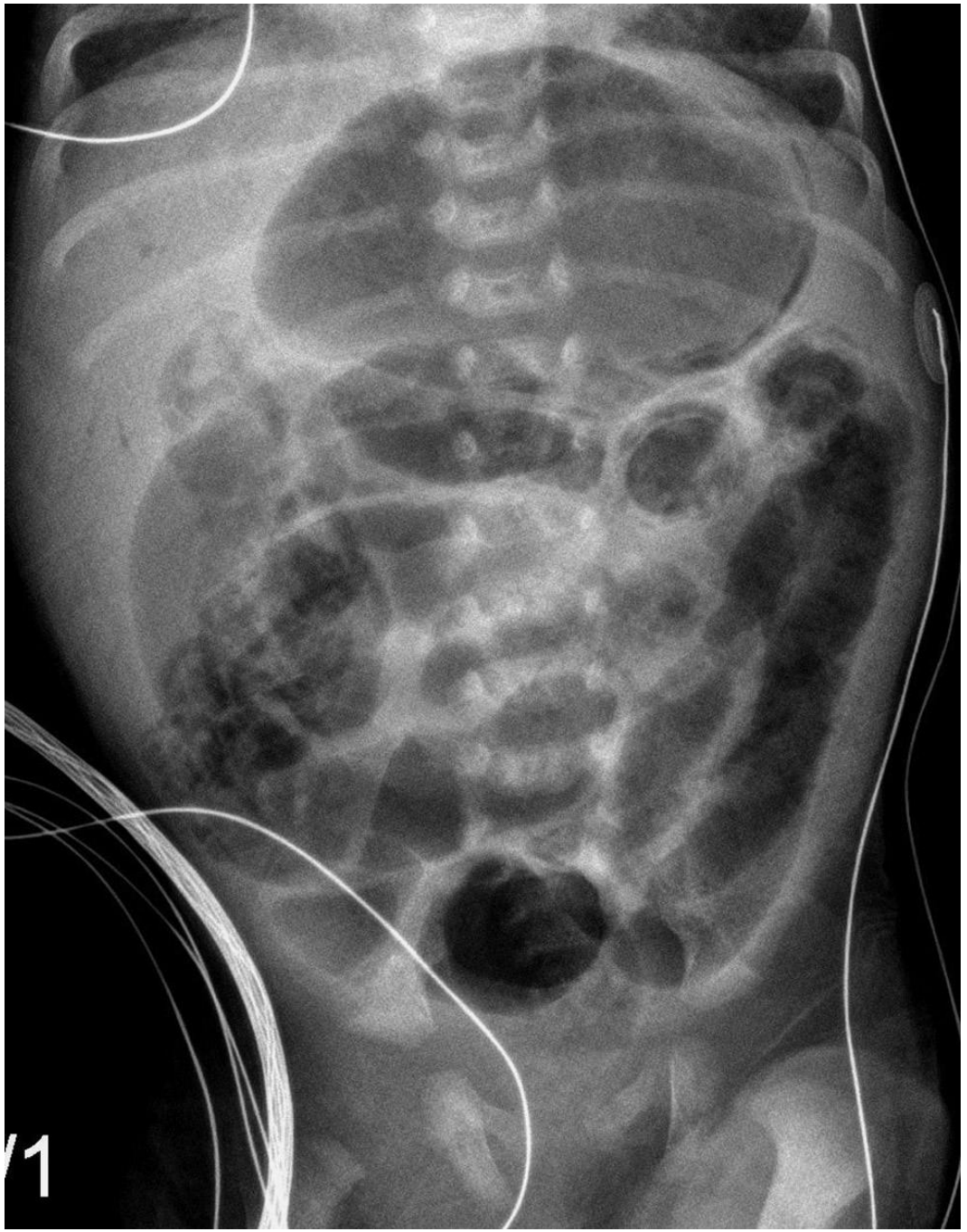
- • Spontaneous or induced labour
- • Meconium or bile- stained amniotic fluid
- • Mode of delivery
- • Determine hip stability.
- • Check for skin abnormalities such as hemangioma, pigmented nevi, or dermal sinus.

Chest and abdominal radiographs

- • Confirm location of the Replogle or single-lumen gastric tube tip within the stomach.
- • With esophageal atresia, the gastric tube typically ends at an air- filled pouch, mid-chest
- • Air pattern in the bowel often indicates the level of obstruction. A classic ‘double bubble’ (dilation of
- the stomach and proximal duodenum)

- • Bowel wall thickening, an asymmetric bowel gas pattern, bubbly looking or linear lucencies (‘string of
- pearls’) in the bowel wall (pneumatosis) or branching lucencies in the liver (portal air), suggest NEC.
- • A cross- table lateral radiograph with the infant in a supine position can best identify air- fluid
- levels within the bowel loops (seen in intestinal obstruction) and free air in the peritoneal cavity.
- • Calcifications in the peritoneal cavity suggest intrauterine bowel perforation, sometimes in association
- with meconium ileus (meconium peritonitis).
- • Assessment of the vertebral column is important with neural tube defects but also when other
- congenital anomalies are present. The presence of hemivertebrae or fused vertebrae may suggest
- VACTERL association

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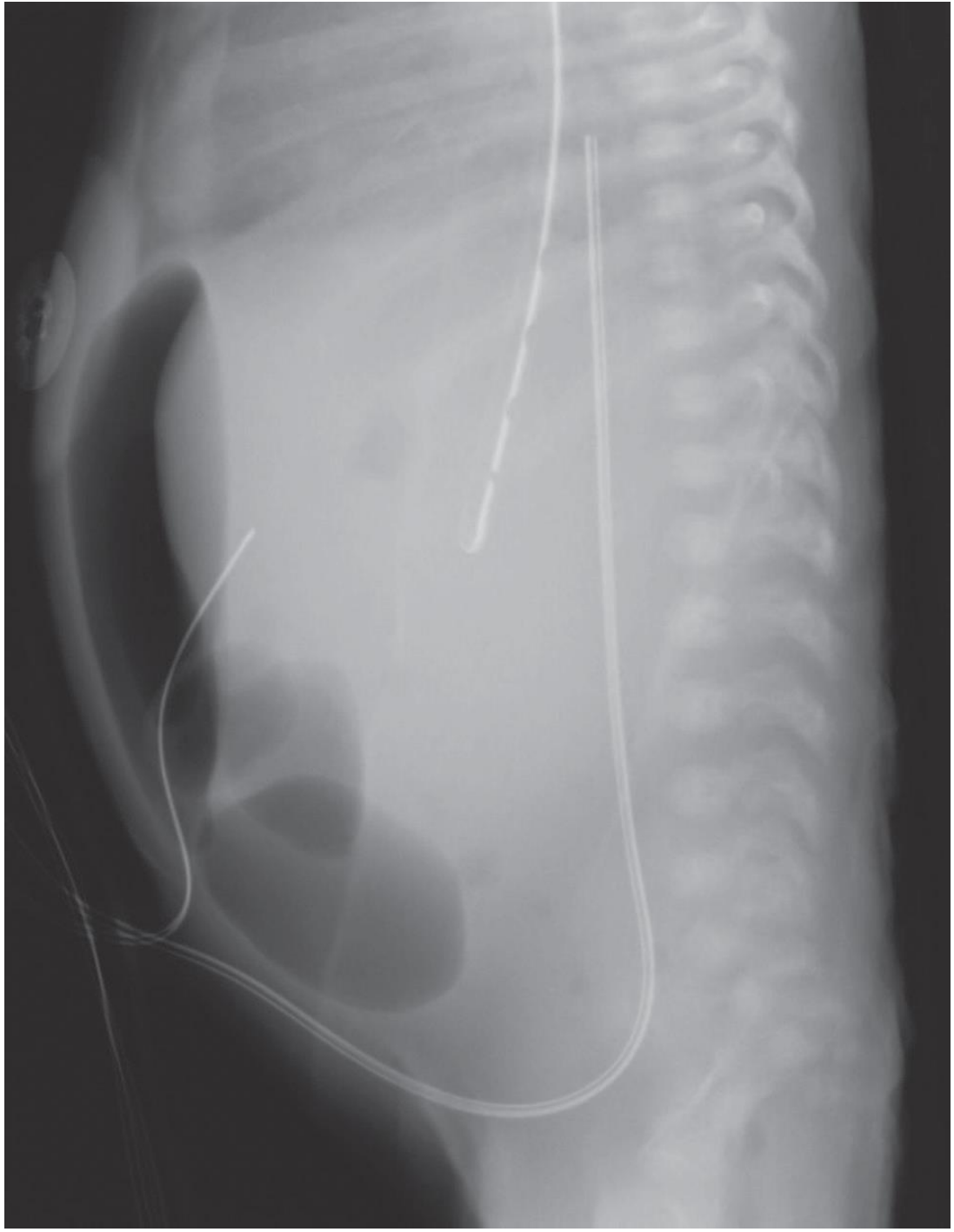


Blood tests

- Order serum sodium, potassium, and chloride to determine whether there are electrolyte abnormalities
- resulting from excessive water or electrolyte losses by evaporation (with an abdominal wall defect), or
- by vomiting, suctioning, or third- spacing (with intestinal obstruction or an acute abdomen). This electrolyte
- assessment should be routine after 12 h of age because measurements earlier than this usually
- reflect maternal values.
- Blood cultures and antibiotics should be initiated when there is an open lesion or evidence of an acute
- abdomen.

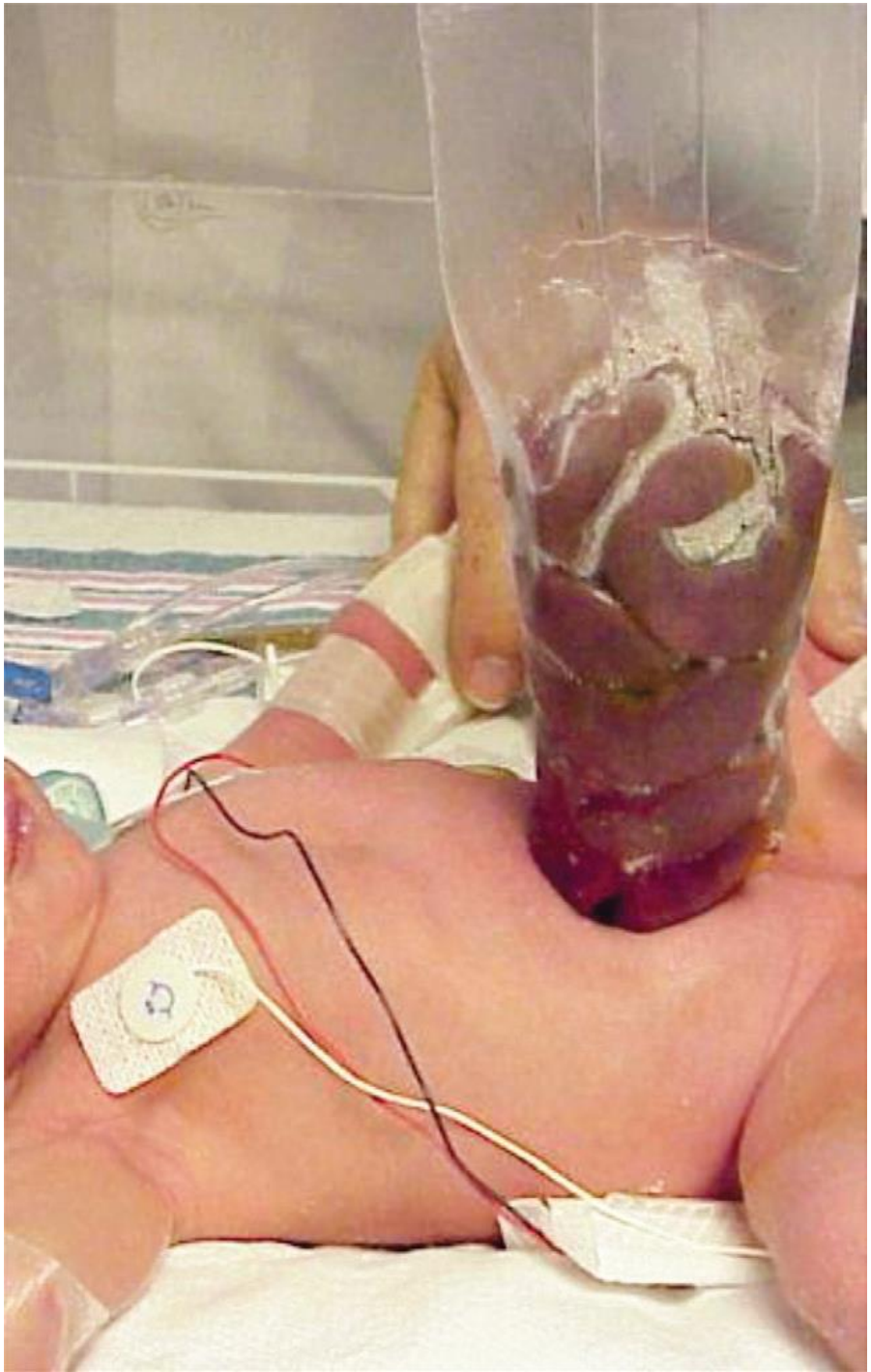
- Newborns with surgical conditions require a fluid balance sheet to monitor intake and output volumes,
- including fluid aspirated through a Replogle or single-lumen gastric tube.
- In gastroschisis, the exposed bowel may lead to excessive fluid loss by evaporation, intestinal edema,
- or both. Using a bowel bag and handling the bowel carefully can reduce fluid losses but paying continuous
- attention to the fluid status of the infant is important. Decisions on IV fluid administration should
- be made in consultation with the tertiary surgical site while awaiting transport

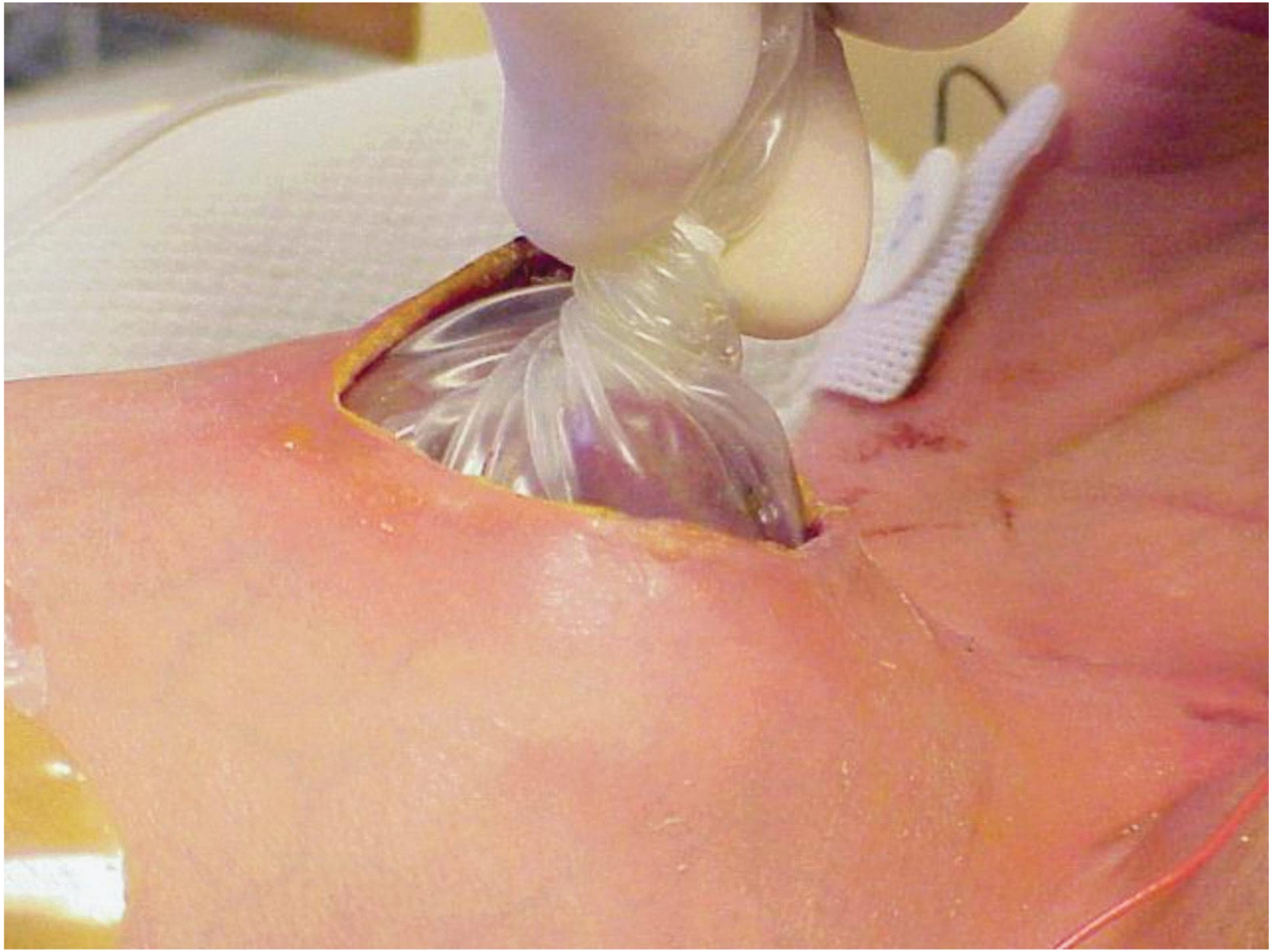
- **Gastroschisis should be managed as soon after birth as possible by tertiary paediatric surgeons, to**
- preserve gut viability and minimize risk of infection. Unless the bowel is easily reduced into the abdomen,
- this will usually involve placing the bowel in a silo, with progressive reduction into the abdomen
- before surgical closure

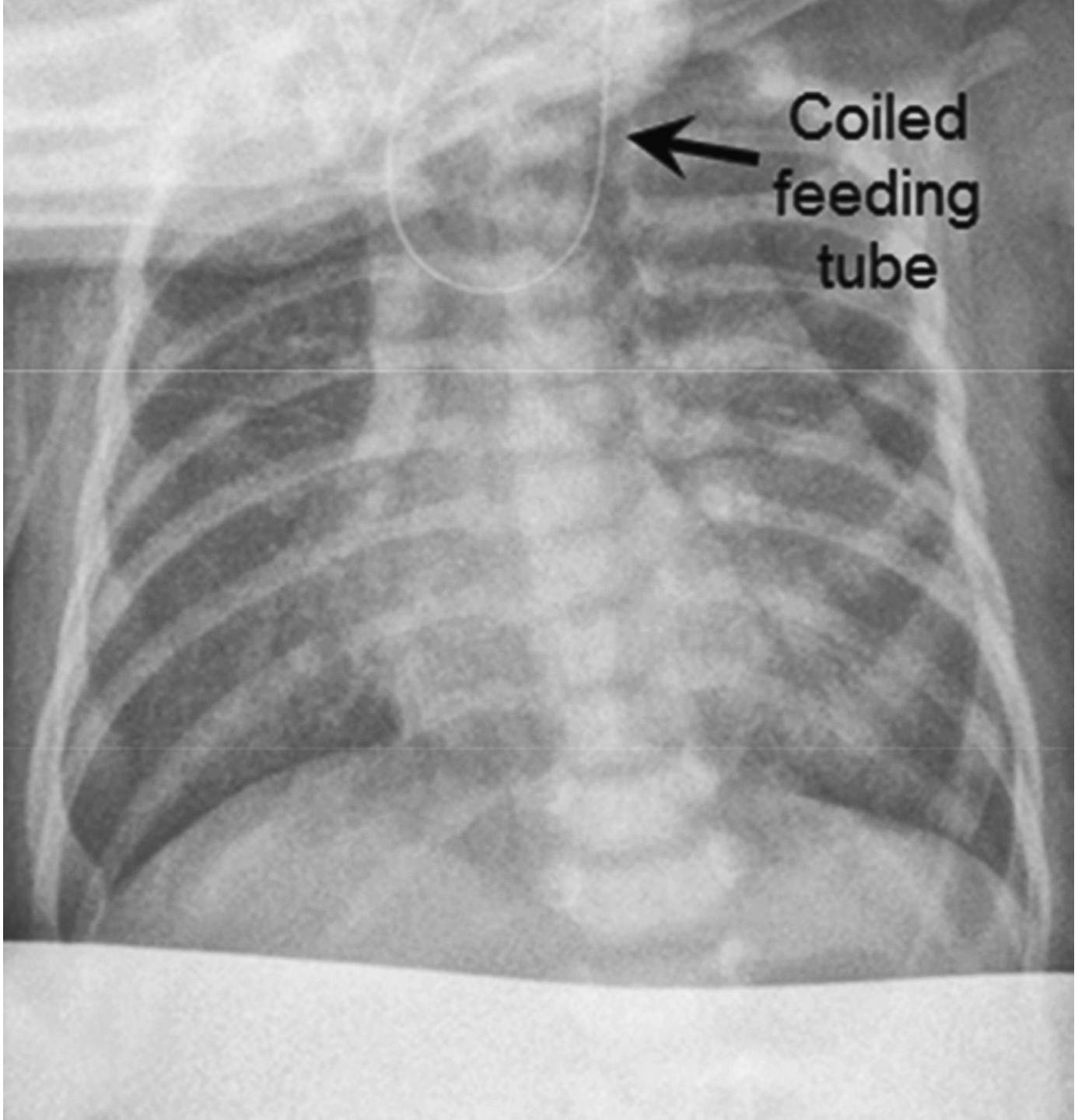
- The urgency to repair an **omphalocele depends on whether the sac is ruptured or not. The timing**
- of the surgery also depends on the dimension of the defect, the infant's size, and the presence or absence
- of other anomalies. In cases of large omphaloceles, the bowel and viscera will not fit within the
- abdomen and the sac will be allowed to epithelialize rather than making an attempt at surgical closure

- Surgical repair of **neural tube defects should occur in the first 24 to 48 h of life, to help prevent** infection and preserve neurologic function. Magnetic resonance imaging will also determine the presence or absence of hydrocephalus or Chiari malformation

- Surgical repair for **esophageal atresia/ tracheoesophageal fistula depends on the results of initial** investigations and type of lesion. Although there are 5 types of **EA/ TEF (Figure 6.7), types A and C are the** most common and most easily diagnosed. Type C (EA with distal fistula) is present in 85% of cases and type A (EA without distal fistula) in 8%.






Coiled
feeding
tube

- • ***Duodenal/ jejunal- ileal atresias: These obstructions require abdominal decompression and a laparotomy***
- for intestinal reanastomosis.
- • ***Malrotation/ volvulus: In the presence of an acute abdomen and cardiorespiratory instability, this is***
- a surgical emergency. Emergent consultation and transport are paramount.
- • ***Meconium ileus: If uncomplicated, this condition can be treated with specialized enemas. For a complicated***
- meconium ileus with in utero perforation resulting from the initial obstruction, there are
- two outcomes:
- ◦ When the perforation has sealed and there is no intestinal obstruction, a laparotomy may not be
- necessary.
- ◦ Laparotomy is required when there is an intestinal obstruction, evidence of persistent peritonitis,
- or an abdominal mass.

- • ***NEC: The absolute indications for surgical intervention include pneumoperitoneum and intestinal***
- gangrene. A laparotomy is performed to inspect the entirety of the GI tract. Necrotic or perforated
- segments of the bowel are resected, and an ostomy is performed.
- • ***Imperforate anus: Surgical management includes a colostomy until the rectum and anus can be***
- reconstructed at a later age.
- • ***Hirschsprung's disease: The usual management is a diverting colostomy proximal to the transition***
- zone, with resection of the aganglionic segment of the bowel. The definitive treatment, when the
- ganglionic bowel is brought down and anastomosed with the anal canal, becomes possible when the
- infant weighs more than 10 kg.

- Surgical Conditions: Case 1— Abdominal wall defect
- *You are called to a 37- week GA baby post-delivery. She was born with loops of small bowel extruding from her*
- *abdominal wall. She is breathing regularly in room air and appears otherwise appropriate.*
- You recognize that this is an unwell baby with a visible anomaly and call for assistance as you move
- the baby to the over-bed warmer for better assessment. As help arrives, you position the baby to open
- the airway and apply the monitors as part of the Consolidated Core Steps. Your colleague begins the
- ACoRN Primary Survey.
- *The baby's breathing is regular and effective at a rate of 48 breaths/min. She is pink in room air, with a heart rate of 145*
- *bpm room air. Her blood pressure is 50/ 38 with a mean of 42. Her temp is 36.9°C. She has normal tone and activity.*
- *The abdominal wall defect has no covering, and you identify the need to protect it as part of the Consolidated Core Steps*

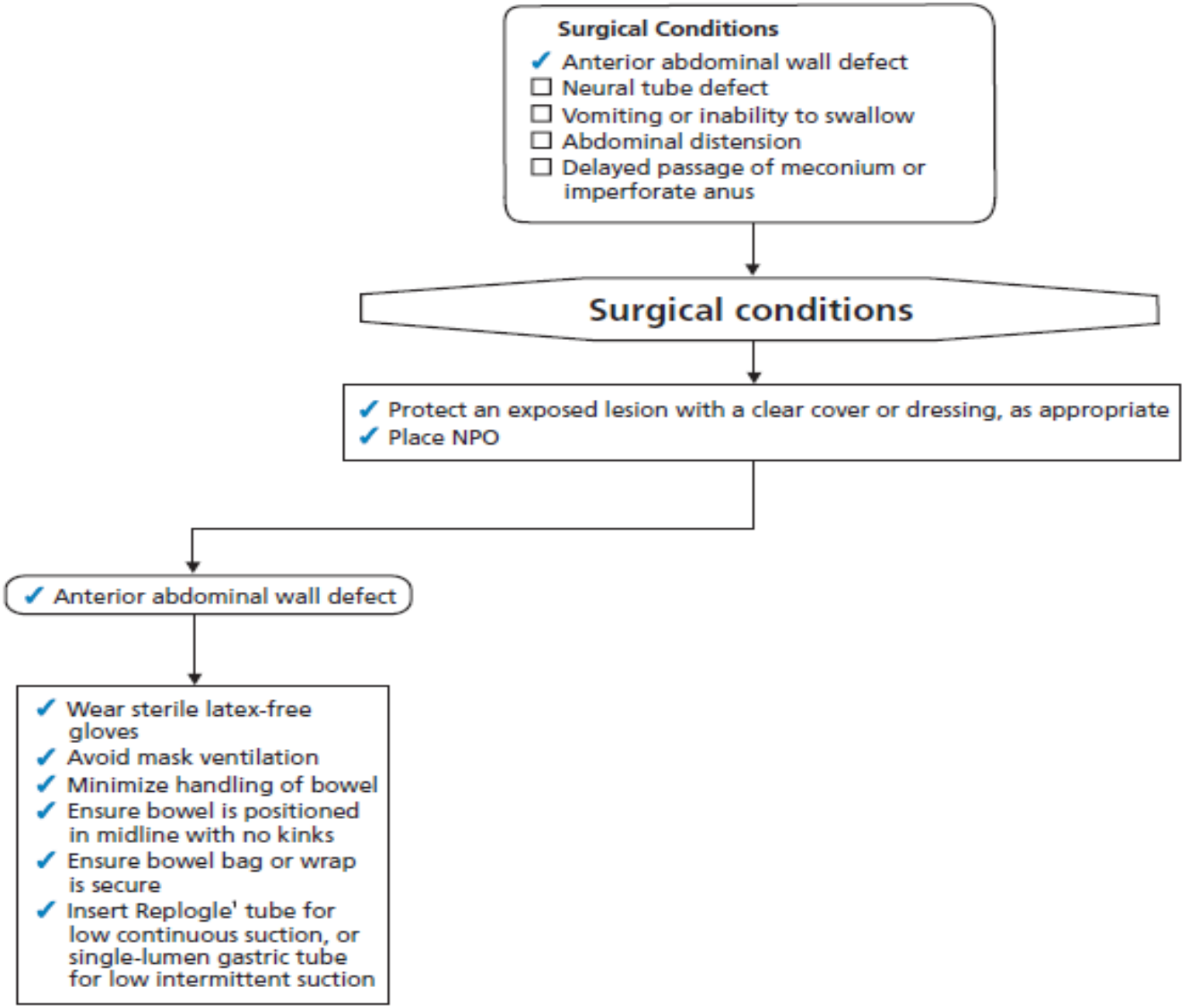
Answer key: Surgical Conditions Case 1— Abdominal wall defect

- **1. How would you protect the exposed bowel during early stage care of this infant?**
- ✓ Use sterile technique.
- ✓ Wear sterile, latex-free gloves.
- ☐ Examine the bowel for areas of stricture or atresia.
- ☐ Let gravity determine the position of the bowel on the bed.
- ☐ Cover the defect with a sterile, dry gauze dressing.
- ✓ Place the baby inside a bowel bag or the bowel inside an occlusive, transparent covering.

What does your Problem List look like?
Problem List

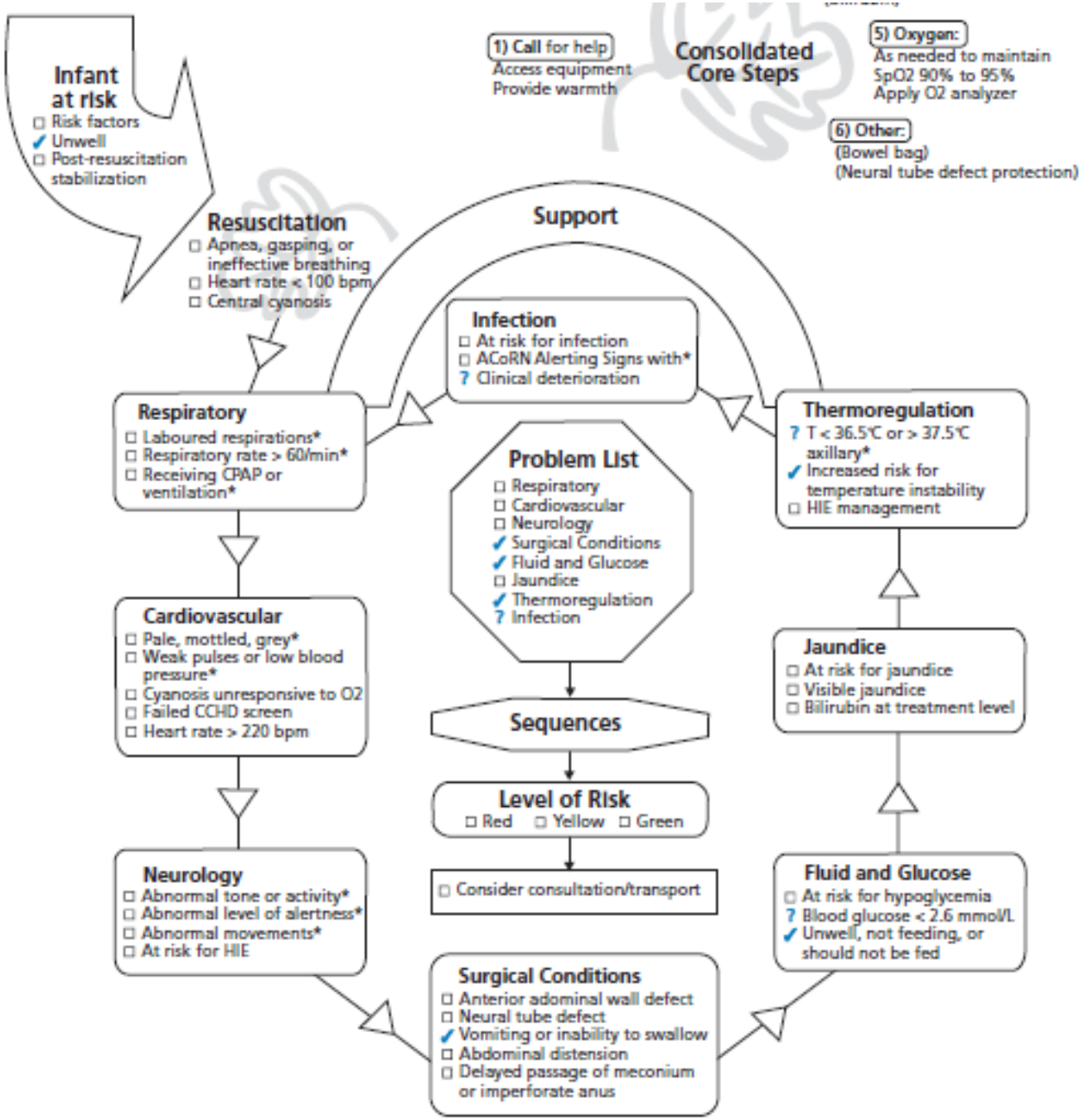
- Respiratory
- Cardiovascular
- Neurology
- Surgical Conditions
- Fluid and Glucose
- Jaundice
- Thermoregulation
- Infection

3. How do you Organize Care for this baby?



4. Based on the Surgical Conditions Sequence, what is this baby's Level of Risk?

✓ Red ☐ Yellow ☐ Green


Infant at risk
☐ Risk factors
✓ Unwell
☐ Post-resuscitation stabilization
Resuscitation
☐ Apnea, gasping, or ineffective breathing
☐ Heart rate < 100 bpm
☐ Central cyanosis
1) Call for help
Access equipment
Provide warmth
Consolidated Core Steps
5) Oxygen:
As needed to maintain SpO2 90% to 95%
Apply O2 analyzer
6) Other:
(Bowel bag)
(Neural tube defect protection)
Support
Infection
☐ At risk for infection
☐ ACoRN Alerting Signs with*
? Clinical deterioration
Respiratory
☐ Laboured respirations*
☐ Respiratory rate > 60/min*
☐ Receiving CPAP or ventilation*
Problem List
☐ Respiratory
☐ Cardiovascular
☐ Neurology
✓ Surgical Conditions
✓ Fluid and Glucose
☐ Jaundice
✓ Thermoregulation
? Infection
Thermoregulation
? T < 36.5°C or > 37.5°C axillary*
✓ Increased risk for temperature instability
☐ HIE management
Cardiovascular
☐ Pale, mottled, grey*
☐ Weak pulses or low blood pressure*
☐ Cyanosis unresponsive to O2
☐ Failed CCHD screen
☐ Heart rate > 220 bpm
Sequences
Jaundice
☐ At risk for jaundice
☐ Visible jaundice
☐ Bilirubin at treatment level
Level of Risk
☐ Red ☐ Yellow ☐ Green
☐ Consider consultation/transport
Neurology
☐ Abnormal tone or activity*
☐ Abnormal level of alertness*
☐ Abnormal movements*
☐ At risk for HIE
Fluid and Glucose
☐ At risk for hypoglycemia
? Blood glucose < 2.6 mmol/L
✓ Unwell, not feeding, or should not be fed
Surgical Conditions
☐ Anterior adominal wall defect
☐ Neural tube defect
✓ Vomiting or inability to swallow
☐ Abdominal distension
☐ Delayed passage of meconium or imperforate anus