PEDIATRIC SURGERY INDEX CASES

CASE 1:
A 2700 gram term male infant is born to a 34 yr. old mother whose pregnancy was complicated by polyhydramnios. Uneventful vaginal delivery. Apgars are 8 at 1 minute and 9 at 5 minutes. As the infant is transported to the nursery, excessive salivation is noted.

PEDIATRIC SURGERY INDEX CASES

A nasogastric tube is inserted but meets an obstruction at 8 cm. beyond the naris. A “babygram” is obtained. He is begun on supplemental oxygen by nasal cannula and a pulse oximeter is placed. Oxygen saturation remains between 92-98% as long as he is suctioned frequently.

PEDIATRIC SURGERY INDEX CASES

On physical examination, he is a vigorous male infant. He is breathing easily without cyanosis. There are no anomalies of the head, neck, trunk, or extremities. Copious secretions are suctioned from his mouth. Breath sounds are audible bilaterally. No cardiac murmur is evident. The abdomen is soft without distention. No masses are palpable. The genitalia are normal and a drop of meconium is visible at the anus.

PEDIATRIC SURGERY INDEX CASES

- **ESOPHAGEAL ATRESIA + TRACHEOESOPHAGEAL FISTULA**
  - Embryology
    - Esophagus + trachea recognized as ventral diverticulum at 22-23 days of gestation
    - Separation of ventral trachea from dorsal foregut occurs first at carina and extends cephalad
    - Division into separate tubes complete at 34-36 days of gestation

PEDIATRIC SURGERY INDEX CASES

- **ESOPHAGEAL ATRESIA + TRACHEOESOPHAGEAL FISTULA**
  - Spectrum of anomalies:
    - C Blind upper pouch + distal TEF 85%
    - A Isolated esophageal atresia (no fistula) 8%
    - E H-type fistula (no atresia) 4%
    - EA + fistula to upper + lower pouches 1%
    - EA + proximal TEF <1%

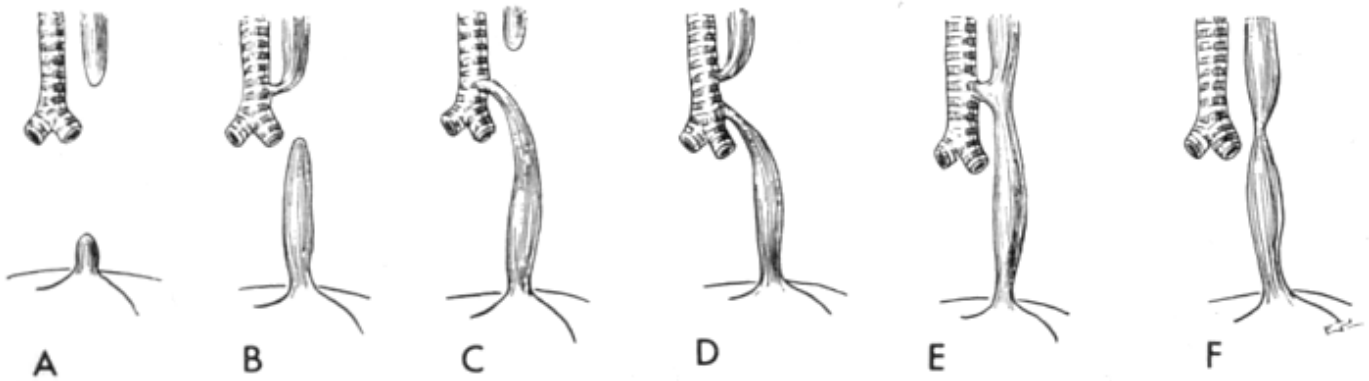
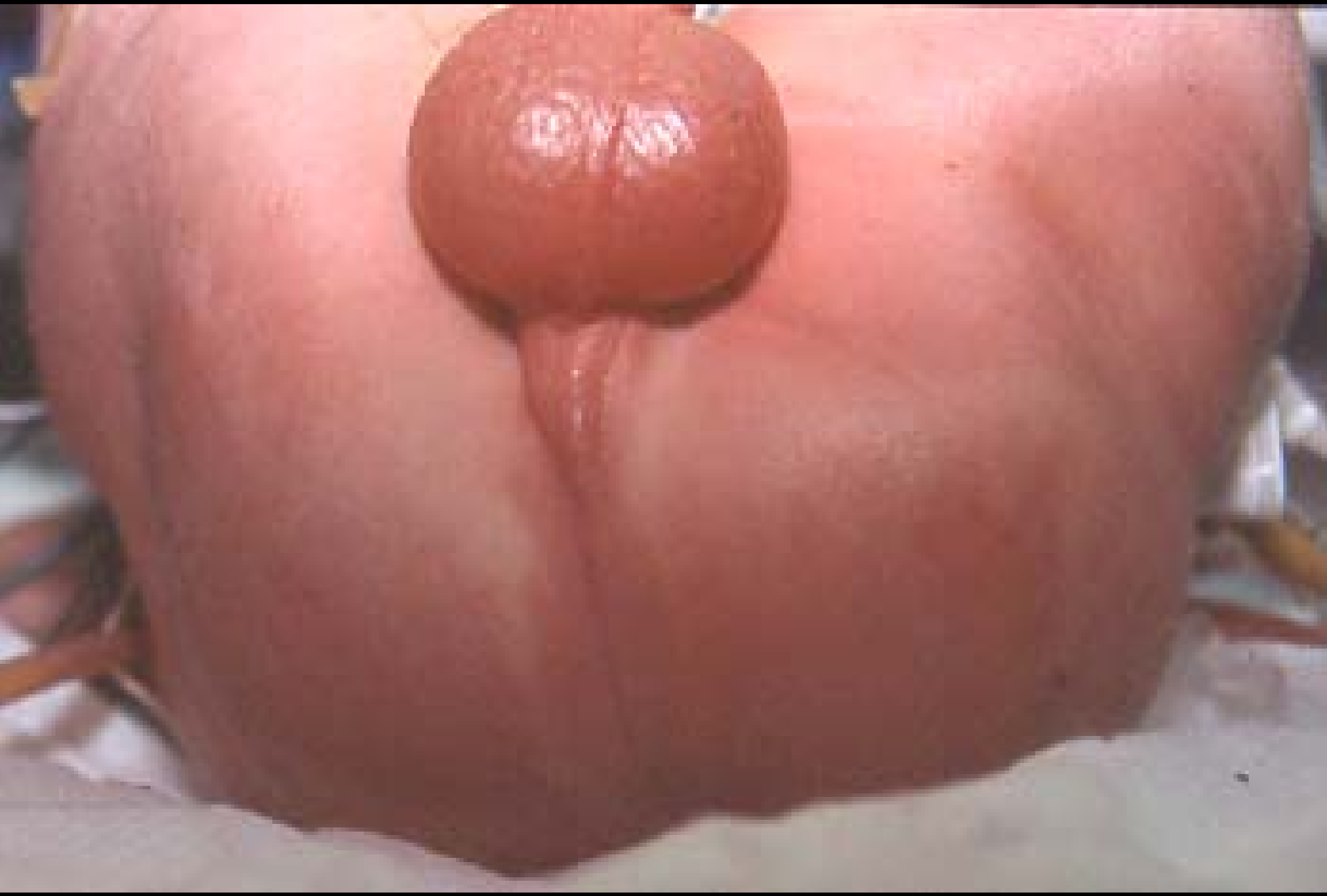
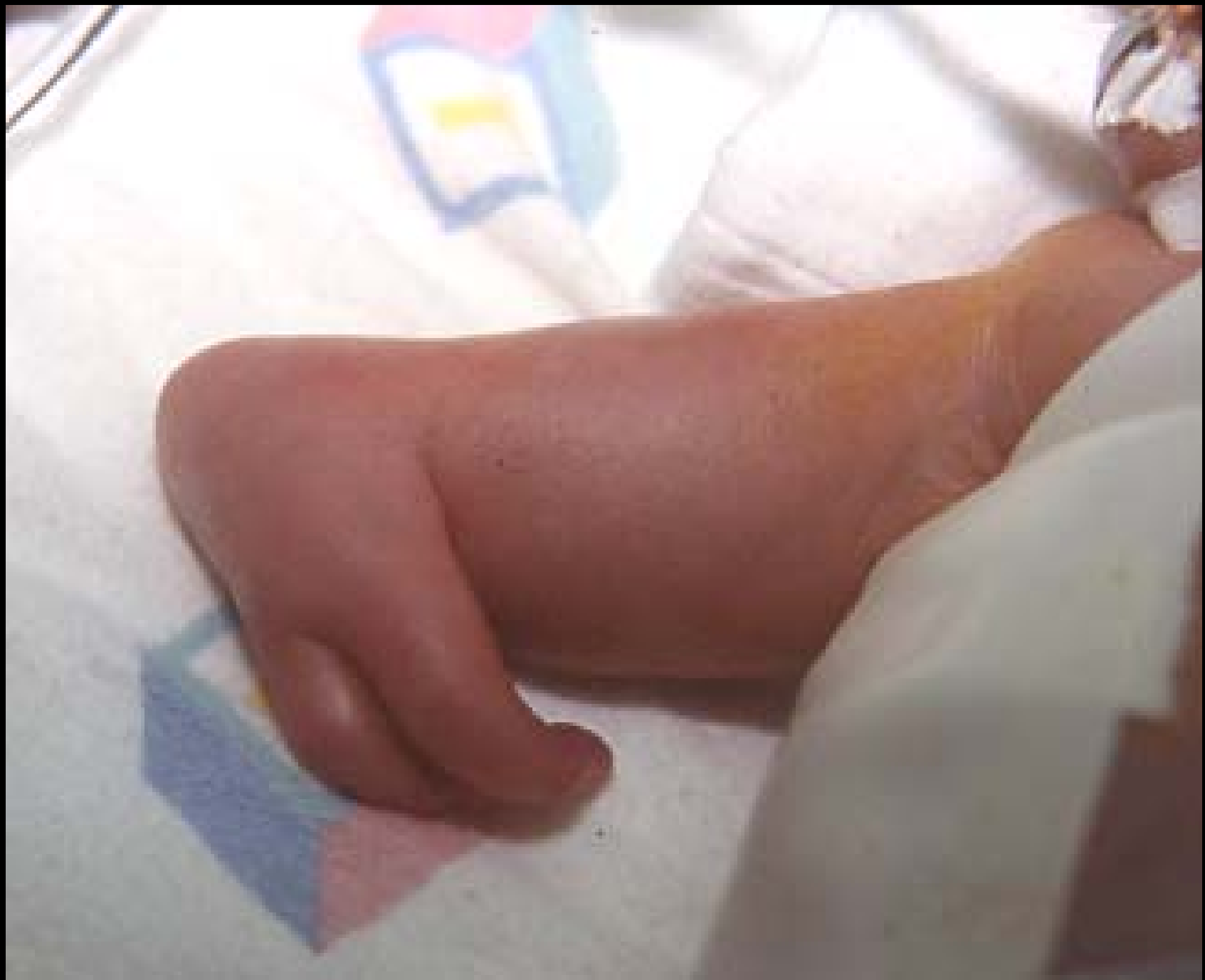


Figure 25–2. Classic diagram showing types of esophageal and related tracheal anomalies (originally described by Vogt and used by Gross). (From Gross, R. E.: The Surgery of Infancy and Childhood. Philadelphia, W. B. Saunders Company, 1953, p. 76.)

PEDIATRIC SURGERY INDEX CASES

- **ESOPHAGEAL ATRESIA + TRACHEOESOPHAGEAL FISTULA**
  - Associated anomalies
    - VACTERL association
    - Overall occurrence of associated anomalies is 50-70%
      - Cardiovascular anomalies in 29-35%
      - Imperforate anus in 24%





PEDIATRIC SURGERY INDEX CASES

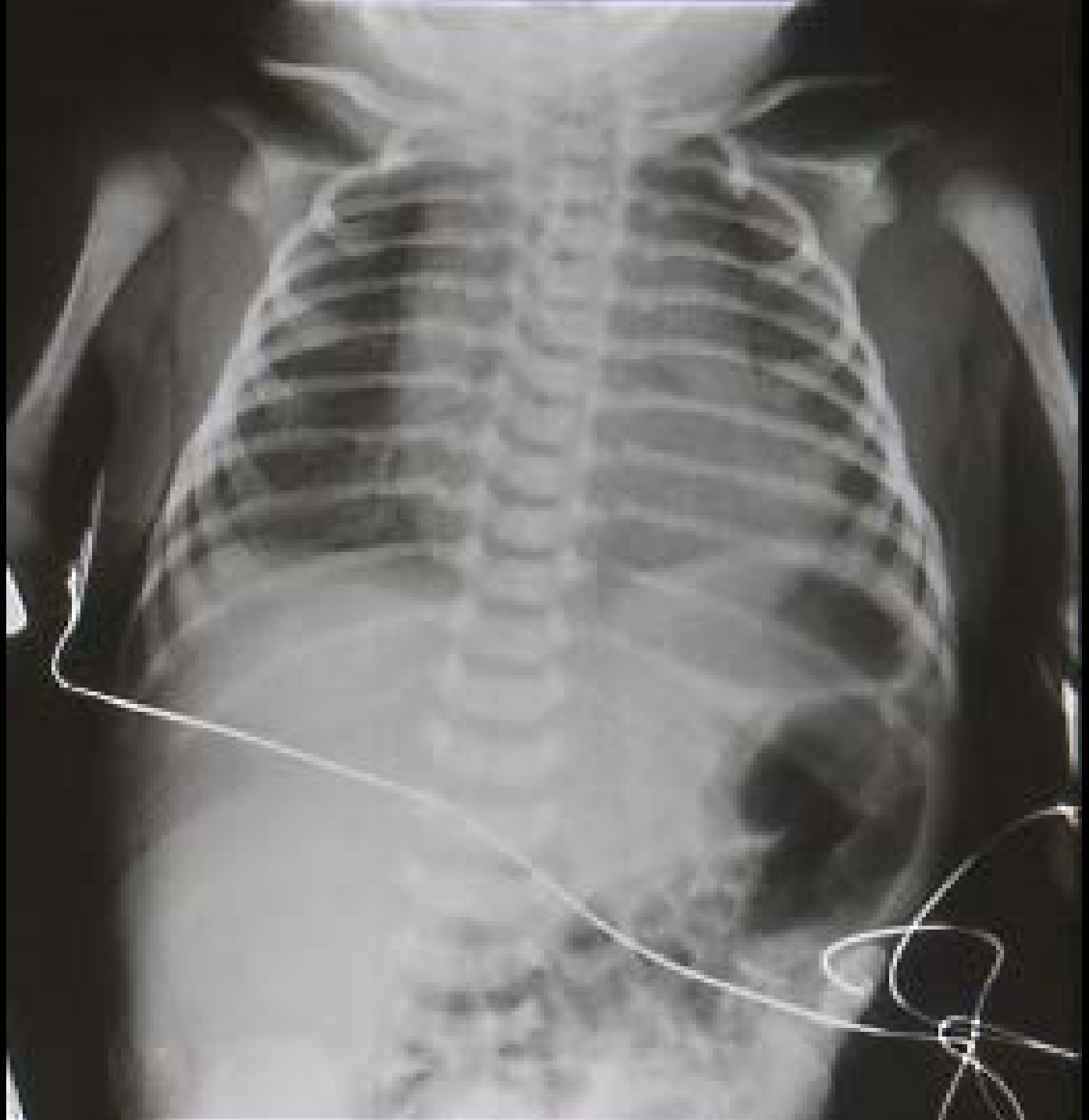
- **ESOPHAGEAL ATRESIA + TRACHEOESOPHAGEAL FISTULA**
  - Incidence: 1/3000 live births
  - Polyhydramnios noted in 32% of infants with EA+TEF; in 85% of infants with isolated EA
  - One of the more common correctable, lethal congenital anomalies.

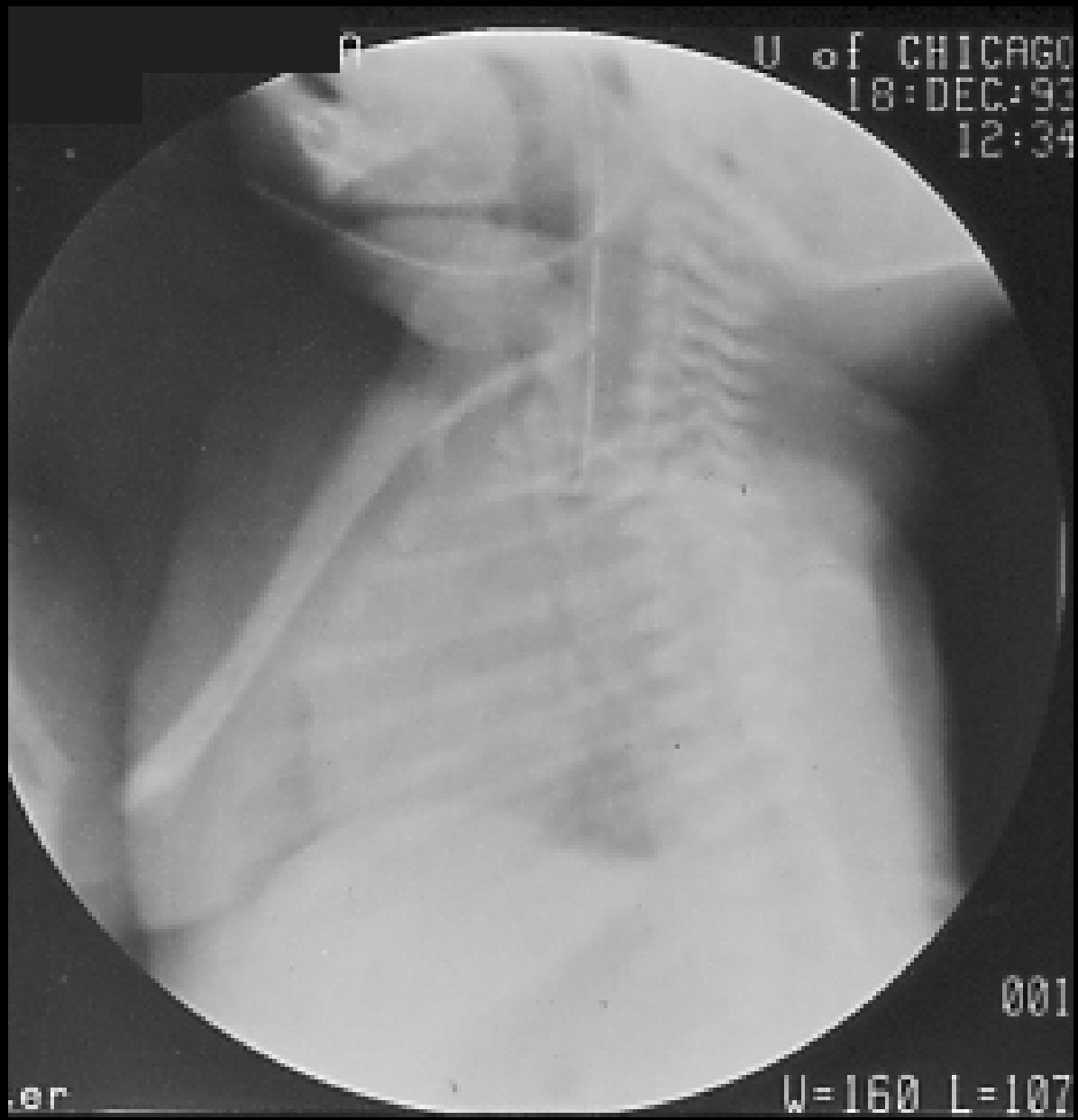
PEDIATRIC SURGERY INDEX CASES

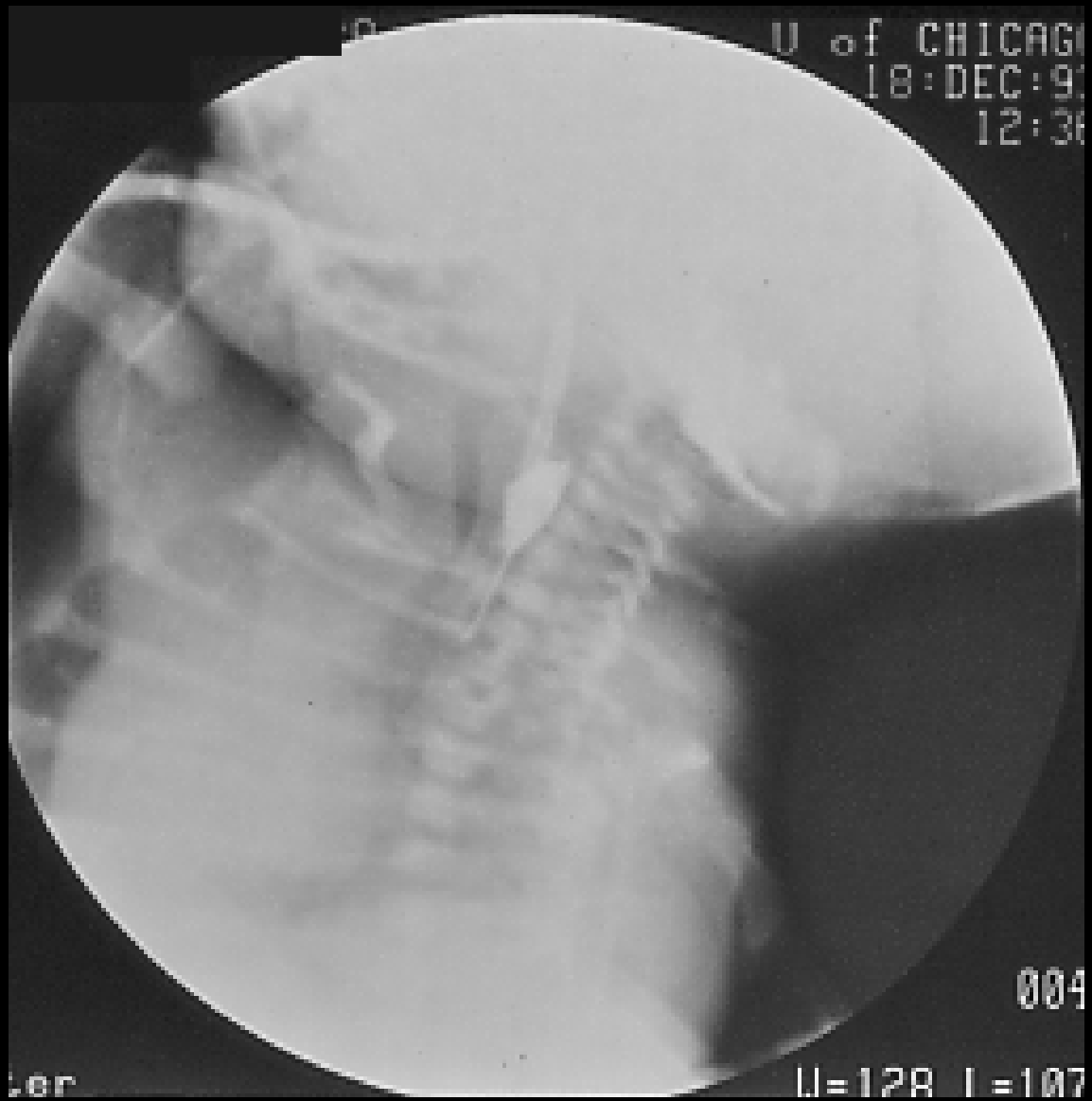
- **ESOPHAGEAL ATRESIA + TRACHEOESOPHAGEAL FISTULA**
  - Presentation
    - Excessive salivation
    - Choking
    - Respiratory distress
    - Cyanosis
    - Pneumonia

PEDIATRIC SURGERY INDEX CASES

- **ESOPHAGEAL ATRESIA + TRACHEOESOPHAGEAL FISTULA**
  - Diagnosis + determination of associated anomalies
    - Pass NG tube
    - Babygram
    - Cardiac ECHO
    - Renal ultrasound
    - Pouchogram

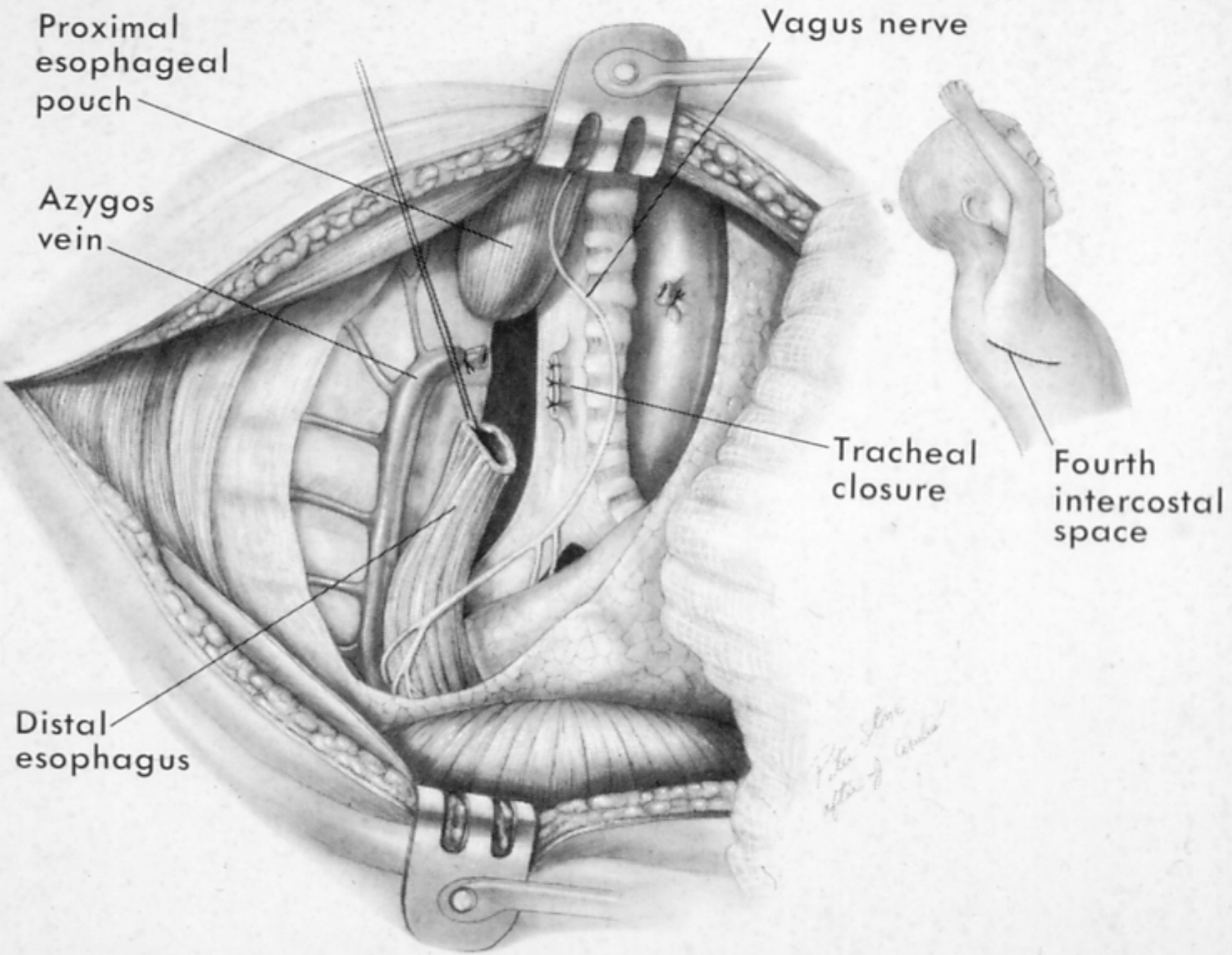


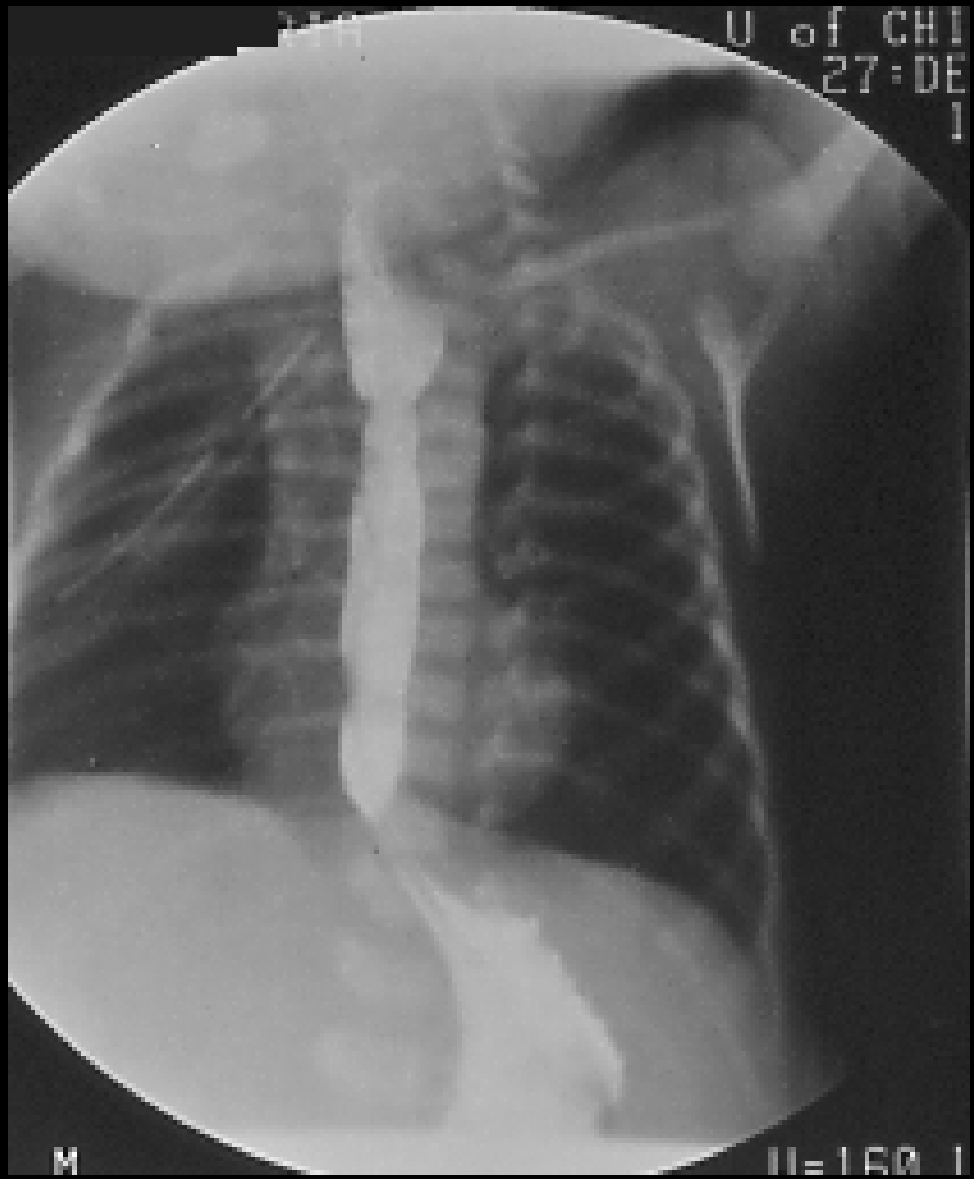

U of CHICAGO
18:DEC:93
12:34
001
W=160 L=107


U of CHICAG
18:DEC:9
12:3
004
U=128 I=107

PEDIATRIC SURGERY INDEX CASES

- **ESOPHAGEAL ATRESIA + TRACHEOESOPHAGEAL FISTULA**
  - Management
    - Division of fistula + esophagoesophagostomy
  - Complications
    - Anastomotic leak
    - Stricture
    - Recurrent TEF


Proximal esophageal pouch
Vagus nerve
Azygos vein
Tracheal closure
Fourth intercostal space
Distal esophagus


U of CHI
27-DE
1
M

PEDIATRIC SURGERY INDEX CASES

CASE 2:

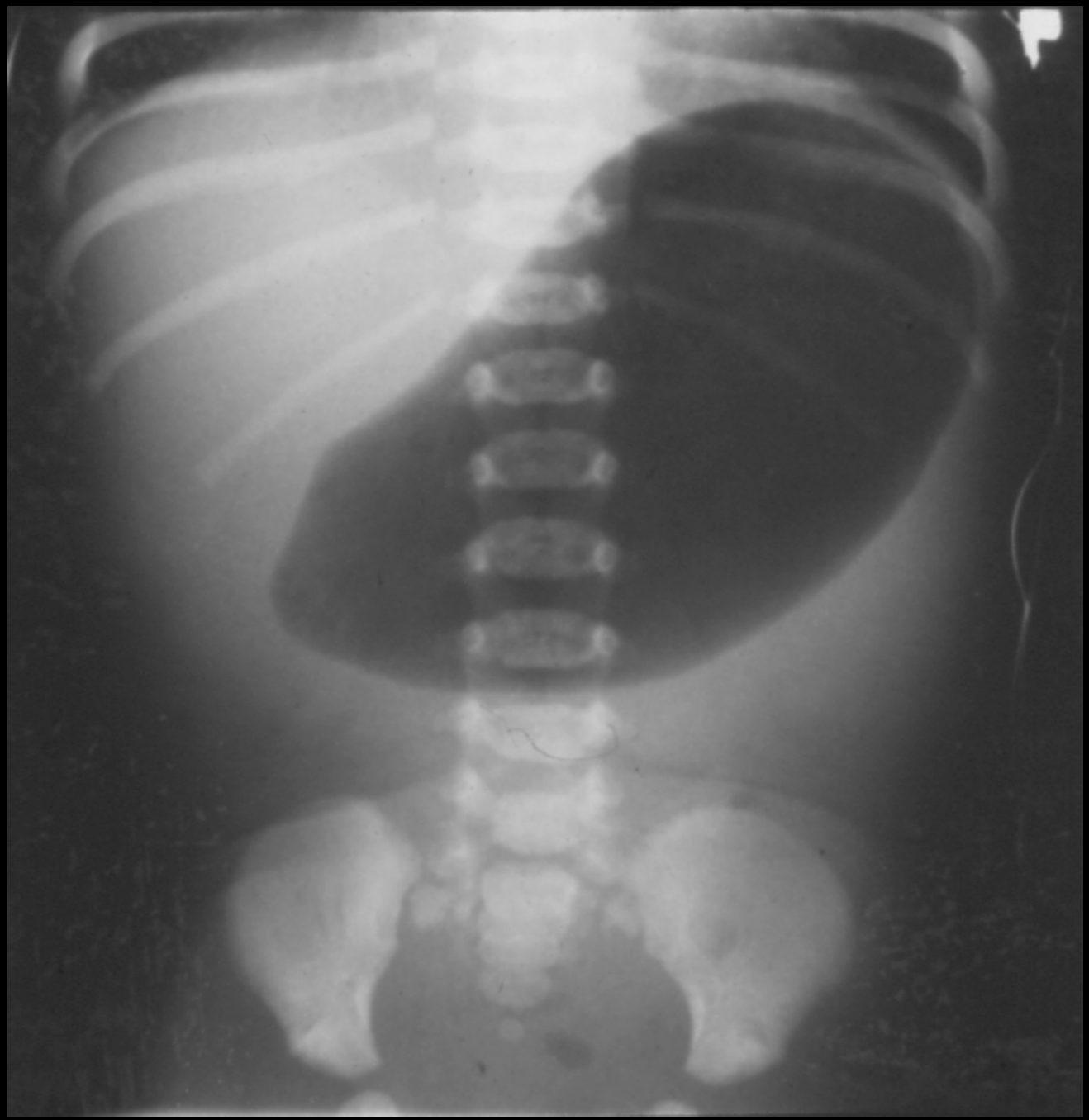
You are working in an emergency room. A mother calls and says that her 5-week old baby is vomiting. He was a term infant without perinatal problems. He is breast-fed. The vomiting began 2 days ago and is now occurring after every feeding.

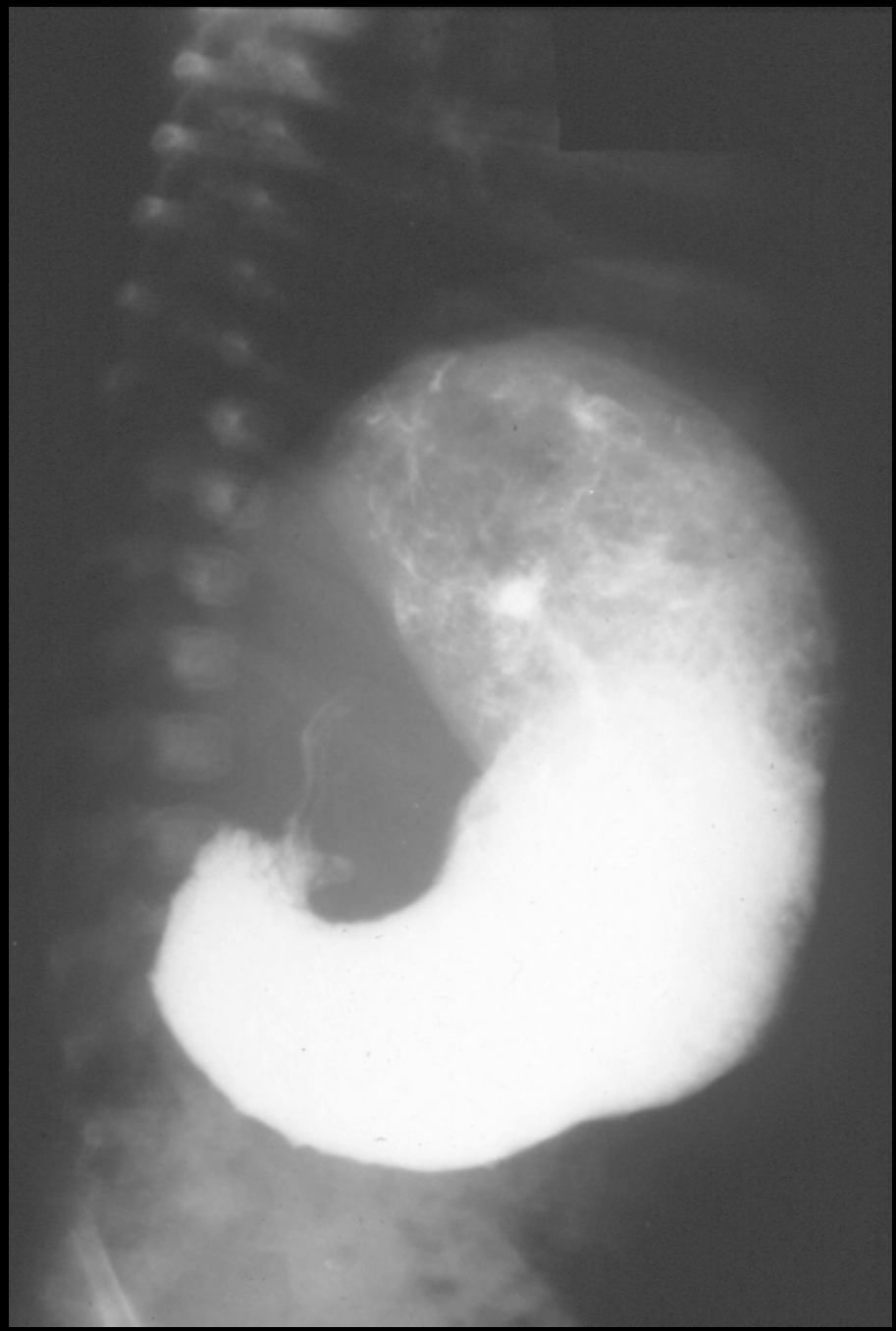
PEDIATRIC SURGERY INDEX CASES

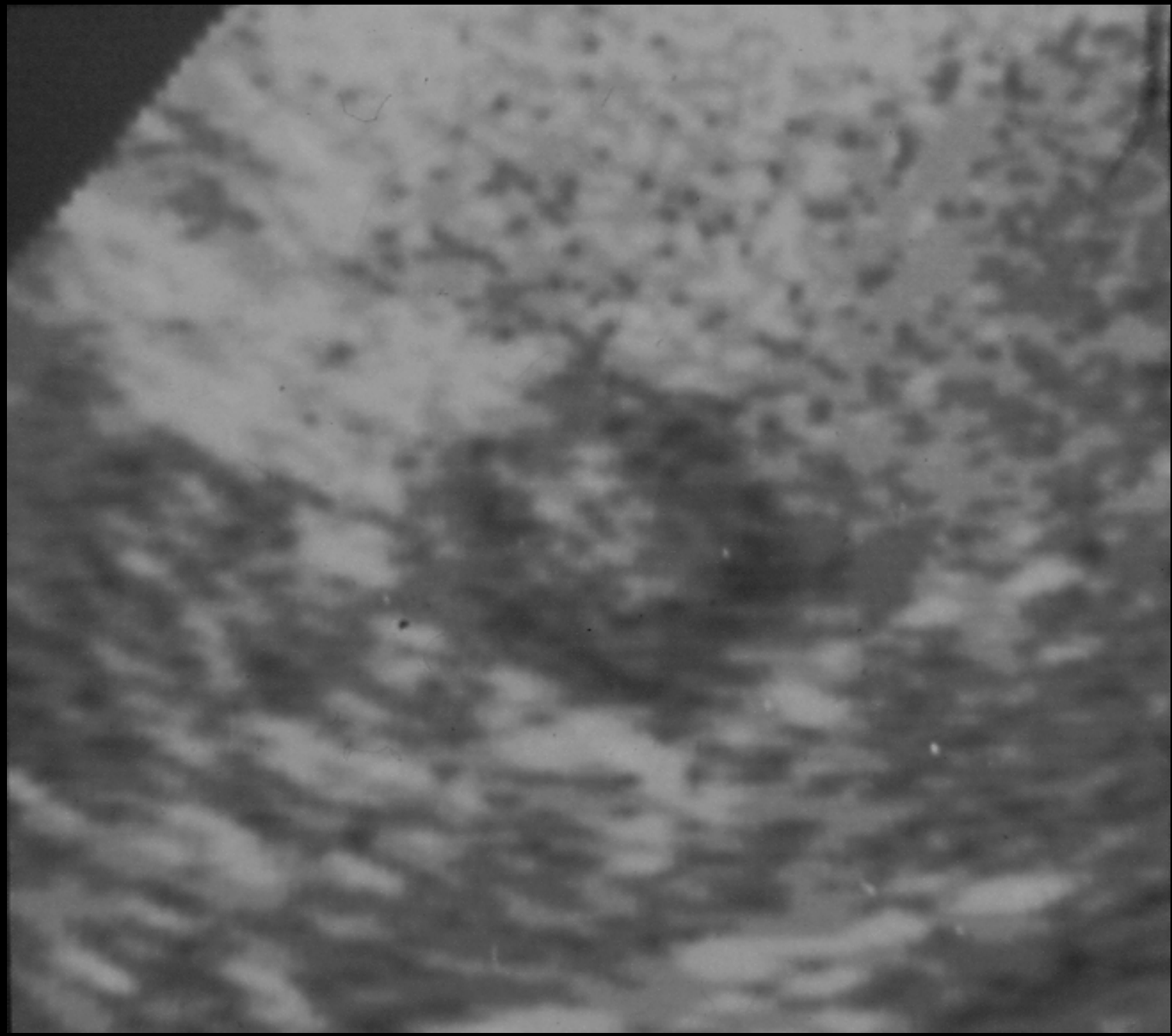
- **HYPERTROPHIC PYLORIC STENOSIS**
  - Presentation
    - Non-bilious emesis
      - Differential diagnosis of non-bilious emesis:
        - Gastroesophageal reflux
        - Pyloric stenosis
        - Antral web
        - Increased intracranial pressure

PEDIATRIC SURGERY INDEX CASES

- **HYPERTROPHIC PYLORIC STENOSIS**
  - Diagnosis
    - Palpable “olive”
    - Ultrasound
    - UGI

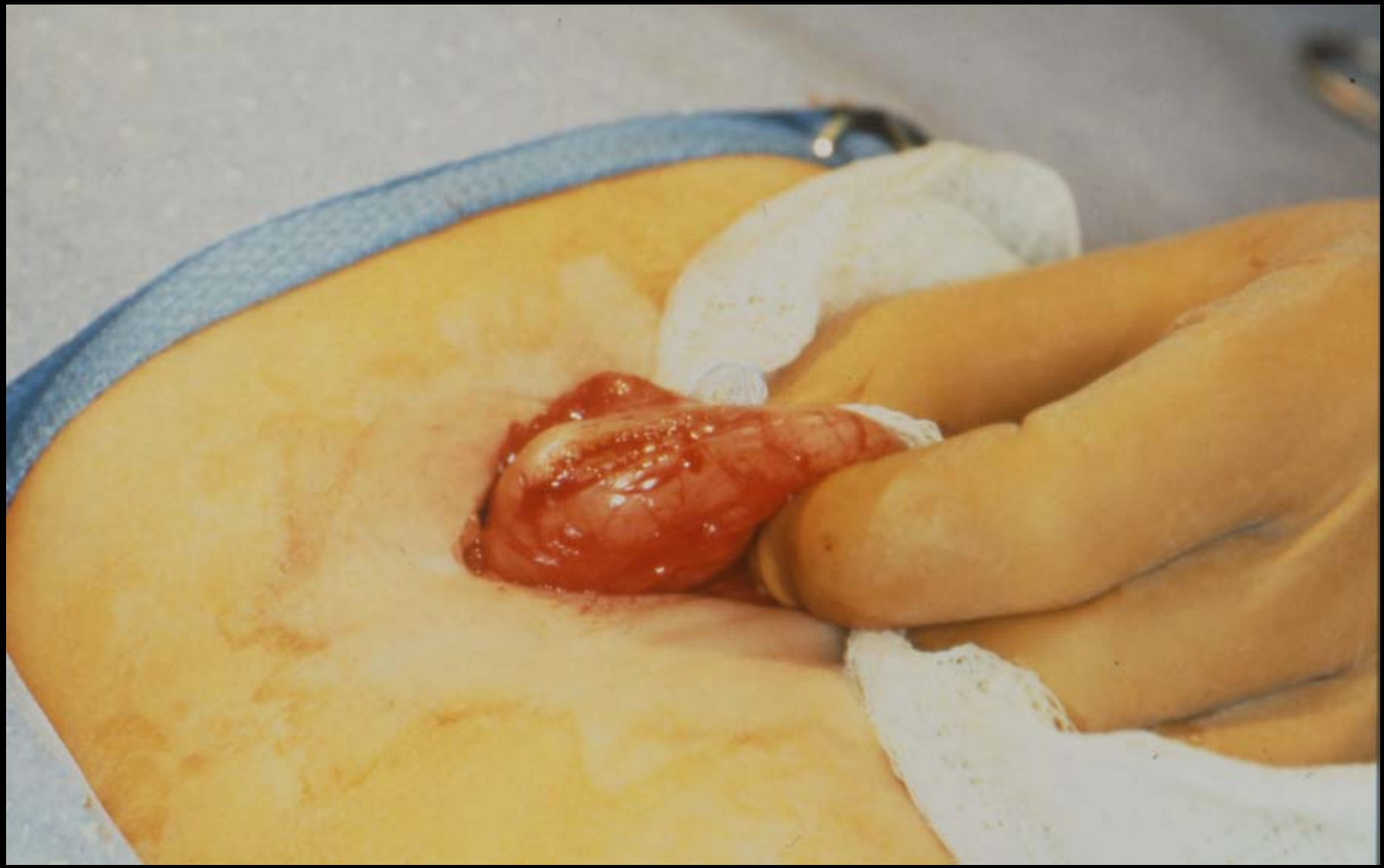






PEDIATRIC SURGERY INDEX CASES

- **HYPERTROPHIC PYLORIC STENOSIS**
  - Pre-operative management
    - IV fluid resuscitation
    - Correction of electrolytes
      - Hypochloremic, hypokalemic metabolic alkalosis
  - Treatment
    - Pyloromyotomy



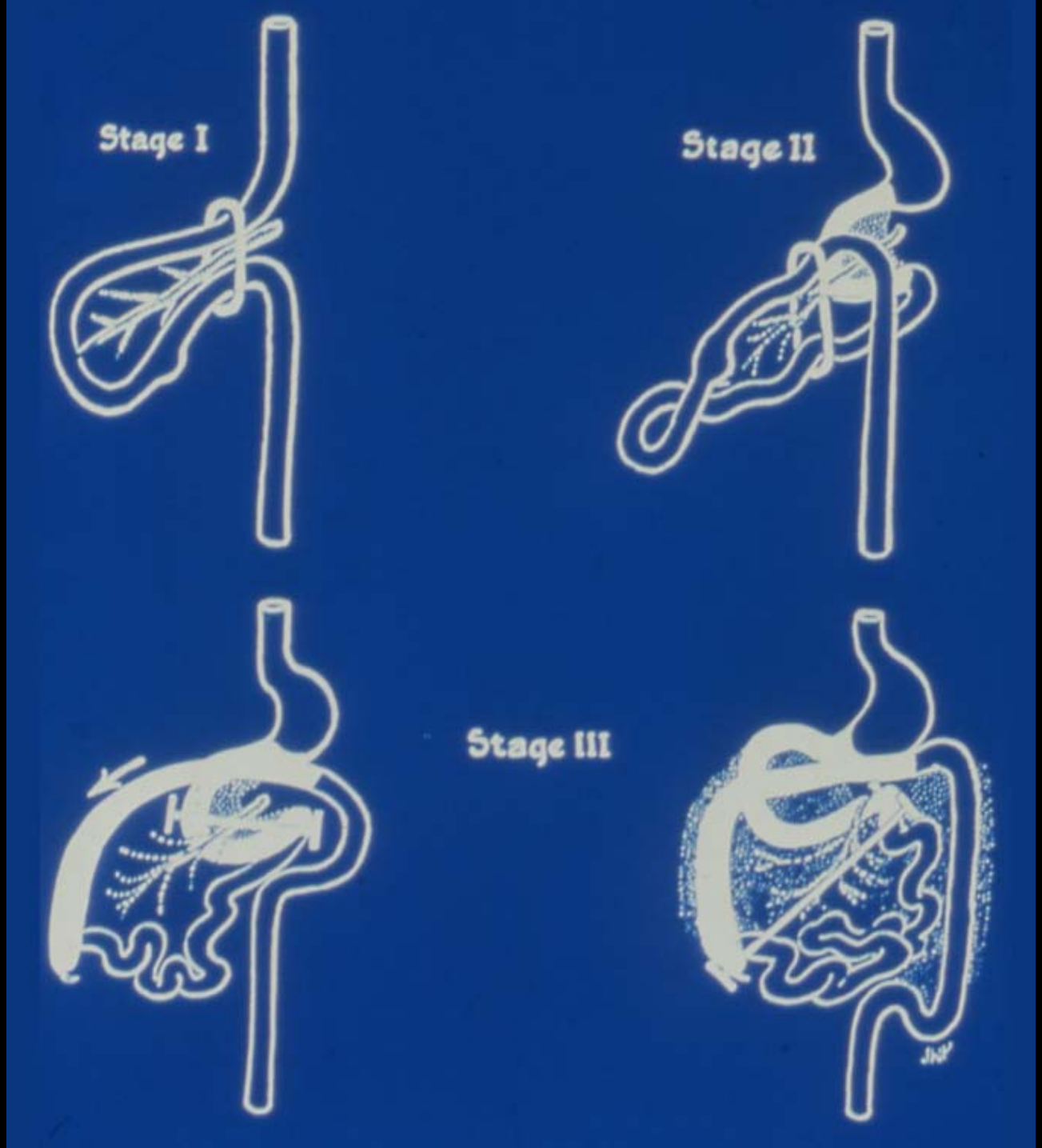
PEDIATRIC SURGERY INDEX CASES

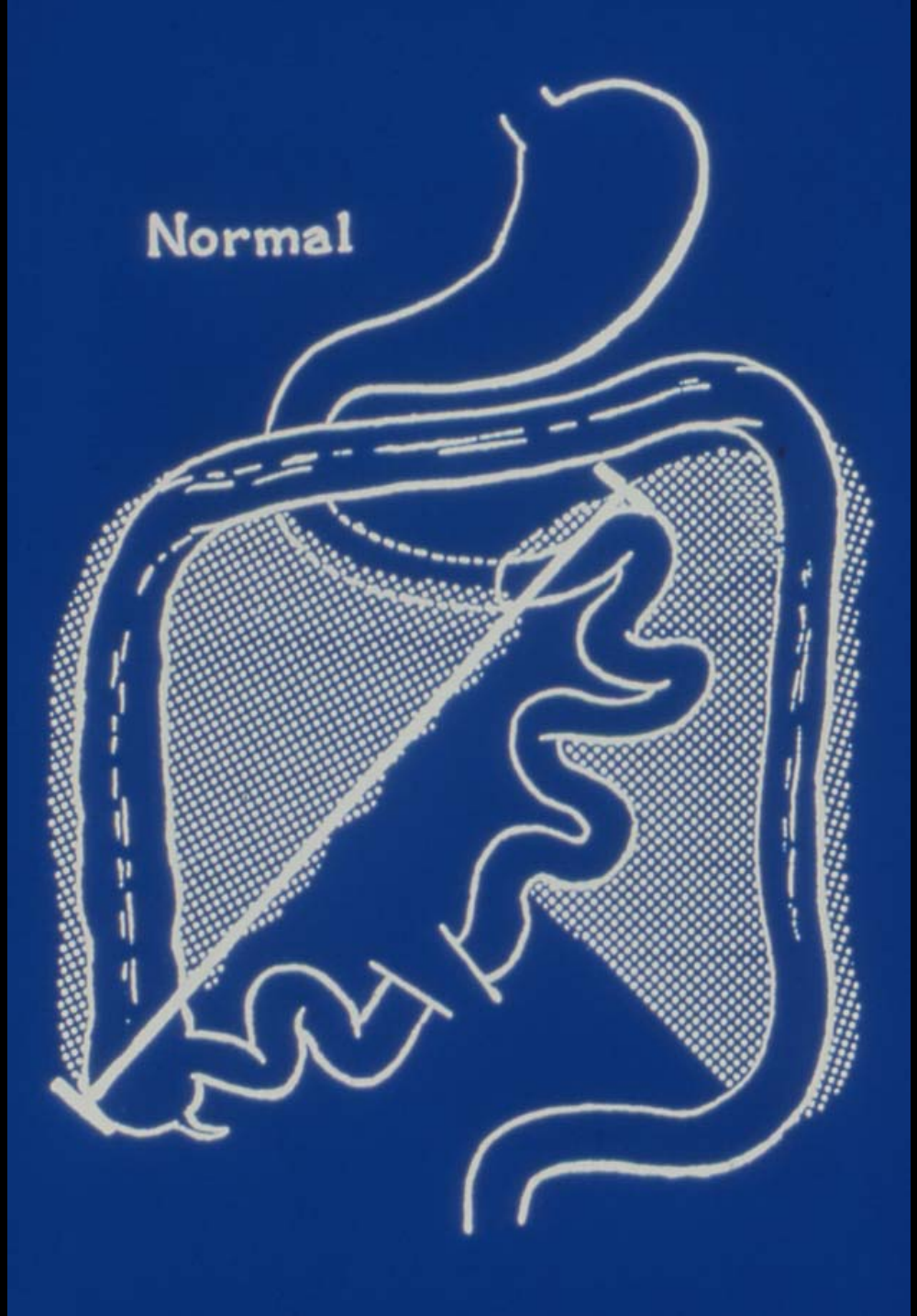
CASE 3:

A healthy, 1-week old, term infant presents with a 1-day history of bilious emesis, decreased urine output. Physical examination reveals delayed capillary refill. The lungs are clear bilaterally; no cardiac murmur is evident. The abdomen is non-distended and non-tender.

PEDIATRIC SURGERY INDEX CASES

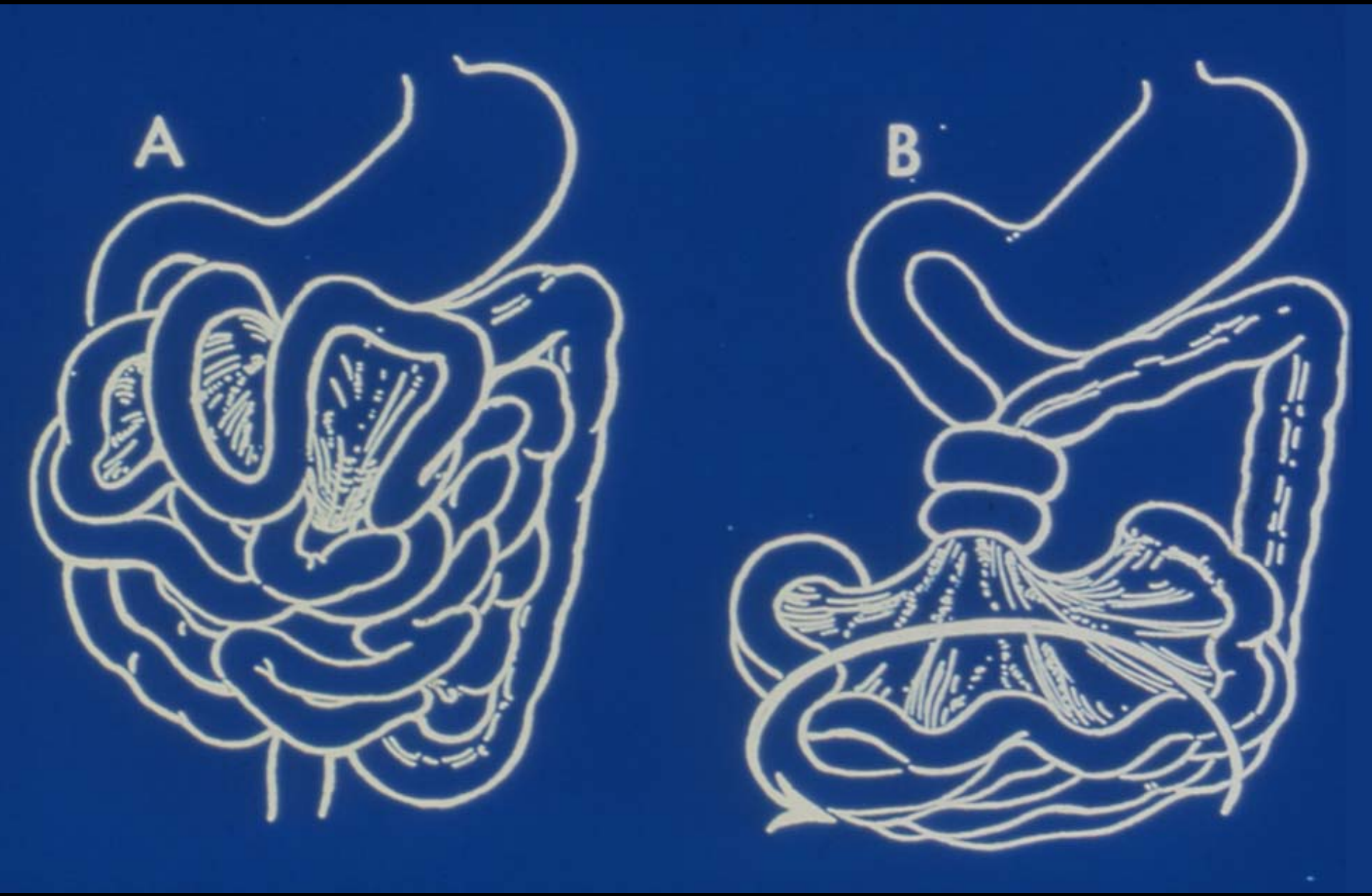
- **MALROTATION**
  - Embryology
    - Rapid elongation of midgut (occurs faster than elongation of embryo)
    - Midgut herniates into umbilical cord (extraembryonic coelom) at 6 weeks
    - During 10^{th} week return of midgut into the abdomen
      - Duodenojejunal limb + cecocolic limb each go through 270^{O} counterclockwise rotation
    - Normal mesentery extends from ligament of Treitz to ileocecal junction


Stage I
Stage II
Stage III


Normal

PEDIATRIC SURGERY INDEX CASES

- **MALROTATION**
  - Results in:
    - Duodenal obstruction
    - Volvulus
    - Internal hernia
    - Failure to thrive

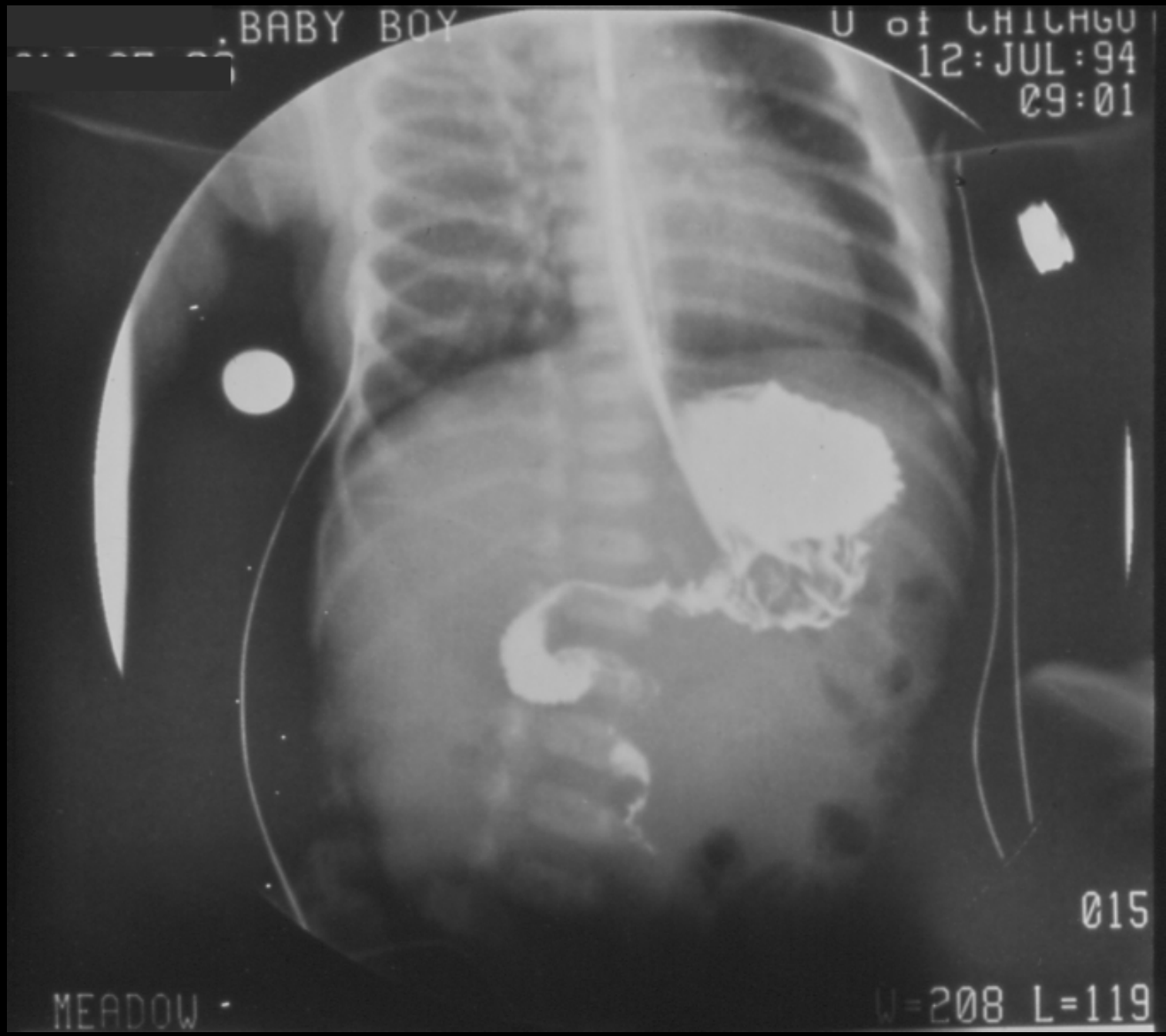

A
B

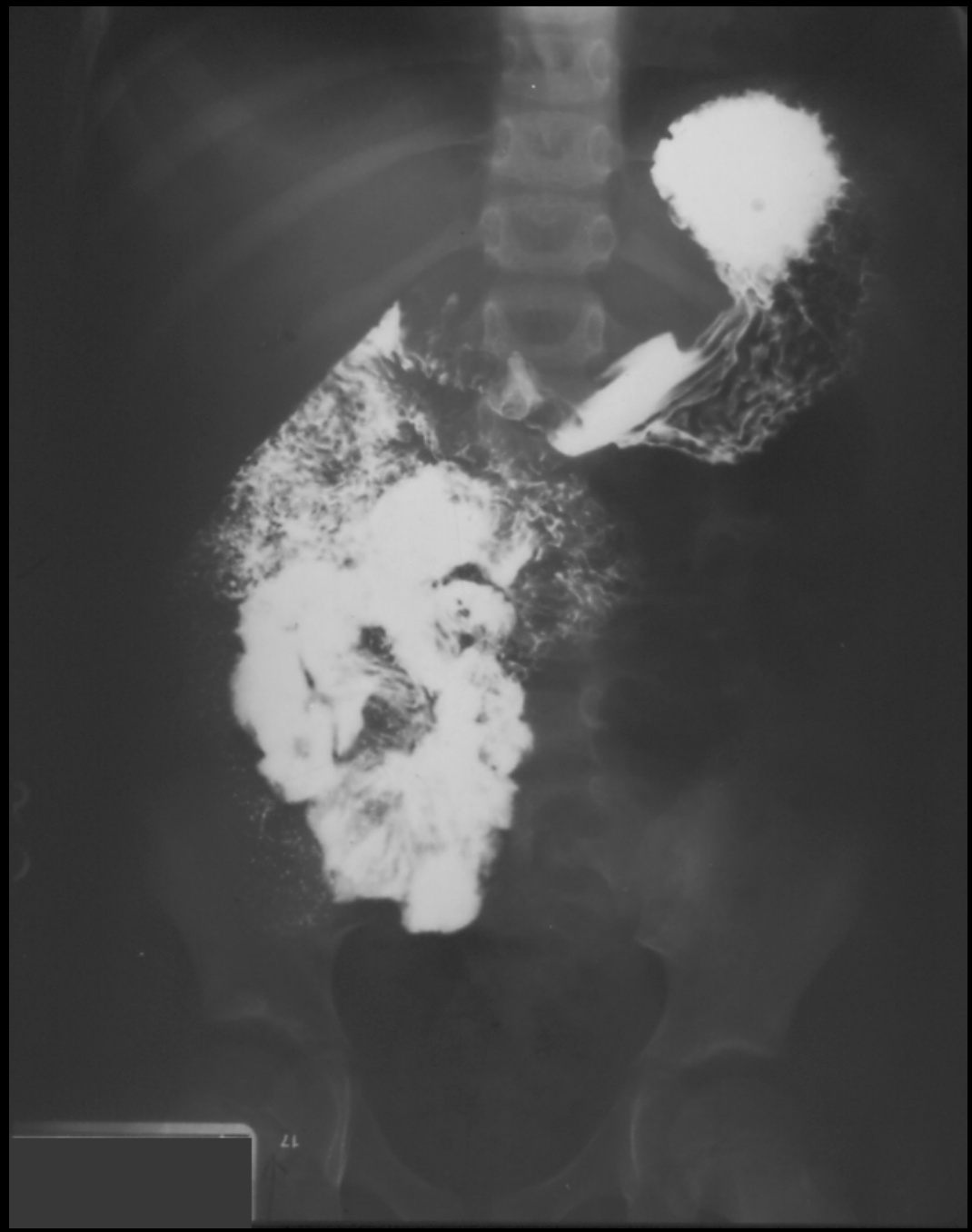
PEDIATRIC SURGERY INDEX CASES

- **MALROTATION**
  - Presentation
    - Bilious emesis indicates intestinal obstruction.
    - The diagnosis is malrotation with midgut volvulus until proven otherwise.
    - Thirty percent of patients with malrotation present within 1st week of life, 50% present within the 1st month, and 90% within the 1st year.

PEDIATRIC SURGERY INDEX CASES

- **MALROTATION**
  - Diagnosis
    - UGI
      - Localization of duodenojejunal flexure (ligament of Treitz) to left of spine
      - Duodenal obstruction
      - Conical termination
    - Doppler ultrasound to determine relationship of superior mesenteric vessels.


BABY BOY
U of CHICAGO
12:JUL:94
09:01
015
MEADOW
W=208 L=119

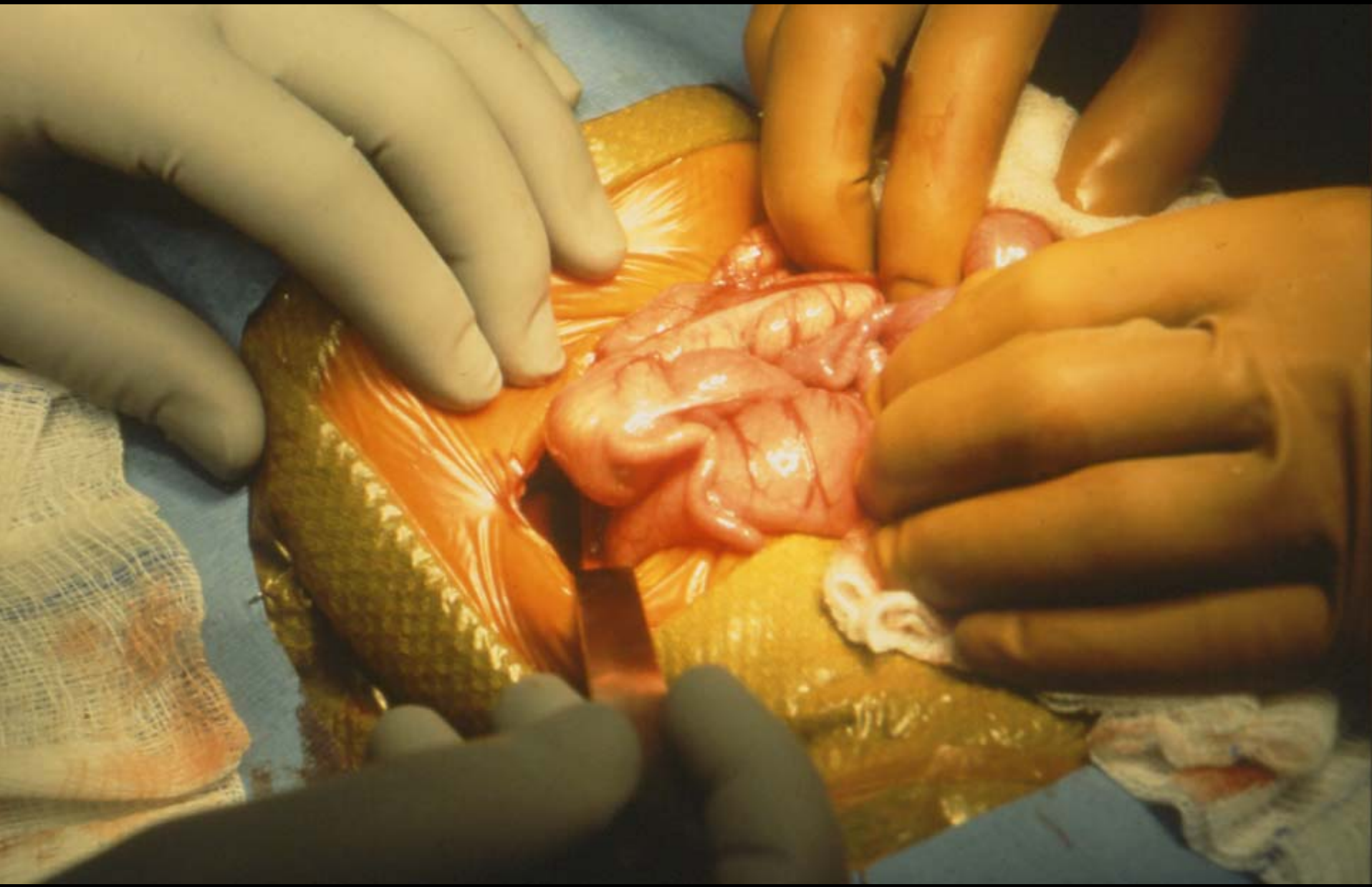


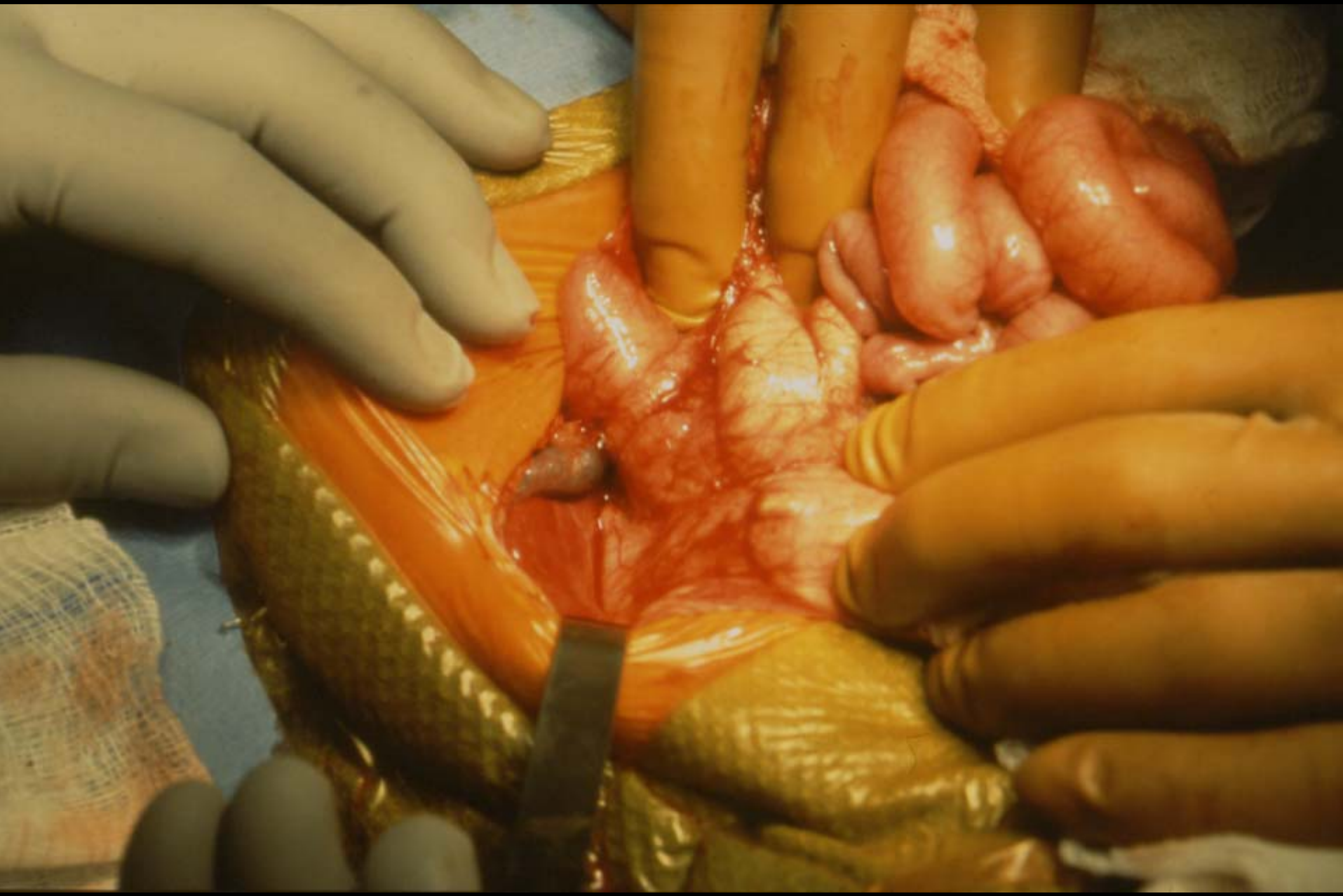
PEDIATRIC SURGERY INDEX CASES

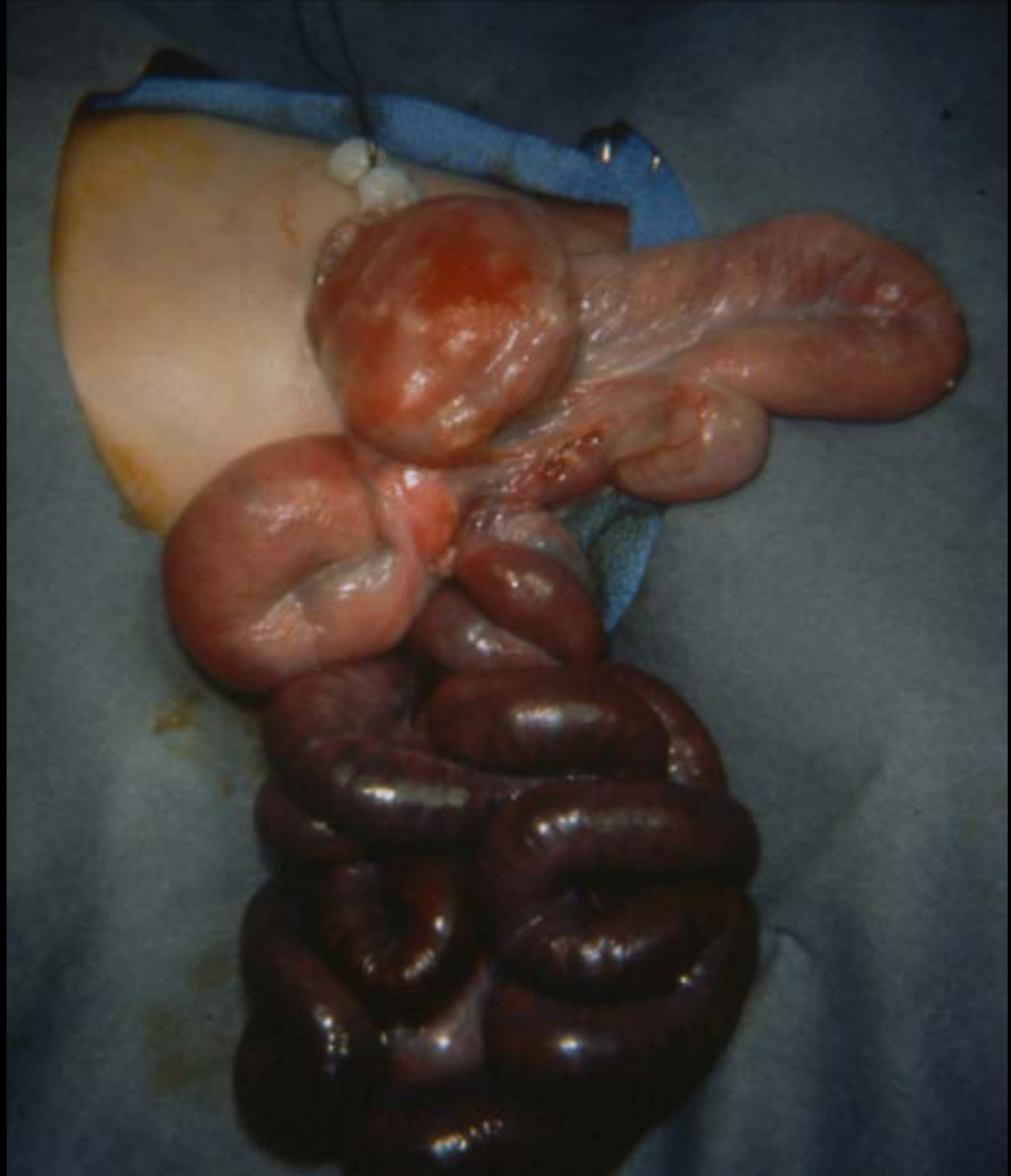
- **MALROTATION**
  - Associated anomalies
    - Congenital diaphragmatic hernia
    - Gastroschisis
    - Omphalocele
    - Duodenal atresia
    - Prune-belly syndrome
    - Heterotaxia syndrome

PEDIATRIC SURGERY INDEX CASES

- **MALROTATION**
  - Treatment
    - Immediate laparotomy
    - Midgut volvulus evident by clockwise twist
      - Untwist bowel in counterclockwise rotation
    - Ladd's procedure with appendectomy







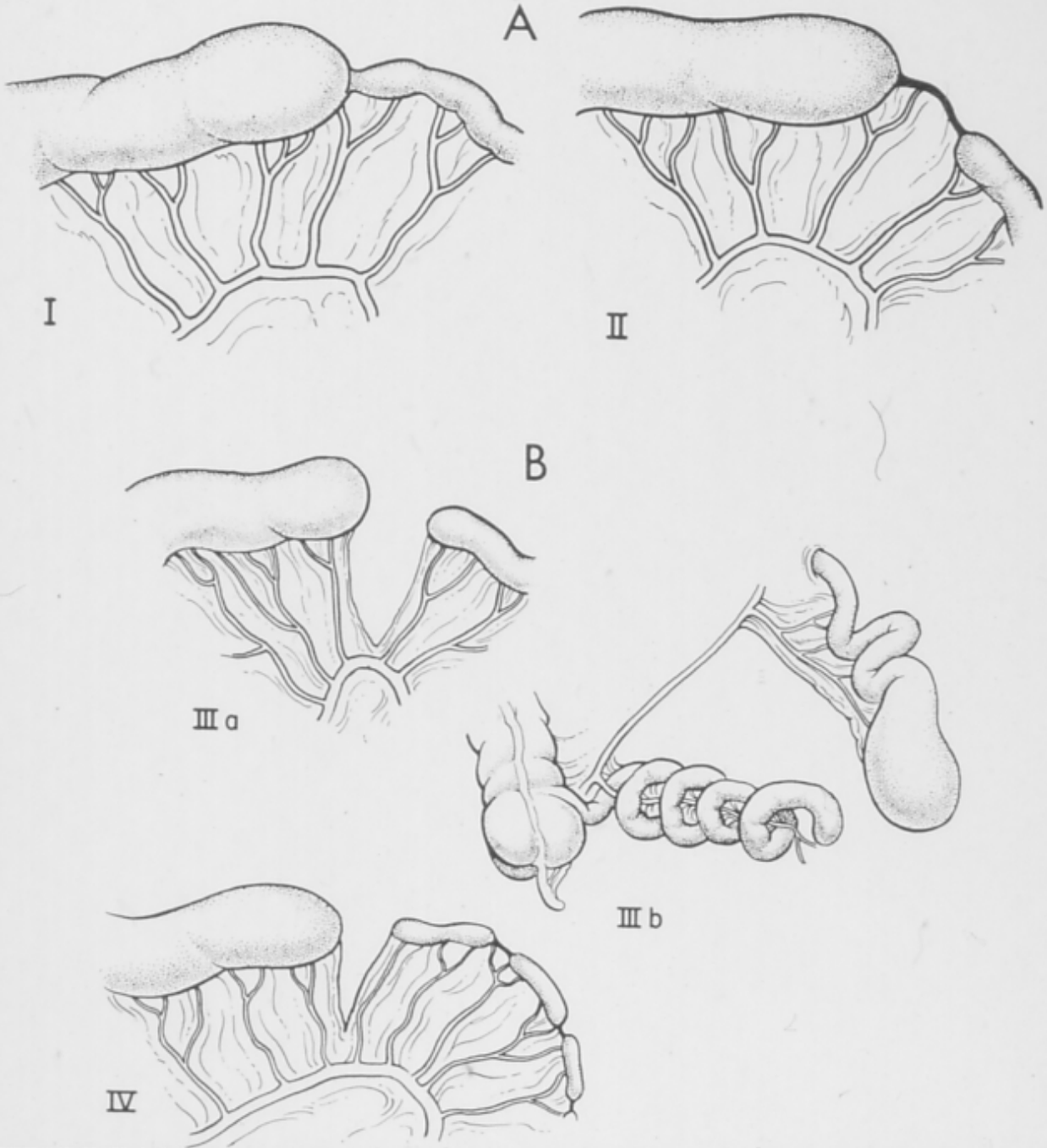
PEDIATRIC SURGERY INDEX CASES

CASE 4:

A neonatologist calls you to evaluate a 36-hour old term male infant with a 12-hour history of bilious emesis after feeding attempts. He has not passed meconium. On physical examination he is distended; the anus is perforate and a nasogastric tube is draining bilious fluid.

PEDIATRIC SURGERY INDEX CASES

- **JEJUNOILEAL ATRESIA**
  - Embryology
    - Intrauterine mesenteric vascular catastrophe
  - Classification
    - Atresia 80%
    - Stenosis 20%
    - Overall distribution equal between jejunum + ileum
    - More than 1 atresia present in 20%

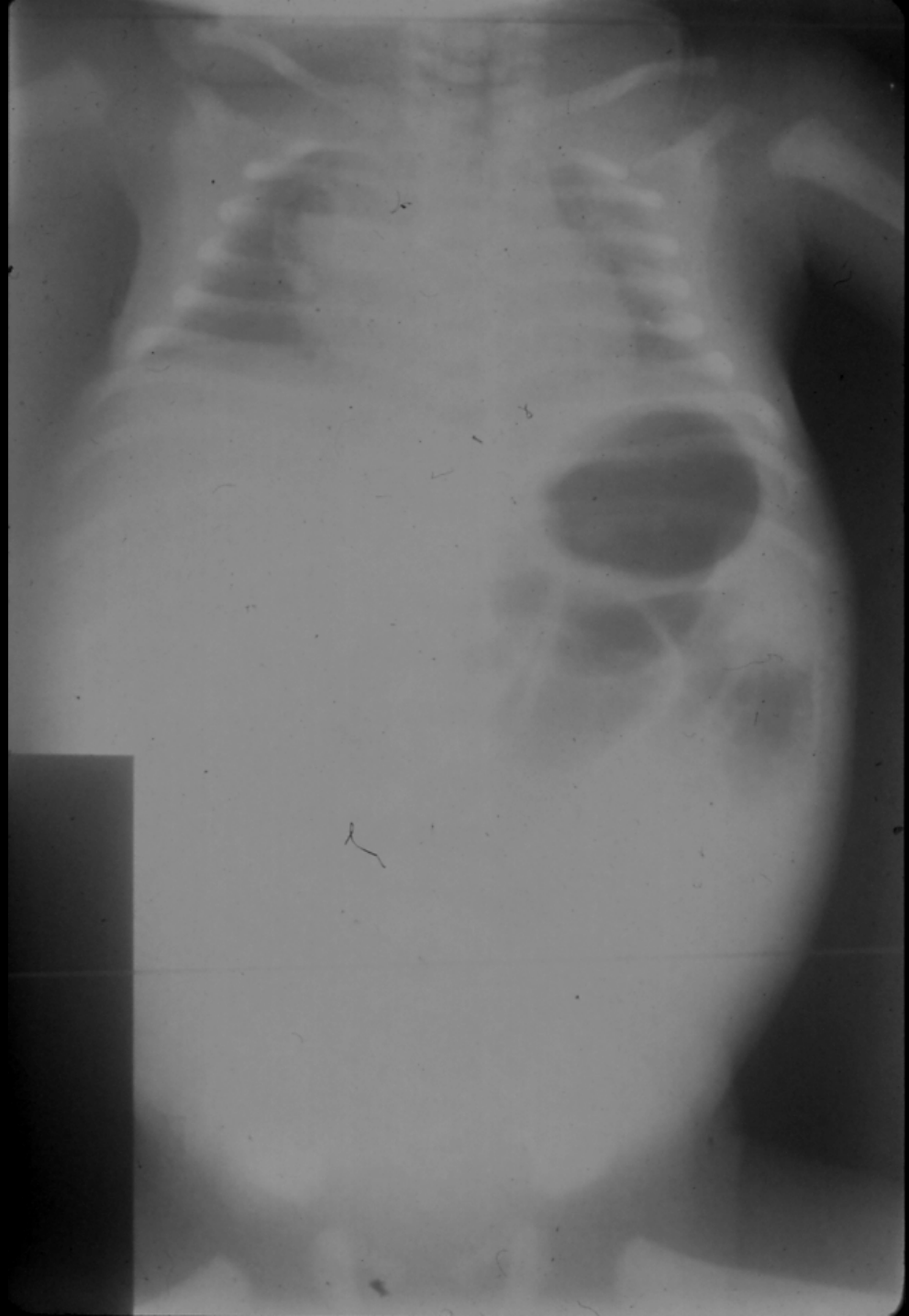

A
I
II
B
IIIa
IIIb
IV

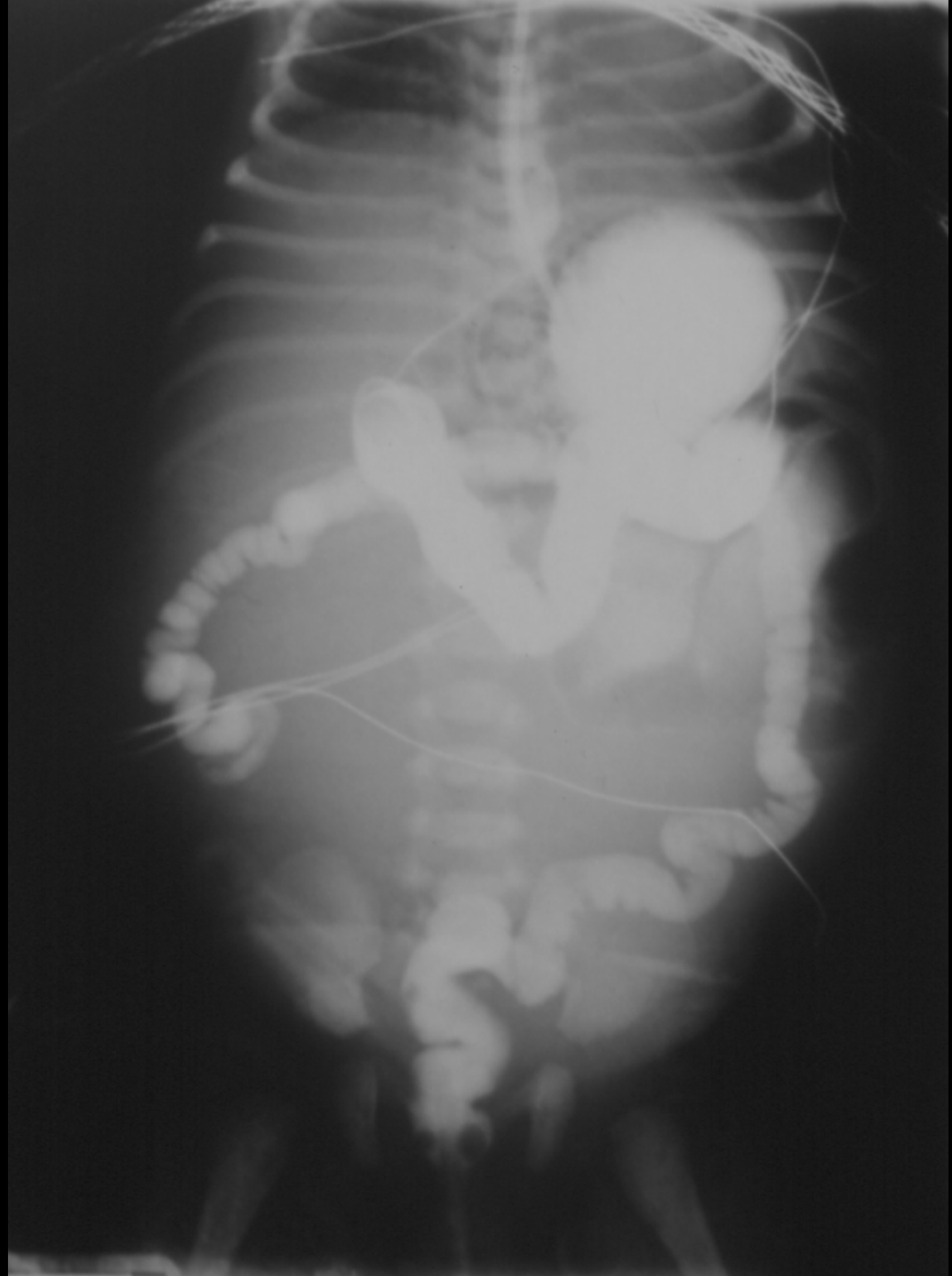
PEDIATRIC SURGERY INDEX CASES

- **JEJUNOILEAL ATRESIA**
  - Presentation
    - Abdominal distention
    - Bilious emesis

PEDIATRIC SURGERY INDEX CASES

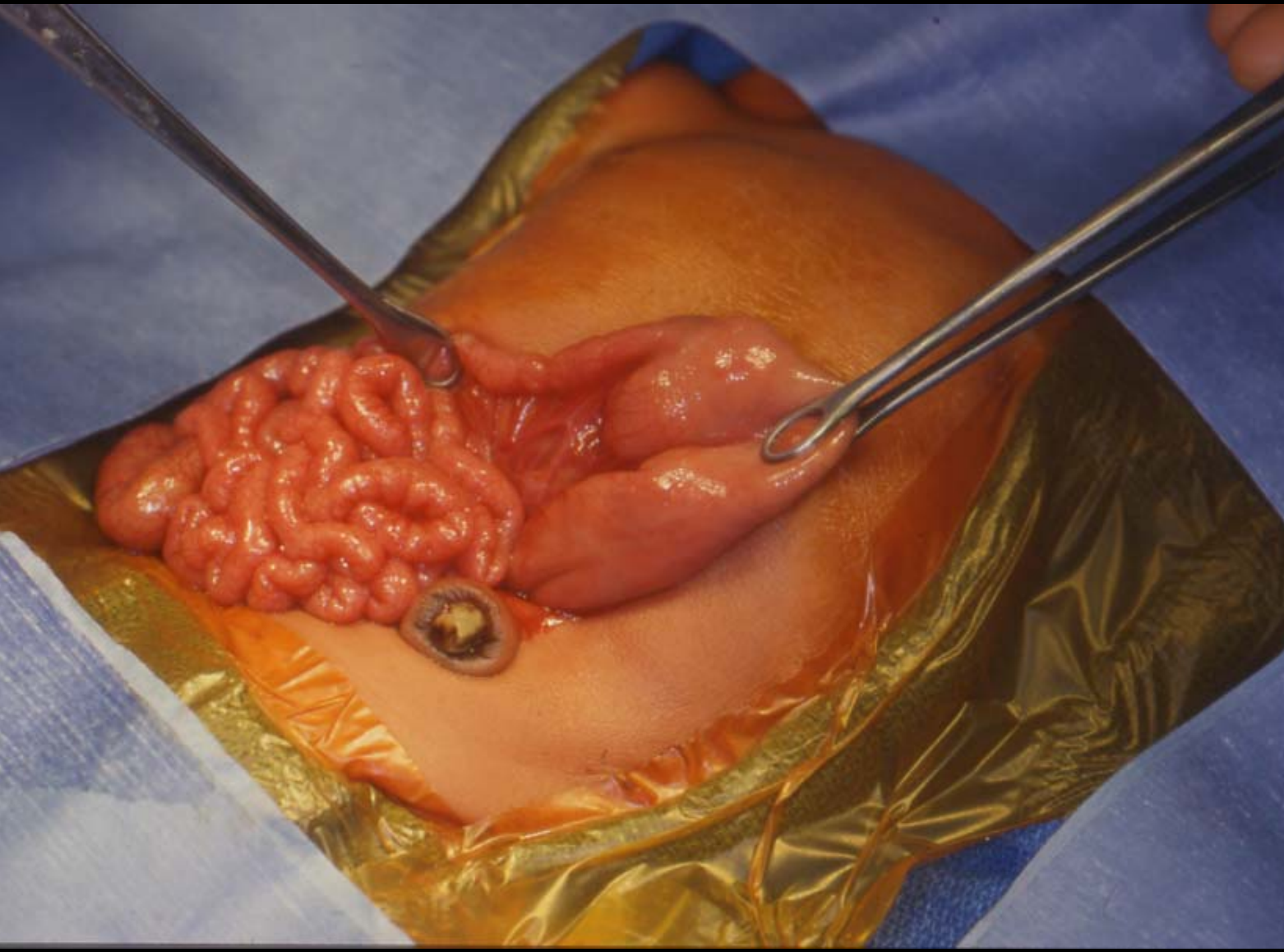
- **JEJUNOILEAL ATRESIA**
  - Diagnosis
    - Plain films
    - Barium enema
      - Demonstration of microcolon

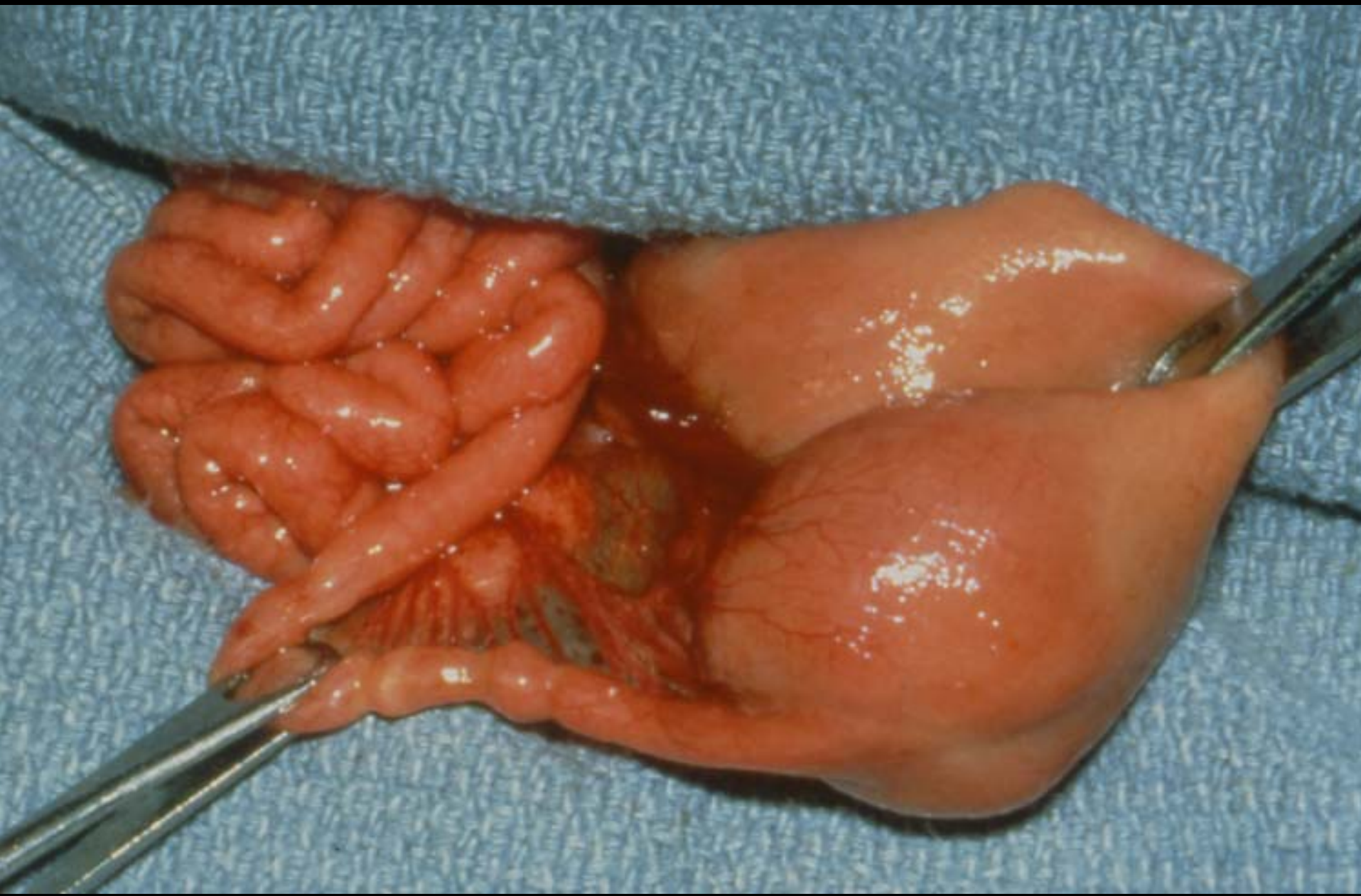


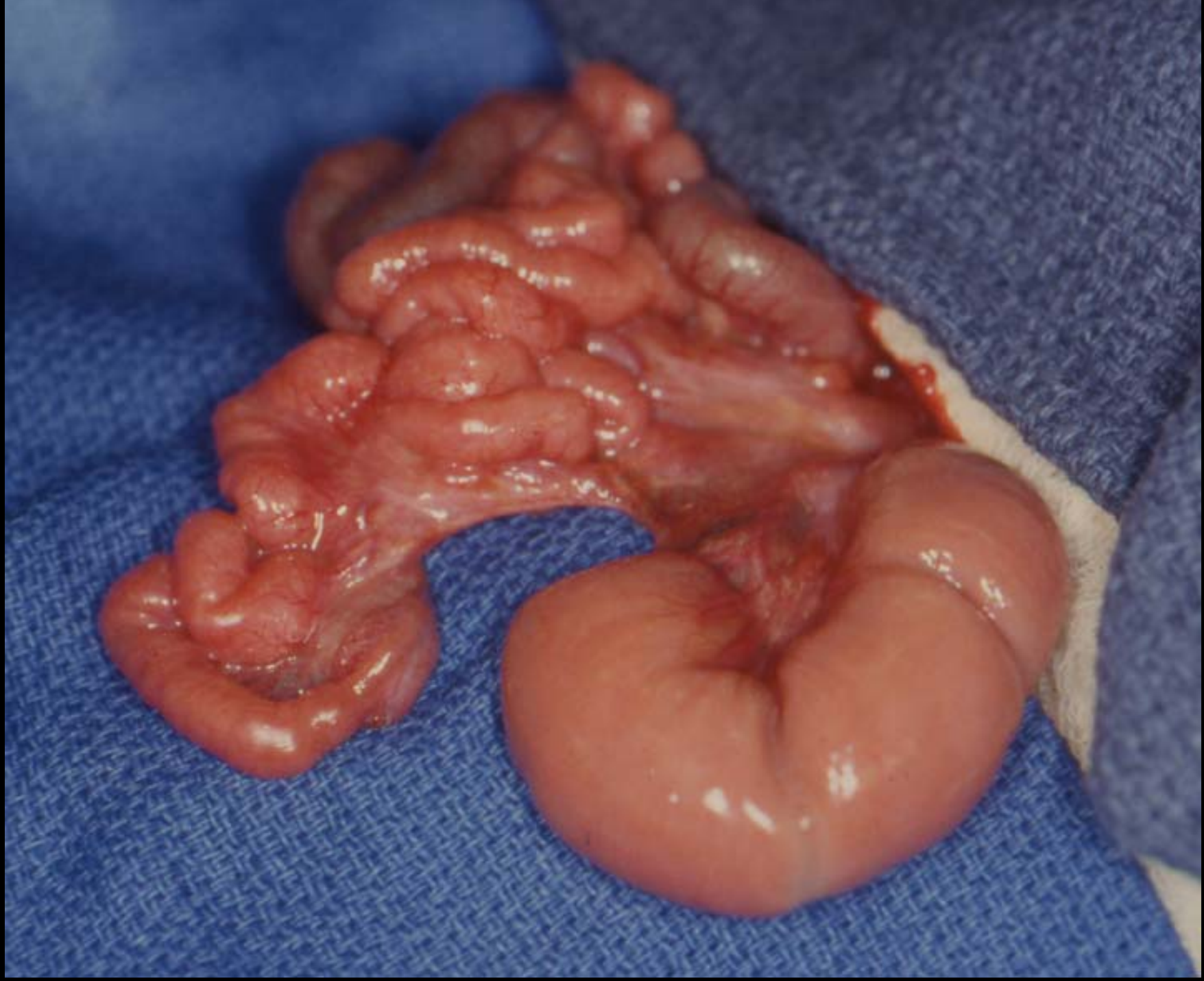


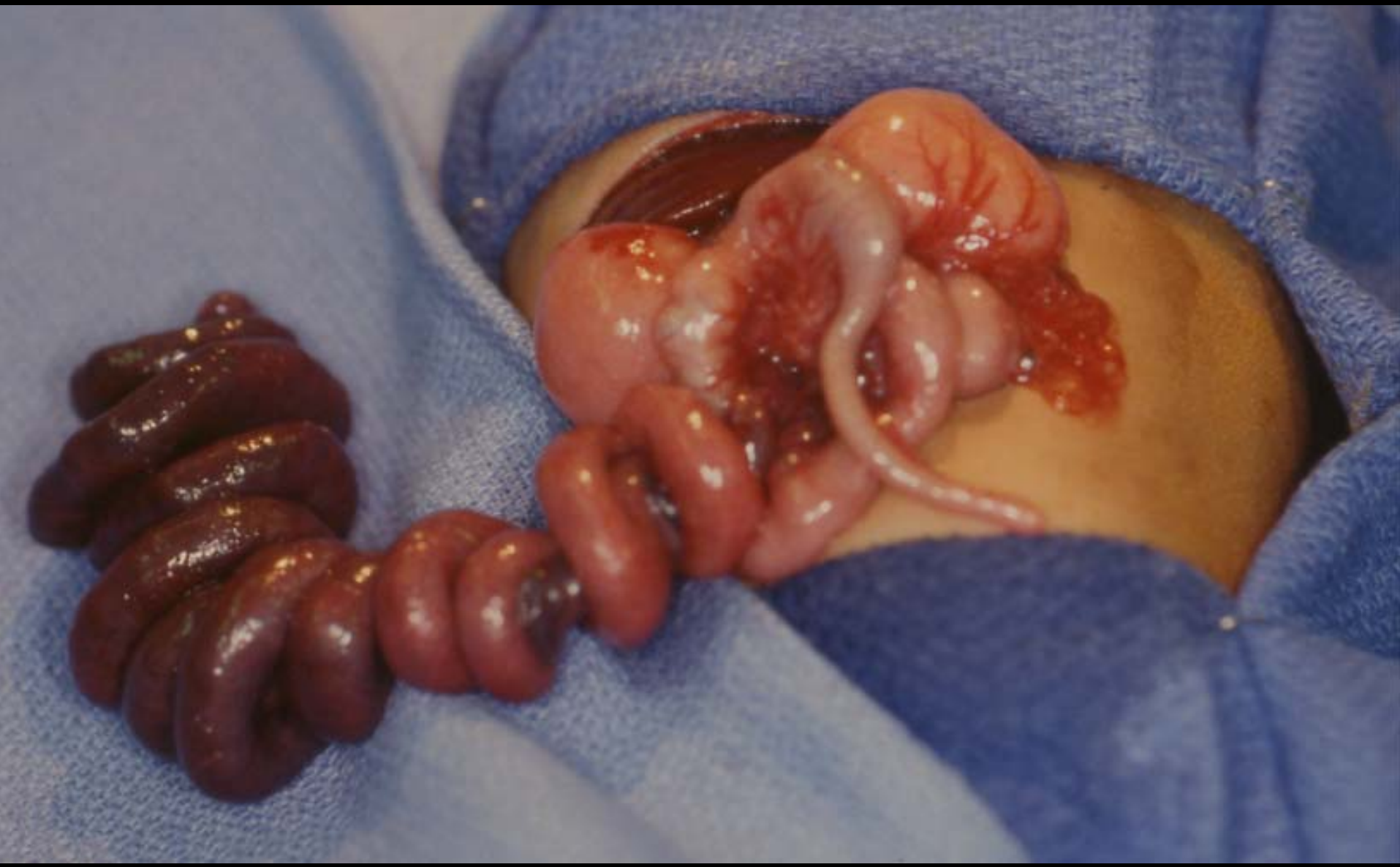
PEDIATRIC SURGERY INDEX CASES

- **JEJUNOILEAL ATRESIA**
  - TREATMENT
    - Resection with anastomois, tapering, or stoma
    - The surgical goal of intestinal reconstruction is to re-establish intestinal continuity while preserving bowel length and normal anatomy.









PEDIATRIC SURGERY INDEX CASES

- **JEJUNOILEAL ATRESIA**
  - Outcome:
    - Prolonged dysfunction of the proximal gut is relatively common and may persist for several months; these patients often require parenteral nutritional support.
    - Functional obstruction may be present due to retention of bowel which was normal in appearance at the original operation but because of ischemia, intrinsic dysmotility, or absorptive problems proves inadequate.

PEDIATRIC SURGERY INDEX CASES

CASE 5:

A 3-day old term male infant had an uneventful perinatal course and was discharged to home at 12 hours of age. His mother notes that he initially fed well but developed bilious emesis on day of life #2. He passed a small amount of meconium for the first time today.

PEDIATRIC SURGERY INDEX CASES

On physical examination he is alert and active; capillary refill is < 2 seconds. Lungs are clear; no cardiac murmur. Abdomen is diffusely distended; non-tender. Rectal examination reveals a normally located anus with a normal location on the interischial line. Digital exam is followed by prompt evacuation of a large amount of meconium.

PEDIATRIC SURGERY INDEX CASES

- **HIRSCHSPRUNG'S DISEASE**
  - Etiology:
    - Characterized by the absence of ganglion cells in the distal bowel extending proximally for varying distances.
    - In 75% of cases, aganglionosis is confined to rectosigmoid; sigmoid/splenic flexure in 17%; total colonic with short segment of ileum in 8%.

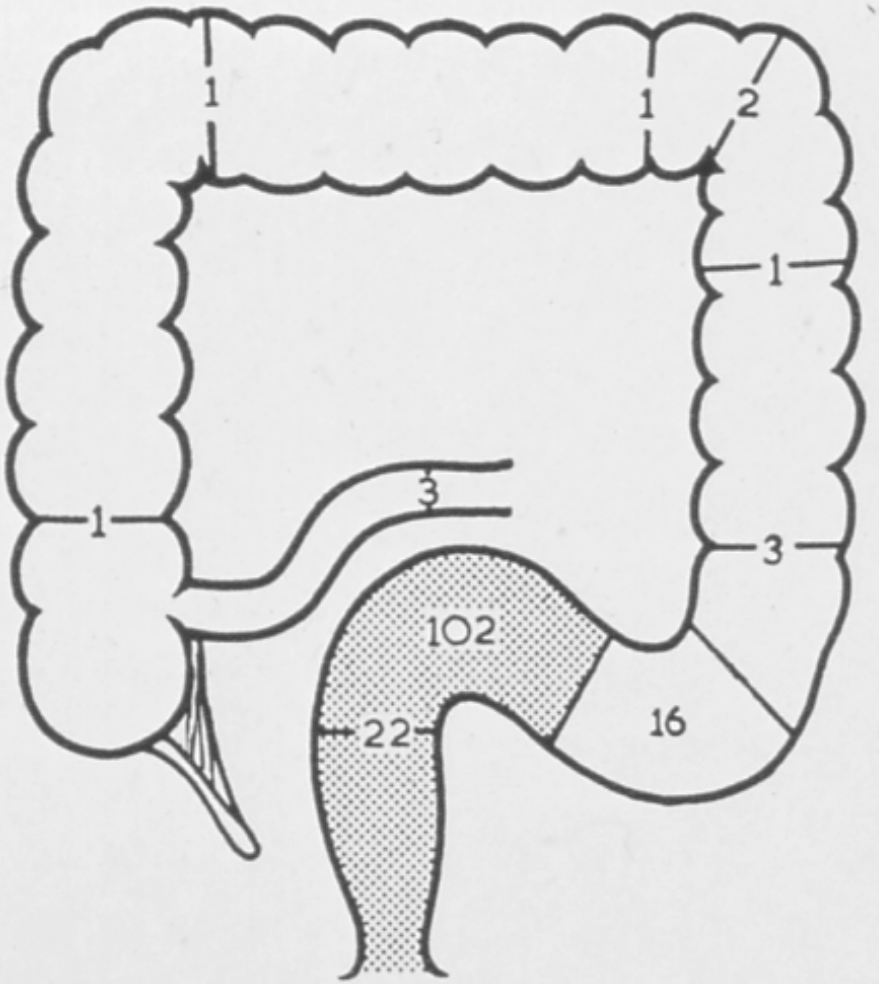


Fig. 17.—Proximal extent of aganglionosis in 152 patients. (From Wyllie, C. G.: Lancet 1:847, 1957.)

PEDIATRIC SURGERY INDEX CASES

- **HIRSCHSPRUNG'S DISEASE**
  - Embryology
    - Enteric ganglion cells derived from neural crest cells.
    - Absence of ganglion cells in Hirschsprung's disease attributed to failure of migration of neural crest cells.
    - Extracellular matrix proteins have been recognized as important microenvironment mediators of the neural processing pathways and important matrix for cell adhesion and movement.

PEDIATRIC SURGERY INDEX CASES

- **HIRSCHSPRUNG'S DISEASE**
  - Presentation
    - Abdominal distention
    - Bilious emesis
    - Failure to pass meconium
    - Failure to thrive
  - Down syndrome in 4-5% of cases

PEDIATRIC SURGERY INDEX CASES

- **HIRSCHSPRUNG'S DISEASE**
  - Diagnosis
    - Barium enema
    - Anorectal manometry
    - Biopsy

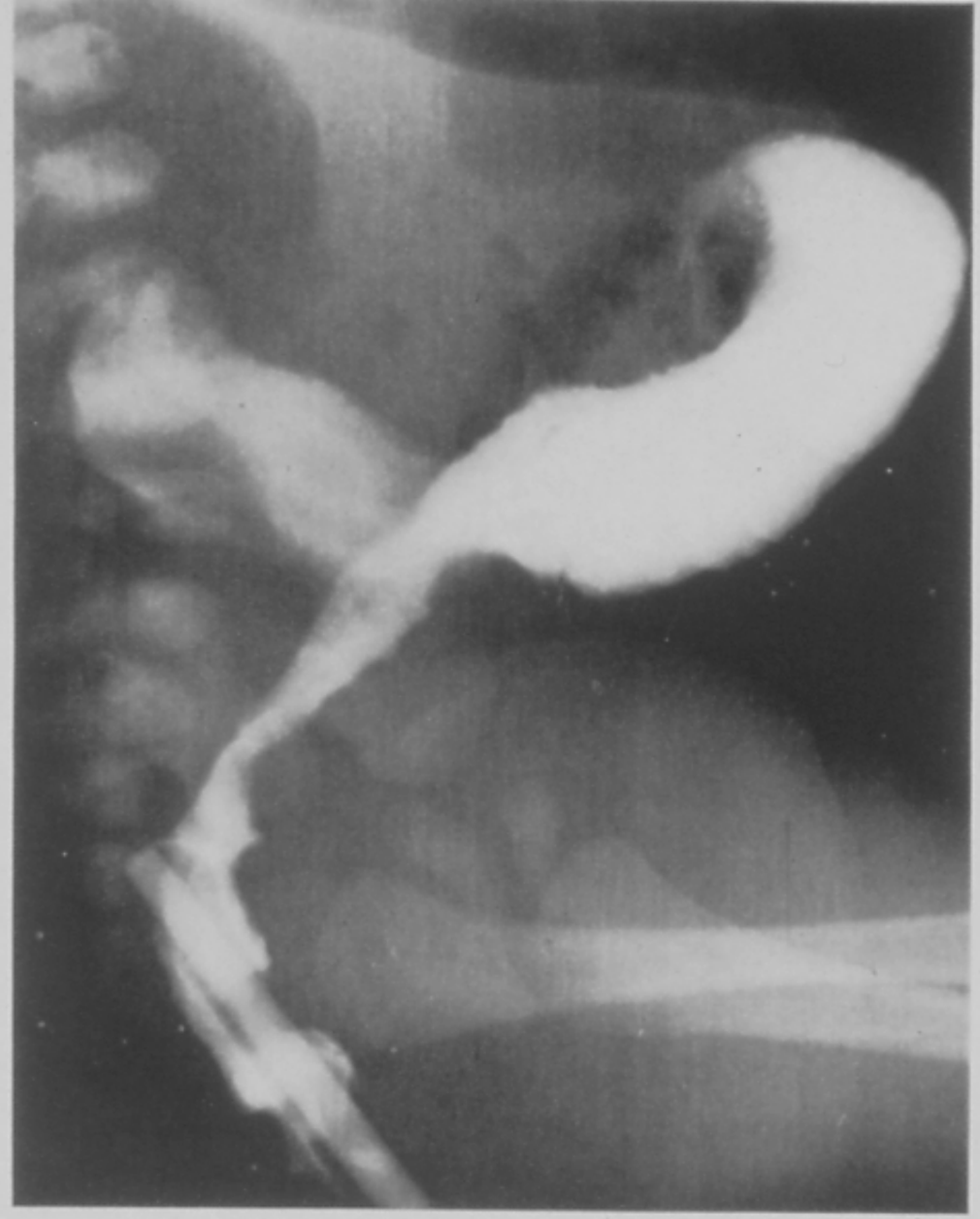
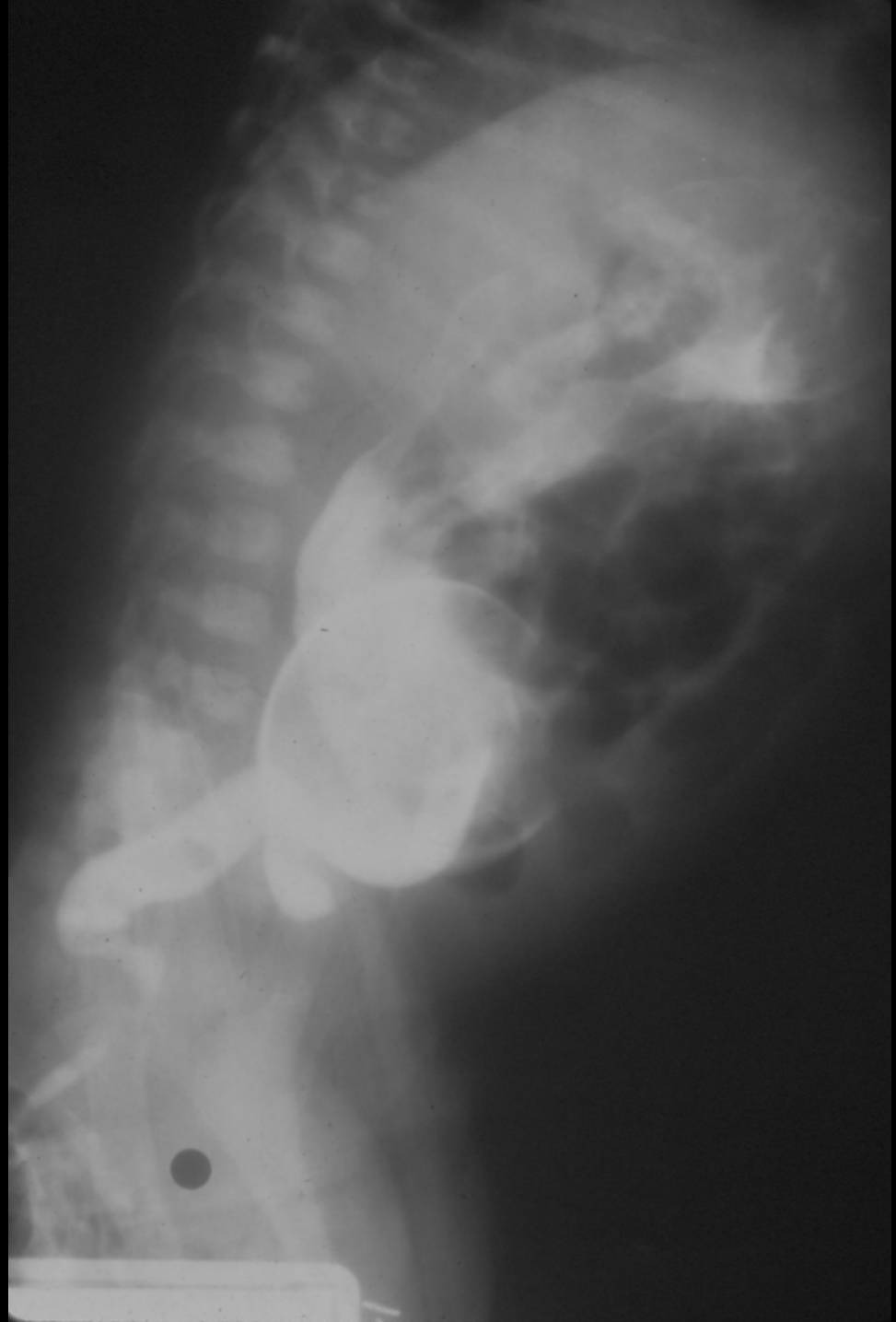
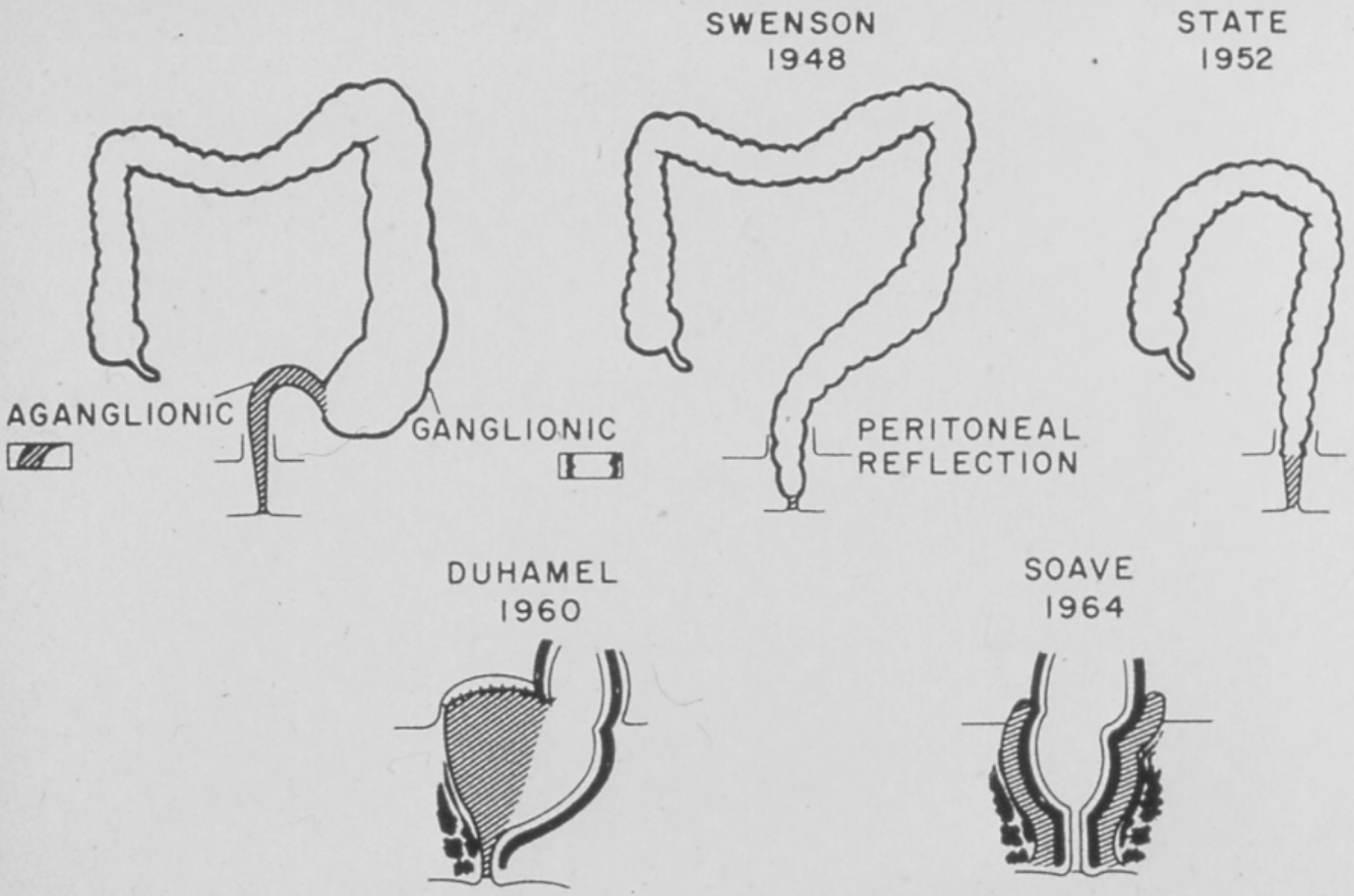


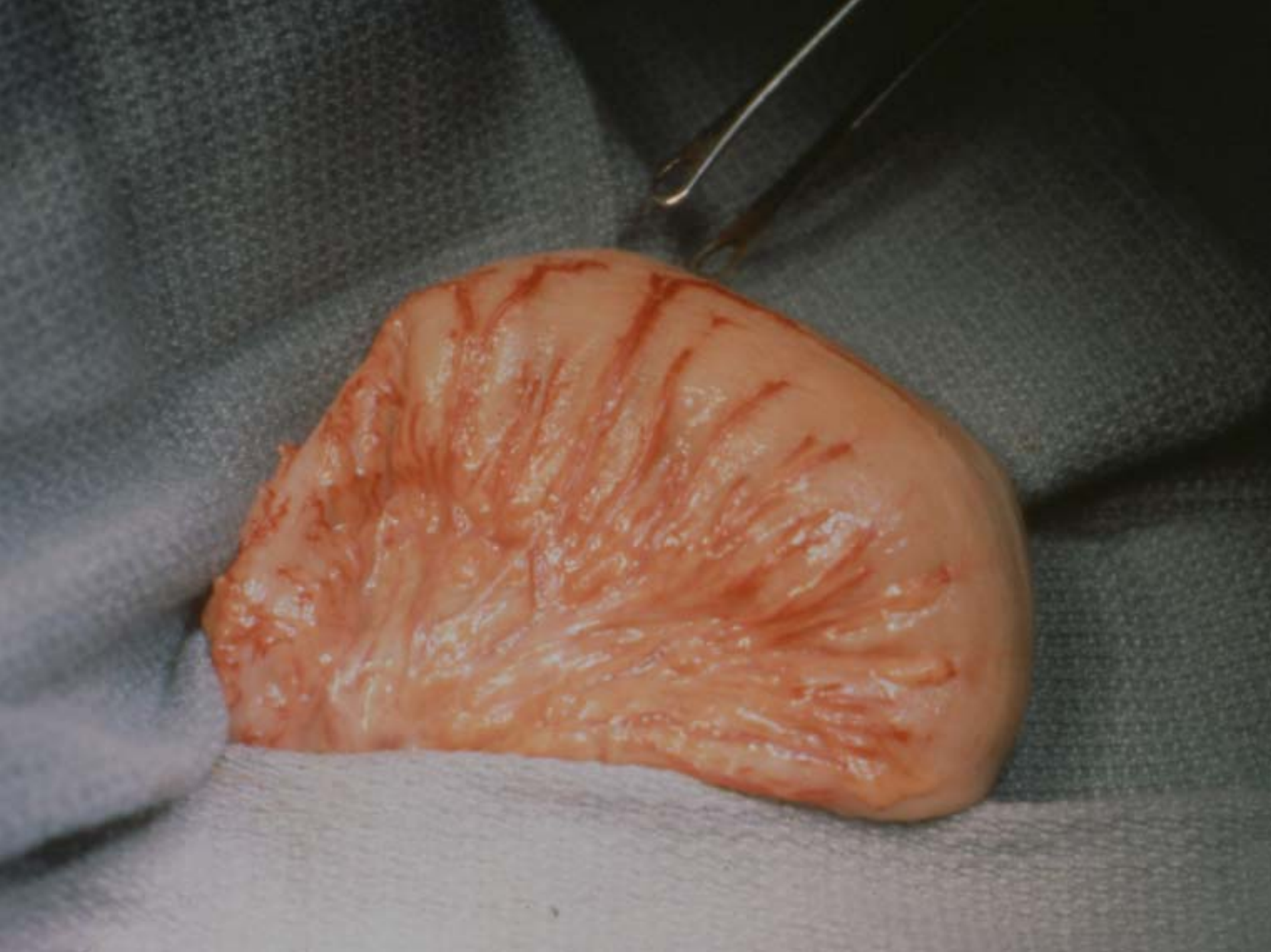
Fig. 23.—Neonatal Hirschsprung's disease: barium enema. Narrow recto-sigmoid segment, transitional zone, and slight dilatation of sigmoid as demonstrated by instillation of small amount of contrast medium. Note slight disparity in size.



PEDIATRIC SURGERY INDEX CASES

- **HIRSCHSPRUNG'S DISEASE**
  - Treatment
    - Leveling colostomy
    - Pull-through procedure—includes resection of aganglionated intestine with restoration of fecal continence by means of anastomosis of ganglionated bowel to the anal verge
      - Swenson
      - Duhamel
      - Soave


SWENSON
1948
STATE
1952
AGANGLIONIC
GANGLIONIC
PERITONEAL
REFLECTION
DUHAMEL
1960
SOAVE
1964



PEDIATRIC SURGERY INDEX CASES

- **HIRSCHSPRUNG'S DISEASE**
  - COMPLICATIONS
    - Enterocolitis
      - May occur prior to colostomy or after a pull-through procedure.
      - Presents with abdominal distention and pain, fever, and explosive watery diarrhea.
      - Spectrum from mild to fulminant gram (-) sepsis
      - Treatment includes rectal irrigations to decompress colon, antibiotics, bowel rest

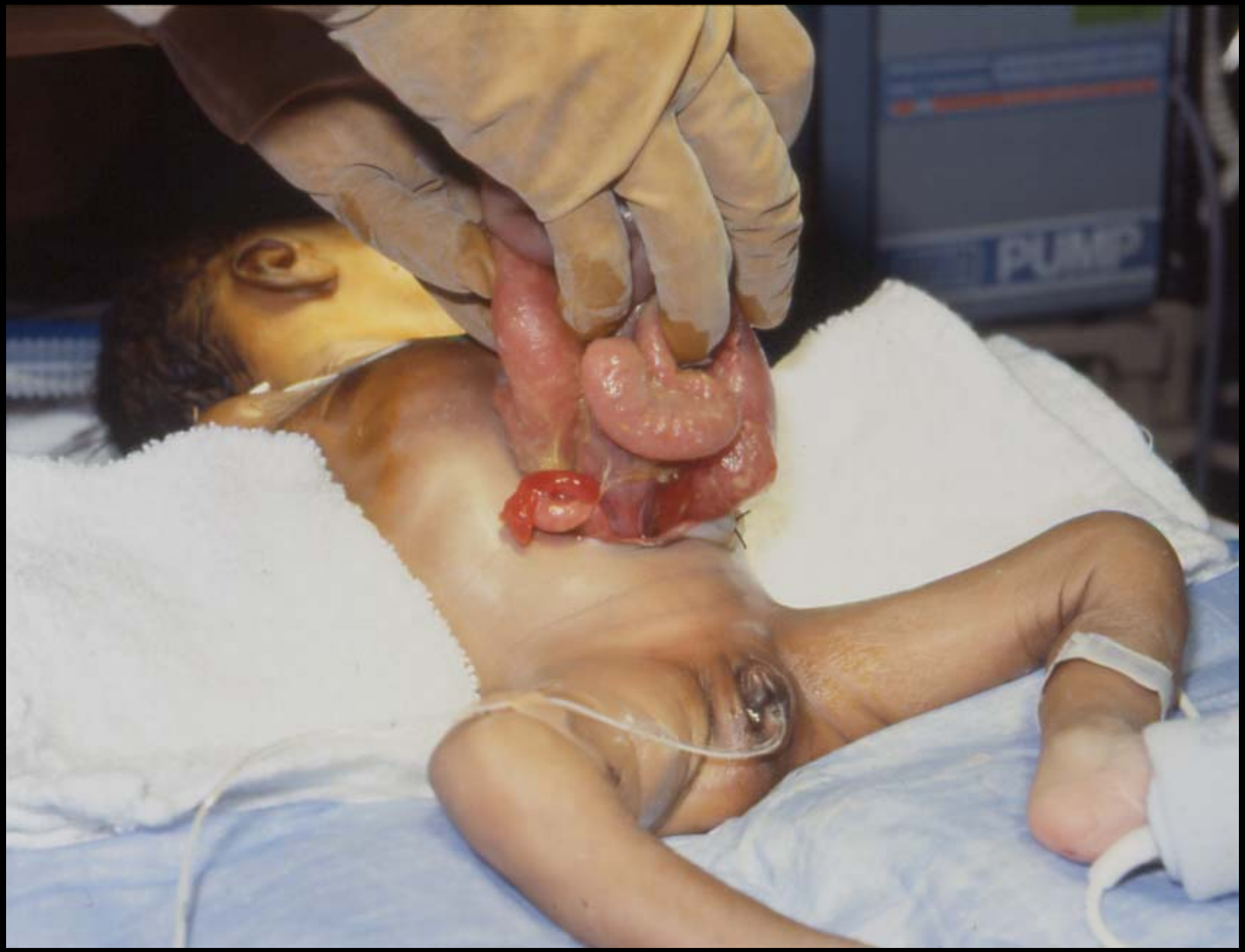
PEDIATRIC SURGERY INDEX CASES

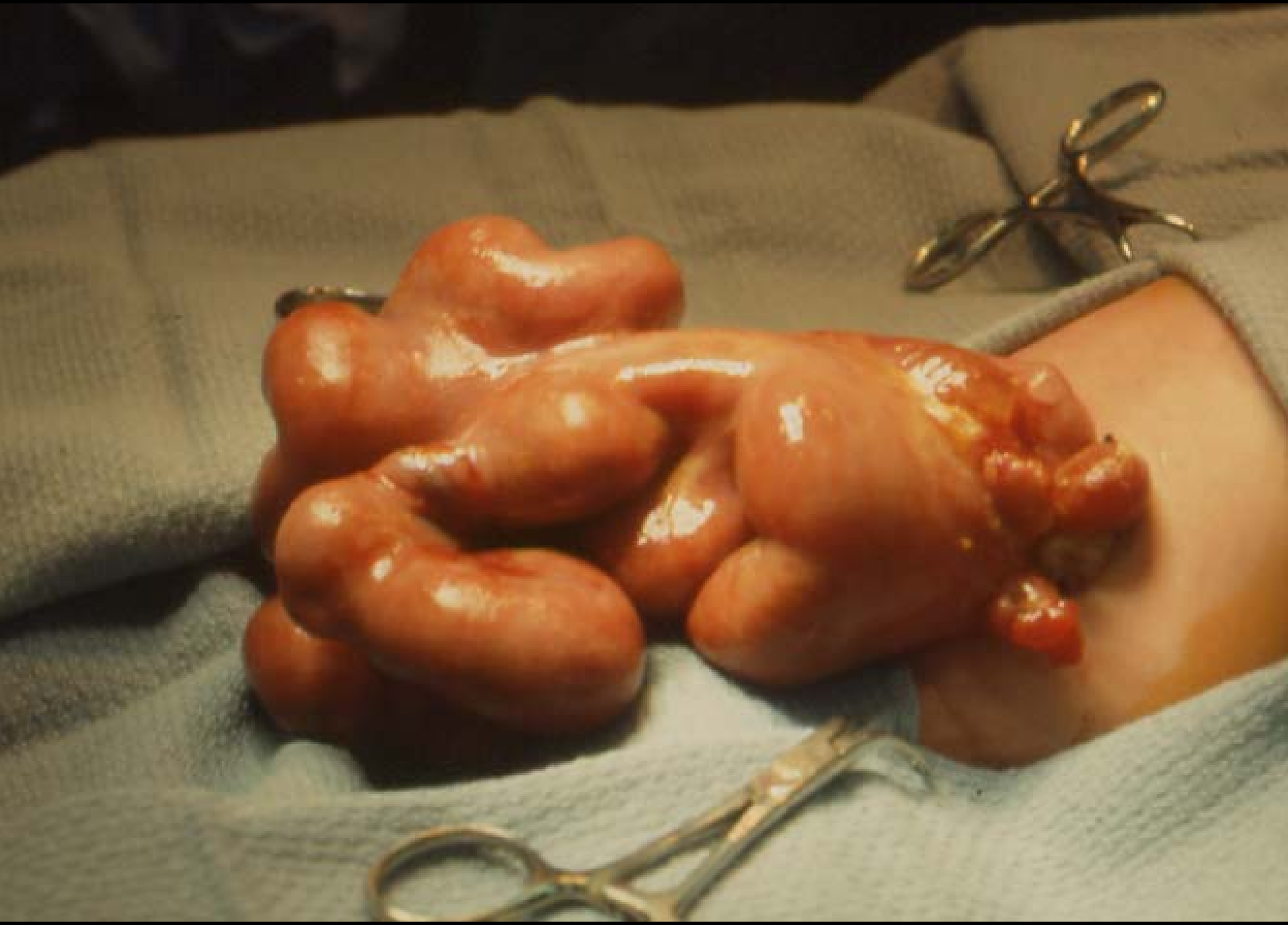
CASE 6:

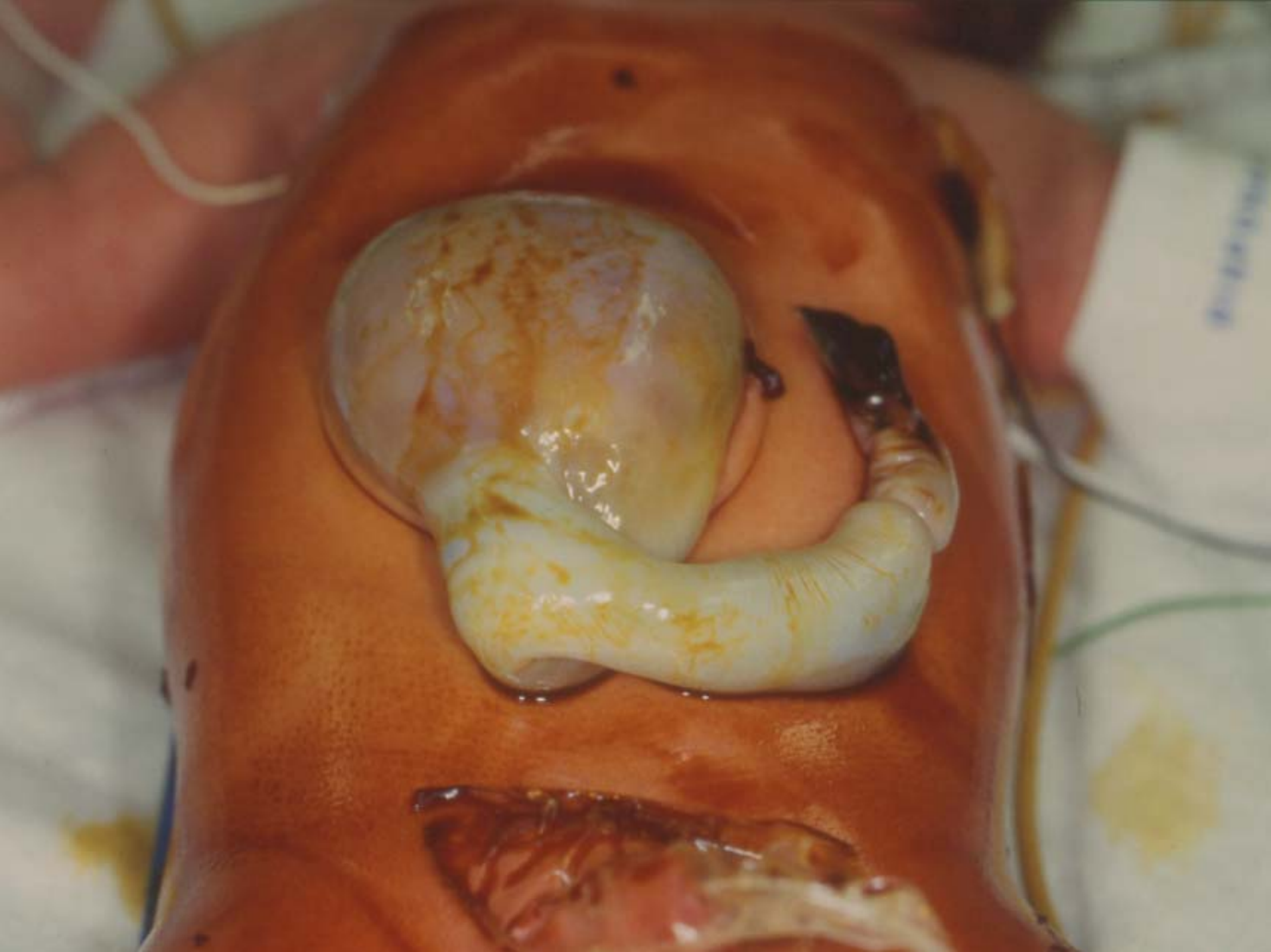
A 2500 gram female infant is born via vaginal delivery at 36 weeks gestation with an abdominal wall defect.

PEDIATRIC SURGERY INDEX CASES

- **GASTROSCHISIS**
  - Diagnosis
    - Inspection
      - Defect generally to right of normal appearing umbilical cord
      - Differentiate gastroschisis from omphalocele
        - Associated anomalies—(30-50%)
      - Appearance of intestine varies; typically is thickened, shaggy appearing and matted.
      - Associated anomalies—ileal atresia (10%), undescended testes (25%)

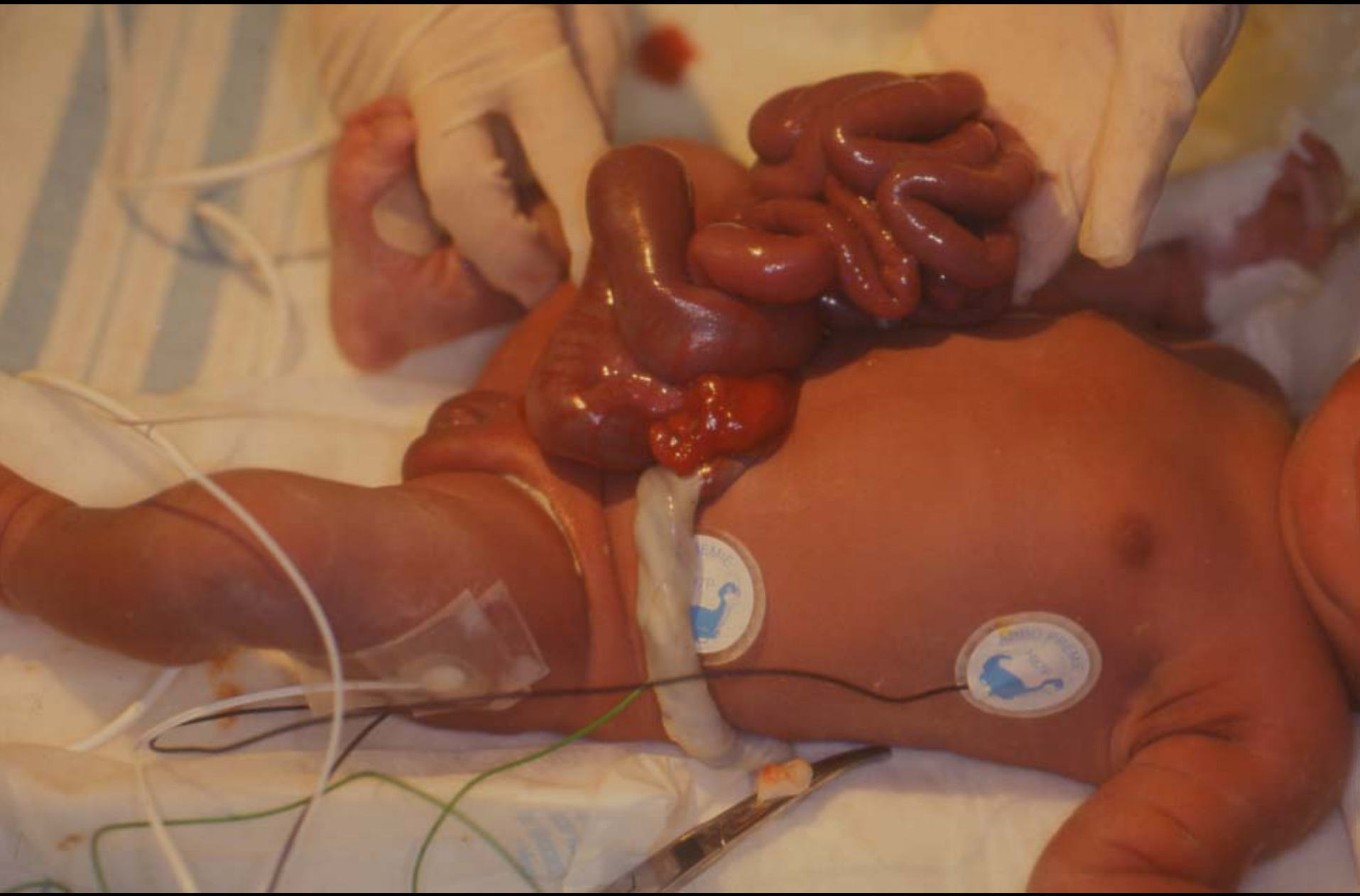


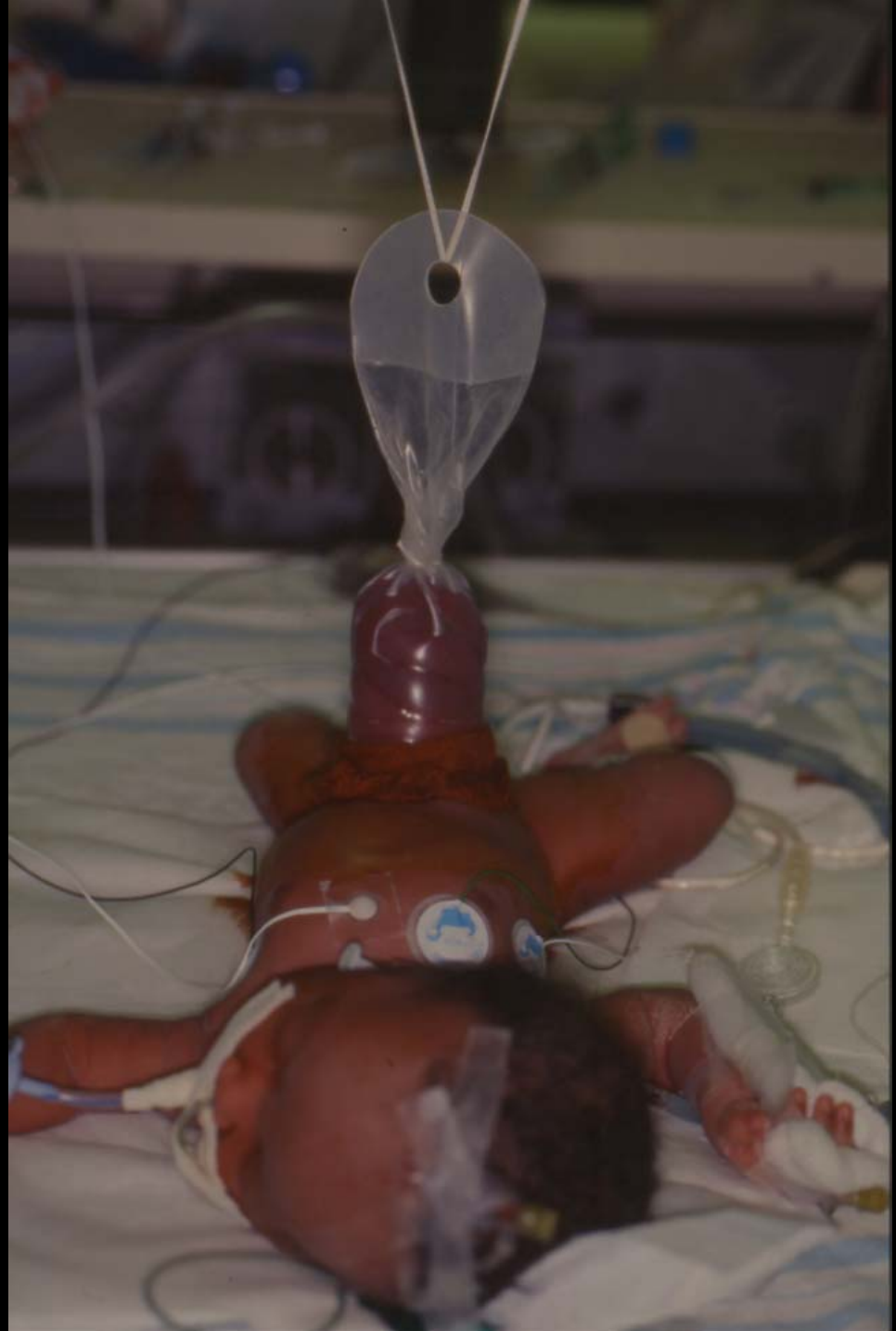




PEDIATRIC SURGERY INDEX CASES

- **GASTROSCHISIS**
  - Treatment
    - Primary reduction and closure
    - Placement of silo with gradual visceral reduction and elective closure
      - Advantages—can be performed in NICU, transparent silo allows continuous inspection of bowel, reduction can be individualized for each newborn, decreased pulmonary injury due to mechanical ventilation





PEDIATRIC SURGERY INDEX CASES

- **GASTROSCHISIS**
  - Overall survival 95%
  - Post-operative complications (10-25%)
    - Intestinal ischemia
    - Bowel infarction
    - Enterocutaneous fistula
    - Necrotizing enterocolitis
    - Prolonged intestinal dysfunction
    - Catheter associated sepsis, venous thrombosis, malposition

PEDIATRIC SURGERY INDEX CASES

CASE 7:

A neonatologist calls you to evaluate a 2-hour old neonate, 36-week EGA, birth weight 2700 grams with persistent, worsening cyanosis. The infant has been intubated and is currently on conventional mechanical ventilation. Capillary refill > 3.5 seconds. Heart tones are difficult to hear and breath sounds are decreased on the left. A babygram has been obtained.

PEDIATRIC SURGERY INDEX CASES

- **CONGENITAL DIAPHRAGMATIC HERNIA**
  - Embryology
    - Forms from septum transversum, dorsal mesentery of the esophagus, pleuroperitoneal canals, and lateral body wall
    - Formation complete at 9 weeks of gestation
    - Lung development divided into 4 stages:
      - Pseudoglandular period (5-17 weeks)
      - Canalicular period (13-25 weeks)
        - Surfactant production starts at 24 weeks
      - Terminal period (24 weeks-birth)
      - Alveolar period (late fetal-8 yrs.)

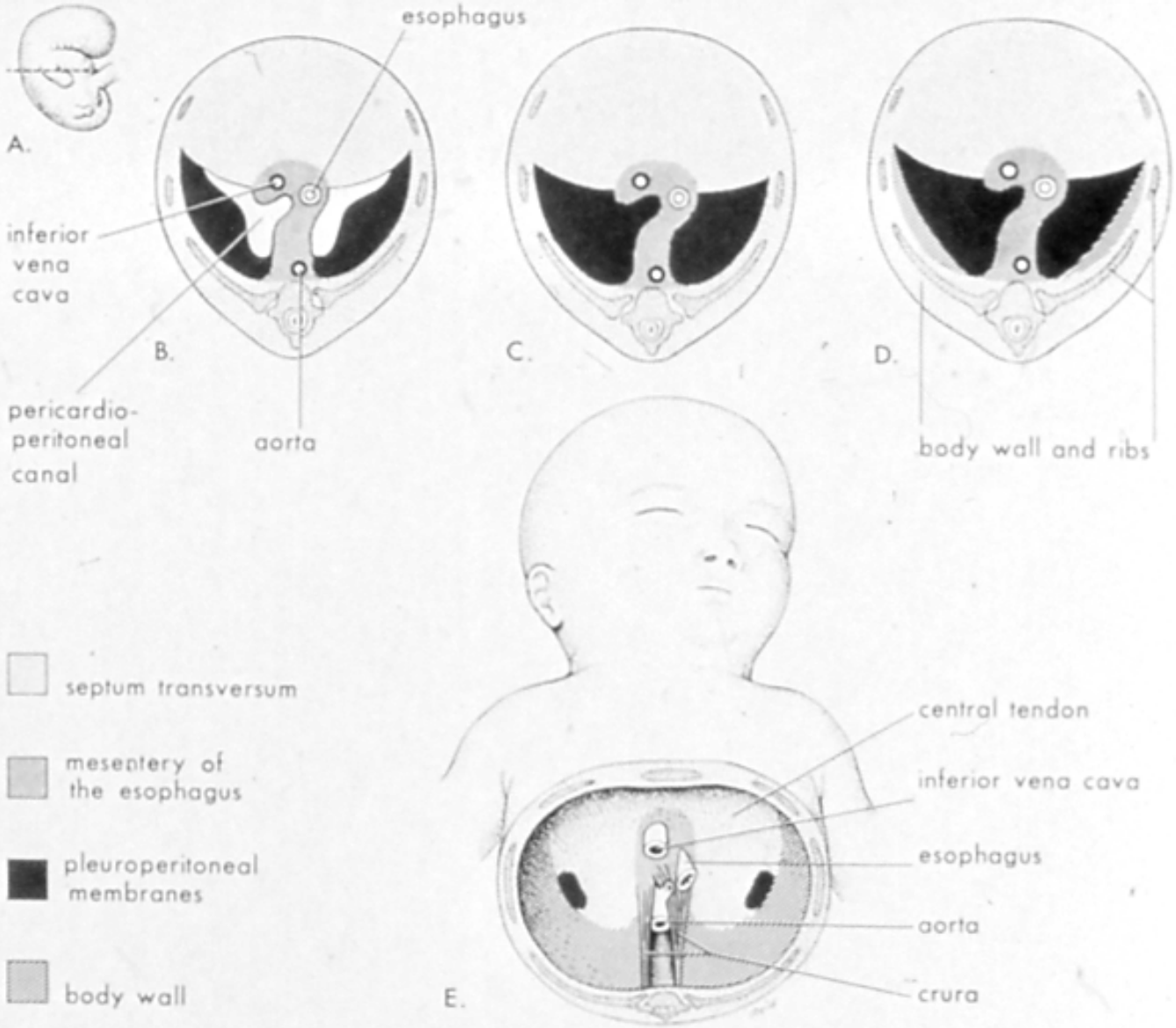


Figure 9–7 Drawings illustrating development of the diaphragm as viewed from below. *A*, sketch of a lateral view of an embryo at the end of the fifth week *(actual size)* indicating the level of section *B*. *B*, transverse section showing the unfused pleuroperitoneal membranes. *C*, similar section at the end of the sixth week after fusion of the pleuroperitoneal membranes with the othes two diaphragmatic components. *D*, transverse section through a 12-week embryo after ingrowth of the fourth diaphragmatic component from the body wall. *E*, view of the diaphragm of a newborn infant, indicating the probable

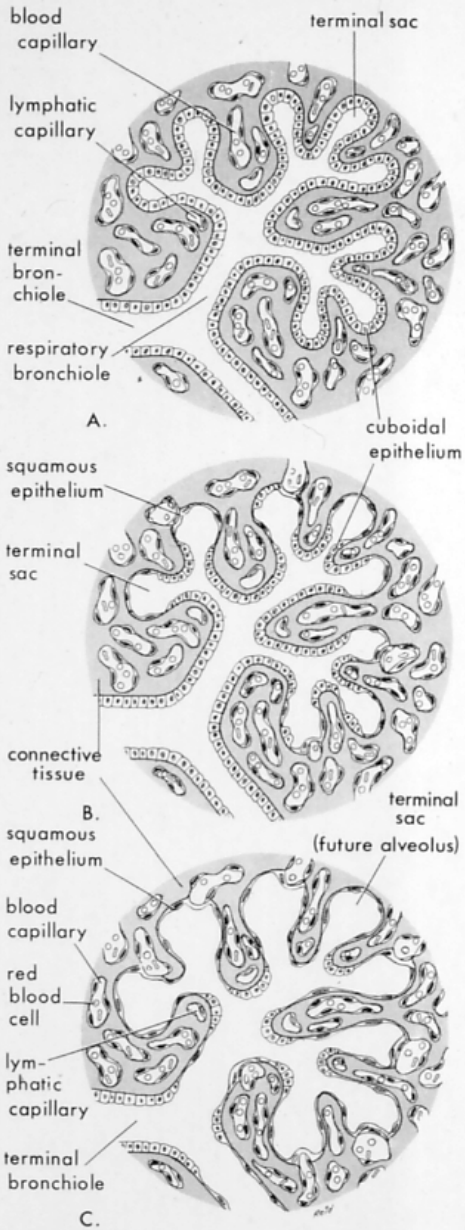


Figure 11–8 Diagrammatic sketches of sections illustrating progressive stages of lung development. *A*, Late canalicular period (about 24 weeks). *B*, Early terminal sac period (about 26 weeks). *C*, Newborn infant. Early alveolar period. Note the thin alveolar-capillary (respiratory) membrane. Note also that some of the capillaries have begun to bulge into the terminal sacs.

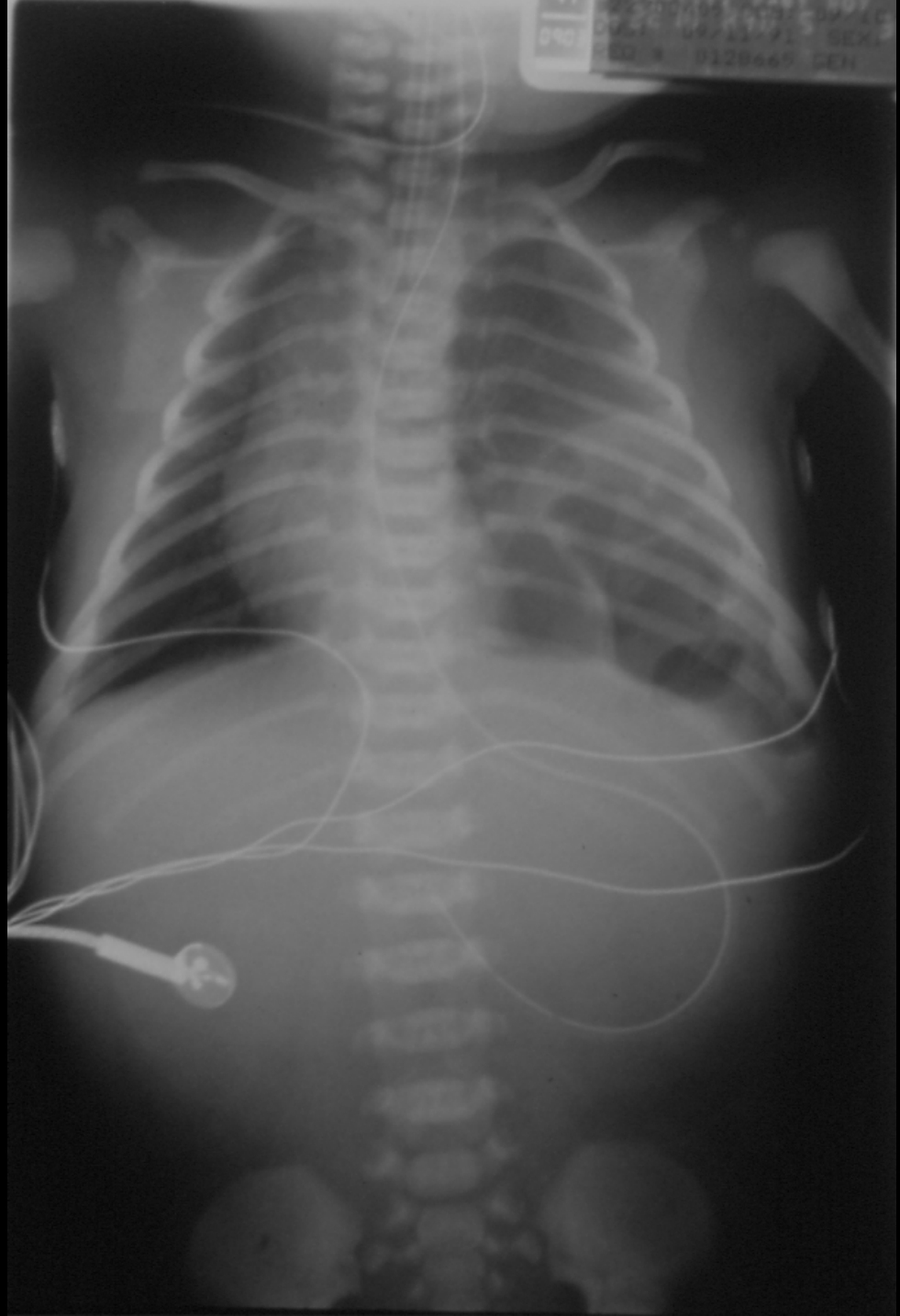
PEDIATRIC SURGERY INDEX CASES

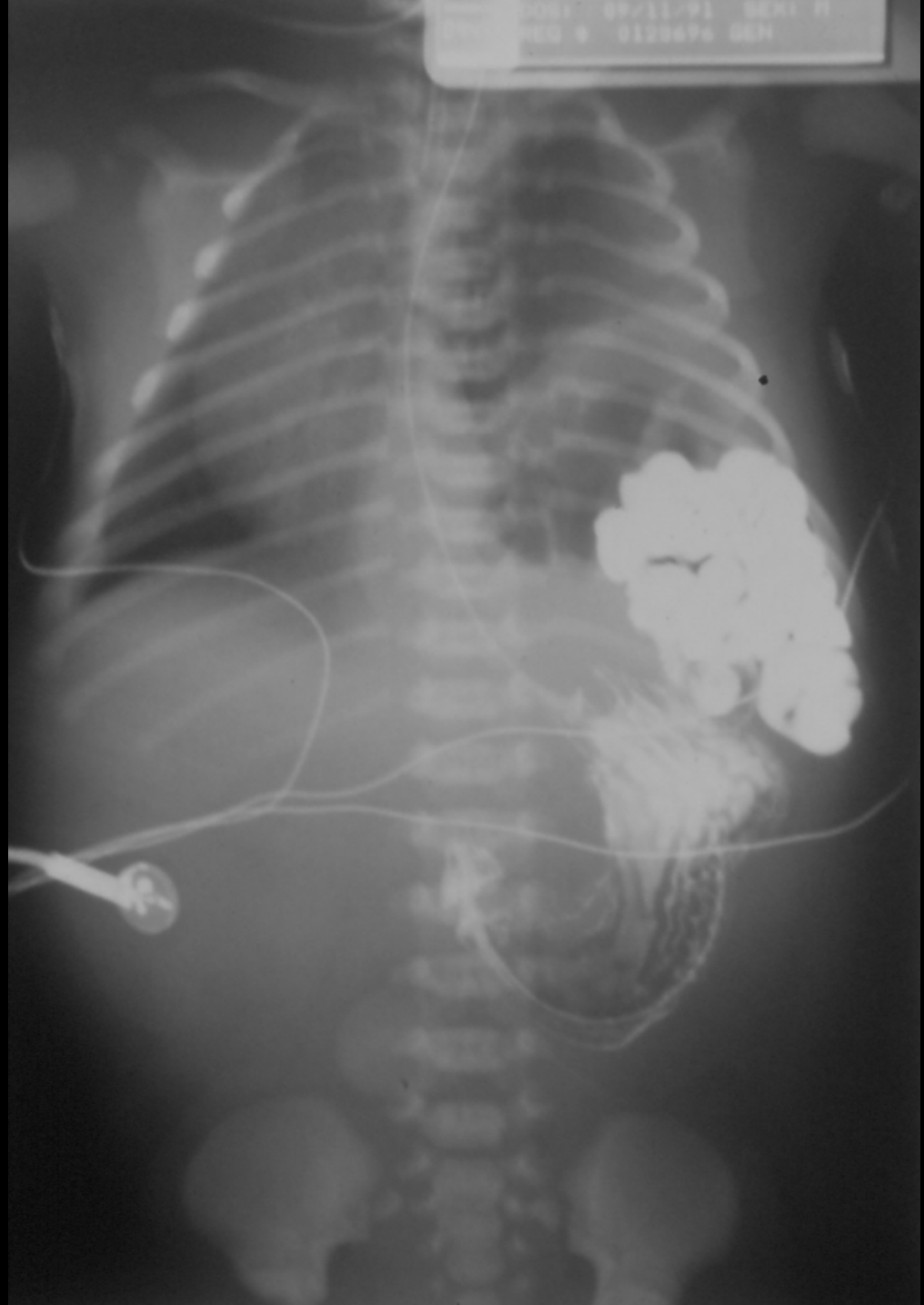
- **CONGENITAL DIAPHRAGMATIC HERNIA**
  - Presentation
    - Respiratory distress
      - Cyanosis + tachypnea
    - Scaphoid abdomen

PEDIATRIC SURGERY INDEX CASES

- **CONGENITAL DIAPHRAGMATIC HERNIA**
  - Differential diagnosis:
    - Cystic adenomatoid malformation
    - Congenital diaphragmatic hernia
    - Eventration of the diaphragm
  - Diagnosis
    - Prenatal ultrasound
    - Babygram

L56





PEDIATRIC SURGERY INDEX CASES

- **CONGENITAL DIAPHRAGMATIC HERNIA**
  - Associated anomalies
    - Pulmonary hypoplasia
      - Decreased number of pulmonary artery branches with thickened muscular wall
    - Malrotation
    - Extralobar sequestration

PEDIATRIC SURGERY INDEX CASES

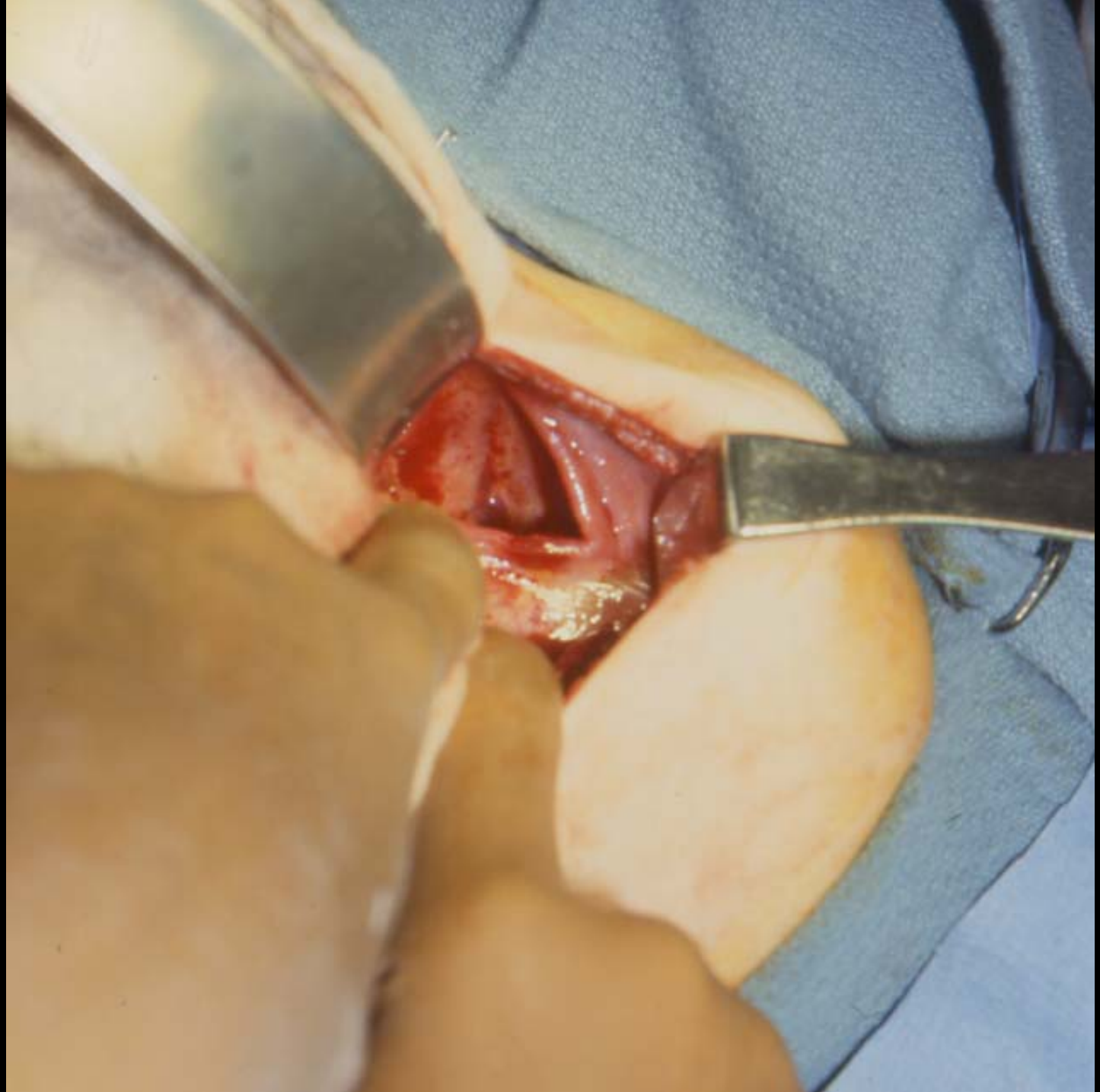
- **CONGENITAL DIAPHRAGMATIC HERNIA**
  - Timing of surgery
    - A number of observations have suggested that emergency repair is unnecessary:
      - Abdominal viscera move easily from thorax to abdomen
      - All children have some degree of bilateral hypoplasia
      - Children with CDH rarely improve after surgery

PEDIATRIC SURGERY INDEX CASES

- **CONGENITAL DIAPHRAGMATIC HERNIA**
  - Timing of surgery:
    - After surgery, several factors have been noted to bring about changes in mechanical forces across the diaphragm
      - Diaphragm frequently distorted after repair
      - Abdominal wall is tense after return of viscera
      - Ipsilateral lung hypoplasia

PEDIATRIC SURGERY INDEX CASES

- CONGENITAL DIAPHRAGMATIC HERNIA
  - Treatment
    - Stabilization
    - Repair
    - ECMO
  - Outcome
    - Mortality 55-58%



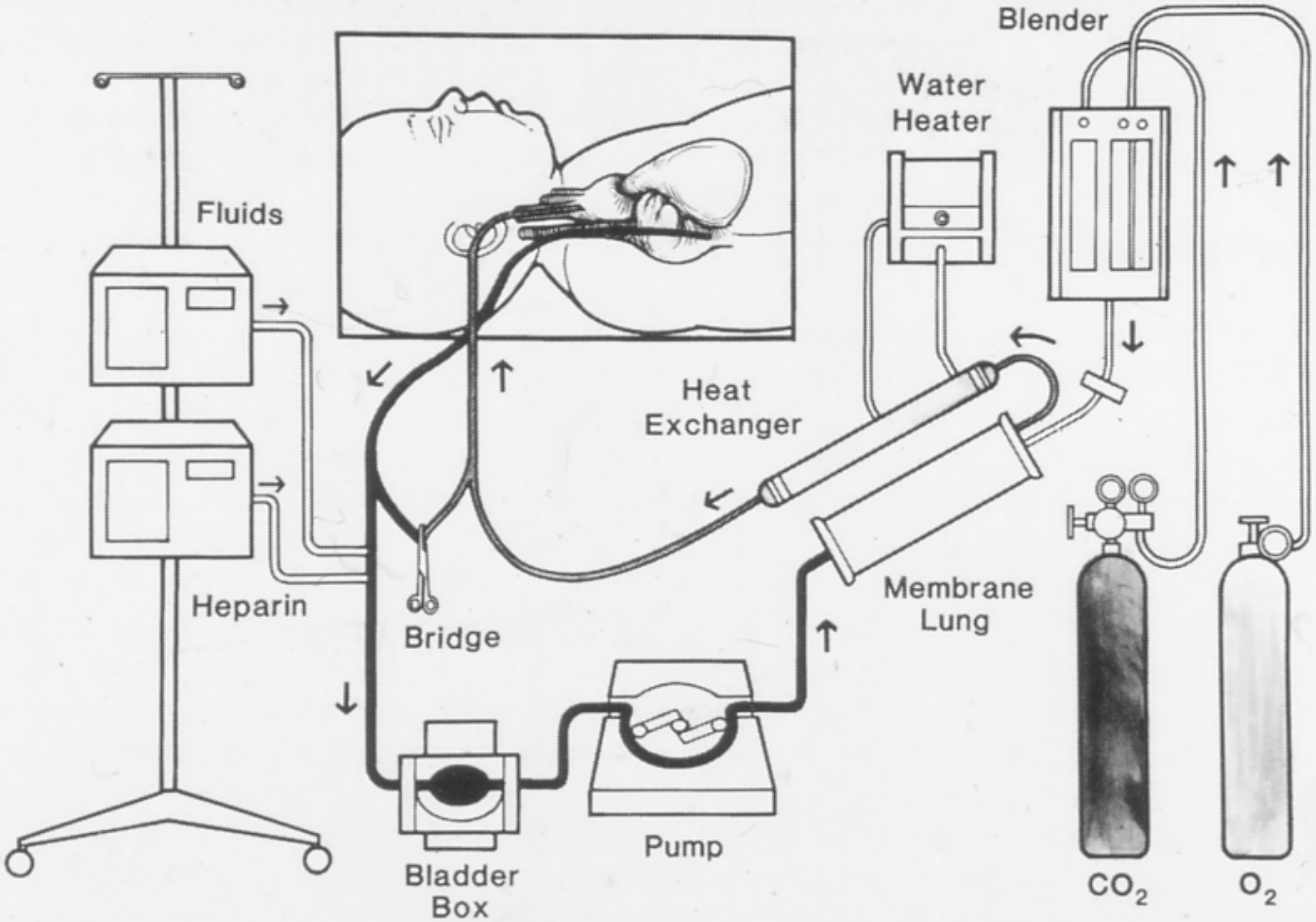


Figure 1. ECLS circuit demonstrating the components of most cardiopulmonary bypass circuits: a capacitance reservoir (bladder box), a pump (roller head or vortex), a membrane oxygenator, and a heat exchanger.