BMJ Best Practice

Omphalocele and gastroschisis

Straight to the point of care

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Summary

Gastroschisis and omphalocele are defects of the abdominal wall that occur in utero, can be detected antenatally using fetal ultrasonography, and result in herniation of abdominal contents.

The significant fluid balance changes and heat loss from exposed intestines in gastroschisis require emergency surgical intervention to establish abdominal wall closure or temporary coverage.

The lack of a protective sac in gastroschisis exposes the intestines to amniotic fluid in utero, leading to a thick inflammatory film or peel overlying the intestine and causing delay of return of normal bowel function after closure.

Intestinal atresia, occurs in 10% to 15% of infants with gastroschisis, and is related to ischaemia of the exposed gut caused by constriction of its mesenteric blood supply at the level of the abdominal wall defect.

The most common cause of mortality in omphalocele relates to associated organ system and chromosomal anomalies such as pentalogy of Cantrell and Beckwith-Wiedemann syndrome, which are characterised by cardiac anomalies and chromosomal defects.

Successful postnatal management of all abdominal wall defects is focused on timely abdominal wall closure, temperature maintenance, fluid resuscitation, and prevention of additional fluid loss from the abdominal contents, as well as prevention of ischaemia and infarction of the intestine.

Definition

Gastroschisis and omphalocele are congenital defects of the abdominal wall resulting in intestinal herniation from the abdominal cavity. In contrast to omphalocele, there is no sac covering the intestines in gastroschisis. They can both be detected antenatally using fetal ultrasonography. Limited studies have attempted to categorise gastroschisis defects into subsets of simple or complex defects, with simple gastroschisis existing as an isolated defect, and complex gastroschisis occurring with other gastrointestinal anomalies such as intestinal atresia, perforation, necrosis, or volvulus.

Epidemiology

The reported incidence of gastroschisis is 1-6 in 10,000 live births.[1] [2] Gastroschisis occurs more commonly in infants of young mothers, with a 10- to 16-fold higher incidence in children born to women aged younger than 20 years compared with children born to women aged 25-29 years, and it is slightly more frequent in male infants.[3] [4] Some studies suggest the incidence of gastroschisis is increasing worldwide with the use of vasoconstrictive medications.[5] [6]

Small omphaloceles occur in 1 in 5000 live births, while large omphaloceles are less common, occurring in 1 in 10,000 live births.[7] The frequency of omphalocele is the same in the US and Europe.[8] In contrast to gastroschisis, omphalocele occurs more often in older mothers, aged >40 years.[9] [10]

Aetiology

The aetiology of gastroschisis and omphalocele is unclear, and many theories have been proposed to explain these abdominal wall defects. Due to the poor understanding of the aetiology of these conditions, it is difficult to propose modifiable factors for prevention. Some studies suggest that maternal use of recreational drugs may be a risk factor for gastroschisis.[11] Others suggest that the incidence of gastroschisis is less in young mothers taking daily multivitamins during pregnancy.[12] Conversely, the incidence of gastroschisis in babies of teenage mothers who smoke is increasing.[3] [4] [9] [10]

Evidence supporting an association between smoking during pregnancy and the development of abdominal wall defects in the fetus is well described for both gastroschisis and omphalocele. It is suggested that cigarette smoke causes vasoconstriction that contributes to placental insufficiency and failed development of the vascular system.[11] [13] [14] [15] Maternal infections and illness during pregnancy are seen in mothers who deliver infants with omphalocele and gastroschisis. It is proposed that maternal illness can contribute to placental insufficiency that may, in turn, contribute to the development of abdominal wall defects.[4] Genitourinary infection during pregnancy has also been shown to be associated with abdominal wall defects.[16]

Pathophysiology

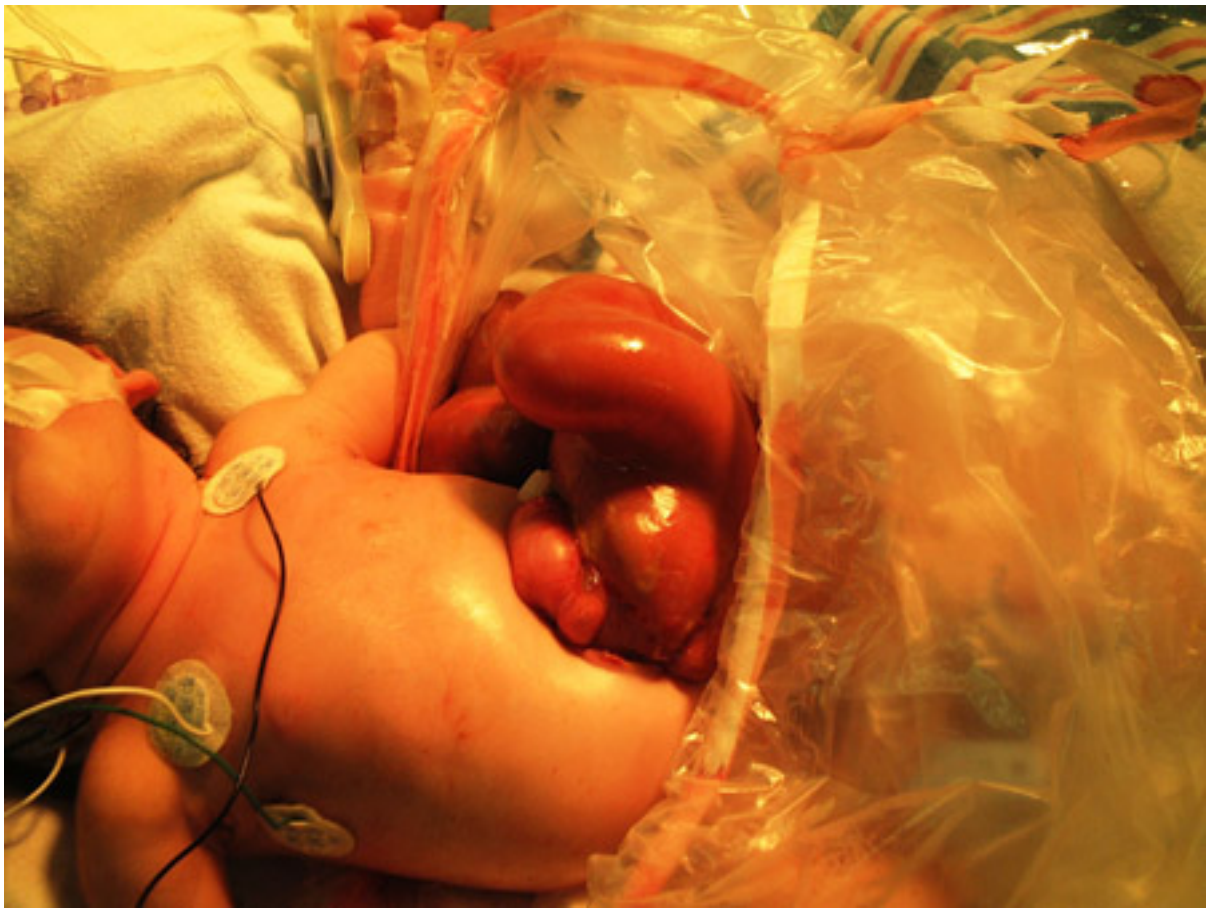
Several theories have been proposed to explain the development of gastroschisis. One theory suggests that the full-thickness defect results from abnormal involution of the right umbilical vein causing rupture of the anterior abdominal wall at the weakened point.[17] Another theory suggests that premature interruption of the right omphalomesenteric artery results in ischaemia of the anterior abdominal wall, allowing herniation of abdominal contents.[1] [18] In utero rupture of an omphalocele leading to the development of gastroschisis has also been suggested.[19] However, the lack of associated anomalies in gastroschisis does not support this theory. The thickened peel covering the intestine in gastroschisis may be due to exposure of the extruded abdominal contents to fetal urine or meconium in utero or mesenteric venous obstruction resulting in oedema.

Many theories have also been suggested to explain the development of omphalocele. These include failure of intestinal migration into the abdomen by 10 to 12 weeks of embryological development, failure of central migration of the lateral body folds of mesoderm, and persistence of a body stalk beyond 12 weeks' gestation.[20]

Case history

Case history #1

A newborn infant is transferred to the neonatal intensive care unit following a spontaneous vaginal delivery at which the neonatal resuscitation team was present and performed an intubation for respiratory distress. The abdomen of the infant is noted to have thickened loops of dark intestine outside of the abdominal cavity and to the right of the umbilical cord. The infant is placed feet first into a clear bowel bag containing warm saline-soaked gauze pads, and vascular access is obtained via the neck or upper extremity.

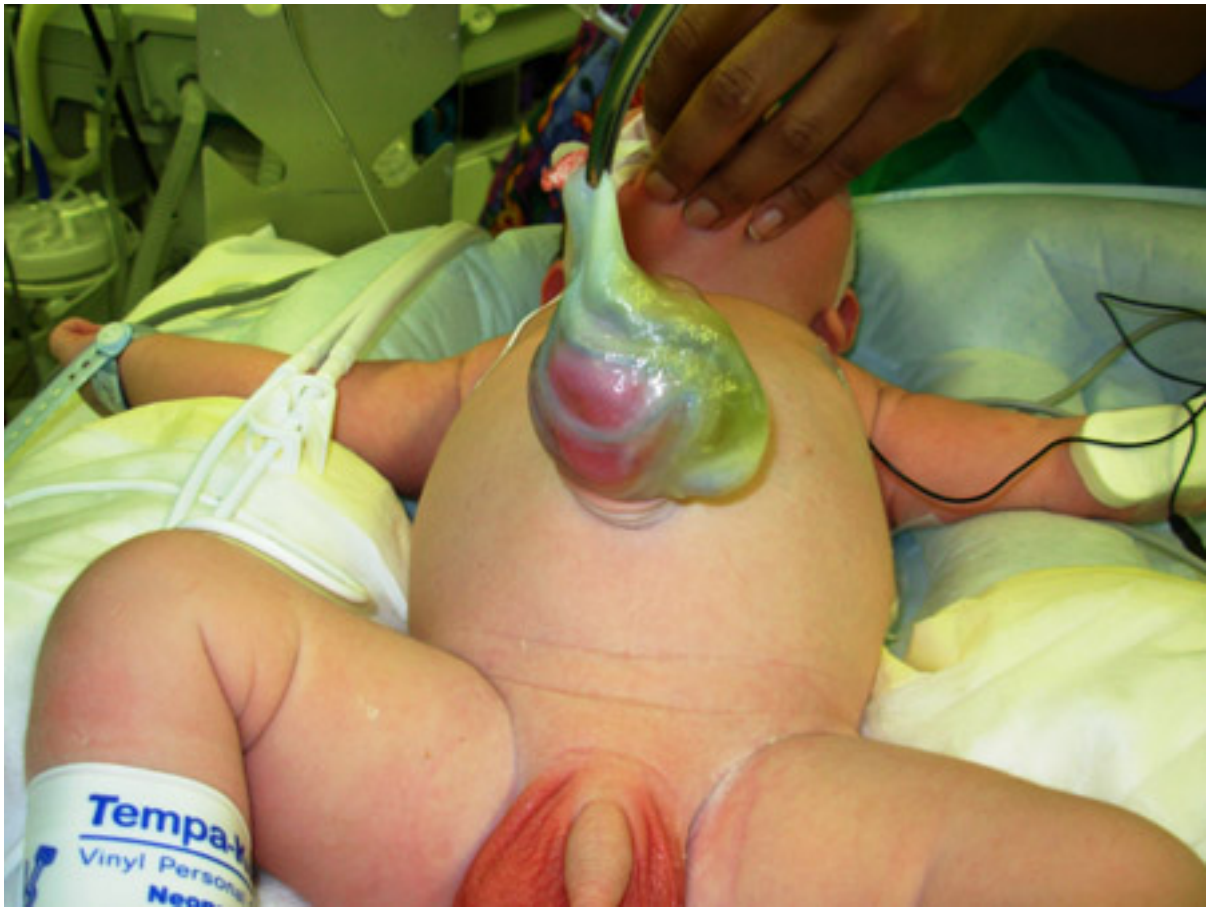


Immediately after delivery, an infant with gastroschisis is placed in a protective bowel bag

From collection of J.J. Tepas III, MD, FACS, FAAP

Case history #2

A newborn boy, with an estimated gestational age of 35 weeks, was delivered by spontaneous vaginal delivery. On initial examination, his Apgar score was noted to be five at 1 minute and nine at 5 minutes. There is a large clear membrane covering his abdomen, through which intestine is visible.



Note the membrane covering the abdominal contents in this omphalocele

From collection of J.J. Tepas III, MD, FACS, FAAP

Approach

Risk factors related to the development of abdominal wall defects are generally thought to be unmodifiable. However, maternal health factors associated with the development of abdominal wall defects include smoking and the use of certain recreational drugs.[3] [4] [9] [10] There are strong associations related to the discovery of these abdominal wall defects, which include maternal age <20 years for the development of gastroschisis and maternal age >40 years for omphalocele. Additionally, gastroschisis is more likely to be seen in male infants than in female infants. Weak factors associated with the development of abdominal wall defects include infections during pregnancy.

Abdominal wall defects of the fetus are routinely detected on antenatal ultrasound in the second trimester, when an abdominal mass may be visualised outside of the abdominal wall. Elevated alpha-fetoprotein levels, measured as part of the routine maternal triple screen at 16 to 18 weeks' gestation, suggest the presence of an abdominal wall defect and warrant further investigation with antenatal ultrasonography.[21] Definitive diagnosis is achieved at birth through a careful physical examination of the infant revealing abdominal contents external to the abdominal wall.

Detection of abdominal wall defects in the antenatal period allows parental counselling and the development of a postdelivery treatment plan, as well as arrangements for the delivery to take place at a tertiary care center equipped to handle the resuscitative and surgical needs of the newborn infant.[9] [17] [20] [22] [23]

Antenatal assessment

Elevated maternal serum alpha-fetoprotein levels reflect protein loss from the intestine floating in the amniotic fluid, and are a reliable but non-specific marker for the presence of gastroschisis. Abnormal maternal serum screening results warrant additional imaging to confirm the diagnosis of an abdominal wall defect, with high-definition ultrasound in addition to routine abdominal ultrasonography to visualise the developing fetus.

If evidence of omphalocele is confirmed on ultrasonography, invasive procedures such as amniocentesis performed at 15 to 20 weeks' gestation or chorionic villus sampling performed at 10 to 12 weeks' gestation may be undertaken to evaluate the possibility of associated chromosomal abnormalities, such as trisomy 13, trisomy 18, trisomy 21, Turner syndrome, Klinefelter syndrome, or triploidy.[9] [17] [20] Chromosomal abnormalities are more common in omphalocele than gastroschisis.

In many pregnancies, despite normal serum screening results, an abdominal wall defect may be detected on a routine antenatal ultrasound. During second-trimester ultrasonography, these defects are characterised by abdominal wall masses and echogenic bowel outside of the abdominal wall.

As associated genetic anomalies can occur in as many as 30% to 40% of infants with antenatal evidence of omphalocele, antenatal genetic testing for chromosomal abnormalities, and an echocardiogram for detection of cardiac anomalies, are routinely performed.[24] [25] Infants with antenatal evidence of gastroschisis do not routinely receive antenatal genetic testing.

In gastroschisis, dilated, fluid-filled loops of bowel floating freely in the amniotic fluid, and in some cases intestinal atresia, may be detected on ultrasound. If omphalocele is suspected, sternal and cardiac defects should be evaluated on ultrasound to determine the potential presence of pentalogy of Cantrell and Beckwith-Wiedemann syndrome, the latter being confirmed by the presence of macroglossia and visceromegaly.[17] [22]

When an abdominal wall defect is visualised on ultrasound, in addition to the diagnosis of gastroschisis or omphalocele, it is important to consider the possibility of cloacal exstrophy, which is characterised by a portion of the large intestine lying outside of the body and splitting the bladder into two halves. In boys, the penis is usually flat and short, with the exposed inner surface of the urethra on top. The penis is sometimes split into a right and left half. In girls, the clitoris is split and there may be one or two vaginal openings. Cloacal exstrophy is a very rare birth defect, affecting 1 in every 200,000 to 400,000 births.[26]

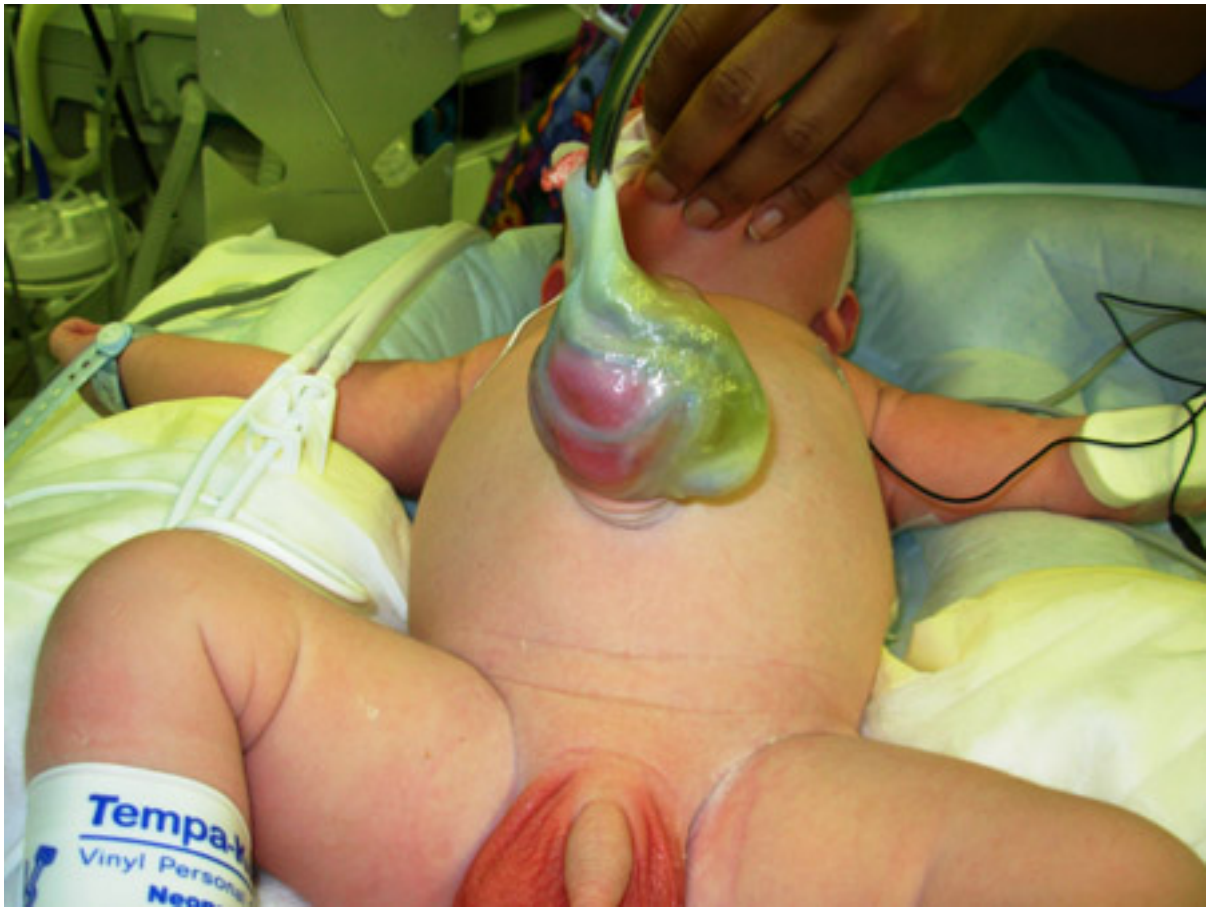
The optimal mode of delivery of infants with a antenatally diagnosed abdominal wall defect is intensely controversial.[17] Anecdotal evidence suggests that vaginal delivery may be contraindicated for infants with abdominal wall defects, particularly gastroschisis. However, other reports have found that avoiding vaginal delivery for these infants confers no benefit.[23] [27] [28] [29] [30]

Postnatal examination

A careful physical examination at birth revealing abdominal contents external to the abdominal wall is diagnostic of omphalocele and gastroschisis.

Omphaloceles are abdominal wall defects ranging from 4 to 12 cm in size and can be located centrally, or in the epigastric or hypogastric regions. In omphalocele, as the abdominal contents have a protective membranous covering in utero, the intestines are usually healthy at birth. This condition may be associated with Beckwith-Wiedemann syndrome and, rarely, with pentalogy of Cantrell. The likelihood of additional developmental abnormalities, most often affecting cardiac anatomy, is high, leading to the descriptive phrase 'bad baby, good bowel'. Antenatal ultrasonography and postnatal physical examination will detect most of the developmental abnormalities associated with omphalocele.

In gastroschisis the lack of a protective membranous covering causes the abdominal contents to be free-floating in utero. This leads to a chemical reaction that creates a thick inflammatory film or peel overlying the intestine. Gastroschisis is commonly associated with intestinal atresia, which occurs in 10% to 15% of infants, and is related to ischaemia of the exposed gut caused by constriction of its mesenteric blood supply at the level of the abdominal wall defect.[31] There are few additional structural abnormalities, although the intestine may require a lengthy period to recover, leading to the descriptive phrase 'good baby, bad bowel'.



Note the membrane covering the abdominal contents in this omphalocele

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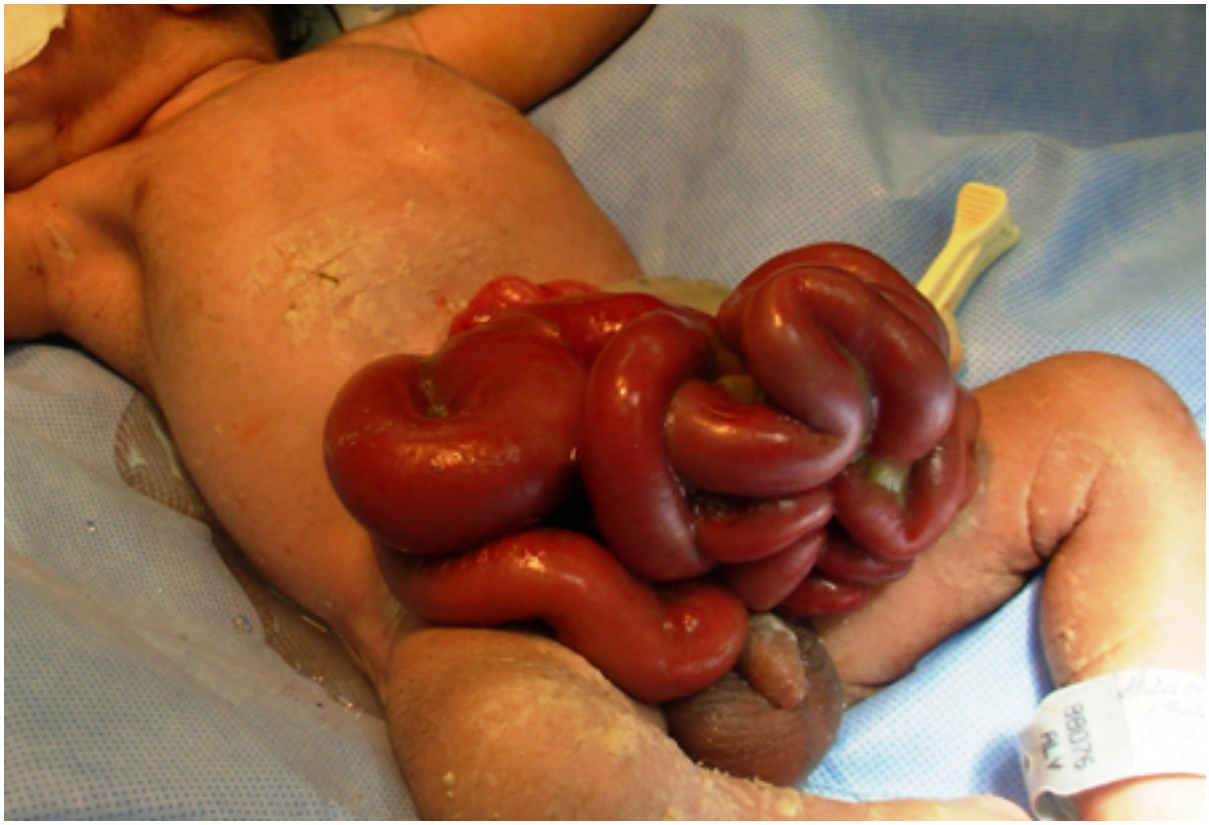


Extruded gut in abdominal wall defect

From collection of J.J. Tepas III, MD, FACS, FAAP

Investigations

During the neonatal period, infants with omphalocele and gastroschisis do not routinely receive any further investigations unless there are signs of dysmorphia. However, genetic counselling is routinely offered because many families request assistance in determining future child-bearing plans.

History and exam

Key diagnostic factors

presence of risk factors (common)

- Risk factors for abdominal wall defects include smoking and maternal infection during pregnancy.

maternal age <20 years (gastroschisis) (common)

- Gastroschisis occurs more commonly in infants of young mothers, with a 10- to 16-fold higher incidence in children born to women <20 years old compared with children born to women of 25 to 29 years.[3] [4] The mechanism behind the relationship is unknown.[13] [14] [15]

maternal age >35 years (omphalocele) (common)

- Omphalocele occurs more often in mothers >35 years of age.[9] [10]

elevated maternal serum alpha-fetoprotein (gastroschisis) (common)

- Elevated alpha-fetoprotein levels are a reliable but non-specific marker for the presence of gastroschisis due to proteins lost from the intestine floating in the amniotic fluid. This finding warrants further evaluation with a antenatal ultrasound.[21] [22]

positive antenatal ultrasound (common)

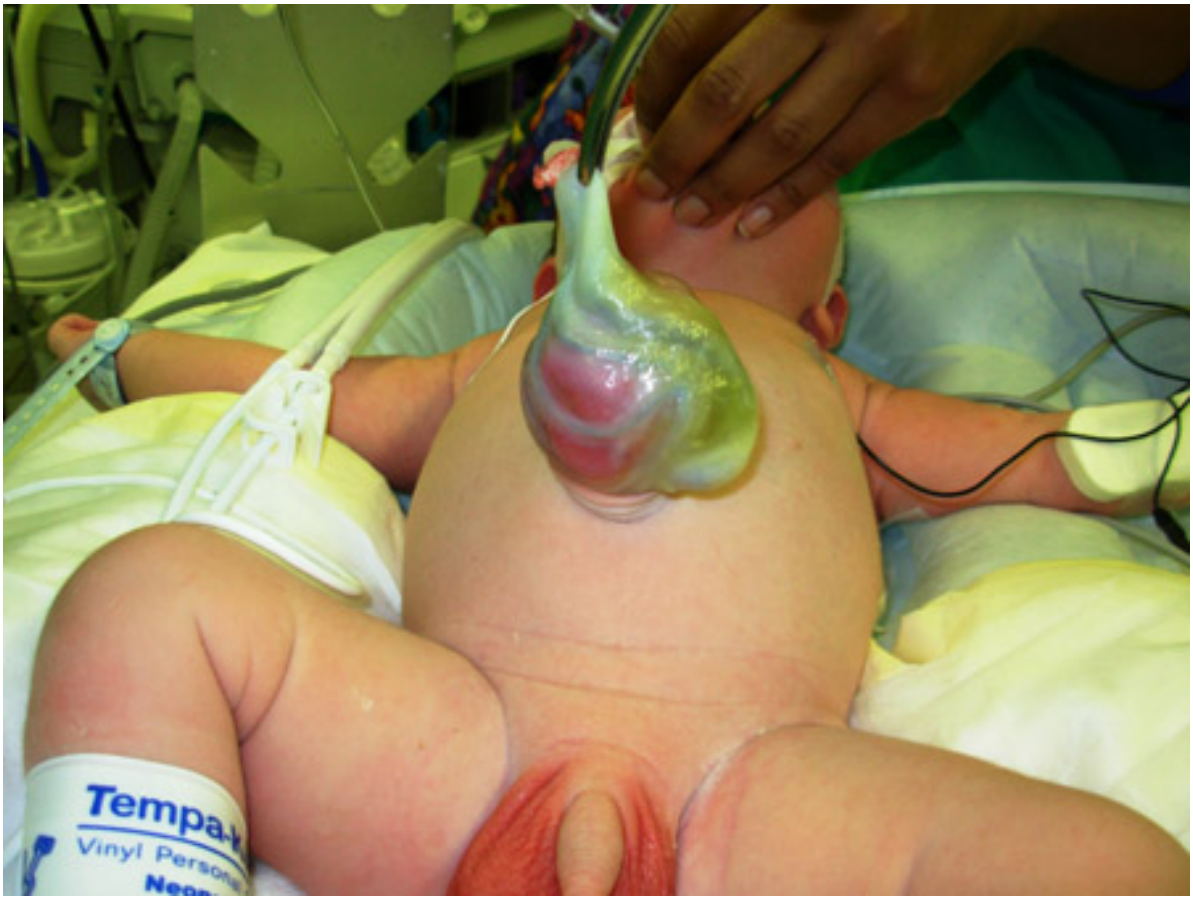
- Abdominal wall defects of the fetus are usually detected on routine antenatal ultrasound in the second trimester, when an abdominal mass may be visualised outside of the abdominal wall.
- During second-trimester ultrasonography, these defects are characterised by abdominal wall masses and echogenic bowel outside of the abdominal wall. In gastroschisis, dilated, fluid-filled loops of bowel floating freely in the amniotic fluid, and, in some cases, intestinal atresia, may be detected.

fetal chromosomal abnormalities (omphalocele) (common)

- If evidence of omphalocele is confirmed on ultrasonography, invasive procedures, such as amniocentesis performed at 15 to 20 weeks' gestation or chorionic villus sampling performed at 10 to 12 weeks' gestation, may be undertaken to evaluate the possibility of associated chromosomal abnormalities such as trisomy 13, trisomy 18, trisomy 21, Turner syndrome, Klinefelter syndrome, or triploidy.[9] [17] [20]

abdominal contents external to the abdominal wall (common)

- Omphaloceles are abdominal wall defects ranging from 4 to 12 cm in size and can be located centrally, or in the epigastric or hypogastric regions. In omphalocele, as the abdominal contents have a protective membranous covering in utero, the intestines are usually healthy at birth.
- In gastroschisis, the lack of a protective membranous covering causes the abdominal contents to be free-floating in utero, leading to a chemical reaction that creates a thick inflammatory film or peel overlying the intestine. Gastroschisis is commonly associated with intestinal atresia, which occurs in 10% to 15% of cases, and is related to ischaemia of the exposed gut caused by constriction of its mesenteric blood supply at the level of the abdominal wall defect.[31]



Note the membrane covering the abdominal contents in this omphalocele

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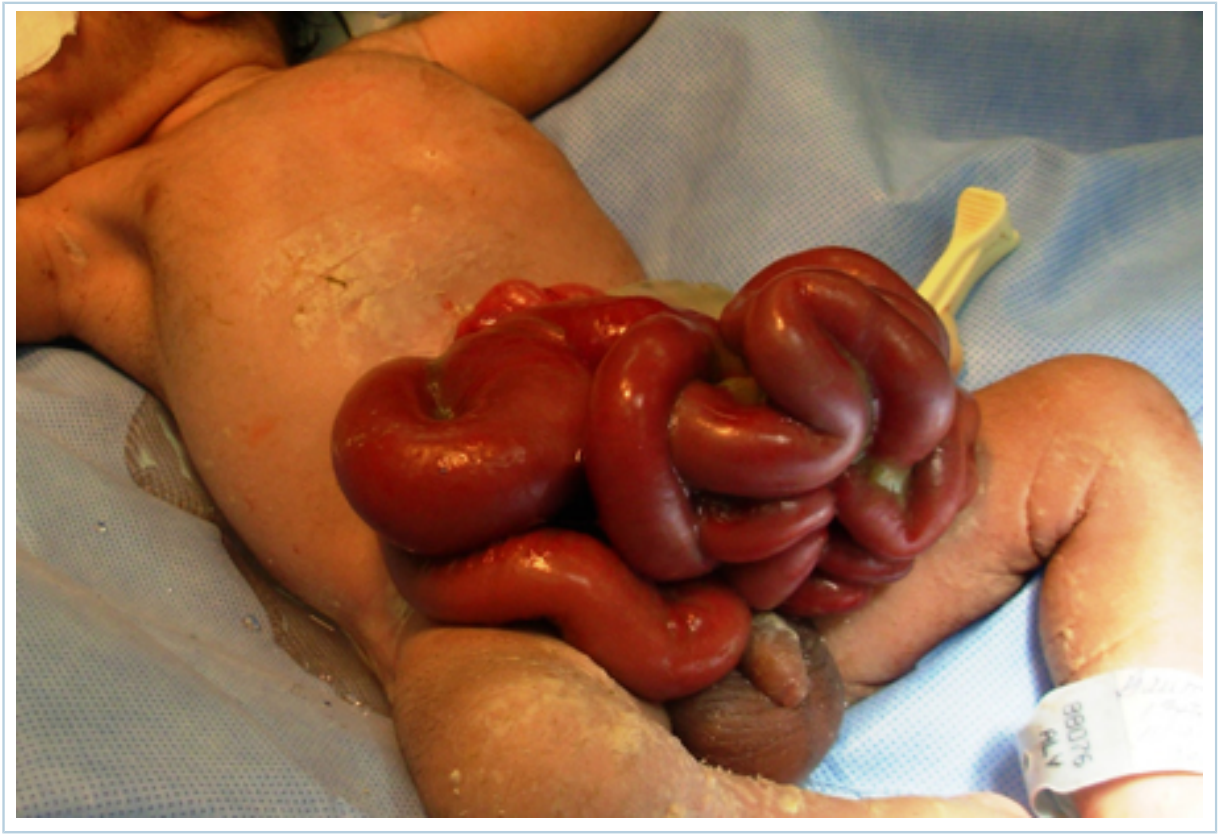

Extruded gut in abdominal wall defect

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Risk factors

Strong

maternal age <20 years (gastroschisis)

- Gastroschisis occurs more commonly in infants of young mothers, with a 10- to 16-fold higher incidence in children born to women <20 years old compared with children born to women of 25 to 29 years.[3] [4] The mechanism behind the relationship is unknown.[13] [14] [15]

male sex of neonate (gastroschisis)

- Gastroschisis is slightly more frequent in male infants than in female infants.[3] [4]

maternal age >35 years (omphalocele)

- Omphalocele occurs more often in infants of mothers >35 years of age.[9] [10]

smoking

- Evidence supporting an association between smoking during pregnancy and the development of abdominal wall defects in the fetus is well described for both gastroschisis and omphalocele. It is suggested that cigarette smoke causes vasoconstriction that contributes to placental insufficiency and failed development of the vascular system.[11] [13] [14] [15] The incidence of gastroschisis in babies of teenage mothers who are smokers is increasing.[3] [4] [9] [10]

Weak

maternal infection during pregnancy

- Maternal infections and illness during pregnancy are seen in mothers who deliver infants with omphalocele and gastroschisis. It is proposed that maternal illness can contribute to placental insufficiency that may, in turn, contribute to the development of abdominal wall defects.[4] Genitourinary infection during pregnancy has also been shown to be associated with abdominal wall defects.[16]

Investigations

1st test to order

Test	Result
clinical diagnosis • During the neonatal period, infants with omphalocele and gastroschisis do not routinely receive any further investigations unless there are signs of dysmorphia.	**omphalocele or gastroschisis**

Differentials

Condition	Differentiating signs / symptoms	Differentiating tests
Cloacal exstrophy	• Rare midline defect of the lower abdominal wall in which the intestines lie outside of the abdomen, often surrounded by two halves of the bladder. Genitalia are often bifurcated, the rectum may communicate with the bladder and an imperforate anus may be present.	• Antenatal ultrasonography: visualisation of defect with the presence of gut or other abdominal viscera outside of the abdominal cavity. • Postnatal physical examination: confirmatory of diagnosis.

Screening

Antenatal screening

Antenatal screening is routine, but extra vigilance may be required if there is a family history of birth defects, the mother is aged >35 years, harmful medications have been used during pregnancy, or there has been exposure to radiation. First-trimester ultrasonography is performed to estimate gestational age, to confirm the pregnancy or determine the presence of a multiple pregnancy, and to rule out the presence of abnormal gestation or molar pregnancy. Second-trimester ultrasonography is employed to assess fetal developmental abnormalities, evaluate fetal wellbeing, and identify potential causes of oligo- or polyhydramnios.

Routine maternal serum testing, known as the triple screen, is performed at 16 to 18 weeks' gestation to screen for the presence of alpha-fetoprotein, human chorionic gonadotropin, and oestriol.[21] Screening for abdominal wall defects routinely occurs when maternal serum alpha-fetoprotein levels are elevated due to

proteins lost from the intestine floating in the amniotic fluid in gastroschisis. Ultrasonography performed in the second trimester can detect abdominal wall defects by visualisation of a mass outside of the abdominal wall and confirm the presence of other associated structural abnormalities.

Approach

No intrauterine interventions are available for the treatment of omphalocele or gastroschisis. Therapy is therefore undertaken after delivery of the infant, with the goals of temperature maintenance, fluid resuscitation, and preventing additional fluid loss from the abdominal contents with appropriate care of the herniated viscera, paying particular attention to the preservation of its blood supply. Prevention of intestinal ischaemia and infarction in gastroschisis is achieved with surgical closure of the defect. Some centres may use prophylactic antibiotics, but antibiotic treatment is not routine and local protocols should be followed.

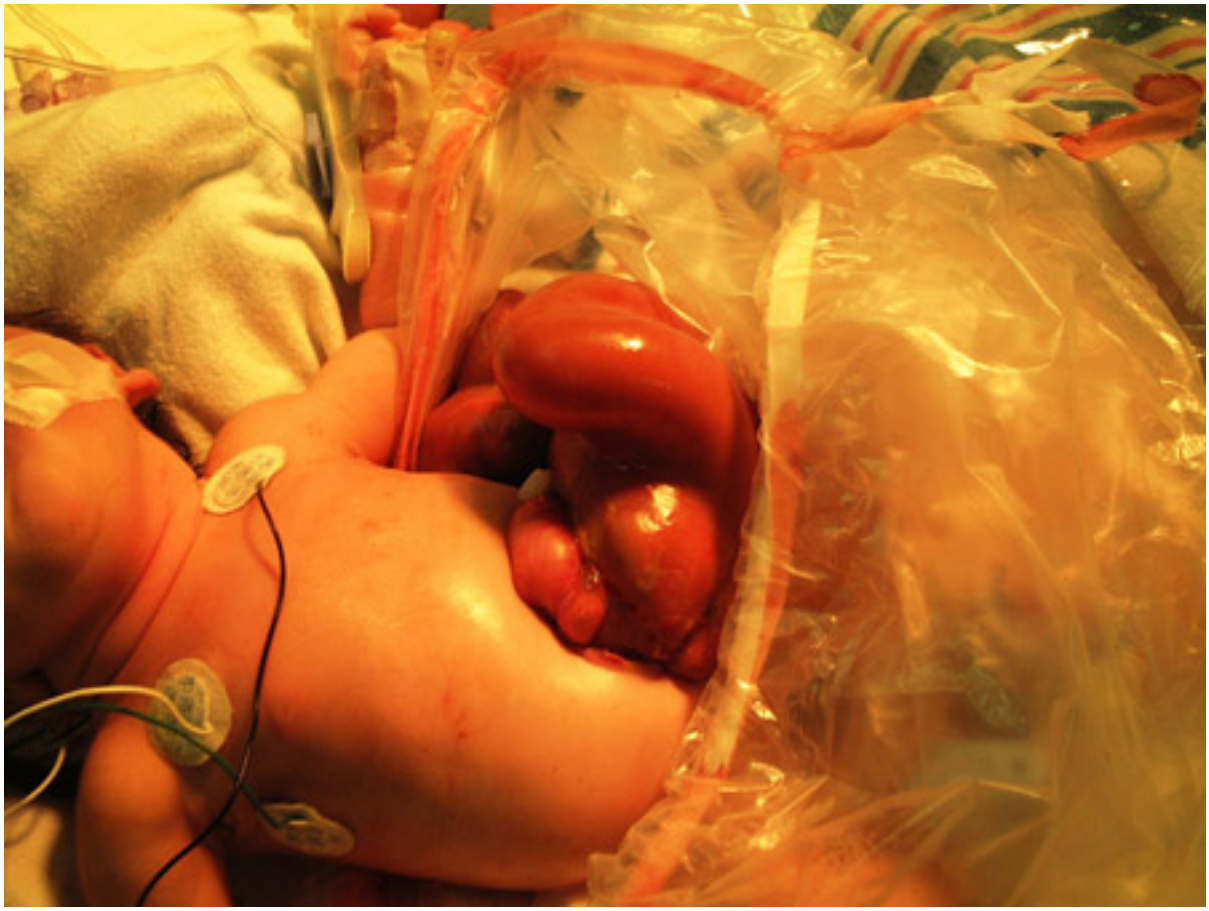
Gastroschisis

Gastroschisis is a surgical emergency requiring immediate closure or coverage. In gastroschisis, significant fluid balance changes require neonatal fluid resuscitation and emergent surgical intervention to prevent fluid loss and restore the viscera to the abdominal cavity.

Immediate postnatal management of gastroschisis involves fluid resuscitation and the placing of a sterile covering or a clear bowel bag over the herniated abdominal contents to prevent evaporation, heat loss, and infection. The infant should then be urgently transferred to a neonatal intensive unit (NICU) with specialised paediatric surgical capacity.[32]

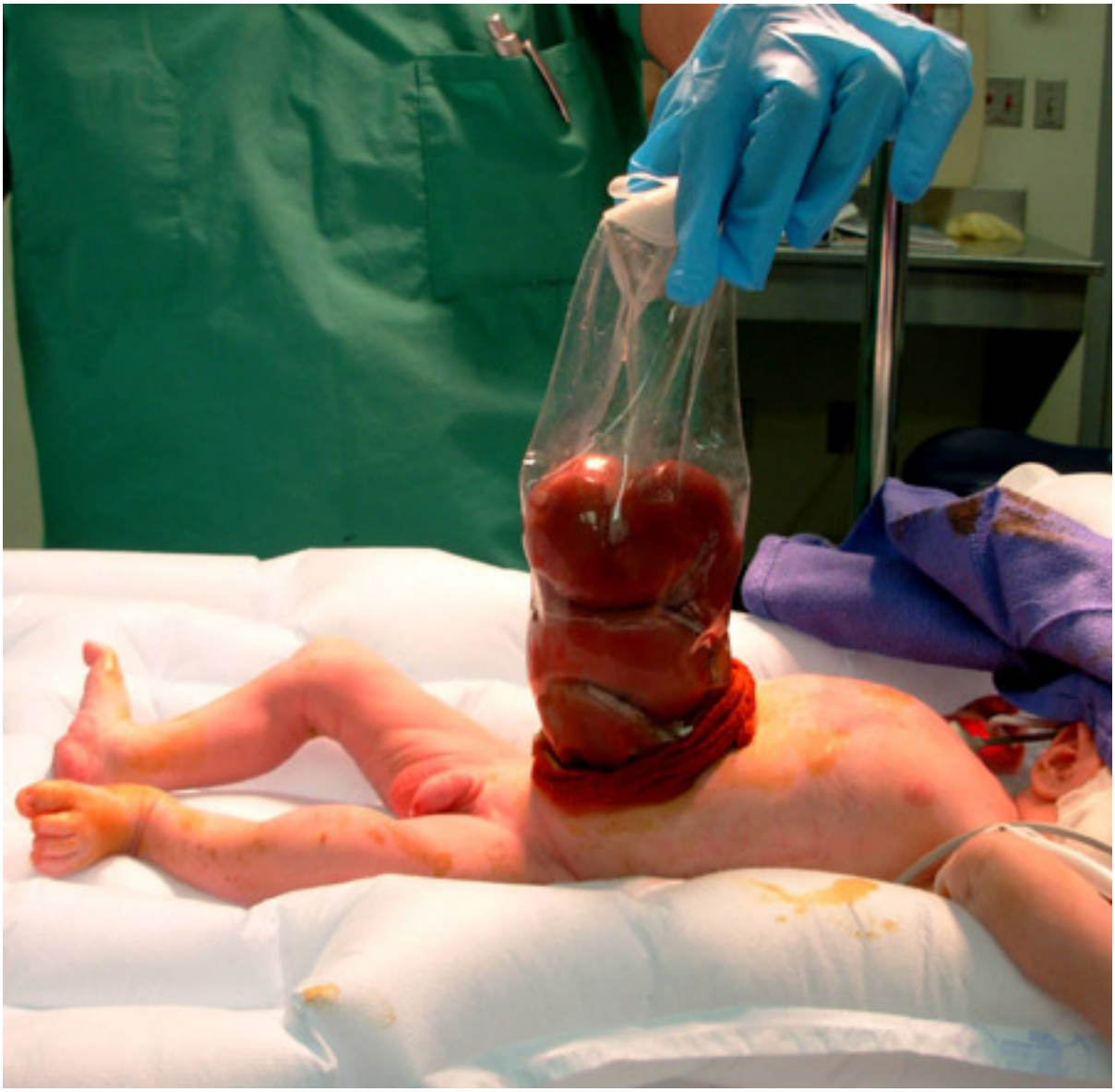
Small defects may be treated with primary surgical repair, while larger defects or those with extensive herniation of abdominal contents require a staged approach with placement of the abdominal contents into a suspended silo and gradual reduction into the abdominal cavity. Ventilatory support is often required during this process. Once the abdominal contents have been fully reduced into the abdominal cavity, surgery is performed to close the fascia and skin. Occasionally, reduction of pliable bowel is possible in the neonatal unit without the need for operative repair. Such bedside repair in the neonatal unit depends on the size of the defect and presence of healthy abdominal contents.

Surgery is followed by a trial of a nasogastric tube for decompression of the bowel and total parenteral nutrition while the inflammatory peel resolves.[33] If an atretic (ischaemic) segment is present, a small bowel obstruction may develop after enteral feeds, necessitating additional surgical intervention. Infants with gastroschisis are often slow to feed due to the intestinal irritation.



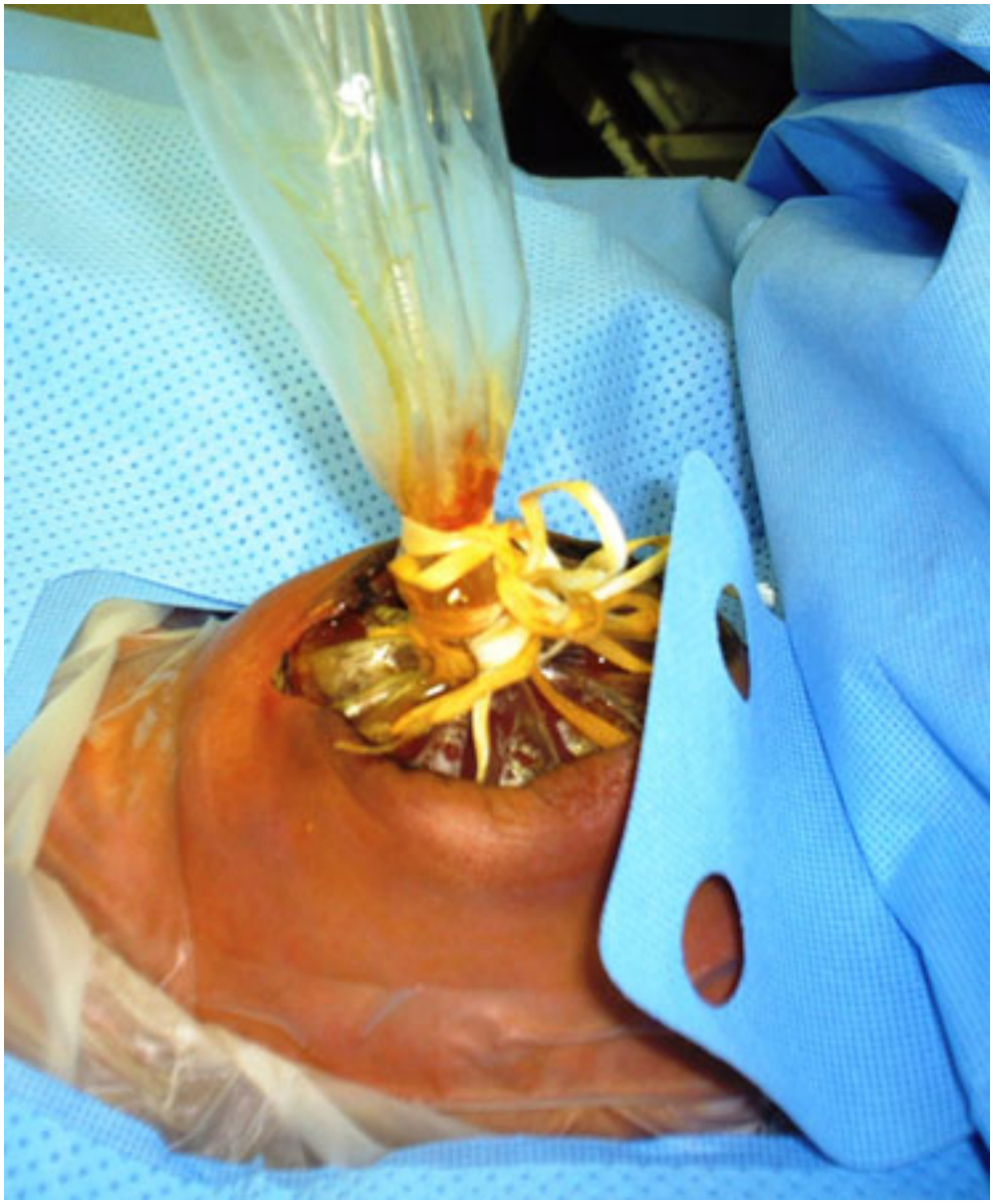
Immediately after delivery, an infant with gastroschisis is placed in a protective bowel bag

From collection of J.J. Tepas III, MD, FACS, FAAP



A staged repair of gastroschisis involves the placement of a silo to reduce contents into the abdomen

From collection of J.J. Tepas III, MD, FACS, FAAP



Once intestinal contents are fully reduced into the abdomen, closure of the abdominal wall follows

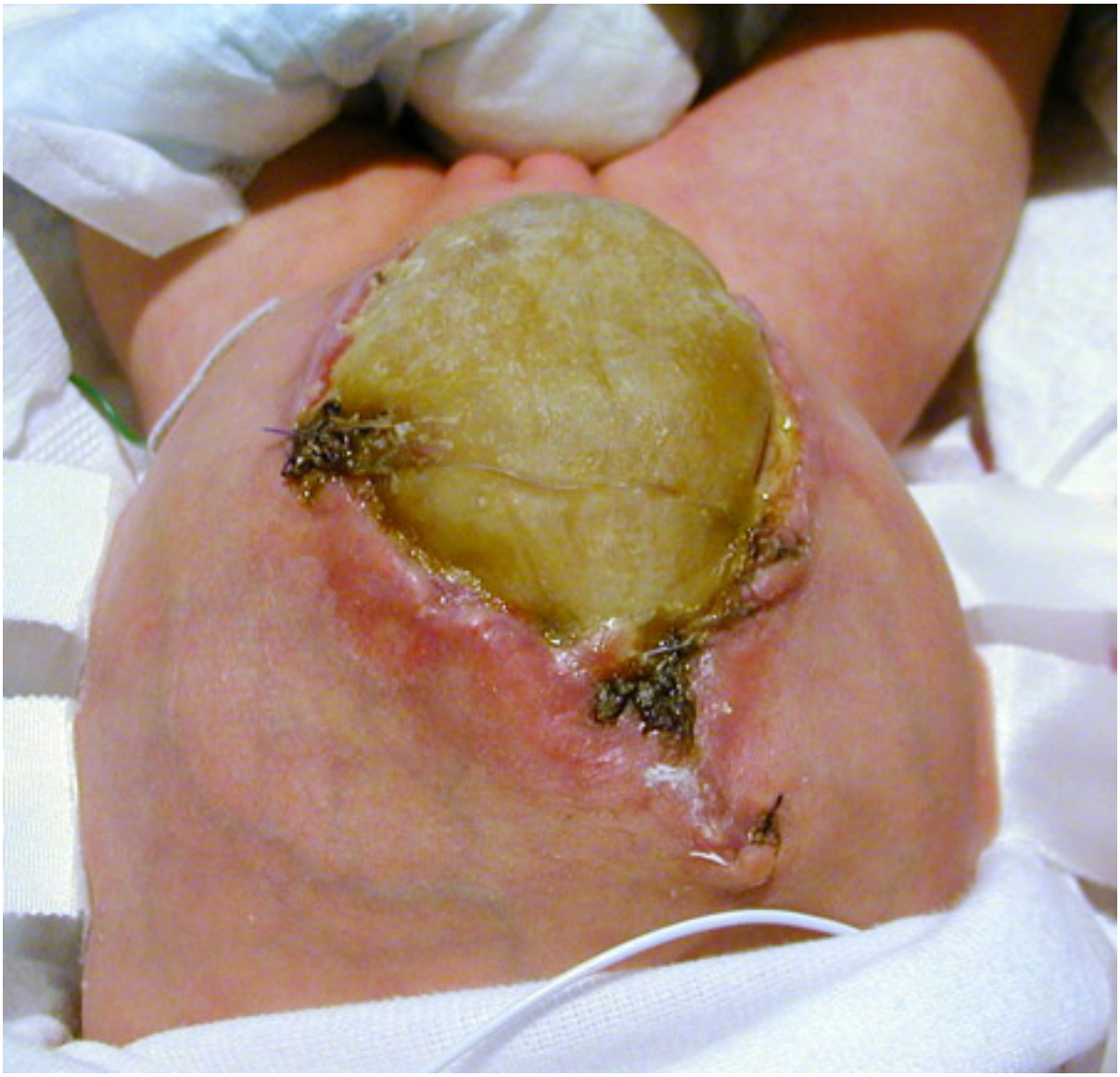
From collection of J.J. Tepas III, MD, FACS, FAAP

Omphalocele

After delivery, infants with omphalocele should receive appropriate fluid resuscitation. If the membranous sac is not intact, a sterile covering or a clear bowel bag must be placed over the herniated abdominal contents to prevent evaporation, heat loss, and infection.[34] The infant should then be urgently transferred to a NICU with specialised paediatric surgical capacity. A nasogastric tube should be inserted to decompress the bowel.

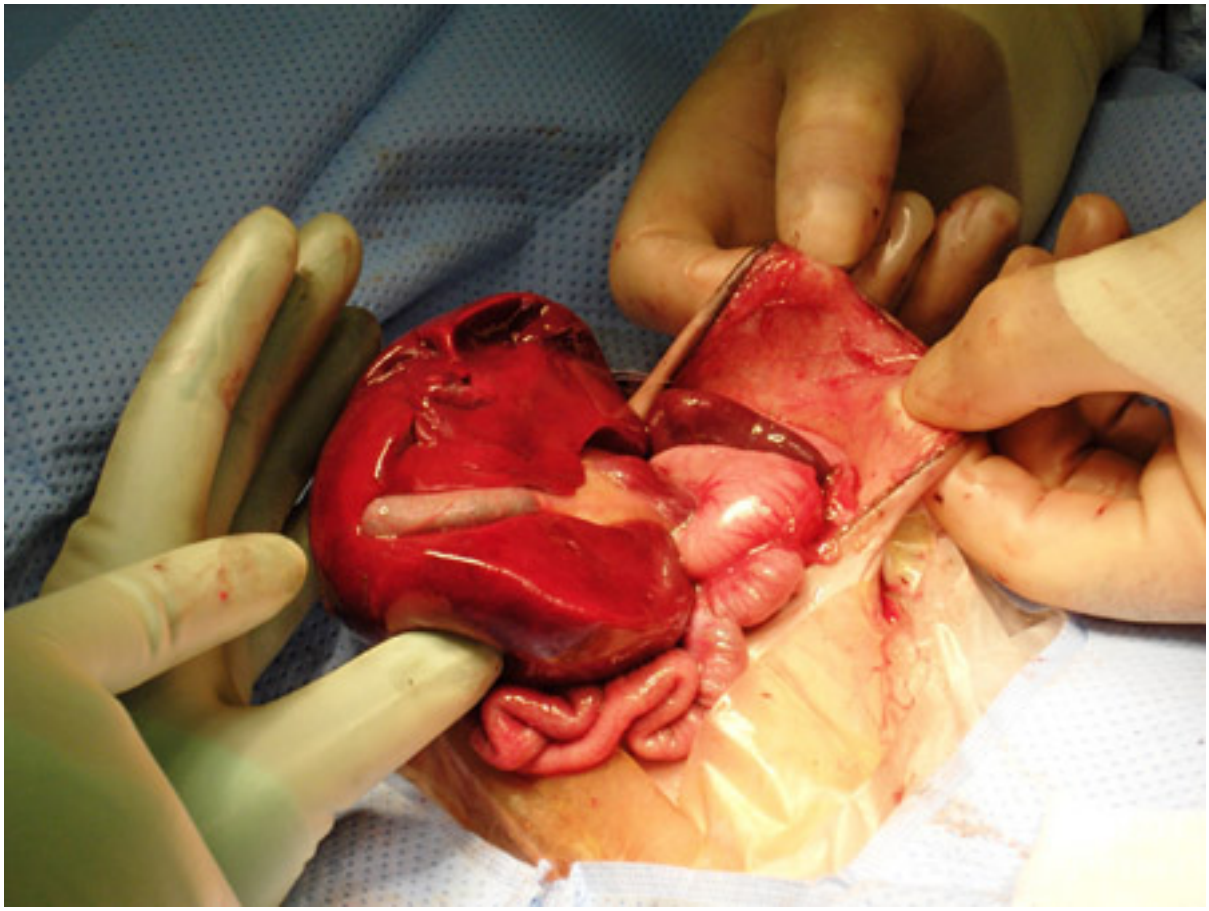
Surgical repair of omphalocele is elective (non-urgent), unless the membranous sac is ruptured. Defects without rupture of the membranous sac may be allowed to epithelialise, especially if associated anomalies render immediate surgical intervention inappropriately risky. The resultant ventral hernia can be repaired at a later date.

If the membranous sac is ruptured, treatment is the same as for gastroschisis. Small defects may be treated with primary surgical repair, while larger defects or those with extensive herniation of abdominal contents require a staged approach with placement of the abdominal contents into a suspended silo and gradual reduction into the abdominal cavity.[35] Ventilatory support is often required during this process. Once the abdominal contents have been fully reduced into the abdominal cavity, the fascia and skin are surgically closed. Surgery is followed by a trial of a nasogastric tube for decompression of the bowel and total parenteral nutrition while the inflammatory peel resolves.



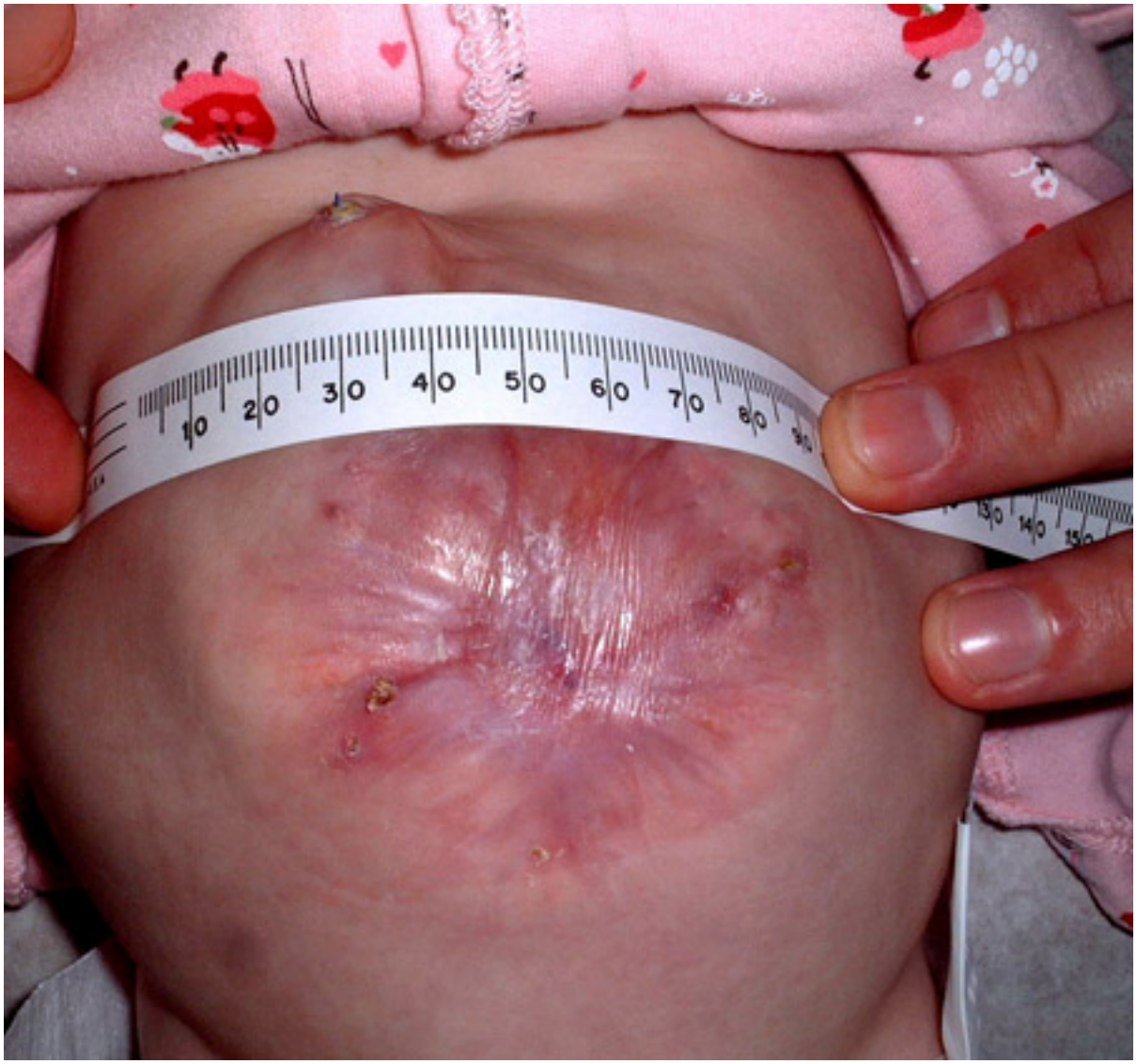
The abdominal wall defect in omphalocele is covered with a synthetic membrane

From collection of J.J. Tepas III, MD, FACS, FAAP



This ruptured omphalocele is similar in appearance to gastroschisis

From collection of J.J. Tepas III, MD, FACS, FAAP



A ventral hernia results from synthetic wall closure of the omphalocele

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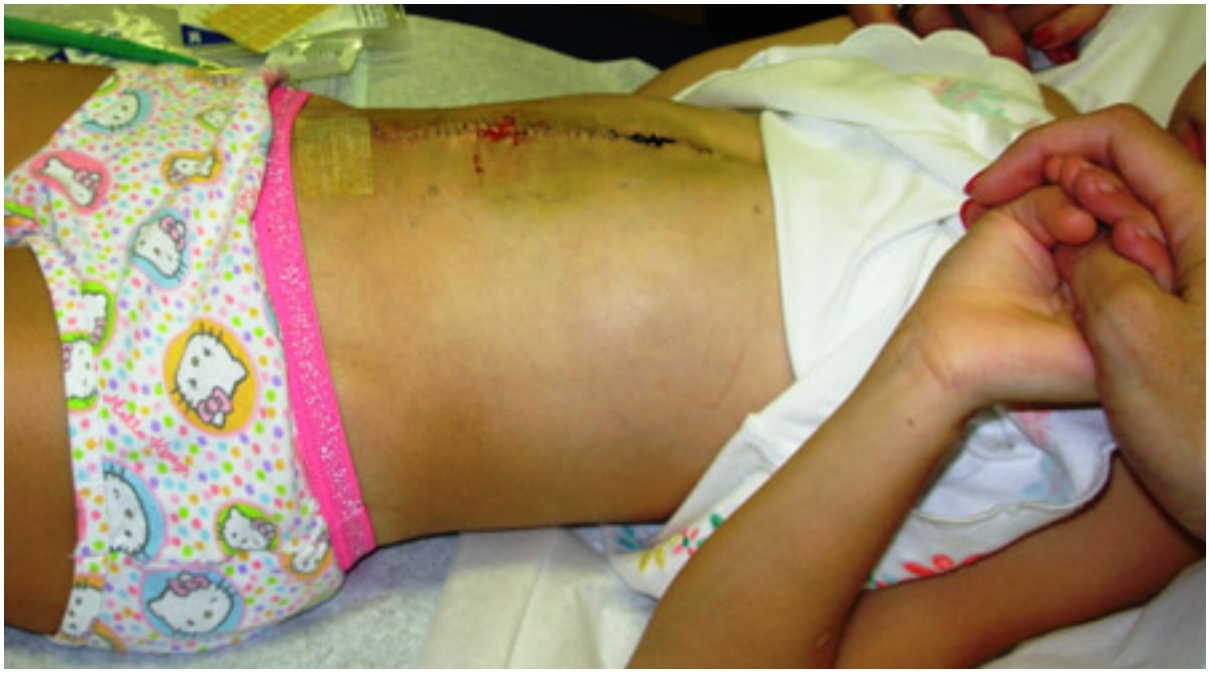


The ventral hernia is repaired in a 6-year-old girl born with omphalocele

From collection of J.J. Tepas III, MD, FACS, FAAP

Treatment algorithm overview

Please note that formulations/routes and doses may differ between drug names and brands, drug formularies, or locations. Treatment recommendations are specific to patient groups: see disclaimer

Acute (summary)		
gastroschisis		
	1st	fluid resuscitation and temperature maintenance
	plus	bowel protection
■ small defect	plus	primary surgical repair
	plus	postsurgical nasogastric tube and parenteral nutrition
■ large defect and/or extensive herniation	plus	staged reduction and surgical repair
	plus	postsurgical nasogastric tube and parenteral nutrition
	adjunct	secondary surgery
omphalocele		
	1st	fluid resuscitation and temperature maintenance
■ intact membranous sac	plus	nasogastric tube
	adjunct	surgical repair
■ ruptured membranous sac: small defect	plus	bowel protection
	plus	primary surgical repair
	plus	postsurgical nasogastric tube and parenteral nutrition
■ ruptured membranous sac: large defect and/or extensive herniation	plus	staged reduction and surgical repair
	plus	post-surgical nasogastric tube and parenteral nutrition

Treatment algorithm

Please note that formulations/routes and doses may differ between drug names and brands, drug formularies, or locations. Treatment recommendations are specific to patient groups: see disclaimer

Acute

gastroschisis

1st **fluid resuscitation and temperature maintenance**

» Immediate postnatal management of gastroschisis involves fluid resuscitation and temperature maintenance.

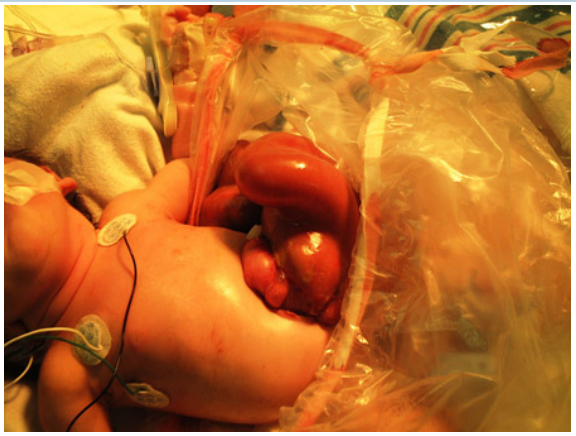
» The infant should then be urgently transferred to a neonatal intensive unit with specialised paediatric surgical capacity.[32]

plus **bowel protection**

Treatment recommended for ALL patients in selected patient group

» A sterile covering or a clear bowel bag should be placed immediately over the herniated abdominal contents to prevent evaporation, heat loss, and infection.

» The infant should then be urgently transferred to a neonatal intensive unit with specialised paediatric surgical capacity.[32]



Immediately after delivery, an infant with gastroschisis is placed in a protective bowel bag
From collection of J.J. Tepas III, MD, FACS, FAAP

small defect **plus** **primary surgical repair**

Treatment recommended for ALL patients in selected patient group

» Small defects may be treated with primary surgical repair.[35]

Acute

plus **postsurgical nasogastric tube and parenteral nutrition**

Treatment recommended for ALL patients in selected patient group

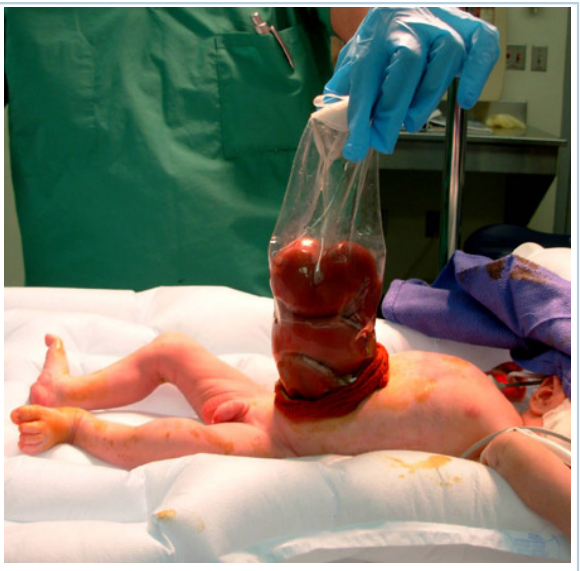
» Surgery is followed by a trial of a nasogastric tube for decompression of the bowel and total parenteral nutrition while the inflammatory peel resolves.

large defect and/or extensive herniation

plus **staged reduction and surgical repair**

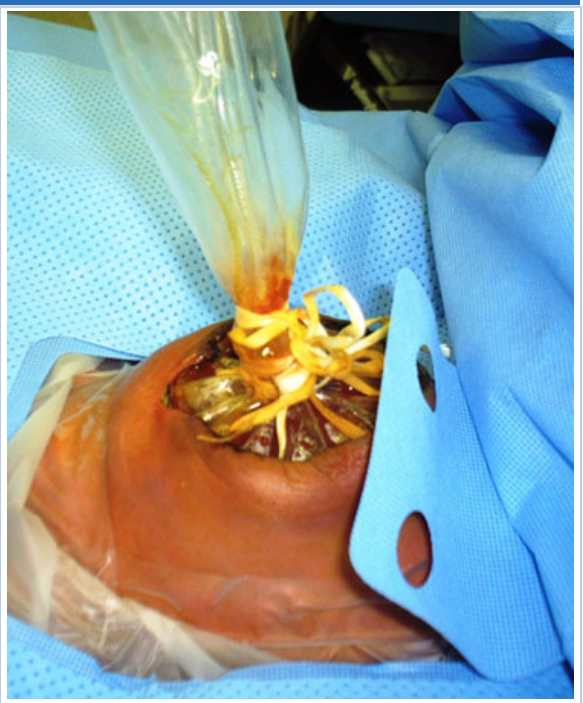
Treatment recommended for ALL patients in selected patient group

» Larger defects or those with extensive herniation of abdominal contents require a staged approach with placement of the abdominal contents into a suspended silo and gradual reduction into the abdominal cavity.[35] Ventilatory support is often required during this process. Once the abdominal contents have been fully reduced into the abdominal cavity, surgery is performed to close the fascia and skin.



A staged repair of gastroschisis involves the placement of a silo to reduce contents into the abdomen
From collection of J.J. Tepas III, MD, FACS, FAAP

Acute



Once intestinal contents are fully reduced into the abdomen, closure of the abdominal wall follows
From collection of J.J. Tepas III, MD, FACS, FAAP

	plus	**postsurgical nasogastric tube and parenteral nutrition**
		Treatment recommended for ALL patients in selected patient group
		» Surgery is followed by a trial of a nasogastric tube for decompression of the bowel and total parenteral nutrition while the inflammatory peel resolves.[33]
	adjunct	**secondary surgery**
		Treatment recommended for SOME patients in selected patient group
		» If an atretic (ischaemic) segment is present, a small bowel obstruction may develop after enteral feeds, necessitating further surgery.

omphalocele

omphalocele	**1st**	**fluid resuscitation and temperature maintenance**
		» After delivery, infants with omphalocele should receive appropriate fluid resuscitation with temperature maintenance.[34]
		» The infant should then be urgently transferred to a neonatal intensive unit with specialised paediatric surgical capacity.
■ **intact membranous sac**	**plus**	**nasogastric tube**

Acute

Treatment recommended for ALL patients in selected patient group

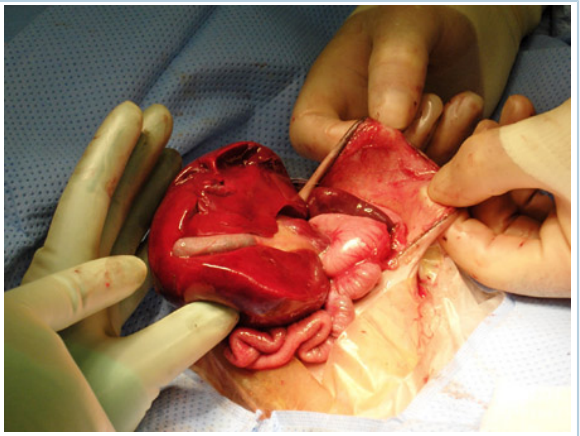
» A nasogastric tube should be inserted to decompress the bowel.

adjunct **surgical repair**

Treatment recommended for SOME patients in selected patient group

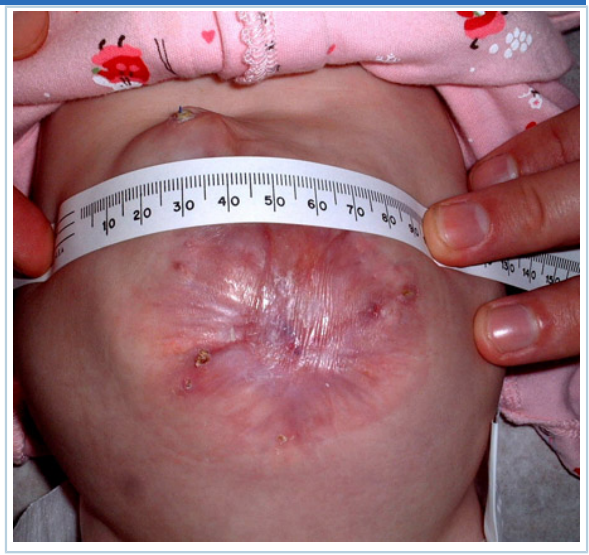
» Surgical repair of omphalocele is elective (non-urgent), unless the membranous sac is ruptured.

» Defects without rupture of the membranous sac may be allowed to epithelialise, especially if associated anomalies render immediate surgical intervention inappropriately risky. The resultant ventral hernia can be repaired at a later date.

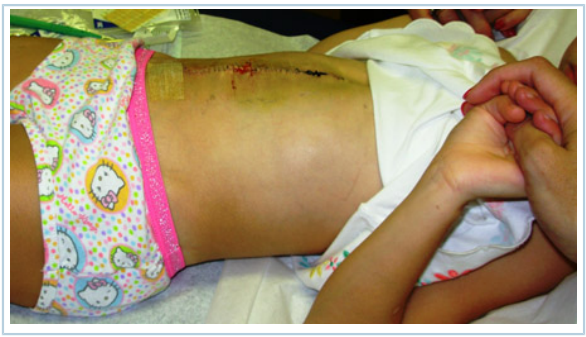


This ruptured omphalocele is similar in appearance to gastroschisis
From collection of J.J. Tepas III, MD, FACS, FAAP

Acute



A ventral hernia results from synthetic wall closure of the omphalocele
From collection of J.J. Tepas III, MD, FACS, FAAP



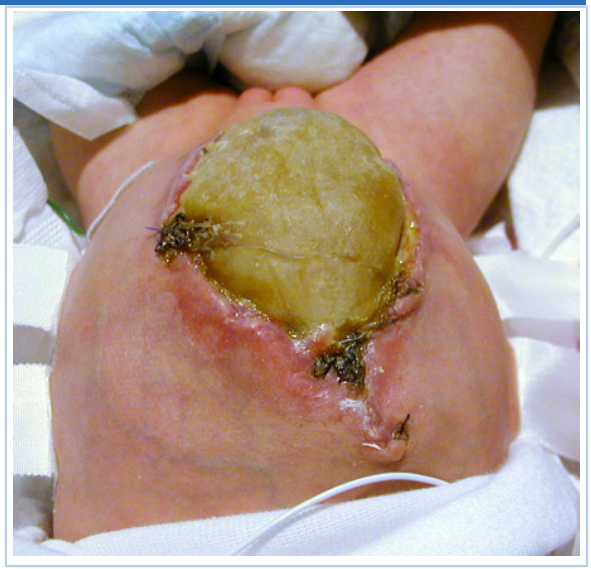
The ventral hernia is repaired in a 6-year-old girl born with omphalocele
From collection of J.J. Tepas III, MD, FACS, FAAP

ruptured membranous sac: small defect **plus** **bowel protection**

Treatment recommended for ALL patients in selected patient group

» If the membranous sac is not intact, a sterile covering or a clear bowel bag must be placed over the herniated abdominal contents to prevent evaporation, heat loss, and infection.[34]

Acute



The abdominal wall defect in omphalocele is covered with a synthetic membrane
From collection of J.J. Tepas III, MD, FACS, FAAP

plus **primary surgical repair**

Treatment recommended for ALL patients in selected patient group

» Small defects may be treated with primary surgical repair.[35]

plus **postsurgical nasogastric tube and parenteral nutrition**

Treatment recommended for ALL patients in selected patient group

» Surgery is followed by a trial of a nasogastric tube for decompression of the bowel and total parenteral nutrition while the inflammatory peel resolves.

ruptured membranous sac: large defect and/or extensive herniation

plus **staged reduction and surgical repair**

Treatment recommended for ALL patients in selected patient group

» Larger defects or those with extensive herniation of abdominal contents require a staged approach with placement of the abdominal contents into a suspended silo and gradual reduction into the abdominal cavity.[35] Ventilatory support is often required during this process. Once the abdominal contents have been fully reduced into the abdominal cavity, the fascia and skin are surgically closed.

plus **post-surgical nasogastric tube and parenteral nutrition**

Treatment recommended for ALL patients in selected patient group

Acute

» Surgery is followed by a trial of a nasogastric tube for decompression of the bowel and total parenteral nutrition while the inflammatory peel resolves.

Primary prevention

As the aetiology of gastroschisis and omphalocele is unclear, it is difficult to develop preventive strategies. Some evidence suggests multivitamins during pregnancy may prevent omphalocele, but this is not strongly supported.[12] Other data suggest that avoiding recreational drugs during pregnancy may prevent the development of these abdominal wall defects.[11] [13] [14] [15] Smoking during pregnancy has also been associated with the development of gastroschisis and omphalocele, and should be strongly discouraged.

Patient discussions

It is important that parents monitor the weight gain and nutritional status of the infant. Lifestyle modification of the infant is usually not indicated. There are many support groups for parents of children with abdominal wall defects.

FOLLOW UP

Monitoring

Monitoring

Monitoring of infants following the repair of abdominal wall defects involves close follow-up to monitor weight gain, nutrition, and feeding patterns. Some infants are slow to feed after repair of gastroschisis and it is important to closely monitor their postoperative nutritional status. Additional imaging studies and functional tests are not required for these conditions, although such investigations may be necessary for the evaluation of anomalies associated with omphalocele, particularly cardiac anomalies.

Complications

Complications	Timeframe	Likelihood
wound infection	**short term**	**medium**
As treatment of gastroschisis and omphalocele often involves staged or repeated surgical procedures with manipulation of incisions at the umbilicus or surrounding skin, localised infection or necrosis of the skin edge may occur. These complications respond quickly to routine wound care, usually healing completely within weeks or even days.		
persistent bowel dysfunction	**short term**	**medium**
The exposure of the extruded gut to amniotic fluid in gastroschisis causes inflammation that often produces a persistent dysmotility. In the vast majority of cases this temporary phenomenon resolves over time.		
bowel obstruction	**variable**	**low**
Approximately 15% of infants with gastroschisis will develop signs of bowel obstruction at some point later in life, especially in the first year after treatment, partly due to the very common occurrence of intestinal malrotation associated with abdominal wall defects.[40] [41] Initial management should involve simple nasogastric decompression; however, occasionally surgical re-exploration is required.		
necrotising enterocolitis	**variable**	**low**
Infants with gastroschisis may have impaired intestinal perfusion and, as such, may be at risk of gut ischaemia that can progress to neonatal necrotising enterocolitis. This occurs in 2% to 18% of infants and is often associated with other risk factors such as very low birth weight, pulmonary insufficiency, and hypotension.[42]		
complications of total parenteral nutrition (TPN)	**variable**	**low**
While the gut recovers, the infant requires intravenous alimentation, which is delivered via a peripherally inserted central catheter or central venous catheter. Both of these routes of administration are at risk of becoming infected if not handled with careful and strict sterile technique. If intestinal function is slow to recover and the infant is dependent on intravenous alimentation, hepatitis, and liver failure may develop as a result of prolonged TPN dependence. Infants who are TPN-dependent may have short gut syndrome.		
cholestatic jaundice	**variable**	**low**
In gastroschisis, as the dyskinetic gut requires parenteral alimentation, the possibility of cholestatic jaundice persists.		

FOLLOW UP

Complications	Timeframe	Likelihood
short bowel syndrome	**variable**	**low**
In gastroschisis, if the acute injury is associated with ischaemic damage that destroys enough of the extruded gut, short bowel syndrome (also known as short gut syndrome) may develop. Infants who are TPN-dependent may have short bowel syndrome.		

Prognosis

The course and prognosis for gastroschisis and omphalocele depend on the severity of the defect and accompanying anomalies. Major malformations and chromosomal anomalies are common with omphalocele but rare in gastroschisis, with the exception of intestinal atresia. Advances in intensive care practices and postnatal resuscitation have decreased the mortality and morbidity associated with abdominal wall defects.

Gastroschisis

Reported survival rates in gastroschisis range from 91% to 100%, although the condition is associated with lengthy hospital stays.[5] [36] Antenatal intestinal insult may contribute to small bowel atresia, bowel infarction, and intestinal motility dysfunction. If the insult to the intestine is severe, short gut syndrome may lead to the need for prolonged hyperalimentation and associated complications of total parenteral nutrition, including hepatitis. After closure, infants may develop intestinal obstruction from atresia, bowel perforations, or peritonitis. Additional operative intervention may prolong the hospital stay and delay opportunities to feed orally prior to discharge. The most important prognostic factor for morbidity is the presence of intestinal atresia; preterm delivery and very low birthweight are also associated with worse clinical outcomes.[37]

Omphalocele

If omphalocele is not accompanied by additional structural abnormalities or chromosomal defects, the prognosis is good, with a low mortality.[38] However, associated congenital heart disease, genetic syndromes, and intestinal problems are common with omphalocele, and the condition is associated with anomalies in other organ systems in 30% to 80% of cases, with the severity of the anomalies dictating prognosis.[39] Most omphaloceles are associated with cardiac and chromosomal anomalies, and more than one half of the fetuses will die. Infections, surgical complications, and low birth weight are responsible for many deaths. Infections, immaturity, hernia rupture, and intestinal obstruction rates are high.

Treatment guidelines

Europe

Gastroschisis: management prior to transfer to surgical centre (https://www.hse.ie/eng/about/who/cspd/ncps/paediatrics-neonatology/resources)

Published by: Health Service Executive Ireland **Last published:** 2020

Oceania

Gastroschisis (https://www.cahs.health.wa.gov.au/For-health-professionals/Resources/Neonatology-guidelines)

Published by: Government of Western Australia **Last published:** 2021

Key articles

- Torfs CP, Velie EM, Oechsli FW, et al. A population-based study of gastroschisis: demographic, pregnancy, and lifestyle risk factors. Teratology. 1994 Jul;50(1):44-53. Abstract (http://www.ncbi.nlm.nih.gov/pubmed/7974254?tool=bestpractice.bmj.com)
- Langer JC, Harrison MR, Adzick NS, et al. Perinatal management of the fetus with an abdominal wall defect. Fetal Ther. 1987;2(4):216-21. Abstract (http://www.ncbi.nlm.nih.gov/pubmed/2978282?tool=bestpractice.bmj.com)
- Glick PL, Harrison MR, Adzick NS, et al. The missing link in the pathogenesis of gastroschisis. J Pediatr Surg. 1985;20:406-9. Abstract (http://www.ncbi.nlm.nih.gov/pubmed/2931510?tool=bestpractice.bmj.com)
- Snyder CL. Current management of umbilical abnormalities and related anomalies. Semin Pediatr Surg. 2007 Feb;16(1):41-9. Abstract (http://www.ncbi.nlm.nih.gov/pubmed/17210482?tool=bestpractice.bmj.com)
- Sipes SL, Weiner CP, Sipes DR 2nd, et al. Gastroschisis and omphalocele: does either antenatal diagnosis or route of delivery make a difference in perinatal outcome? Obstet Gynecol. 1990 Aug;76(2):195-9. Abstract (http://www.ncbi.nlm.nih.gov/pubmed/2142521?tool=bestpractice.bmj.com)

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Images

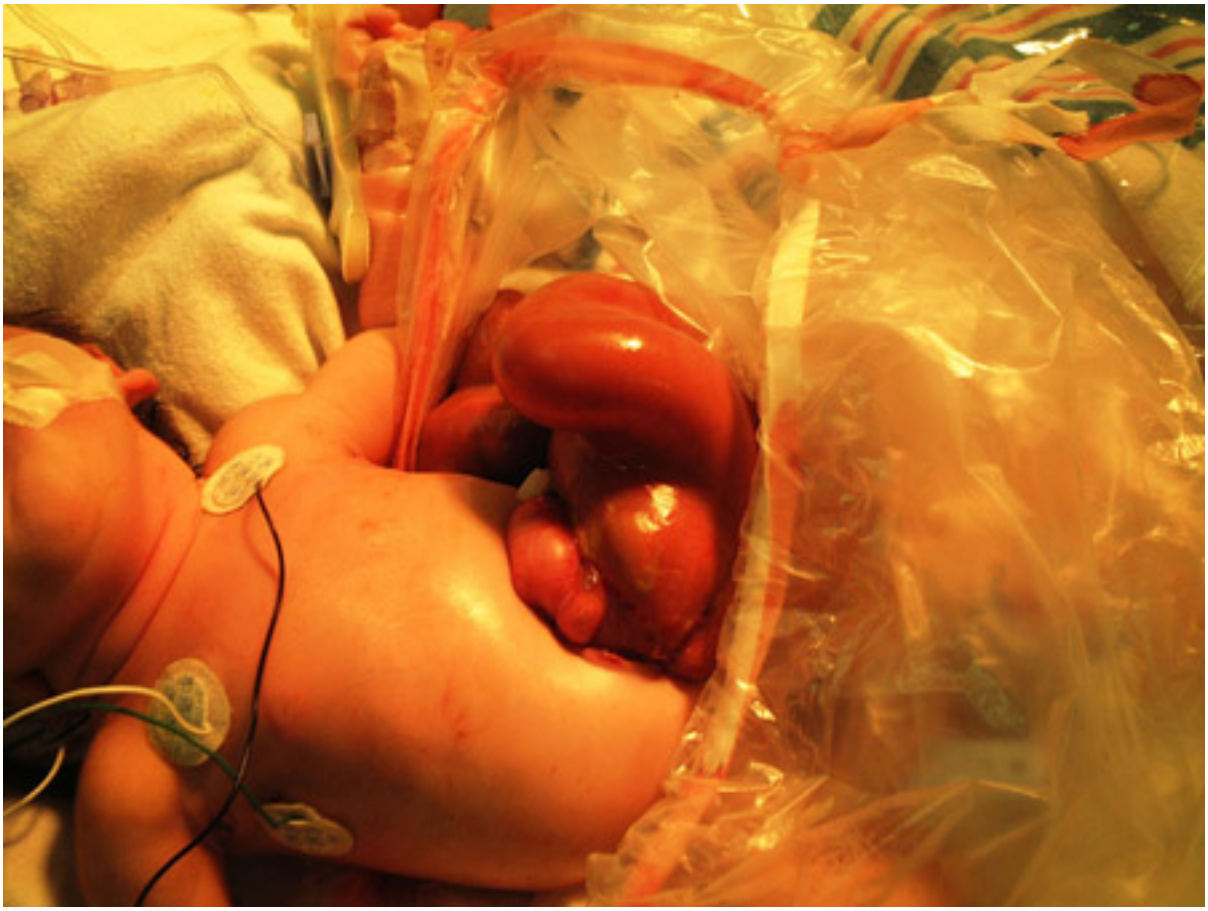

Figure 1: Immediately after delivery, an infant with gastroschisis is placed in a protective bowel bag

From collection of J.J. Tepas III, MD, FACS, FAAP

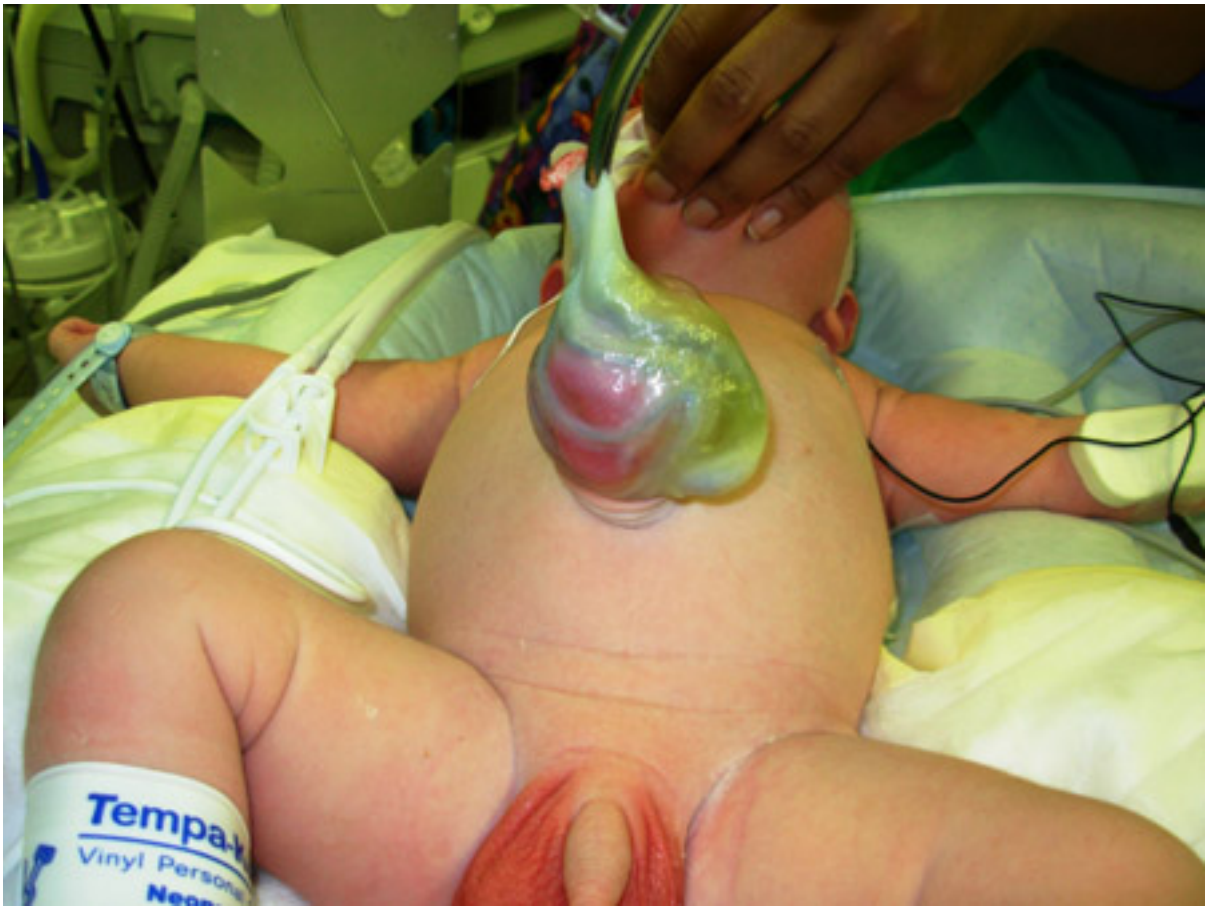

Figure 2: Note the membrane covering the abdominal contents in this omphalocele

From collection of J.J. Tepas III, MD, FACS, FAAP

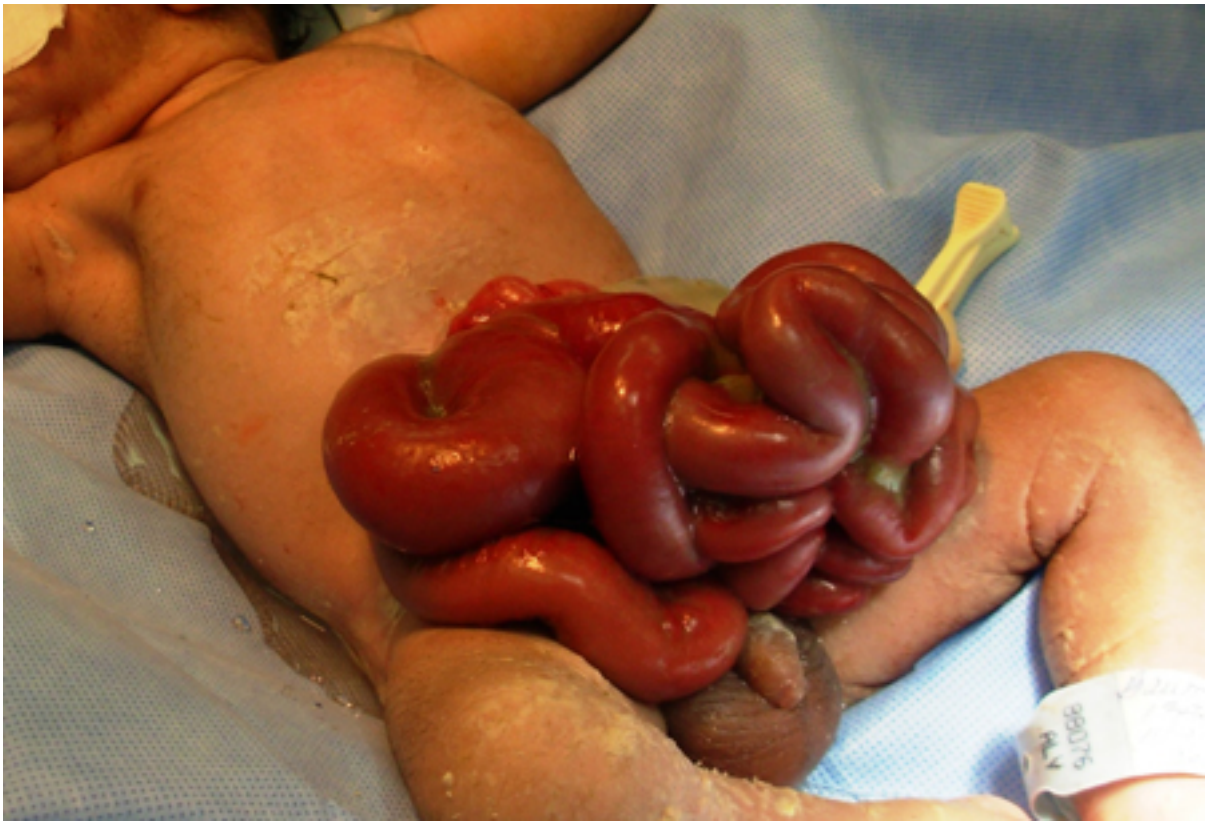

Figure 3: Extruded gut in abdominal wall defect

From collection of J.J. Tepas III, MD, FACS, FAAP

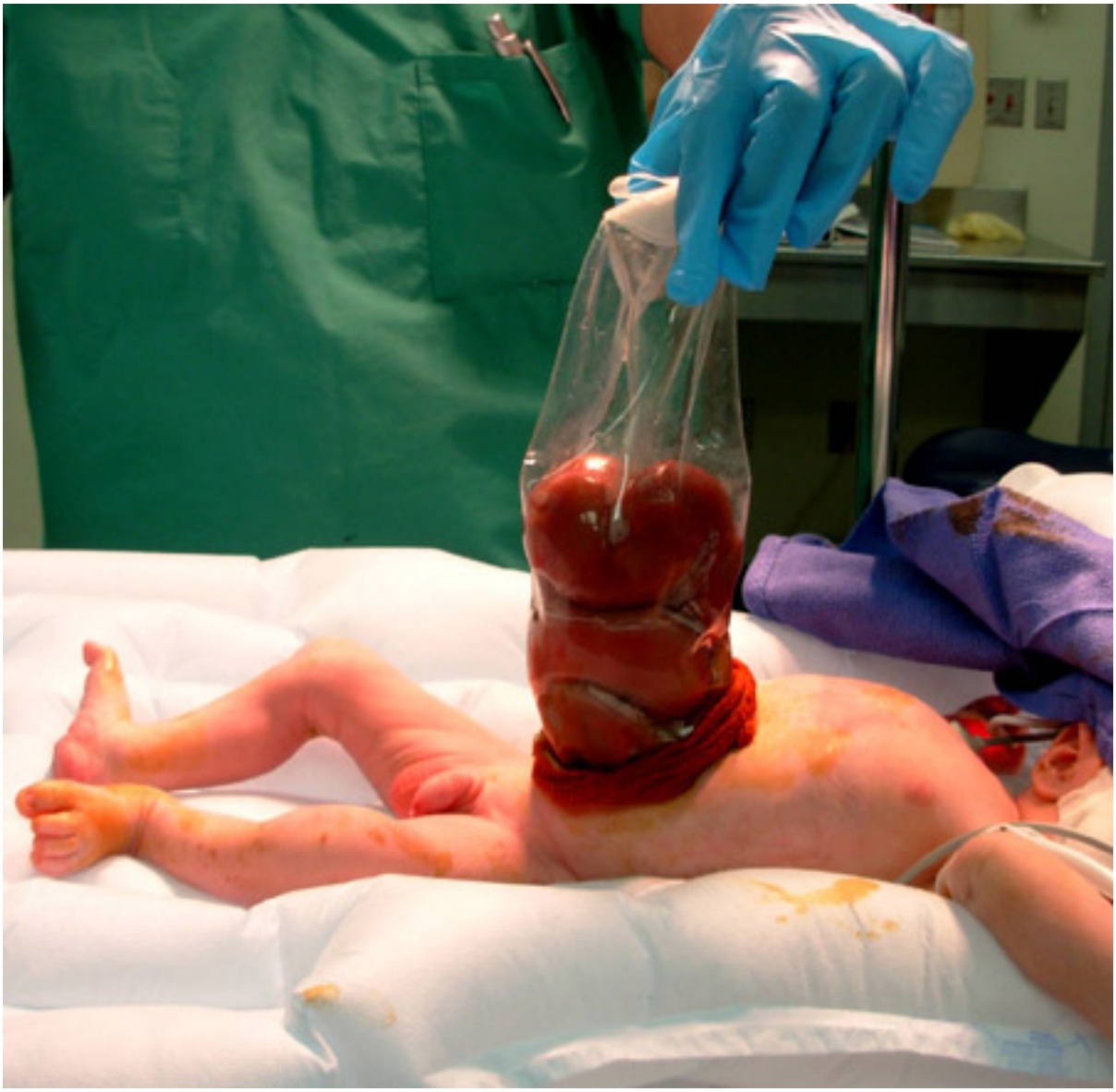

Figure 4: A staged repair of gastroschisis involves the placement of a silo to reduce contents into the abdomen

From collection of J.J. Tepas III, MD, FACS, FAAP

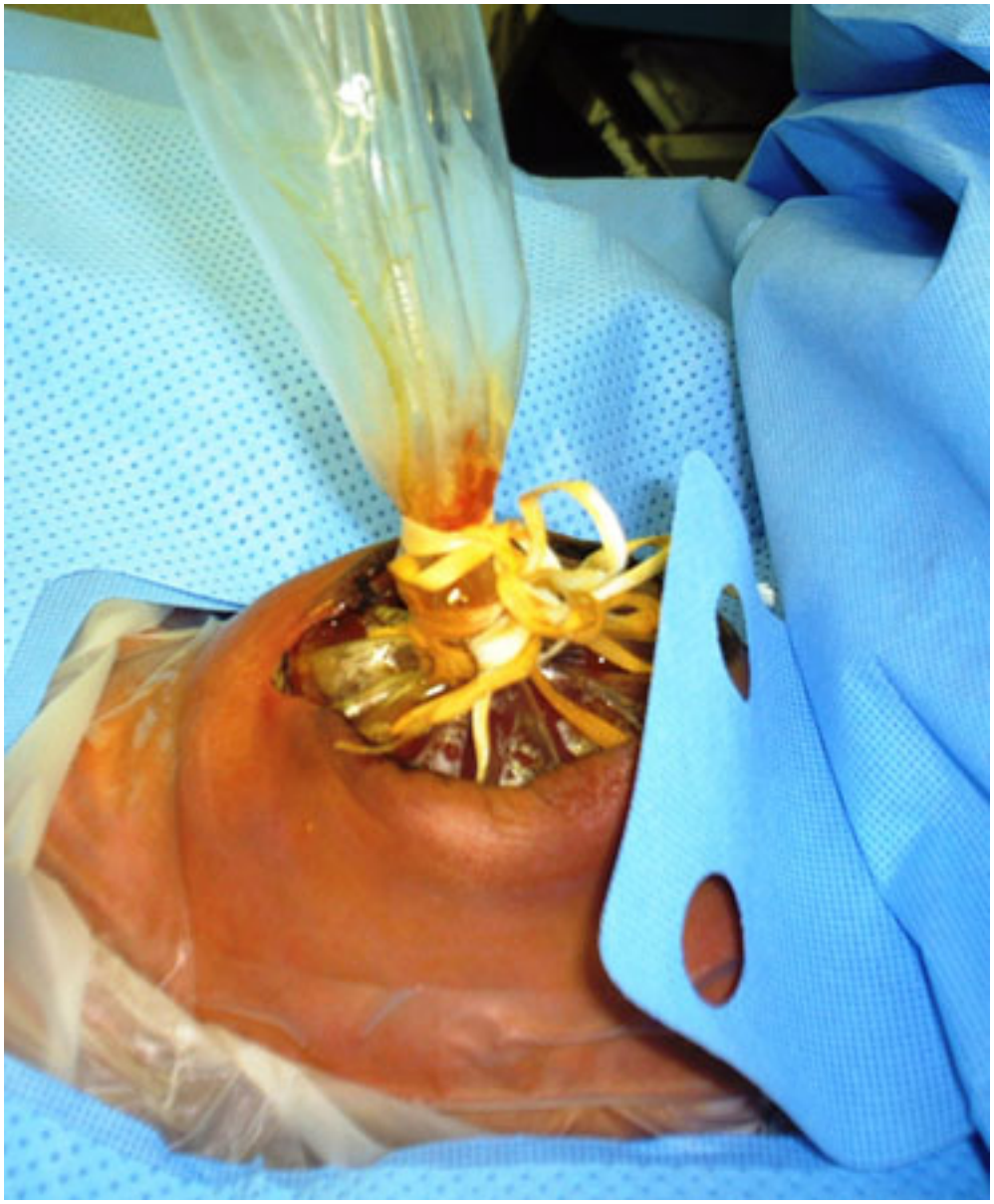

Figure 5: Once intestinal contents are fully reduced into the abdomen, closure of the abdominal wall follows

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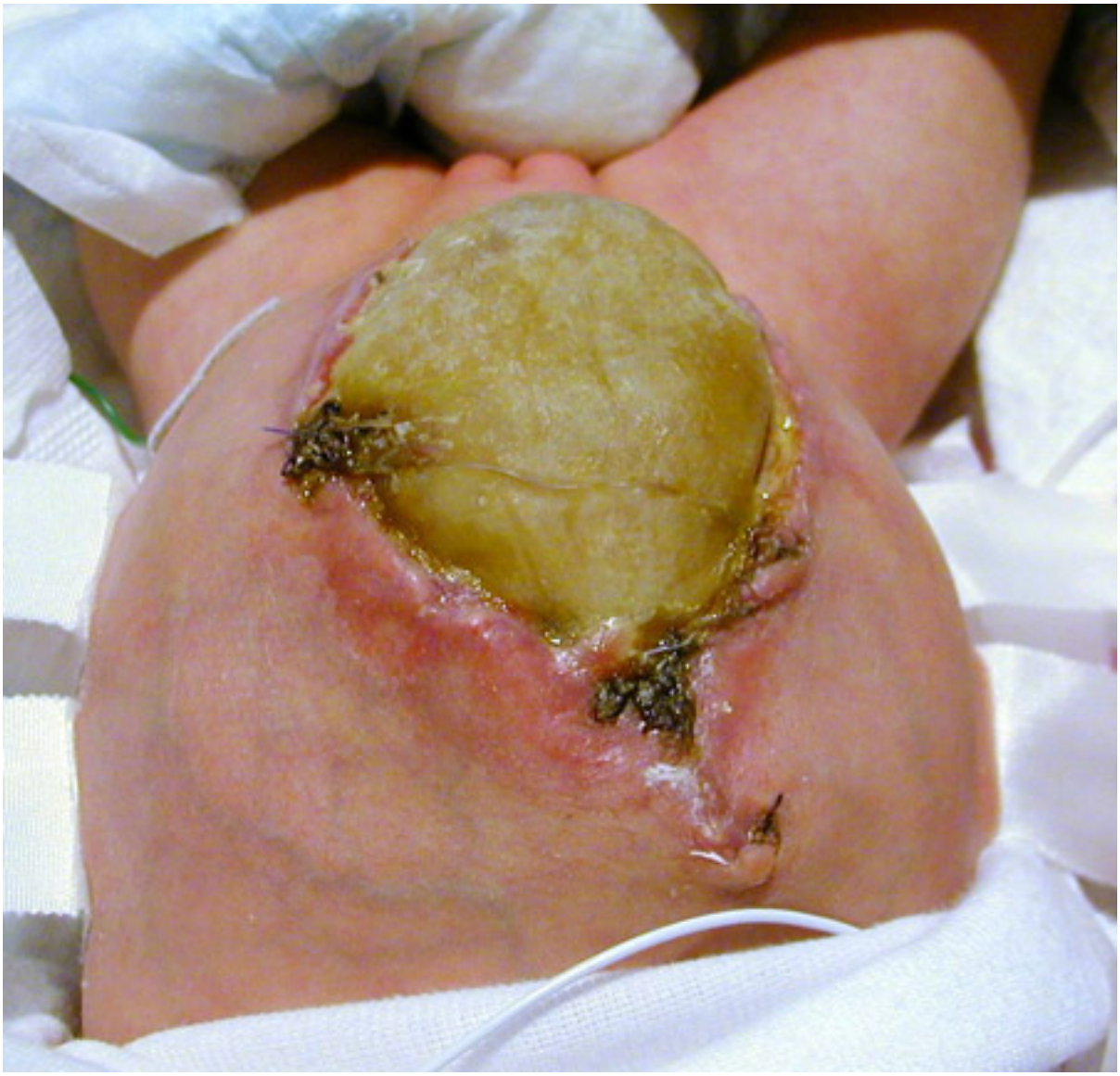

Figure 6: The abdominal wall defect in omphalocele is covered with a synthetic membrane

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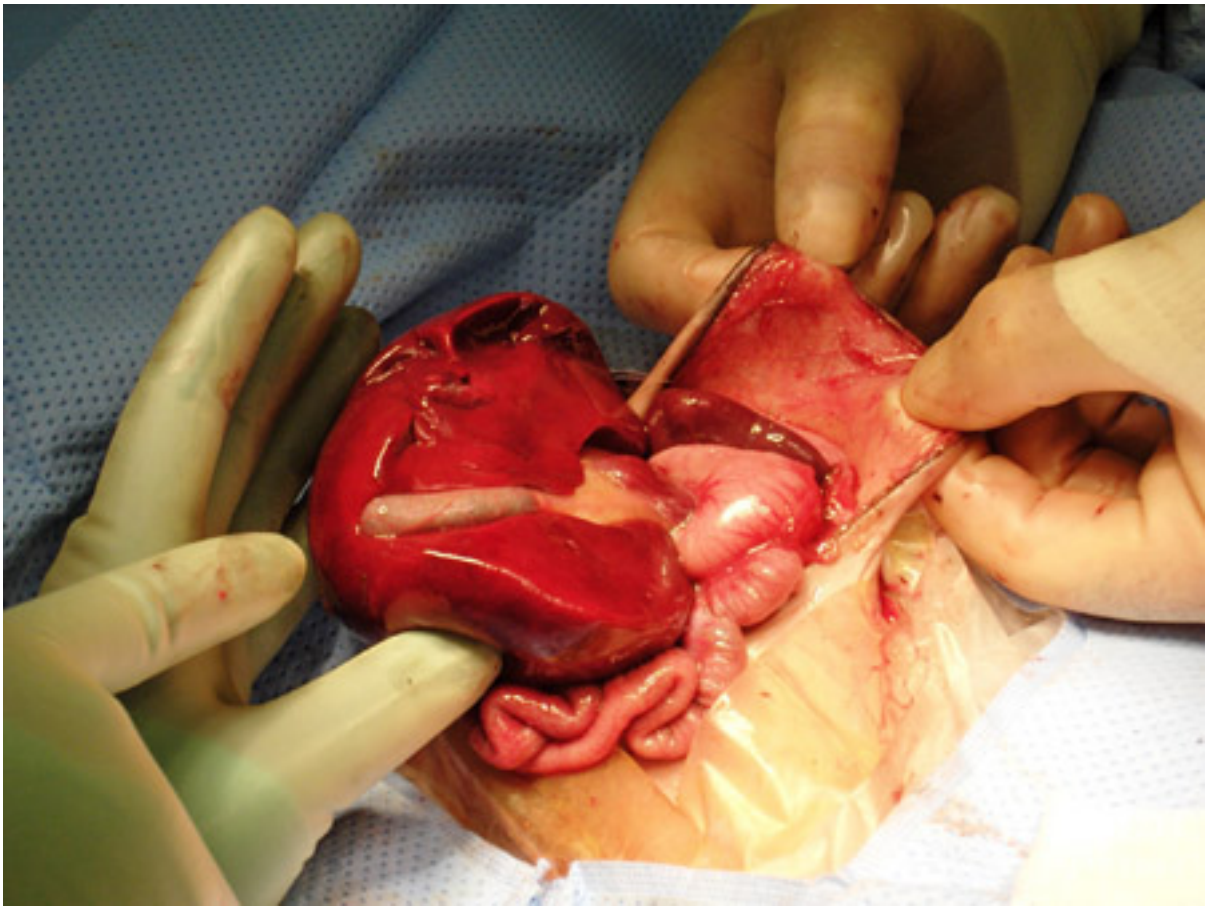

Figure 7: This ruptured omphalocele is similar in appearance to gastroschisis

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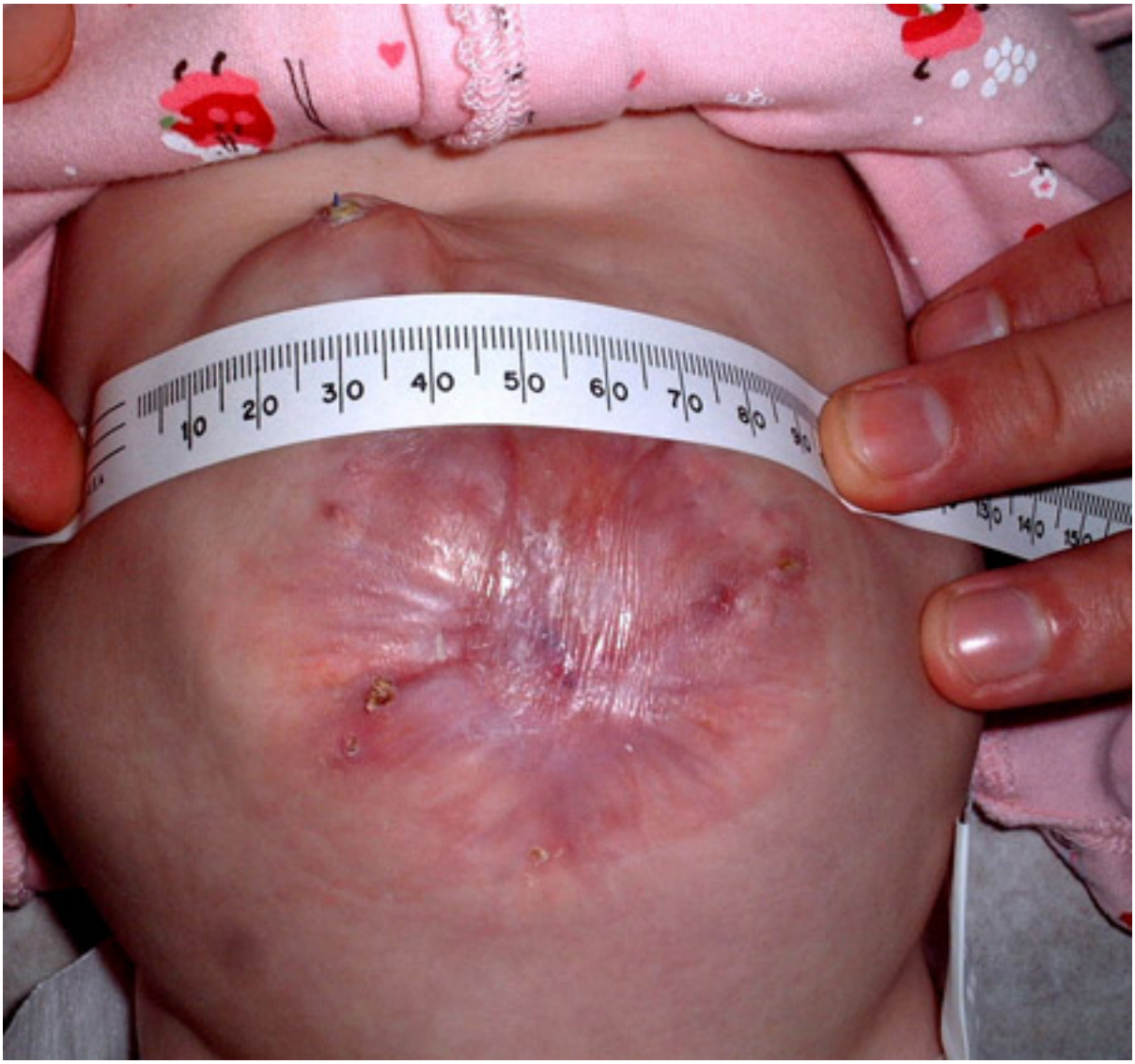

Figure 8: A ventral hernia results from synthetic wall closure of the omphalocele

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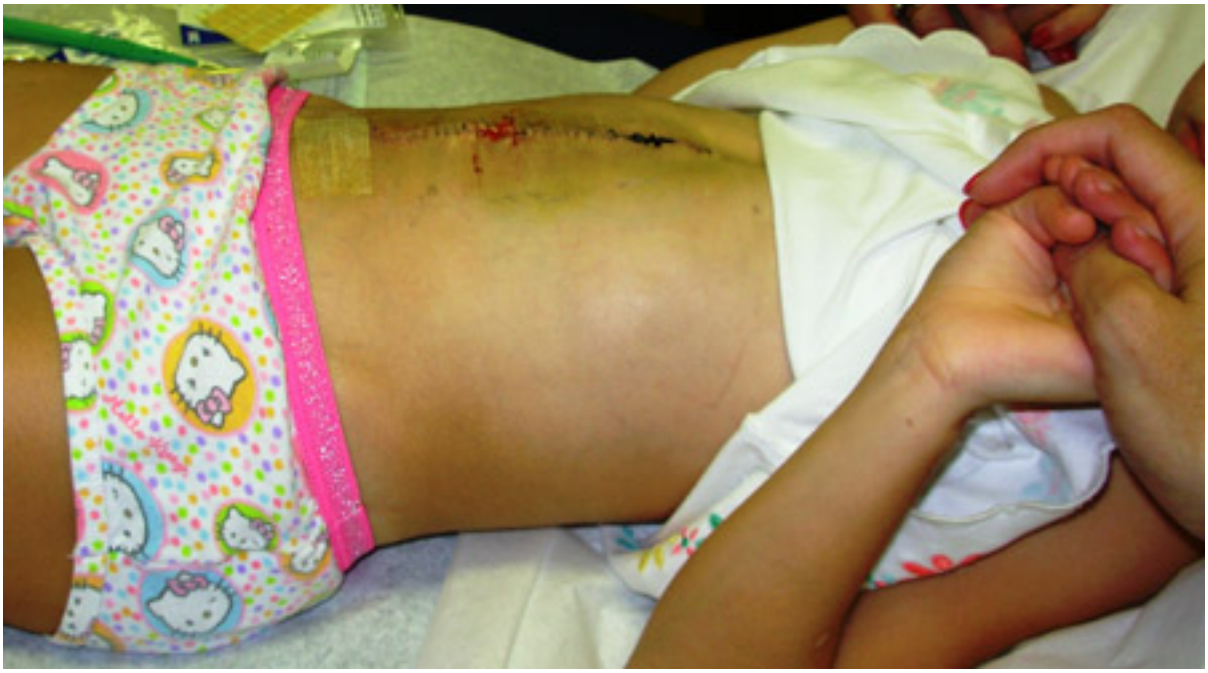

Figure 9: The ventral hernia is repaired in a 6-year-old girl born with omphalocele

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This approach is in line with the guidance of the International Bureau of Weights and Measures Service.

Figure 1 – BMJ Best Practice Numeral Style

5-digit numerals: 10,000

4-digit numerals: 1000

numerals < 1: 0.25

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