The Fetal Anterior Abdominal Wall

Chapter 62

- Ultrasound is proven to be very effective in detecting anterior abdominal wall defects
- Defects occur during the first trimester as the midgut elongates and migrates into the umbilical cord
- Midgut usually returns into the abdominal cavity by the 11th week of gestation
  - Failure to occur
    - Abdominal wall defect is formed

- Two most common defects:
  - Omphalocele
  - Gastroschisis
- Less common defects:
  - Ectopia cordis
  - Limb–body wall complex
  - Cloacal exstrophy
  - Allantoic cyst

Embryology of the Abdominal Wall

- Embryo is a flat disk consisting of three layers by the end of the fifth week
  - Ectoderm
  - Mesoderm
  - Endoderm
- Sixth week process called *folding* helps the embryo transform itself into a cylindrical shape
  - Becomes a critical process in the closure of the abdominal wall

- Fusion of the midline begins during the seventh week and is completed by the eighth week
- Umbilication hernia of the bowel occurs during the eighth week
  - Midgut extends to the extraembryonic coelom in the proximal portion of the umbilical cord

- Midgut grows faster than the abdominal cavity because of the increased size of the liver and kidneys
  - This is where the herniation develops
- Intestines return to the abdominal cavity by the 12th week of gestation

Sonographic Evaluation of the Fetal Abdominal Wall

- Important to image to evaluate the following for presence of defects
  - Cord insertion
  - Fetal anterior abdominal wall
- If the urinary bladder and pelvis are evaluated closely
  - Diagnosis of the following may be made with sonography
    - Bladder and cloacal exstrophy

Abnormalities of the Anterior Abdominal Wall

- Three common abdominal wall defects:
  - Omphalocele
  - Umbilical hernia (a form of omphalocele)
  - Gastroschisis

Things To Do List

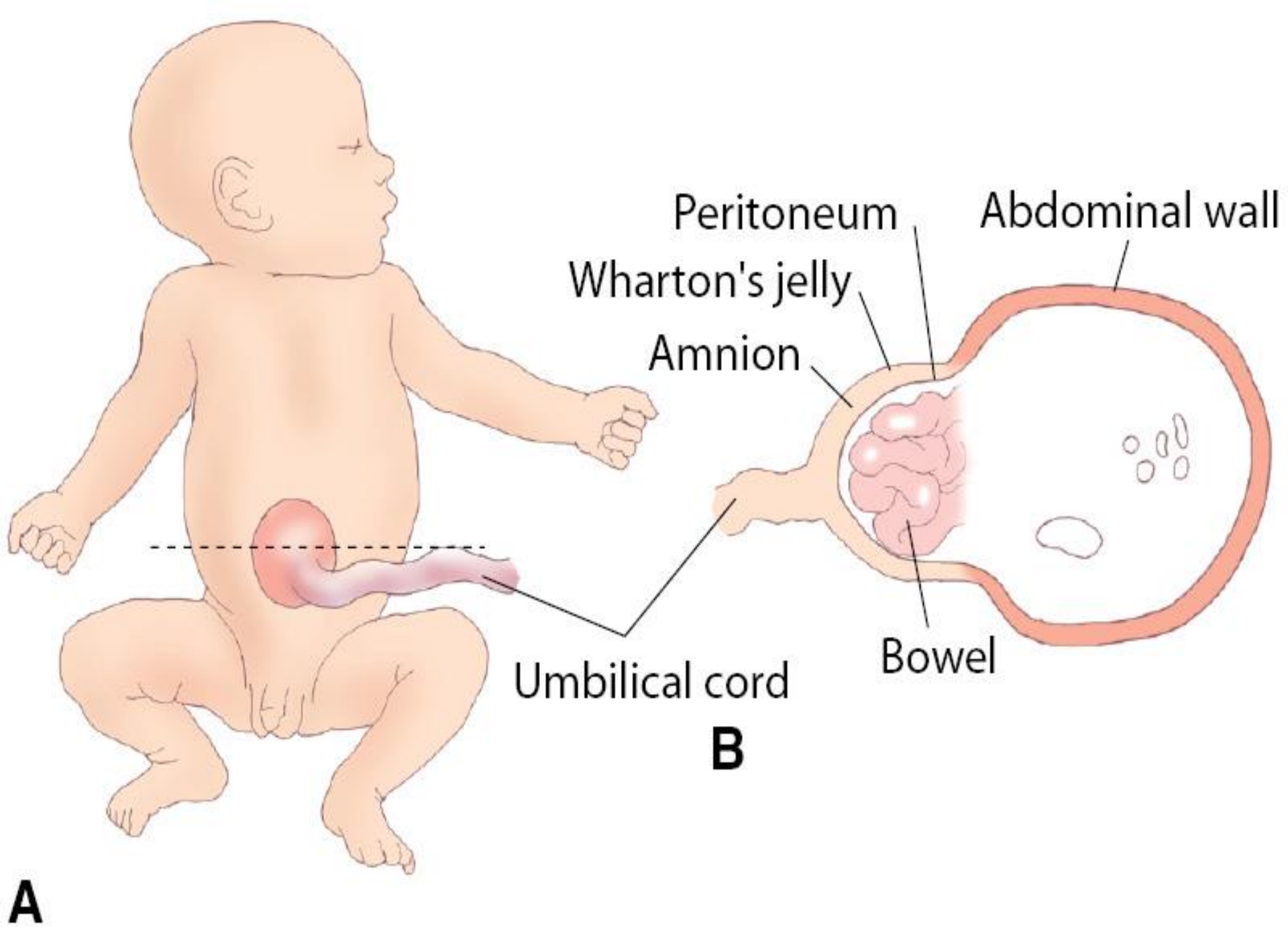
- Look for the presence of a membrane
  - Gastroschisis does not have one
- Look at the umbilical cord
  - Cord goes through the omphalocele
  - Cord usually to the left of the gastroschisis
- Determine which organs are eviscerated
- Determine if the bowel is normal in texture
- Look for other anomalies
  - Omphalocele has a high percentage of chromosome abnormalities

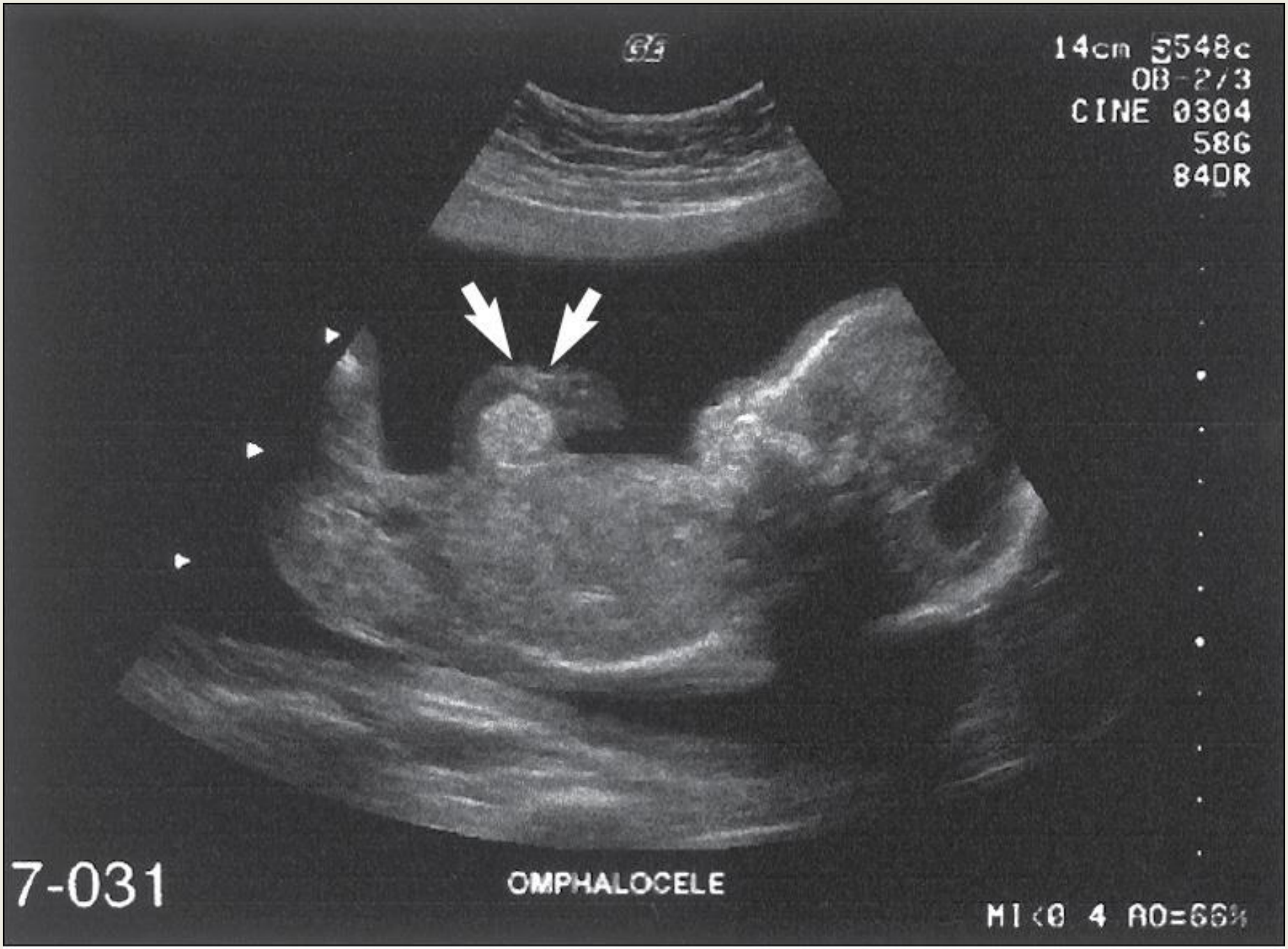
Omphalocele

- During the 8th to 12th weeks in development
  - Fetal bowel normally migrates into the umbilical cord from the abdominal cavity
  - This normal embryological herniation of bowel permits the development of the intraabdominal organs and allows necessary bowel rotation
  - Lack of space within the abdominal cavity because of the liver and kidneys
    - Bowel is forced from the abdomen and into the extraembryonic coelom of the umbilical cord

Omphalocele

- Herniated loops of bowel normally return and rotate into position within the abdominal cavity by the 12th week of pregnancy
- When bowel loops fail to return to the abdomen
  - Bowel-containing omphalocele occurs



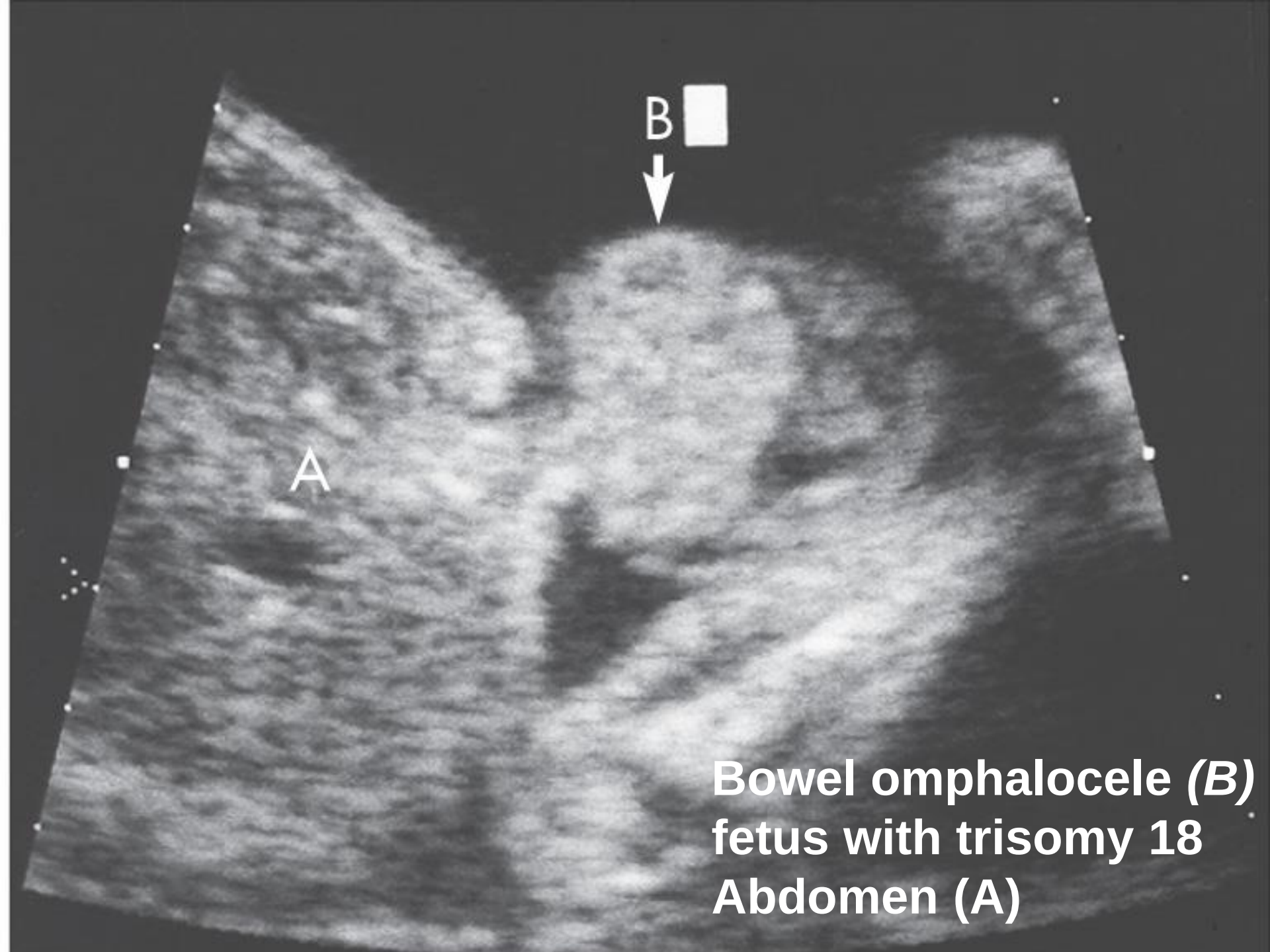


Omphalocele

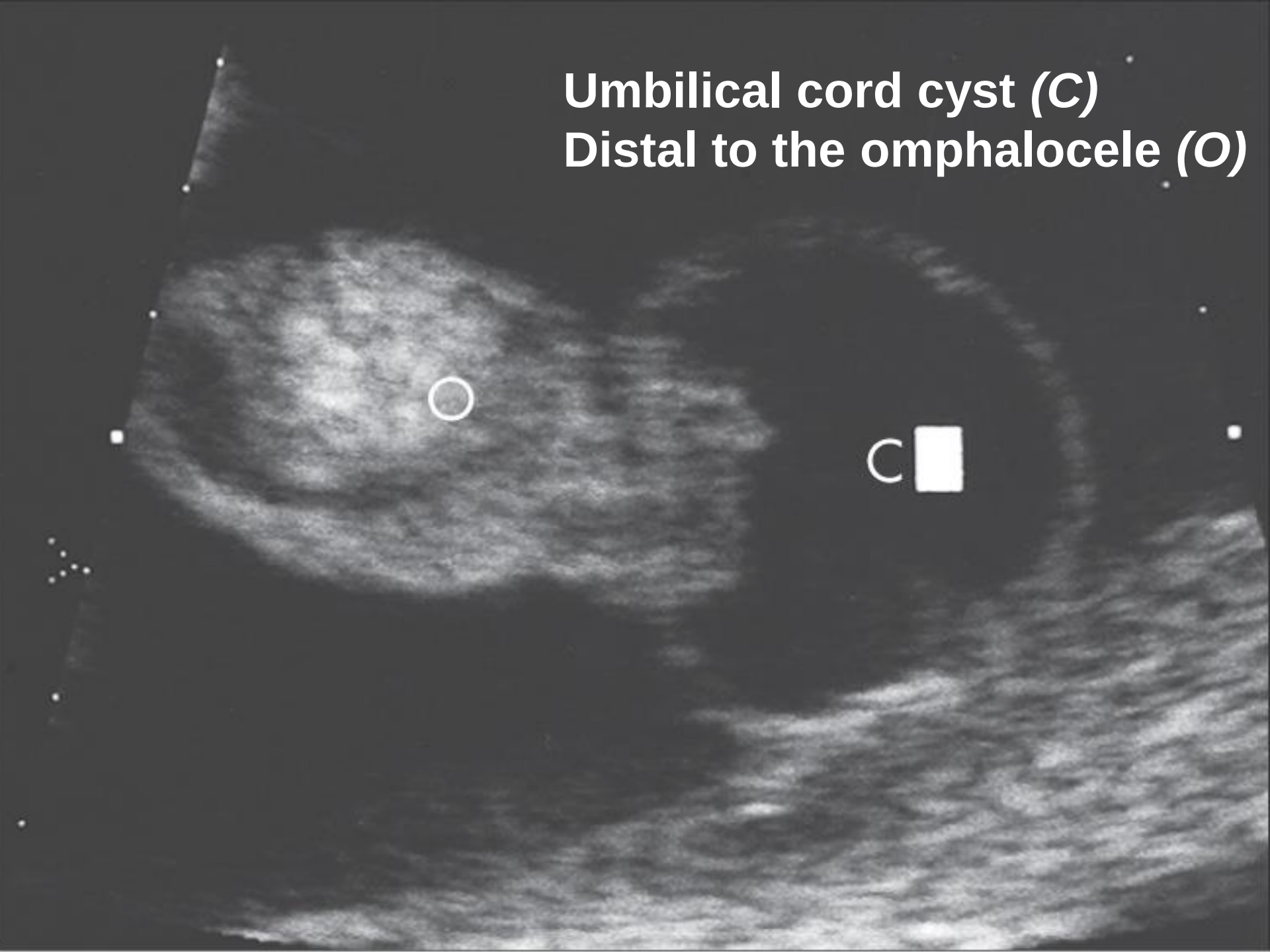
- The alpha-fetoprotein (AFP) level may be slightly elevated or within normal limits
- The omphaloceles are characterized as two types:
  1. Those that contain liver within the sac
  2. Those that contain a variable amount of bowel
     - But no liver

Omphalocele

- Fetuses with omphaloceles that contain bowel only
  - Have a higher risk for
    - Chromosomal abnormalities
    - Other anomalies
- Omphaloceles with bowel develop because the intestine fails to return to the abdomen
  - (primitive body stalk remains)

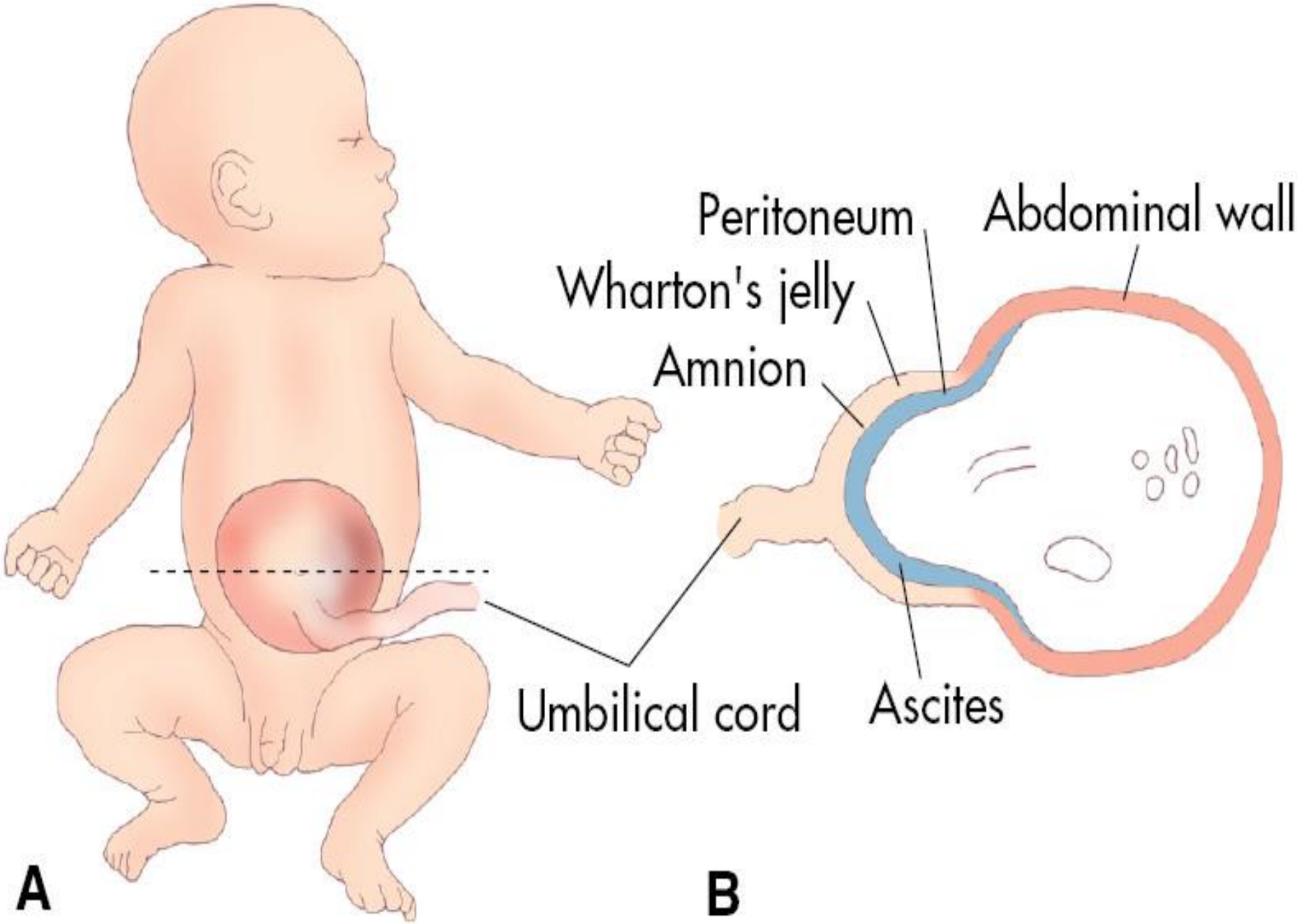


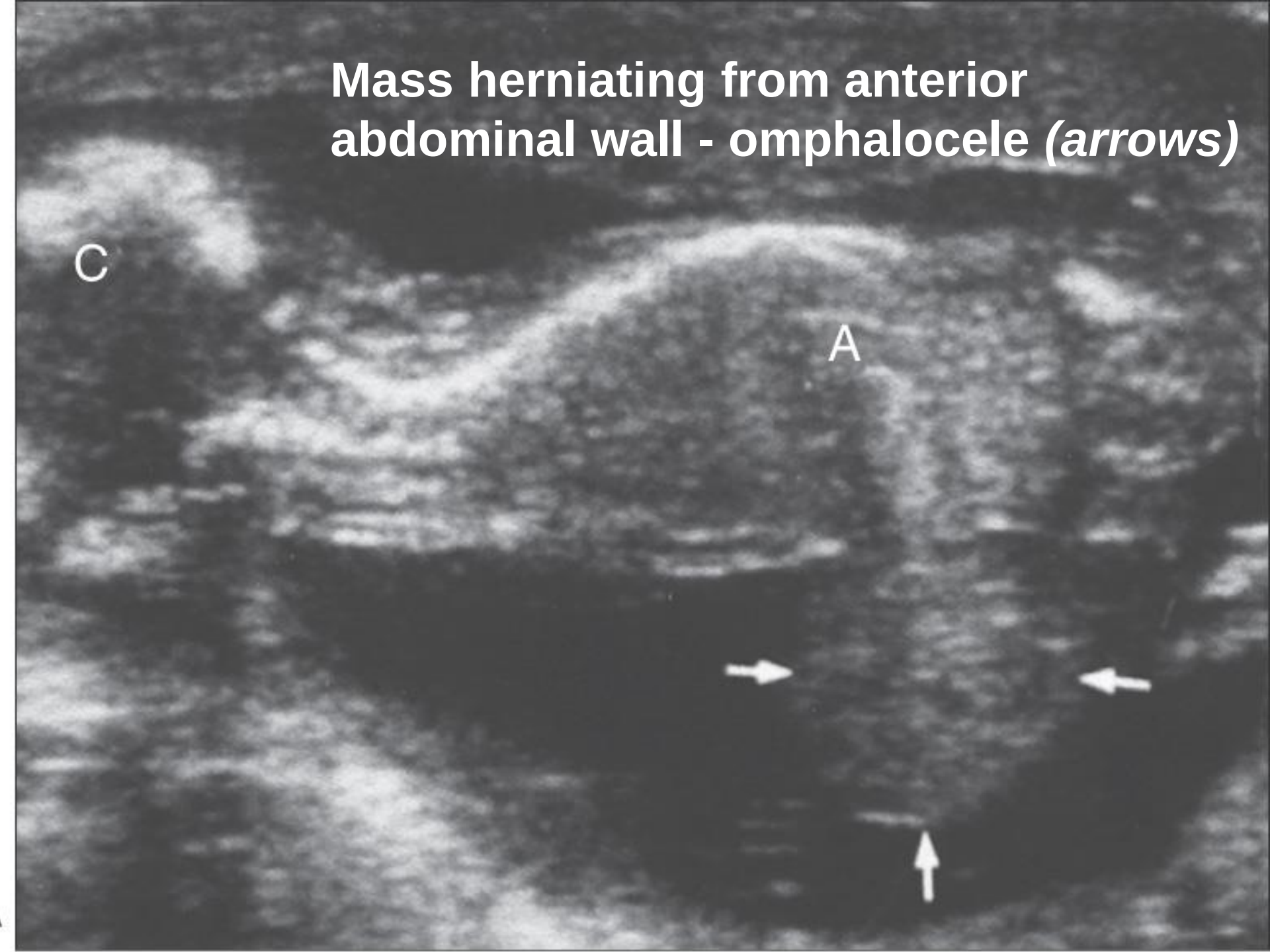
Bowel omphalocele *(B)* fetus with trisomy 18 Abdomen (A)

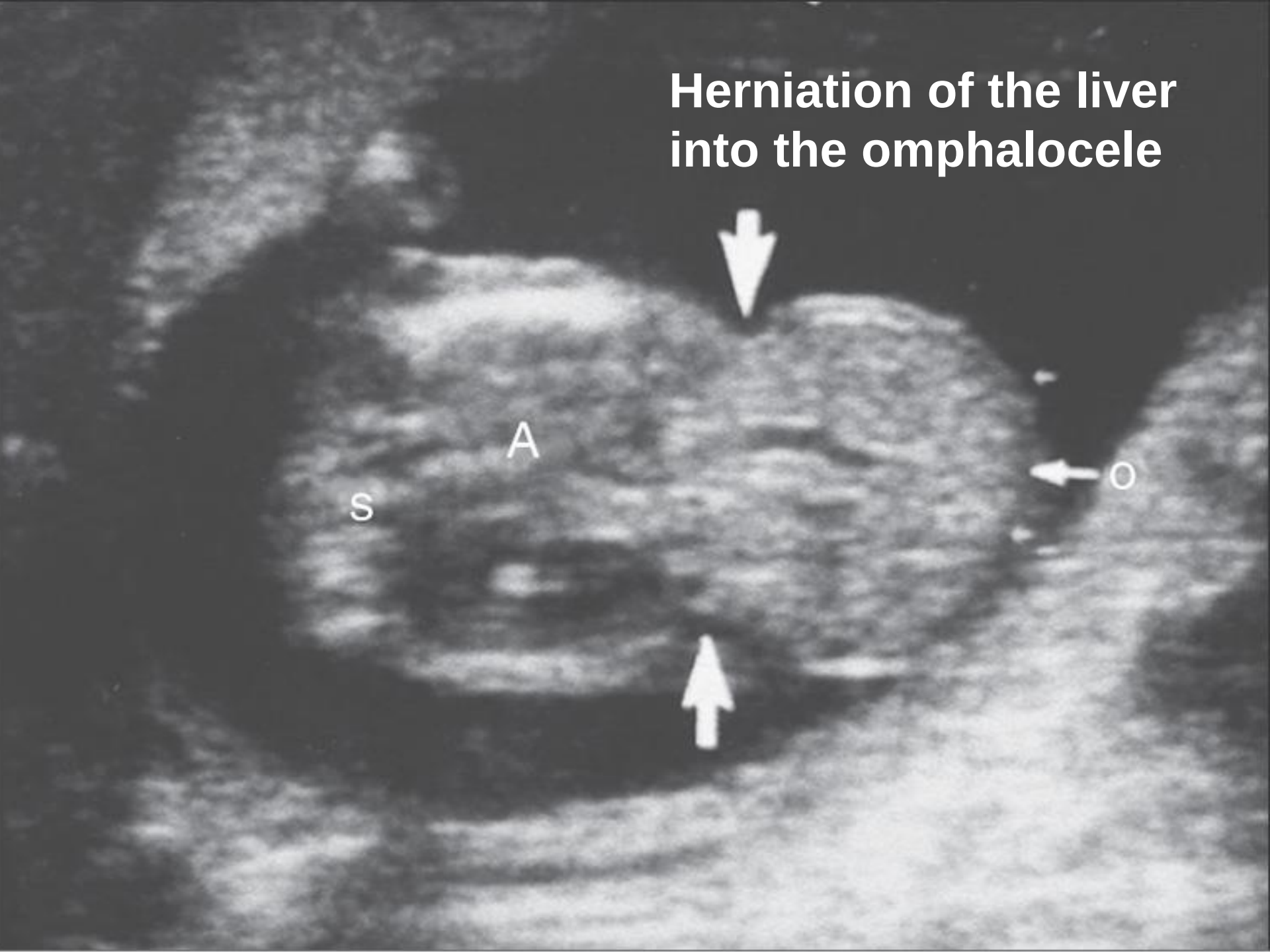

Umbilical cord cyst *(C)*
Distal to the omphalocele *(O)*
C

Liver Omphaloceles

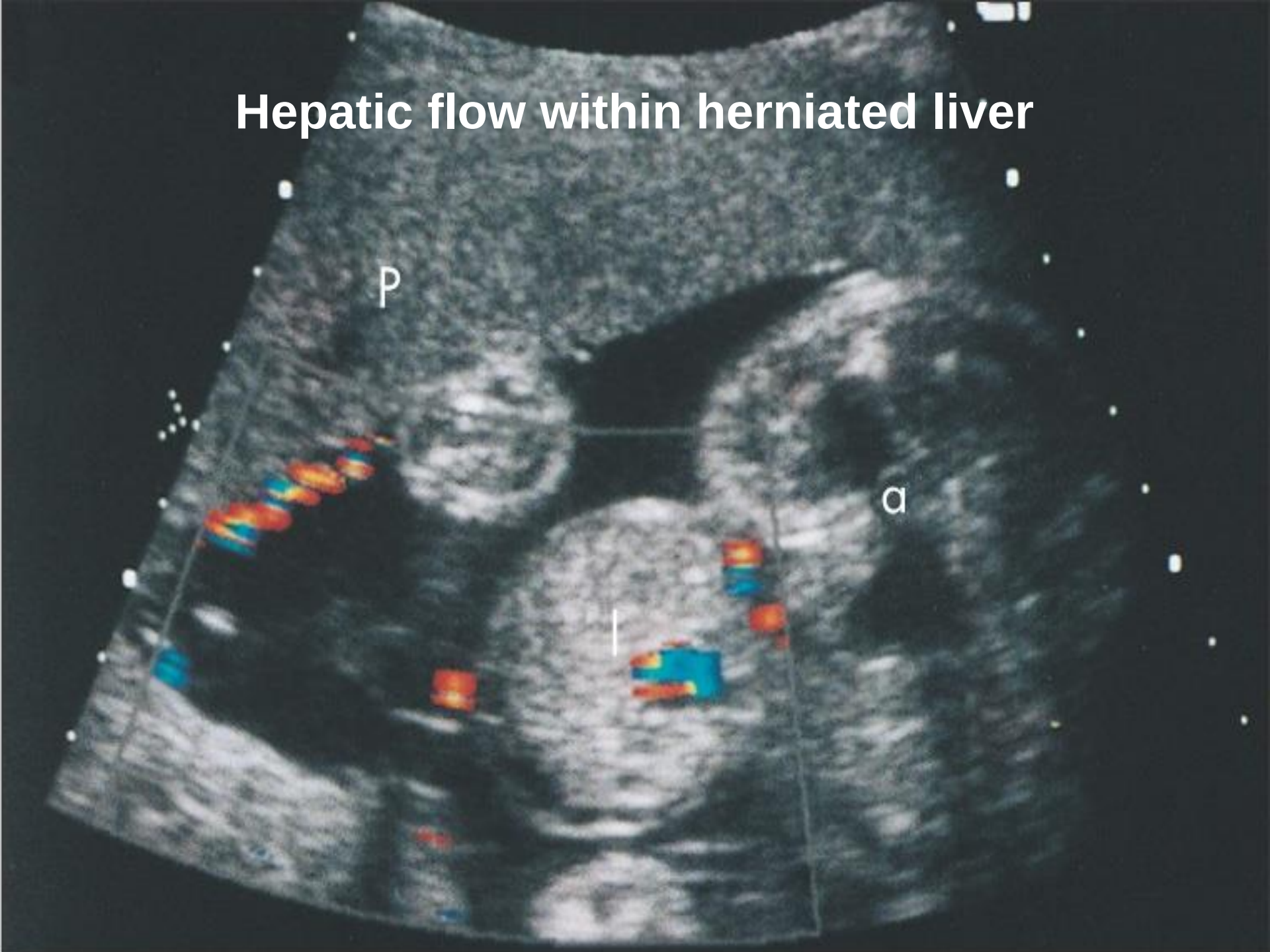
- Represents a developmental defect in abdominal wall closure
  - This type of omphalocele affects the abdominal wall muscles, fascia, and skin
- May contain bowel
- May demonstrate a large abdominal wall defect in comparison to the abdominal diameter

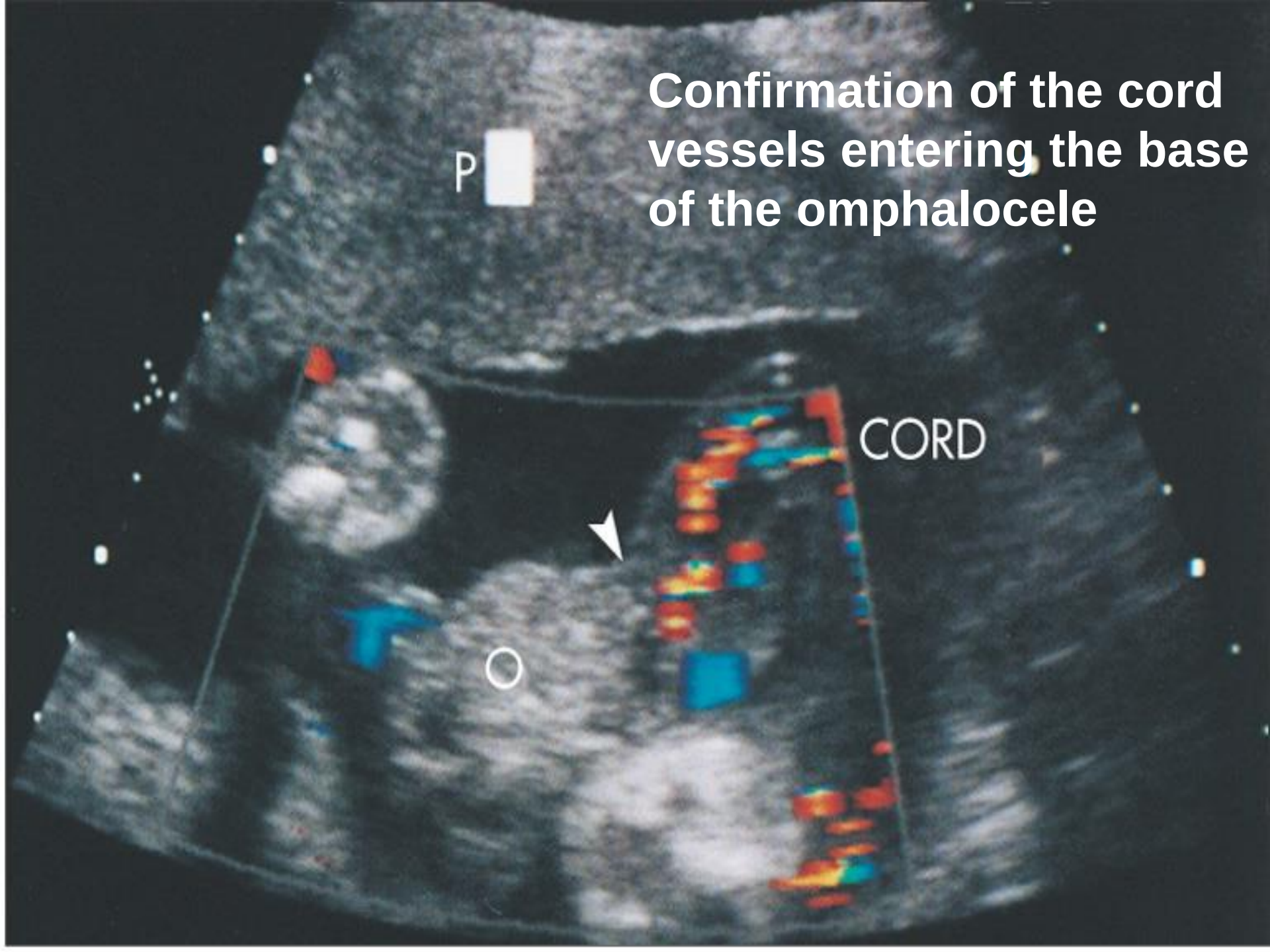



Mass herniating from anterior abdominal wall - omphalocele *(arrows)*
C
A


Herniation of the liver
into the omphalocele
A
S
O

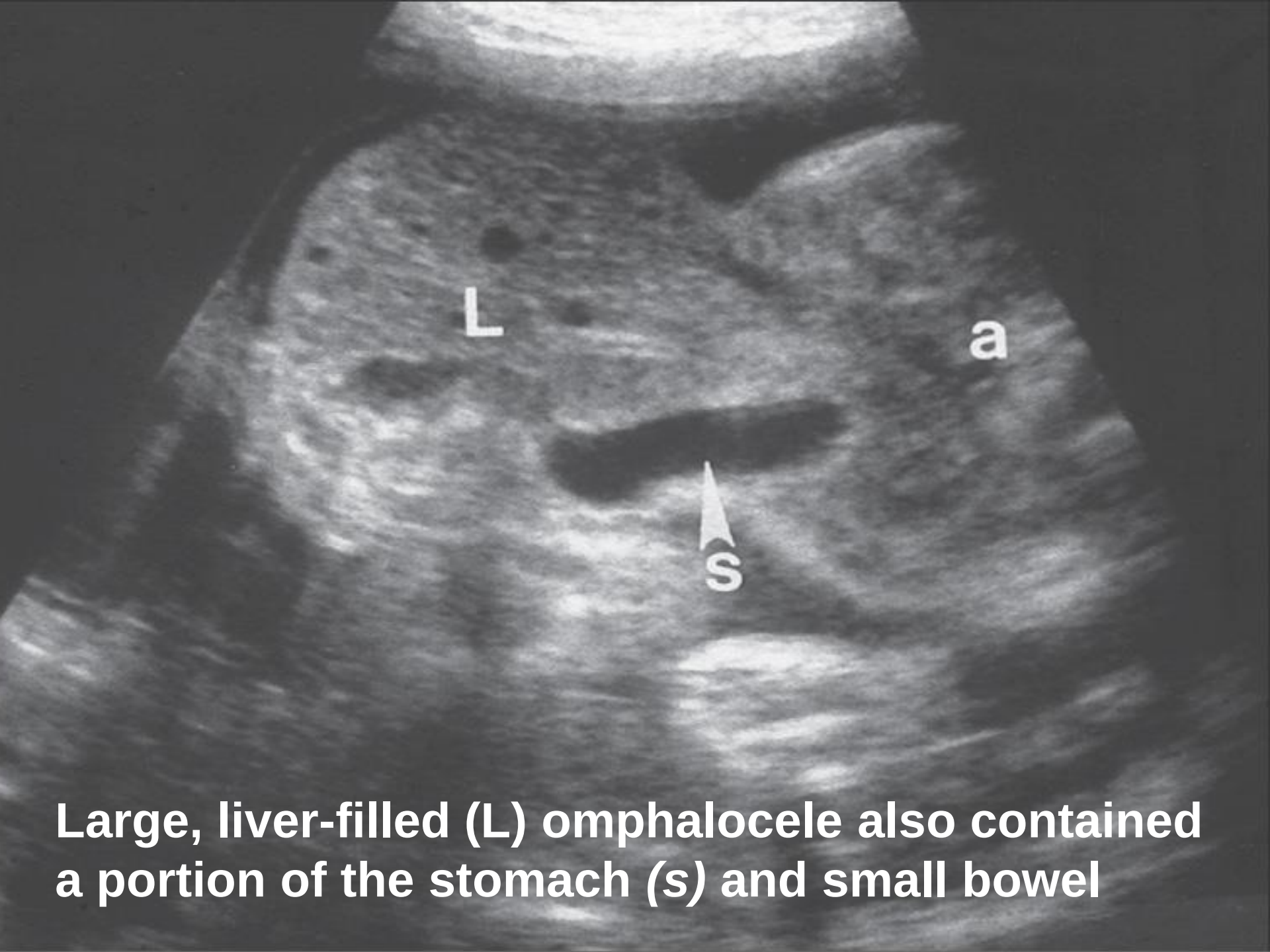
Hepatic flow within herniated liver



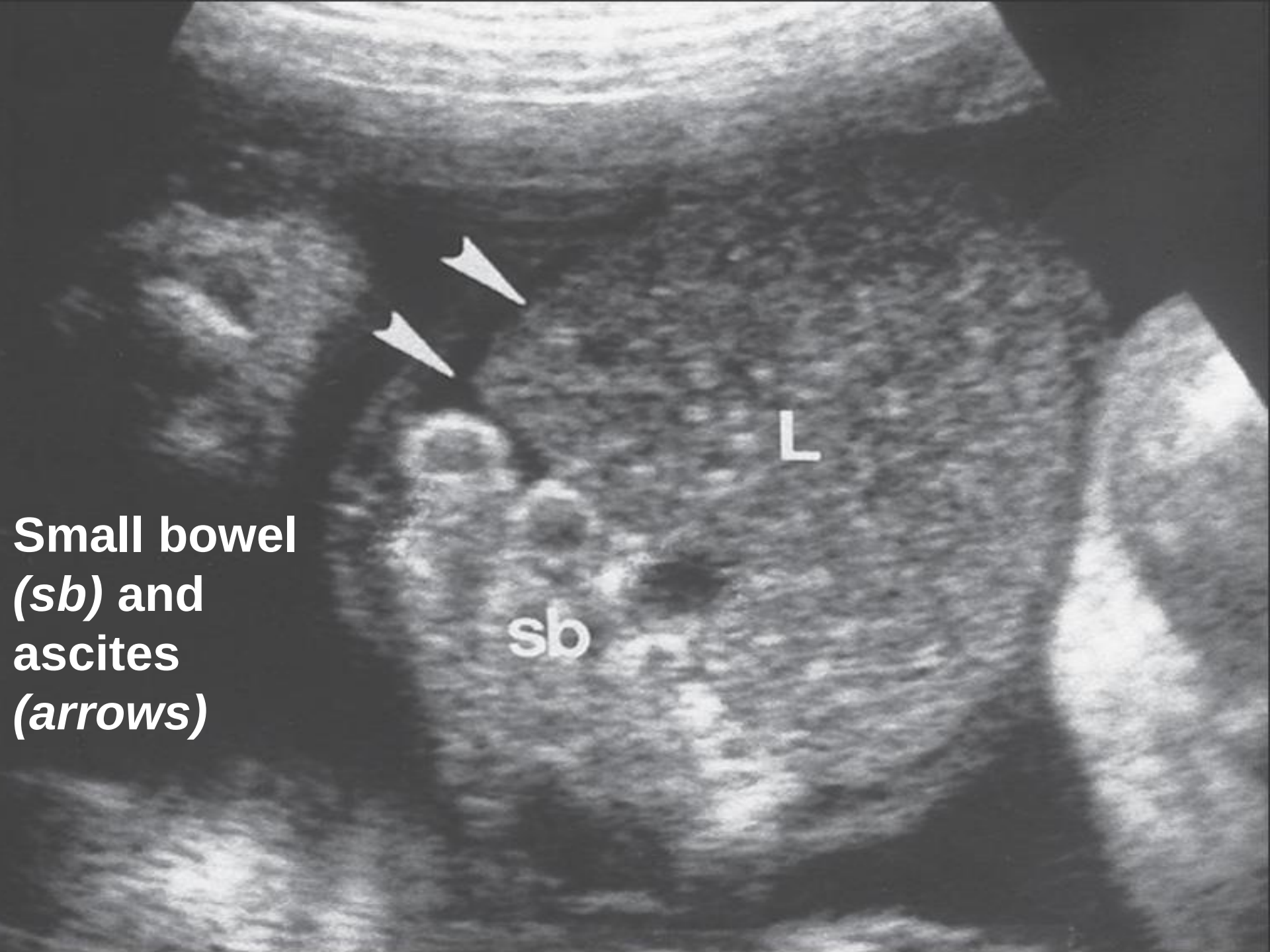

Confirmation of the cord vessels entering the base of the omphalocele
P
CORD

Omphaloceles
Sonographic Findings

- Central abdominal wall defect with evisceration of the bowel or a combination of liver and bowel into the umbilical cord
  - Color-flow imaging may aid in viewing the continuity of the umbilical cord into the omphalocele
  - Stomach may be involved
  - Bowel omphaloceles appear echogenic and must be distinguished from umbilical hernia (covered by skin and fat)



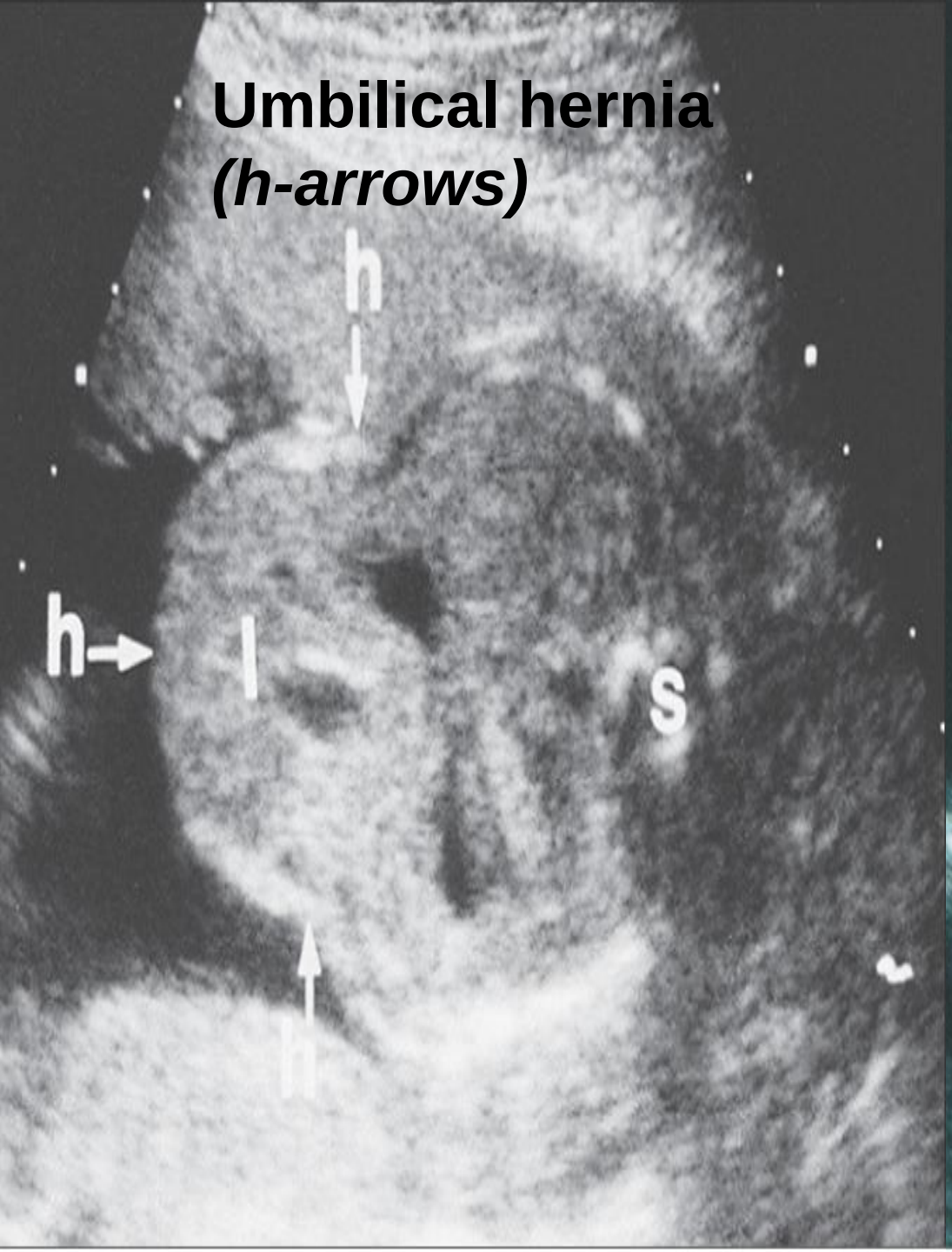
Large, liver-filled (L) omphalocele also contained a portion of the stomach *(s)* and small bowel



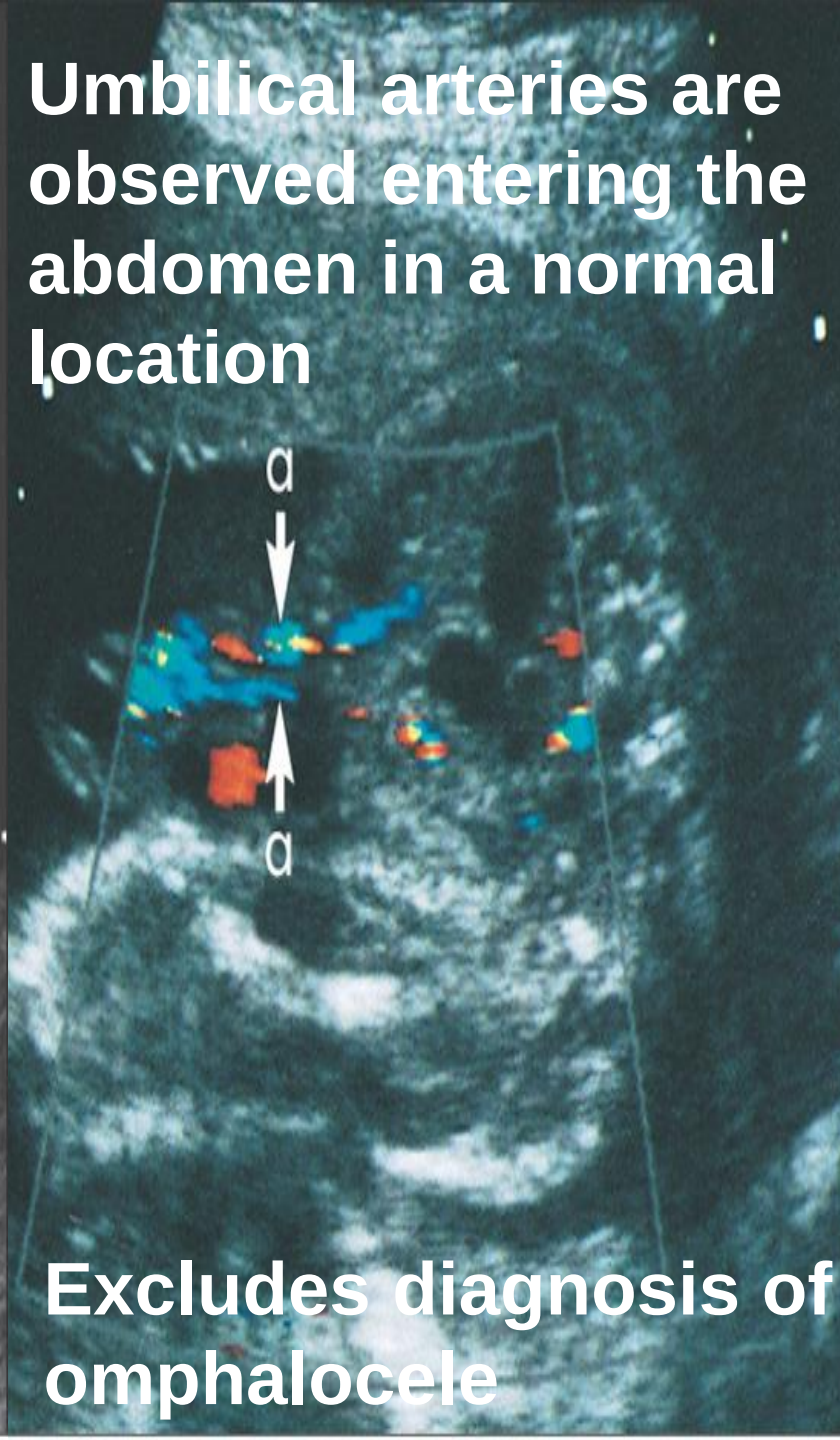
Small bowel *(sb)* and ascites *(arrows)*

Omphaloceles
Sonographic Findings

- Umbilical hernias may be confused with liver omphaloceles
  - Normal cord insertion suggests hernia
- Ascites may coexist with omphalocele
- Polyhydramnios found in one third of fetuses

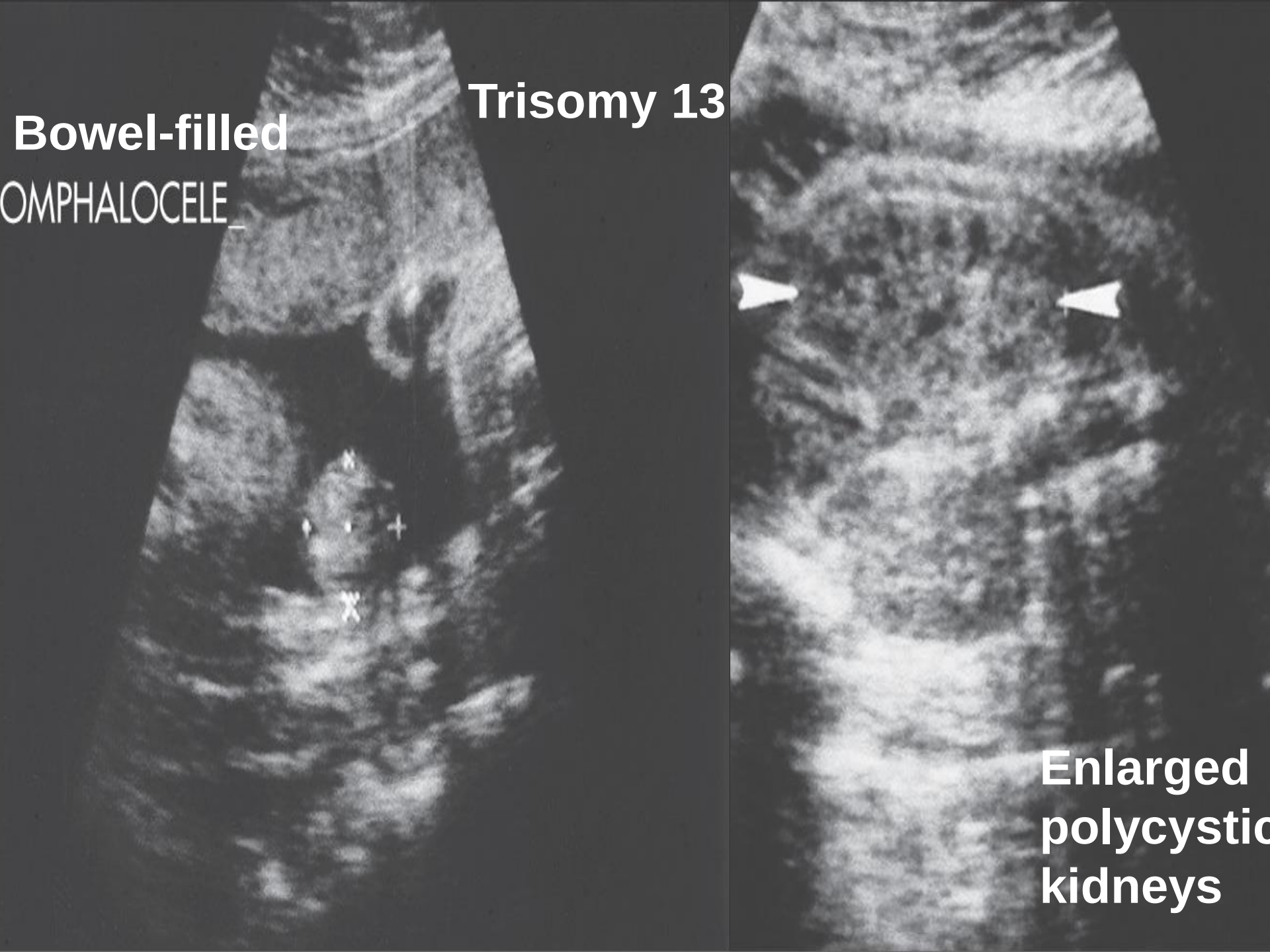


Umbilical hernia *(h-arrows)*



Umbilical arteries are observed entering the abdomen in a normal location

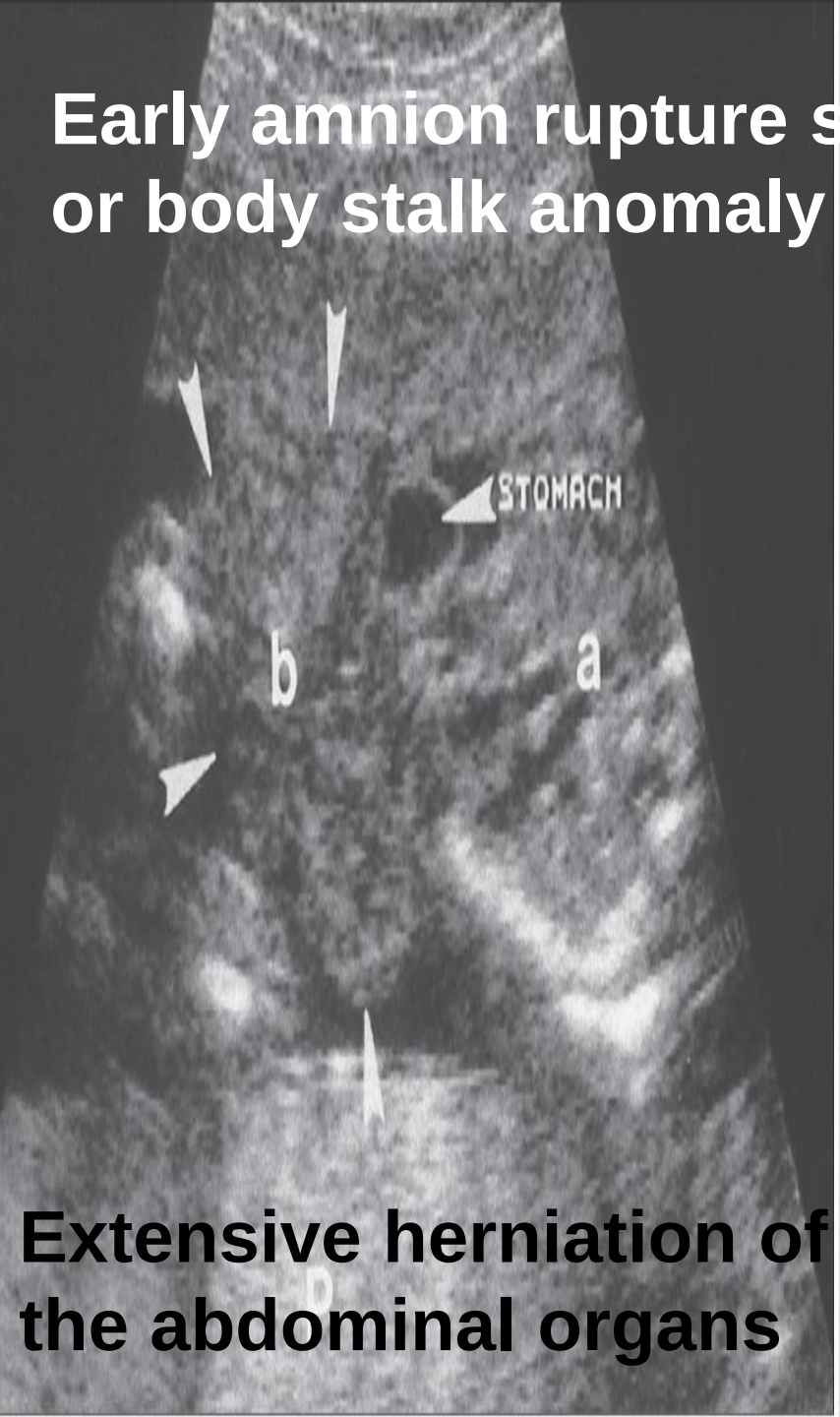
Excludes diagnosis of omphalocele


Trisomy 13
Bowel-filled
OMPHALOCELE_
Enlarged
polycystic
kidneys

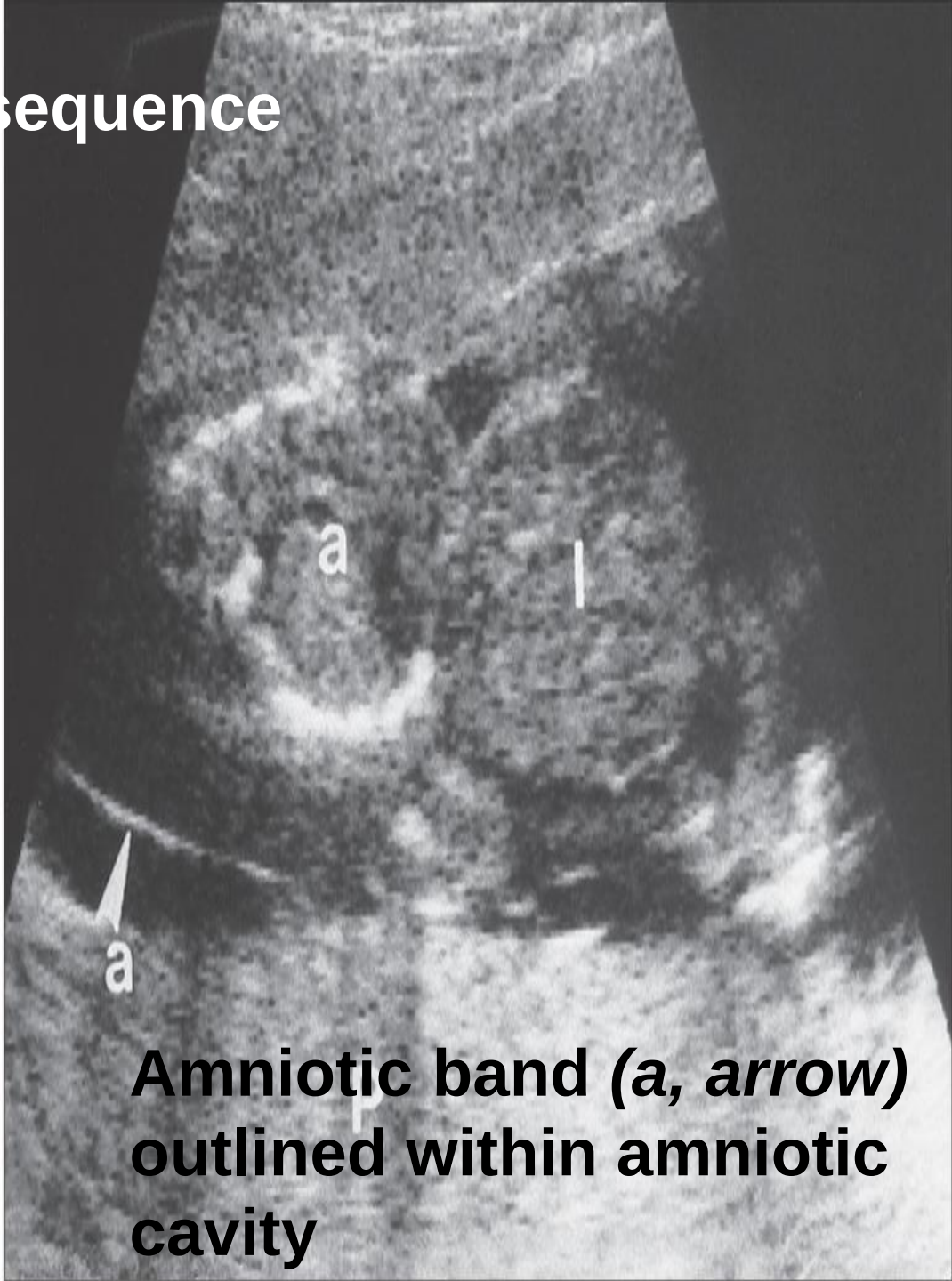
Omphaloceles
Sonographic Findings

- When scoliosis is found
  - Consider limb–body wall complex
    - Lethal disorder
      - Also includes severe defects
        - Acrania
        - Encephalocele
        - Facial clefts
        - Extensive abdominal wall defect of the chest
        - Abdomen and limb defects
    - Abnormal fusion of the amnion and chorion extends as a sheet from the cord and adheres to the fetus and placenta

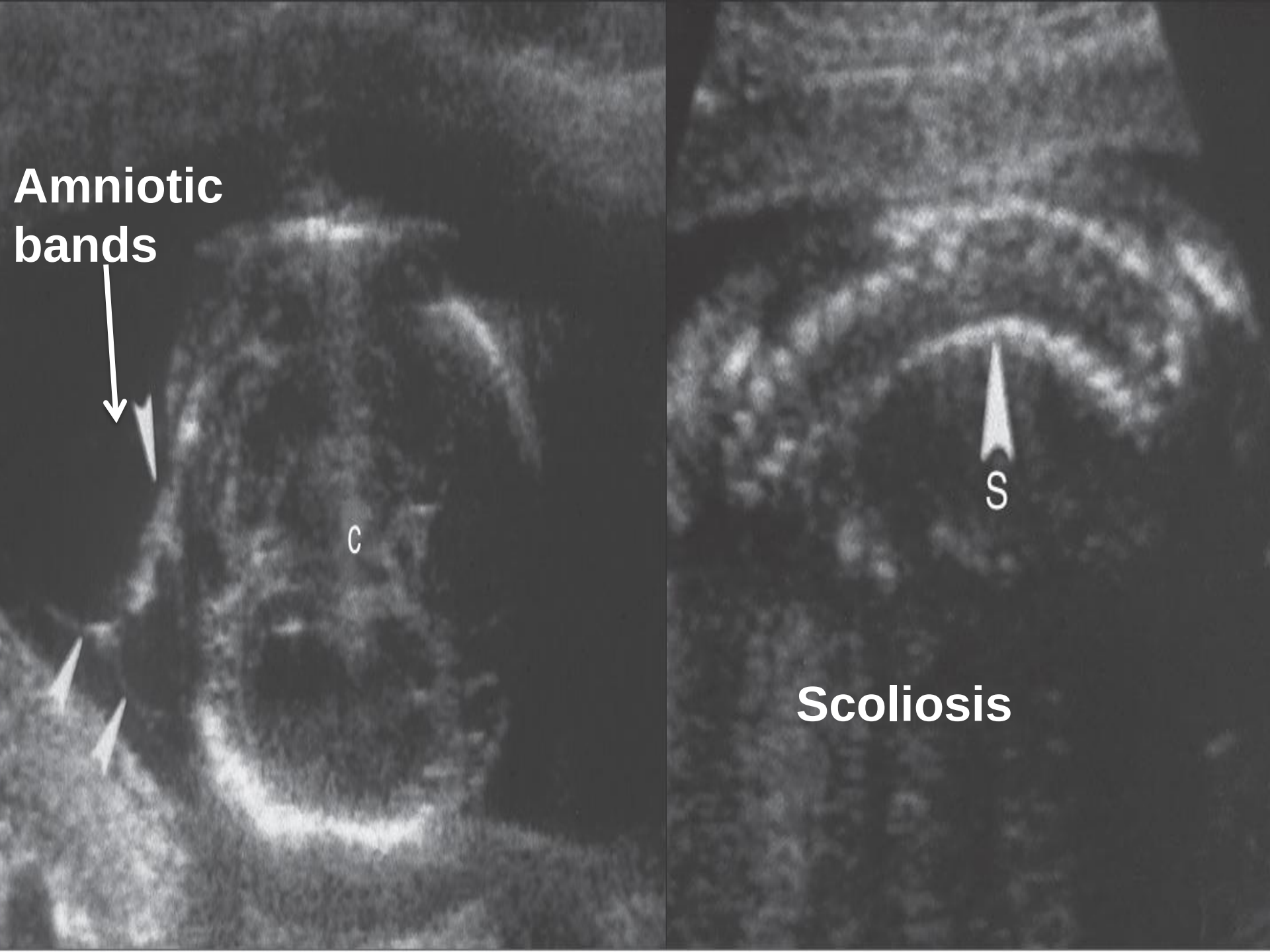
Early amnion rupture sequence or body stalk anomaly



Extensive herniation of the abdominal organs



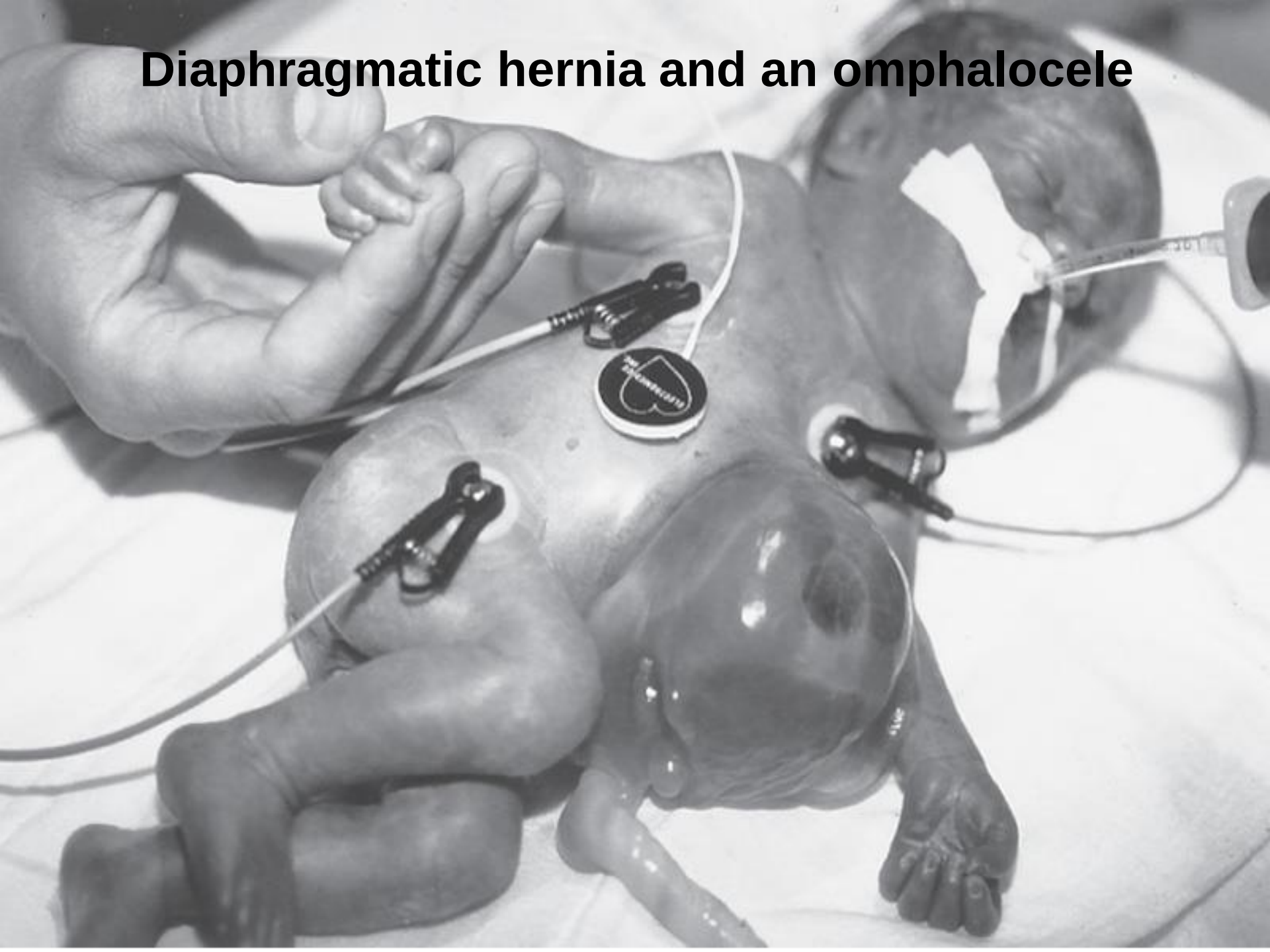
Amniotic band *(a, arrow)* outlined within amniotic cavity

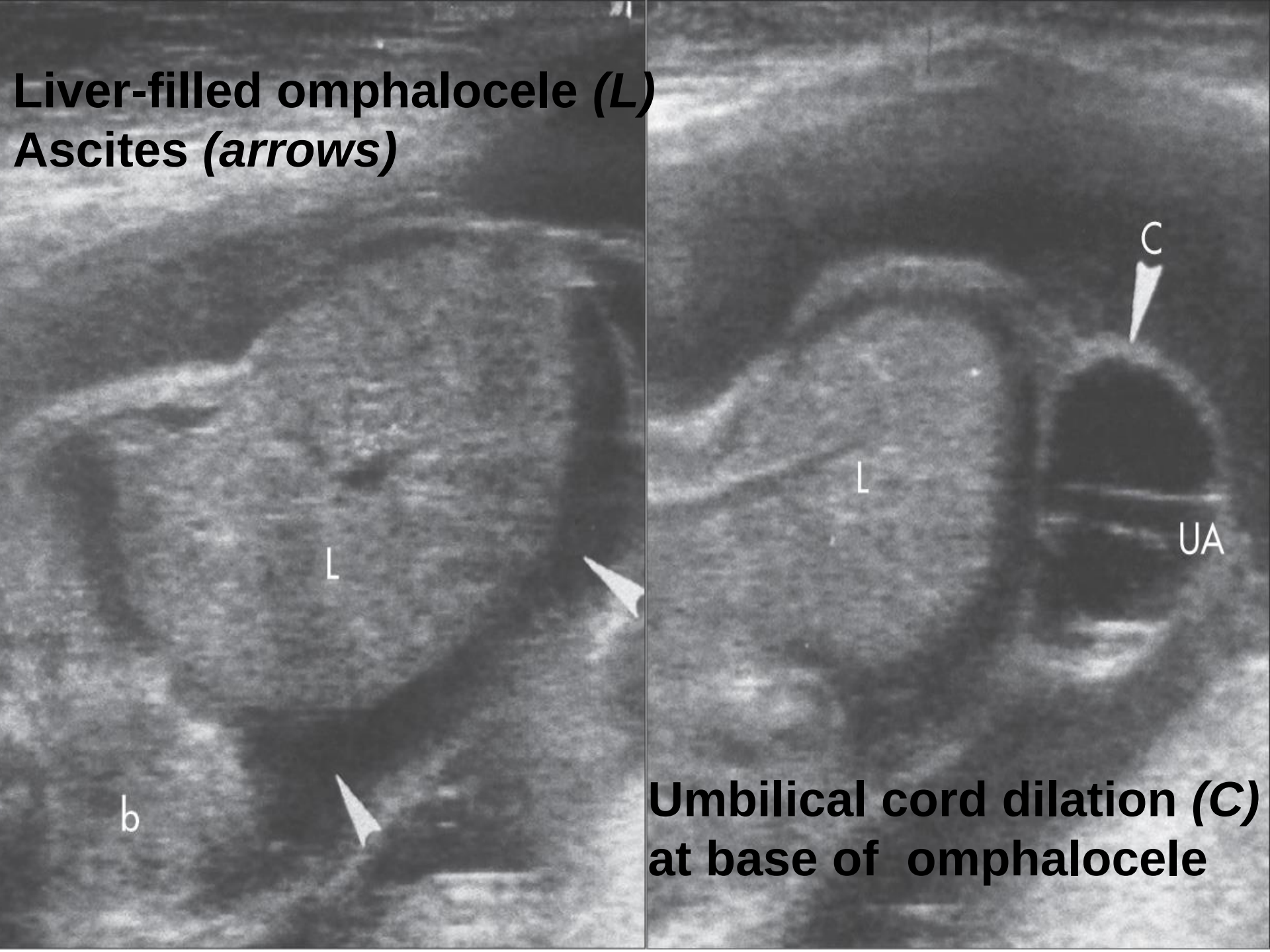

Amniotic bands
C
S
Scoliosis

Omphaloceles Sonographic Findings

- Amniotic band syndrome may represent a milder form of limb–body wall complex
  - Predicted by amniotic bands (fibrous tissue strands) that entangle or amputate fetal parts
  - Common findings
    - Facial clefts
    - Asymmetric encephaloceles
    - Constriction or amputation defects of the extremities
    - Clubfoot deformities

Diaphragmatic hernia and an omphalocele




Liver-filled omphalocele *(L)*
Ascites *(arrows)*
L
b
C
L
UA
Umbilical cord dilation *(C)*
at base of omphalocele

Omphaloceles Sonographic Findings

- Consider bladder or cloacal exstrophy when a low omphalocele is observed
- Other anomalies may include
  - Anal atresia
  - Spina bifida
  - Lower limb defects

Omphaloceles Sonographic Findings

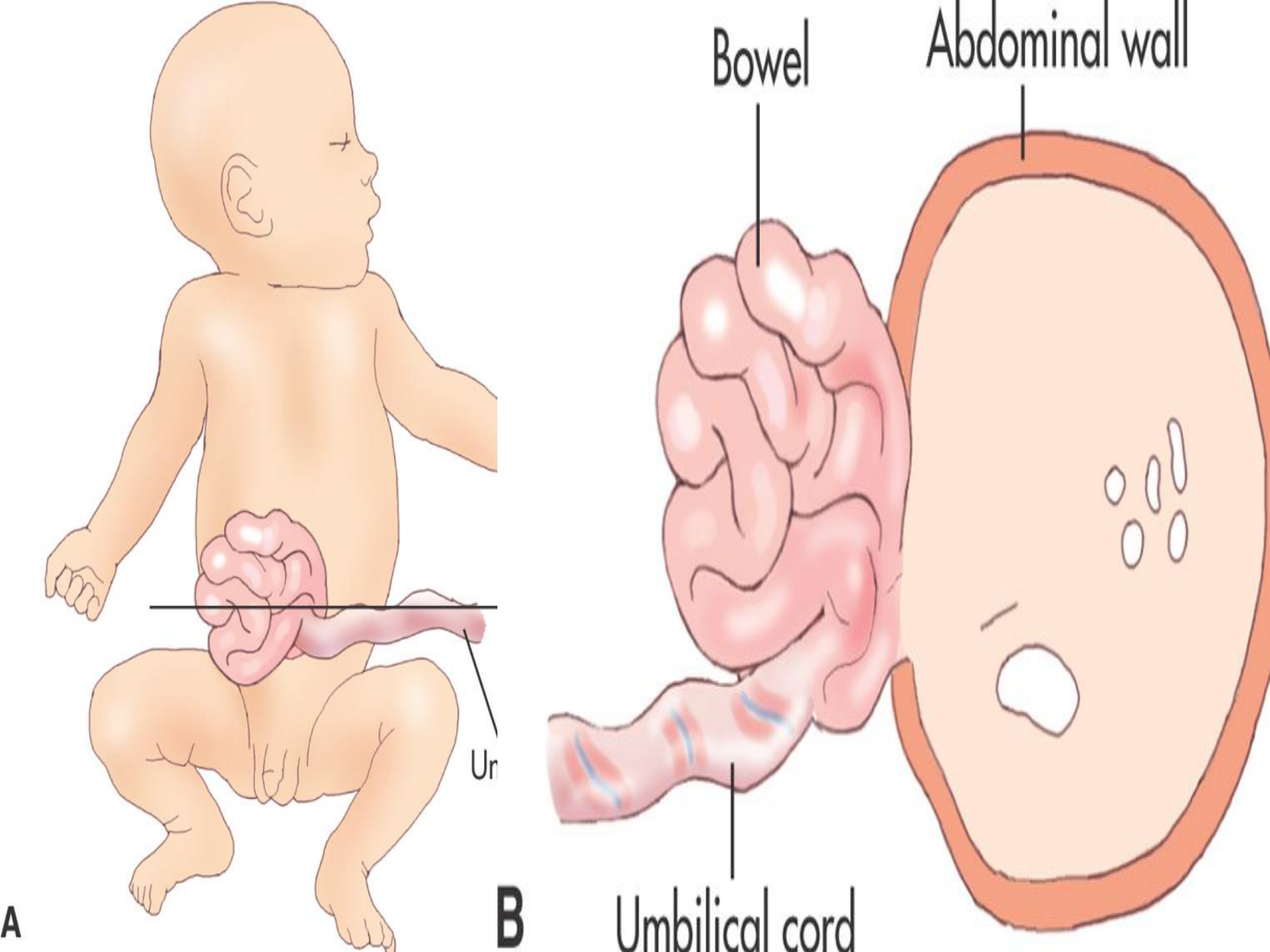
- When organomegaly and macroglossia are observed
  - Beckwith-Wiedemann syndrome is suspected
    - (occurs in 12% of infants with an omphalocele)

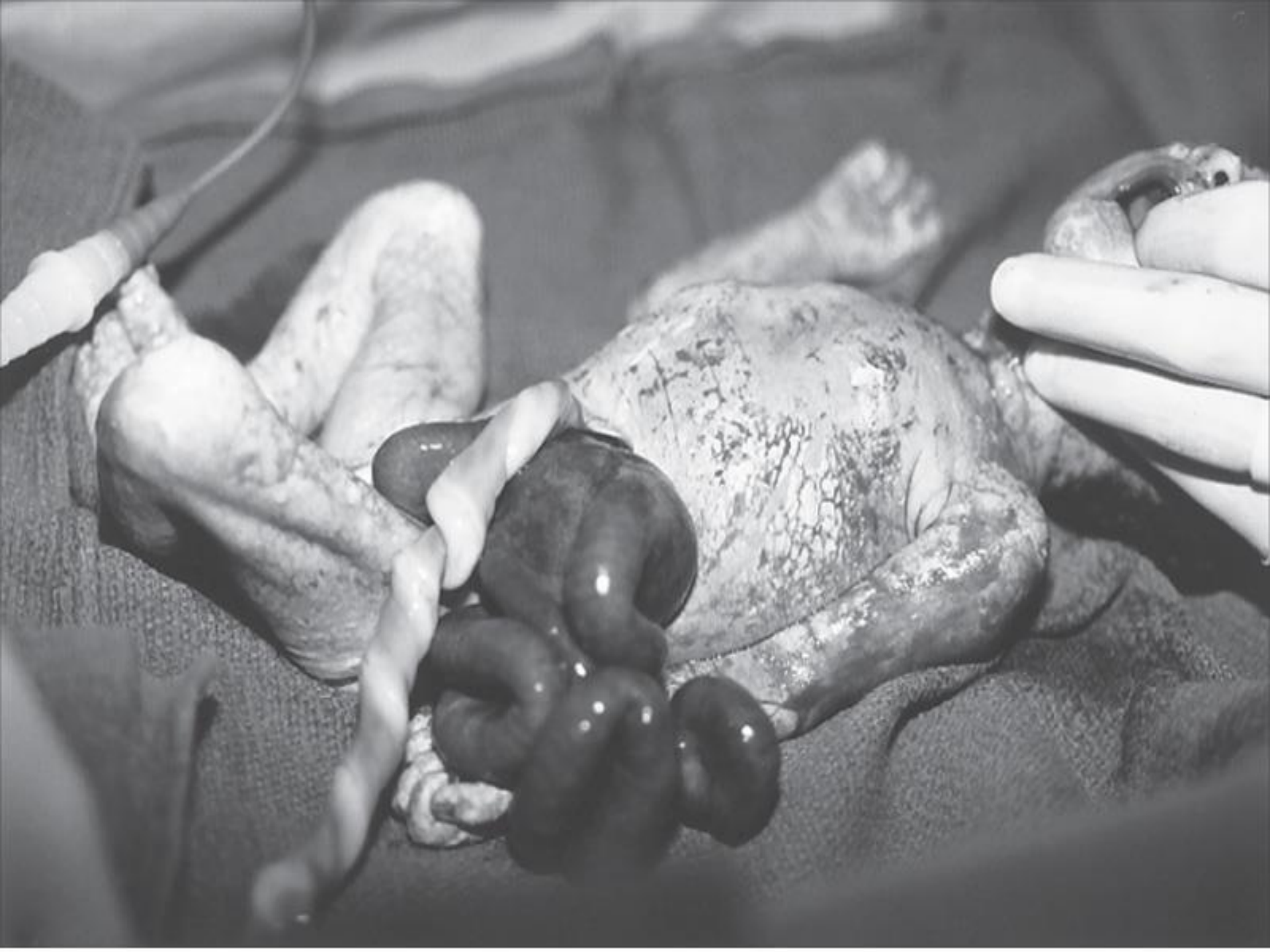
Omphaloceles

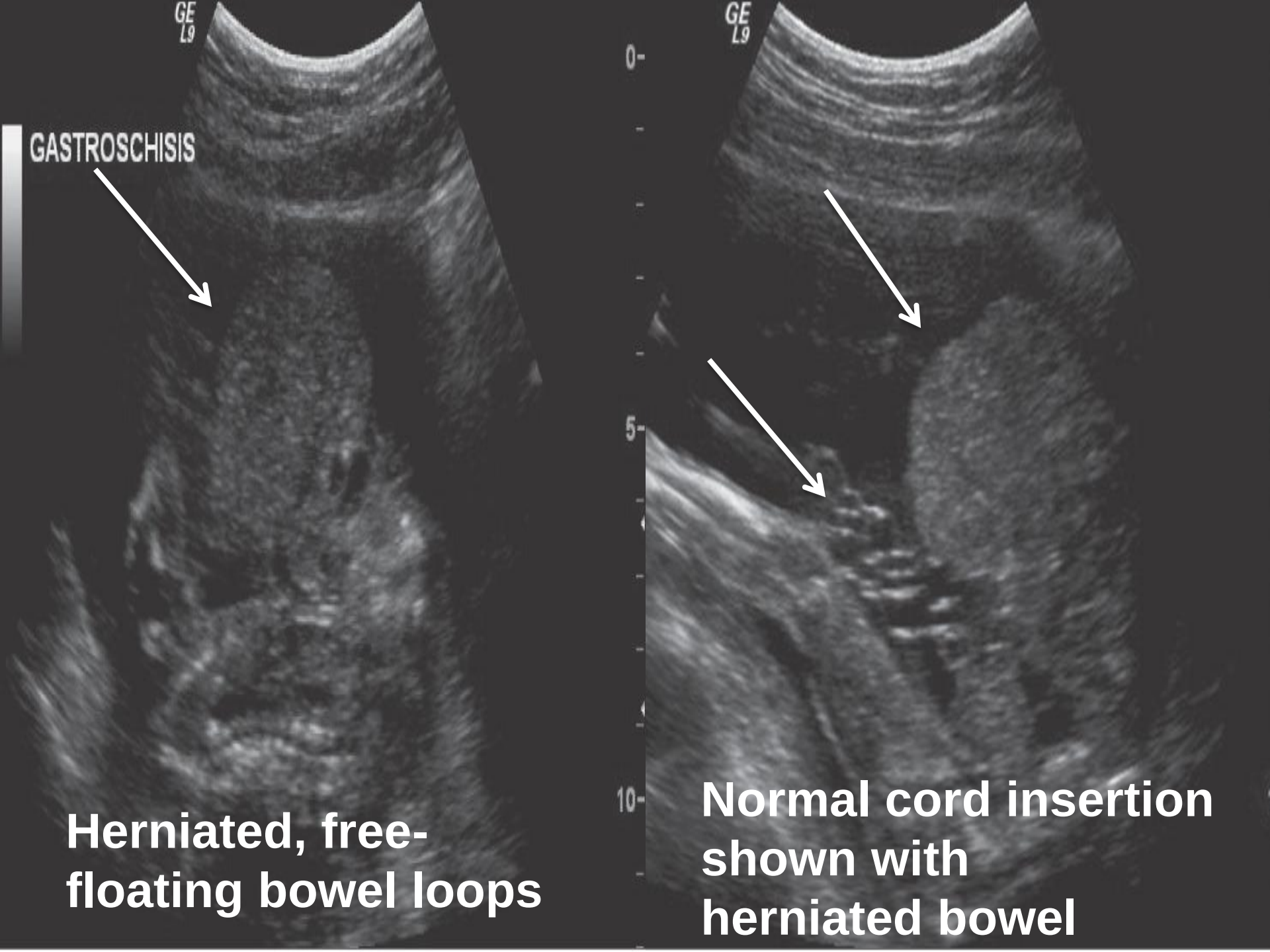
- Prognosis varies according to the extent of the primary defect and associated structural and chromosomal abnormalities
- Perinatal mortality approaches 80% when more than one fetal abnormality exists
  - Almost all infants die when there is a chromosomal or major heart defect
- Mortality rate is approximately 10% with an isolated omphalocele (no other anomalies)

Gastroschisis

- An opening in the layers of the abdominal wall with evisceration (herniation) of the bowel
  - Infrequently eviscerated
    - Stomach
    - Genitourinary organs


Bowel
Abdominal wall
Umbilical cord
A
B




GE
L9
GASTROSCHISIS
GE
L9
0
5
10
Herniated, free-floating bowel loops
Normal cord insertion shown with herniated bowel

Gastroschisis

- It is thought that gastroschisis is a consequence
  - Atrophy of the right umbilical vein
  - Disruption of the omphalomesenteric artery
- Usually not known to be genetically transmitted
  - Recurrence risk for gastroschisis estimated at 3.5%

Gastroschisis

- Defects are small (2–4 cm in size)
- Located next to the normal cord insertion
- Majority of cases
  - Defect is to the right of the umbilical cord
- Insertion of the umbilical cord is normal
- Small bowel is always found in the herniation

Gastroschisis

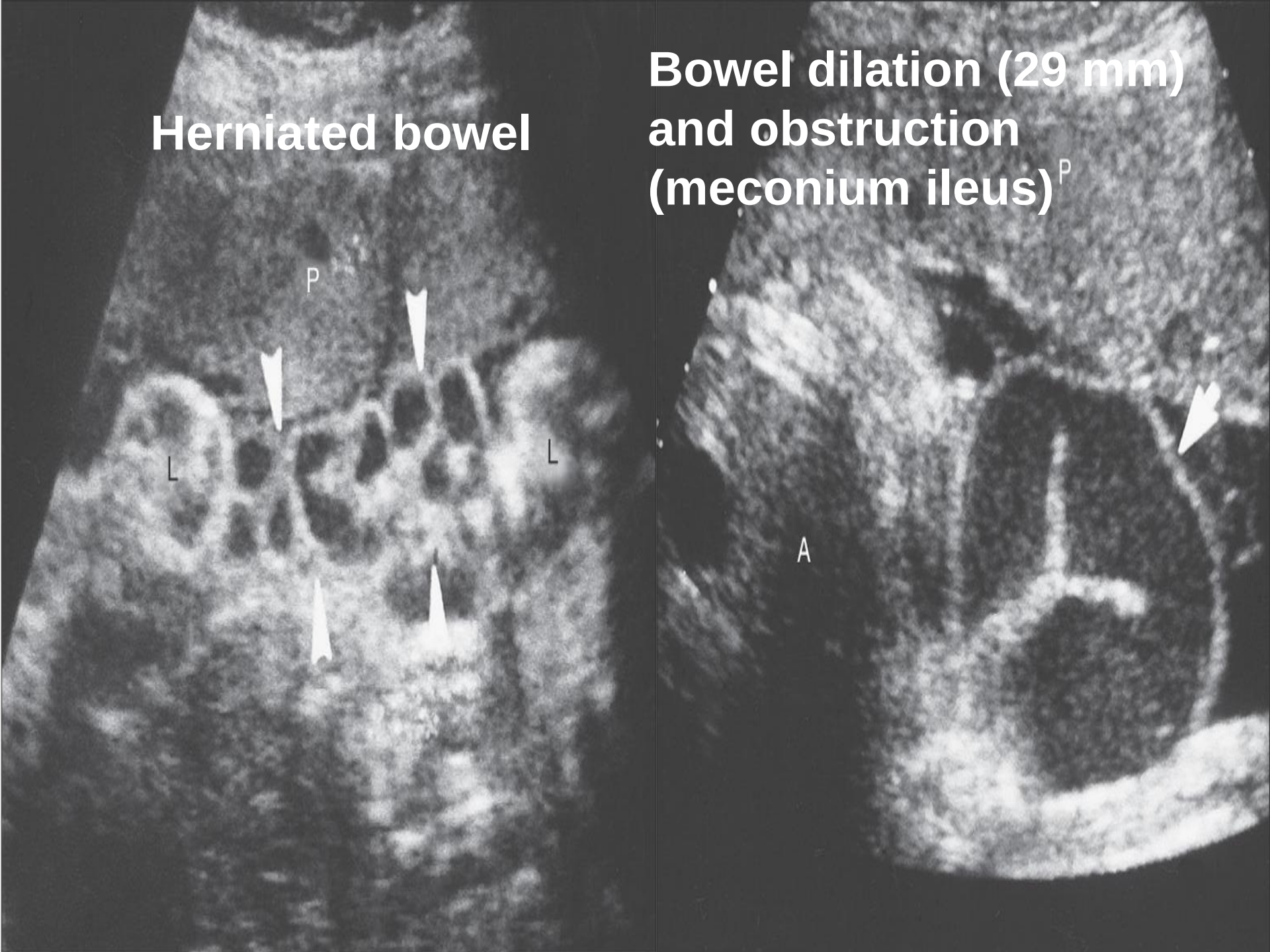
- Other organs that may be involved in the herniation include
  - Large bowel
  - Stomach
  - Occasionally portions of the genitourinary system
  - Rarely liver

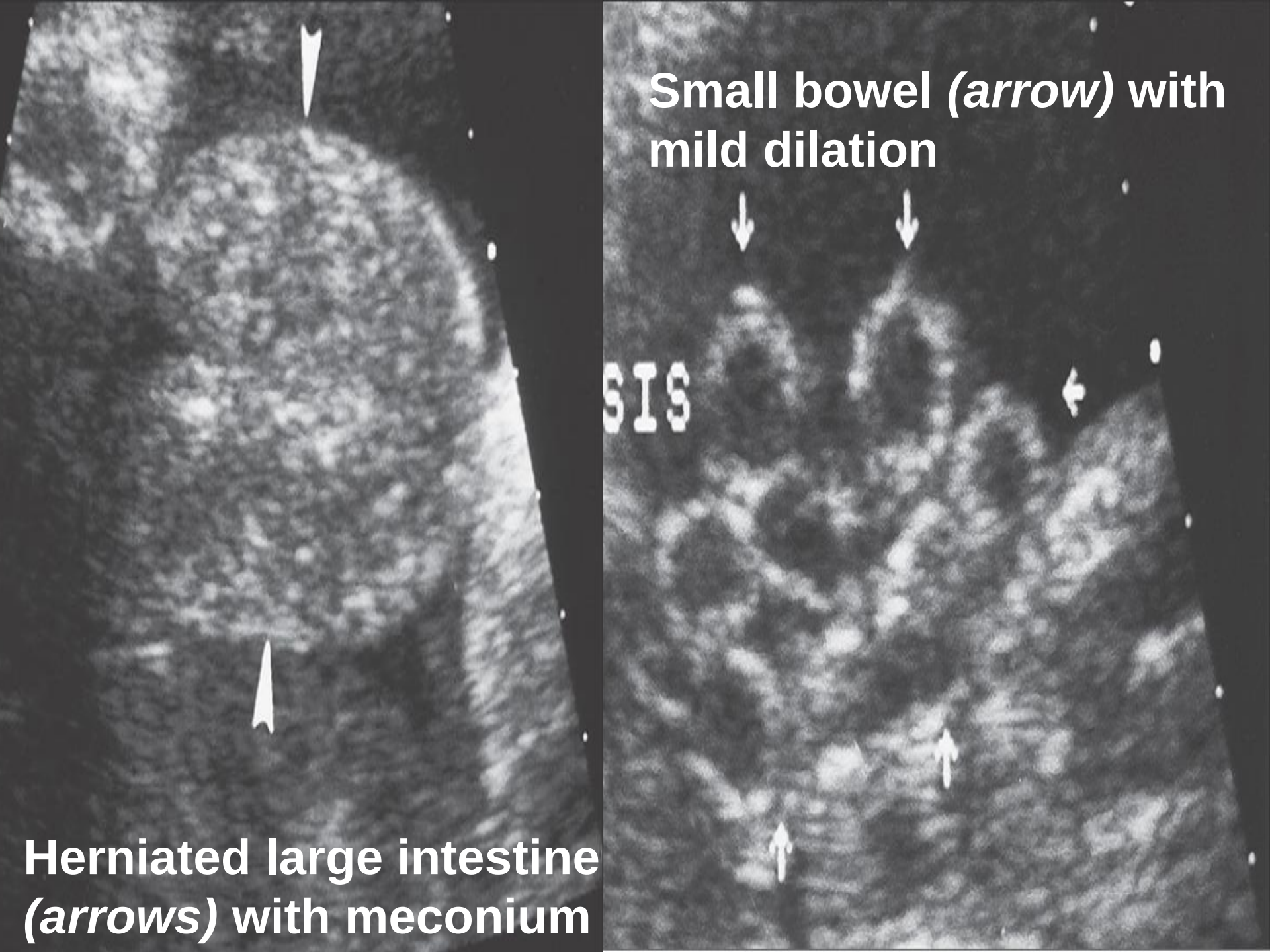
Gastroschisis

- AFP levels are significantly higher in gastroschisis compared with omphalocele
  - Due to the exposed bowel without a covering membrane
- May be able to detect after 12 weeks of gestation
- Patient presents with a markedly elevated maternal serum AFP level

Gastroschisis

- Evaluate
  - Area of umbilical cord insertion
    - Multiple loops of bowel (small bowel and often colon) may be seen outside the abdominal cavity in the area of the cord
- Cord is normally inserted into the abdominal wall and the defect is most always to the right of the umbilical cord insertion


Herniated bowel
P
L
L
Bowel dilation (29 mm) and obstruction (meconium ileus)
P
A


Small bowel *(arrow)* with mild dilation
SIS
Herniated large intestine *(arrows)* with meconium

Gastroschisis

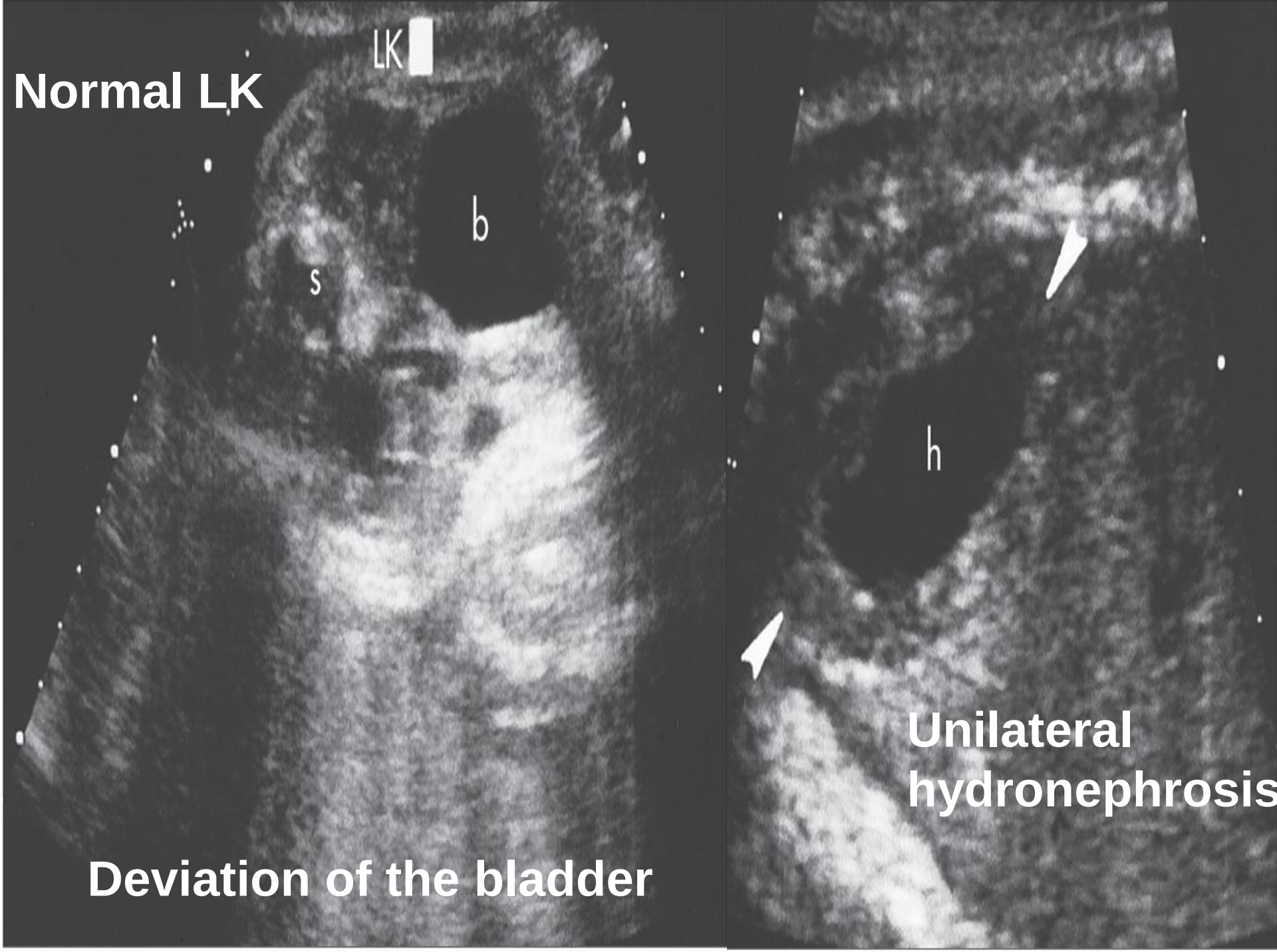
- Edges of bowel are irregular and free floating
  - without a covering membrane
- Ascites is not present in the abdominal cavity

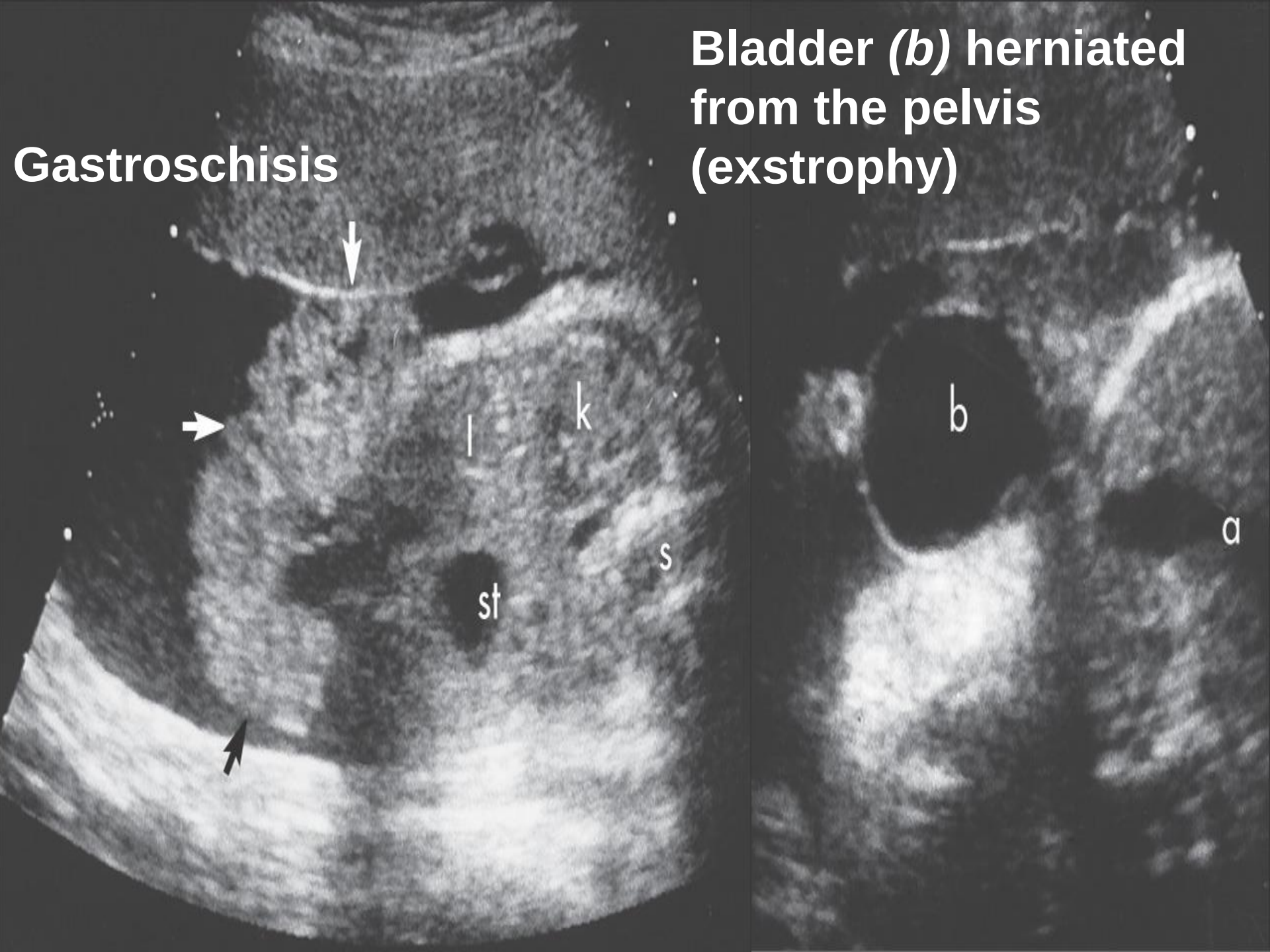
Gastroschisis Sonographic Findings

- Right paraumbilical defect of abdominal wall
  - Rarely a left-sided defect
- Free-floating herniated small bowel
  - Also may be involved
    - Large bowel
    - Stomach
    - Gallbladder
    - Urinary bladder
    - Pelvic organs

Gastroschisis Sonographic Findings

- Markedly dilated bowel may suggest
  - Infarction
  - Bowel atresia
- May also be observed
  - Hydronephrosis
  - Bladder deviation
  - Exstrophy


Normal LK
LK
b
s
h
Unilateral hydronephrosis
Deviation of the bladder

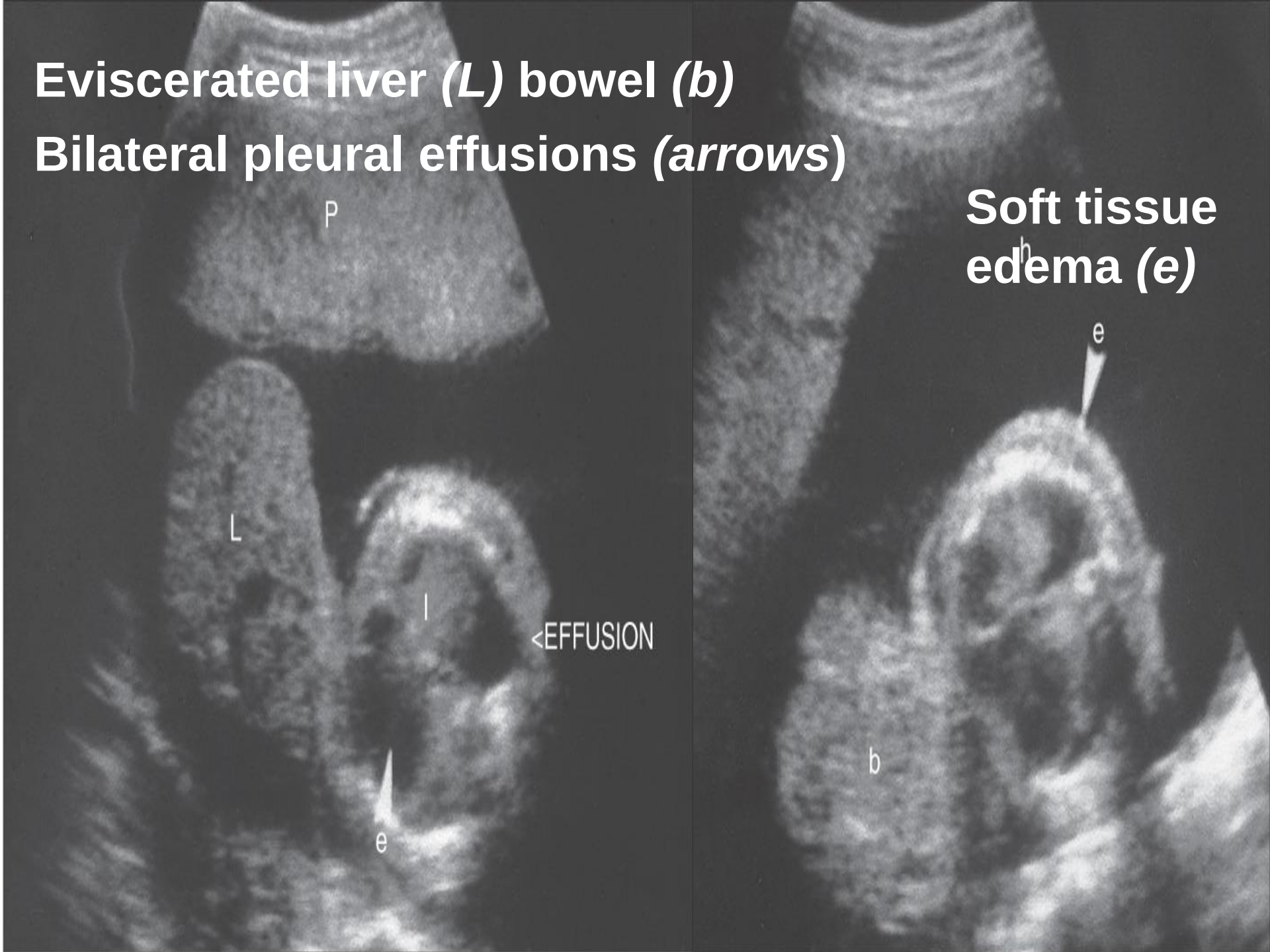

Gastroschisis
Bladder *(b)* herniated from the pelvis (exstrophy)
l
k
st
s
b
a

Gastroschisis Sonographic Findings

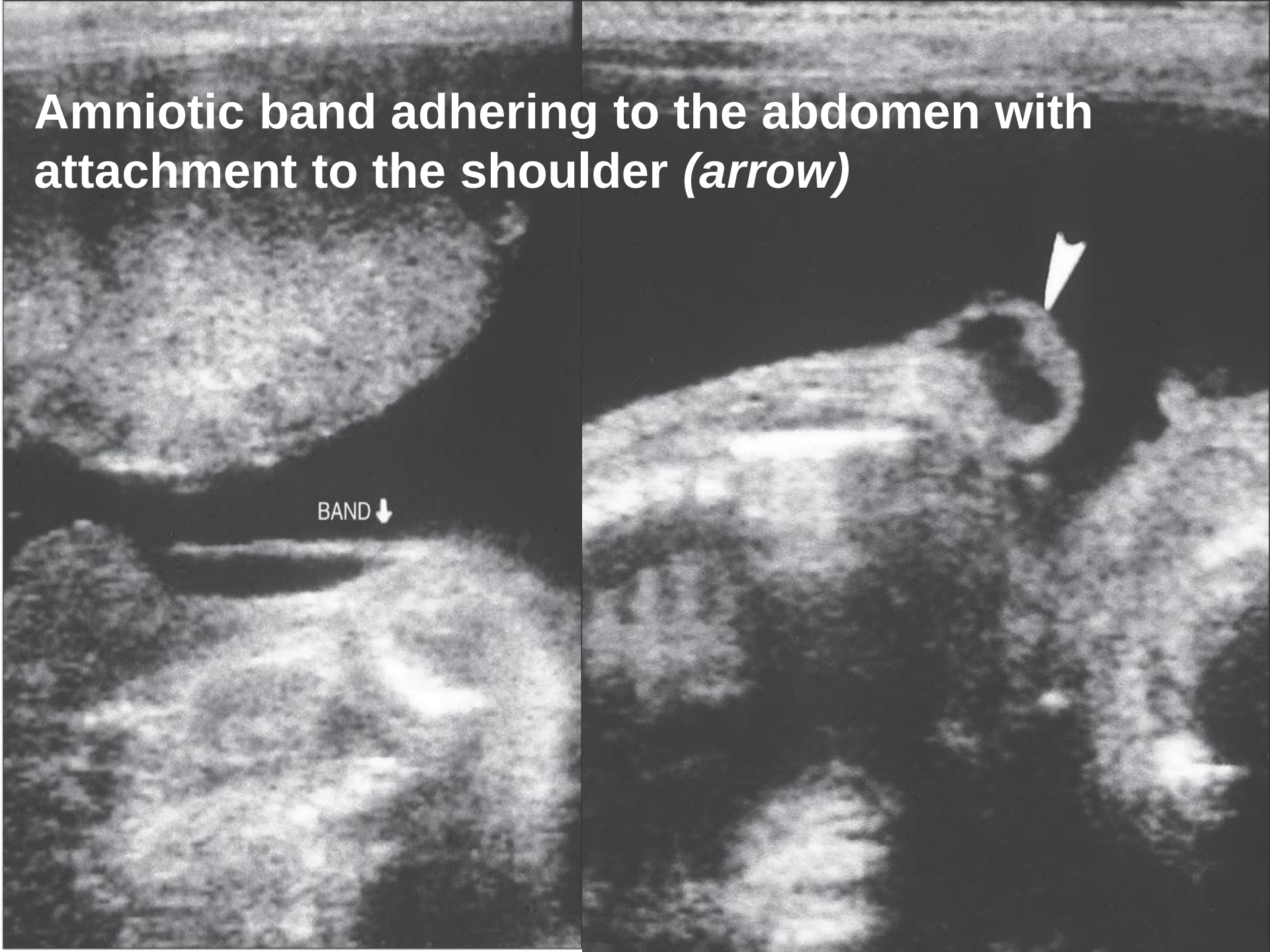
- Consider amniotic band syndrome amputations when clefting of the face and/or encephalocele is found
- Severe body wall defects may be seen in gastroschisis with secondary band formation

Gastroschisis and secondary amniotic band rupture

Next two following images


Eviscerated liver (L) bowel (b)
Bilateral pleural effusions (arrows)
Soft tissue edema (e)
P
L
I
EFFUSION
e
e
b

Amniotic band adhering to the abdomen with attachment to the shoulder *(arrow)*



Amniotic Band Syndrome

- Rupture of the amnion
  - Leads to entrapment or entanglement of the fetal parts by the “sticky” amnion
- May cause amputation or defects
- Early entrapment by the bands may lead to
  - Severe craniofacial defects
  - Internal malformations

Amniotic Band Syndrome

- Late entrapment leads to amputations or limb restrictions
- Prevalence is low
  - 7.8 in 10,000 births
- Associated anomalies include anomalies of
  - Limbs
  - Cranium
  - Face
  - Thorax
  - Abdominal wall
  - Perineum

Beckwith-Wiedemann Syndrome

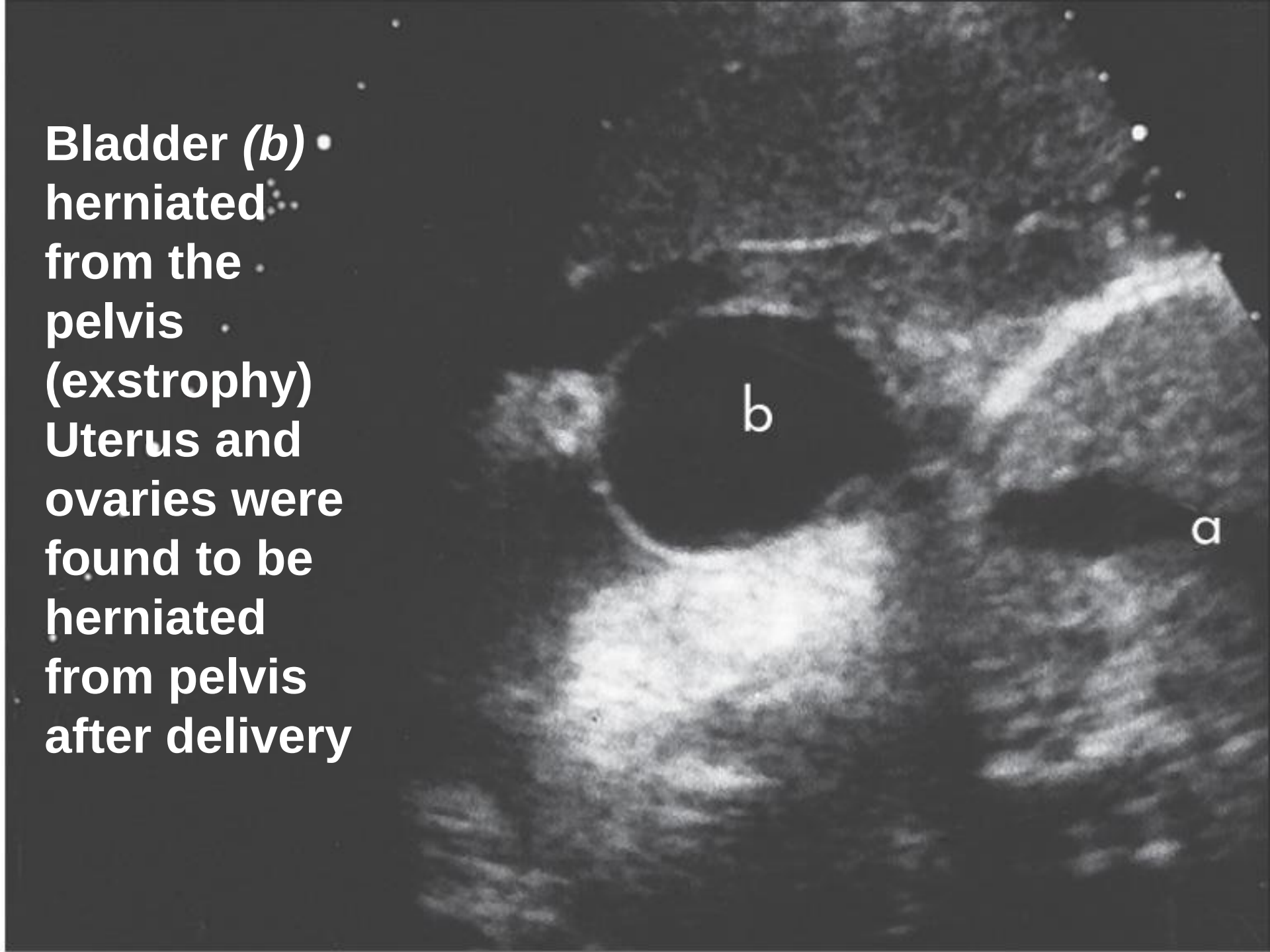
- Group of disorders common with the coexistence of an
  - Omphalocele
  - Macroglossia
  - Visceromegaly
- Most cases are sporadic

Beckwith-Wiedemann Syndrome

- Characterized by:
  - Macrosomia
  - Macroglossia
  - Visceromegaly
  - Omphalocele
  - Neonatal hypoglycemia
- Hydramnios may be present in third trimester

Exstrophy

- **Bladder exstrophy is characterized a defect in the lower abdominal wall and anterior wall of the urinary bladder**
  - **Bladder becomes exposed on the lower abdominal wall**
- **Cloacal exstrophy is rare and occurs early with involvement of the primitive gut**
  - **Results in exstrophy of bladder which two hemi-bladders are separated by intestinal mucosa**



Bladder *(b)* herniated from the pelvis (exstrophy) Uterus and ovaries were found to be herniated from pelvis after delivery

Sonographic Findings

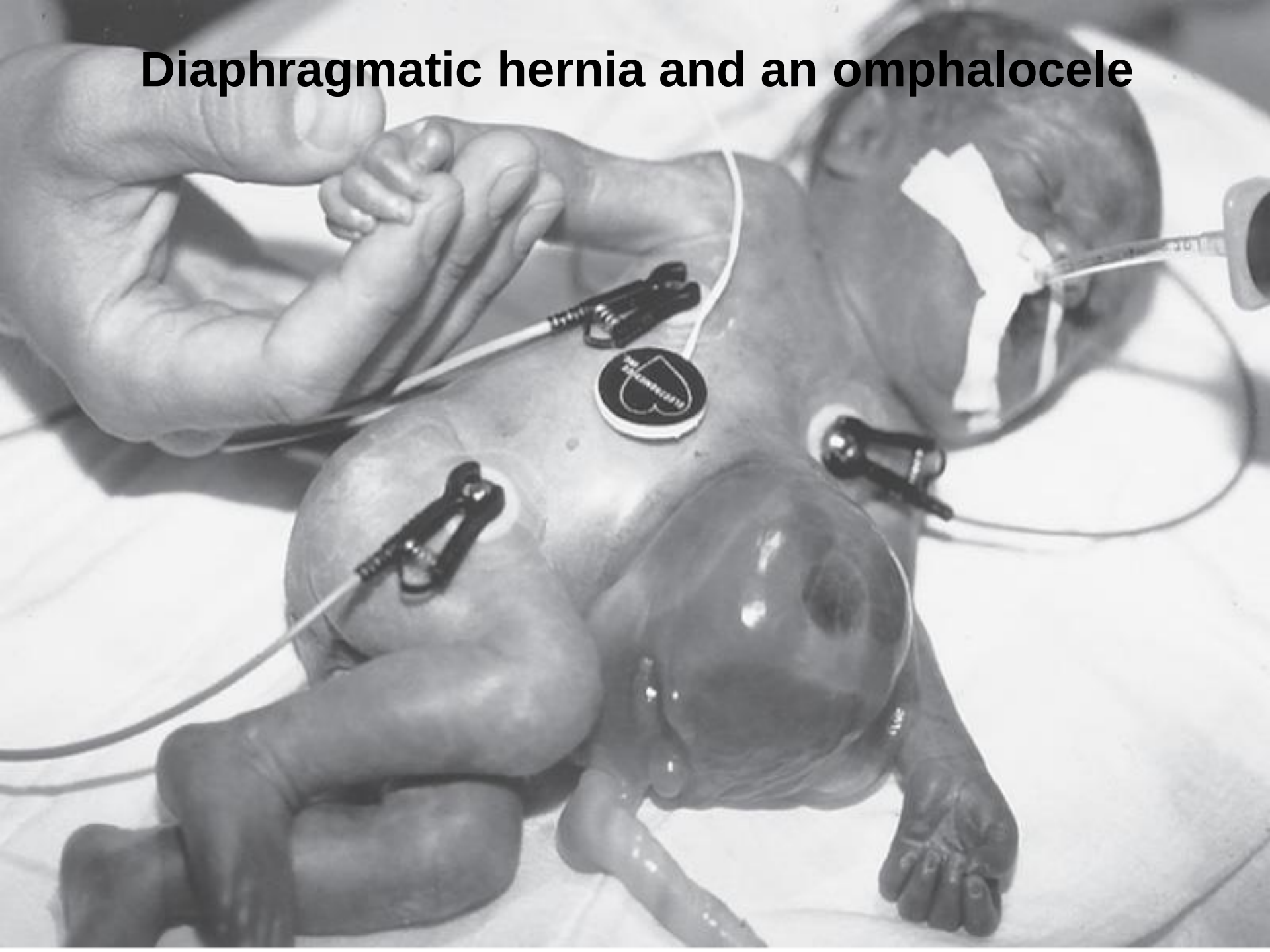
- Bladder exstrophy
  - Failure to see the normal urinary bladder
    - Soft tissue mass seen may
      - Represent the exposed bladder mucosa
      - Be seen on the surface of the lower abdominal wall
- Cloacal exstrophy
  - Anterior abdominal wall defect may be the primary finding

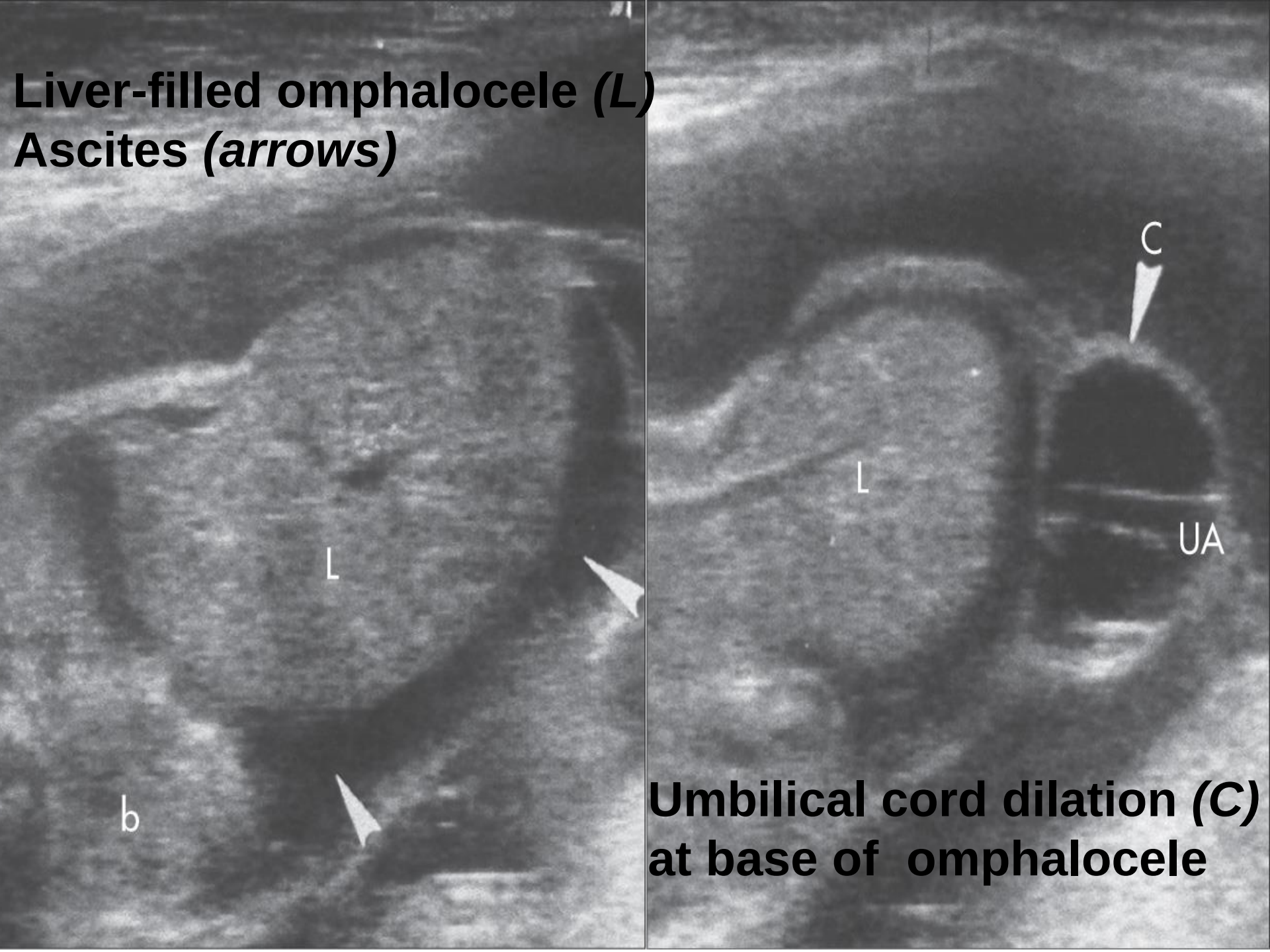
Pentalogy of Cantrell, Ectopic Cordis, and Cleft Sternum

Pentalogy of Cantrell

- Rare
- In association with two major defects
  - Omphalocele (high/superumbilical location)
  - Ectopic heart
    - Heart may be seen outside the normal thoracic cavity or bulge through the defective sternum
- Common to see
  - Pericardial effusions
  - Pleural effusions
  - Diaphragmatic hernia

Diaphragmatic hernia and an omphalocele




Liver-filled omphalocele *(L)*
Ascites *(arrows)*
L
b
C
L
UA
Umbilical cord dilation *(C)*
at base of omphalocele

Ectopia Cordis

- Exposed heart presents outside the chest wall through a cleft sternum
- Heart may present outside the thoracic cavity when a portion or all of the heart protrudes through the defect in the sternum

Cleft Sternum

- May be either partial or absent without ectopia cordis and is typically a superior or total cleft
- Dramatic pulsations of the anterior chest wall occur from the heart beating against the chest without the presence of the sternum to protect it

Limb–Body Wall Complex

- Anomaly associated with large cranial * defects
  - *Exencephaly* *
  - *Encephalocele* *
  - Facial cleft
  - Body-wall defect involving
    - Thorax
    - Abdomen
    - Limb defects

or

both

Limb–Body Wall Complex

- Occurs with the fusion of the amnion and chorion
  - Amnion does not cover the umbilical cord normally
    - Extends as a sheet from the margin of the cord and is continuous with both the body wall and the placenta
- Left-sided body-wall defects are three times more common than right-sided defects

Limb–Body Wall Complex

- Sonographically
  - Defects are large
    - Involving the abdomen and thorax
- Eviscerated organs form a complex, bizarre-appearing mass entangled with membranes
- Umbilical cord is short and adherent to the placental membranes