Pictorial Essay

Prenatal MRI of Congenital Abdominal and Chest Wall Defects

Pedro Daltro[1], Bradley L. Fricke[2], Beth M. Kline-Fath[2], Heron Werner[1], Leise Rodrigues[3], Tatiana Fazecas[3], Romeu Domingues[3], Lane F. Donnelly[2]

Fetal MRI is being increasingly used in the evaluation of fetal abnormalities [1]. It is particularly useful in evaluating the anatomic details of complex anomalies [1–9]. The additional information beyond that obtained on fetal sonography can be useful for prenatal counseling, planning for delivery, and planning for prenatal or postnatal intervention. We describe and illustrate the fetal MRI appearances of congenital abdominal and chest wall anomalies.

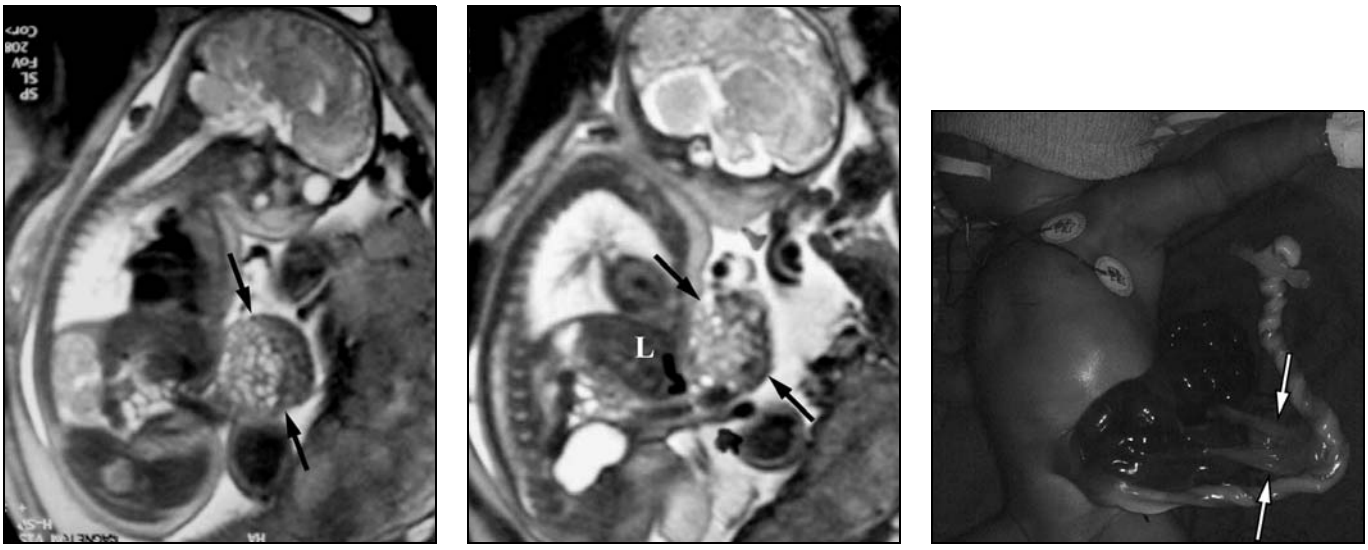


Fig. 1.—Omphalocele in 31-week fetus.
A and **B**, Sagittal MR images show herniation through anterior abdominal wall defect of bowel and peritoneal covering (*arrows*). In **B,** liver (L) is interior to abdominal wall. **C**, Photograph of infant shows omphalocele protruding through anterior abdominal wall defect. Peritoneal covering (*arrows*) has been incised and pulled away to reveal bowel. (Reprinted with permission from [9])

Received June 2, 2004; accepted after revision August 10, 2004.

[1]Clinica de Diagnostico por Imagem, Barra da Tijuca; Instituto Fernandes Figueira-FIOCRUZ-Rio de Janeiro, Brazil.

[2]Department of Radiology, Cincinnati Children's Hospital Medical Center, 3333 Burnet Ave., Cincinnati, OH 45229-3039. Address correspondence to L. F. Donnelly.

[3]Instituto Fernandes Figueira-FIOCRUZ-Rio De Janeiro, Brazil.

AJR 2005;184:1010–1016 0361–803X/05/1843–1010

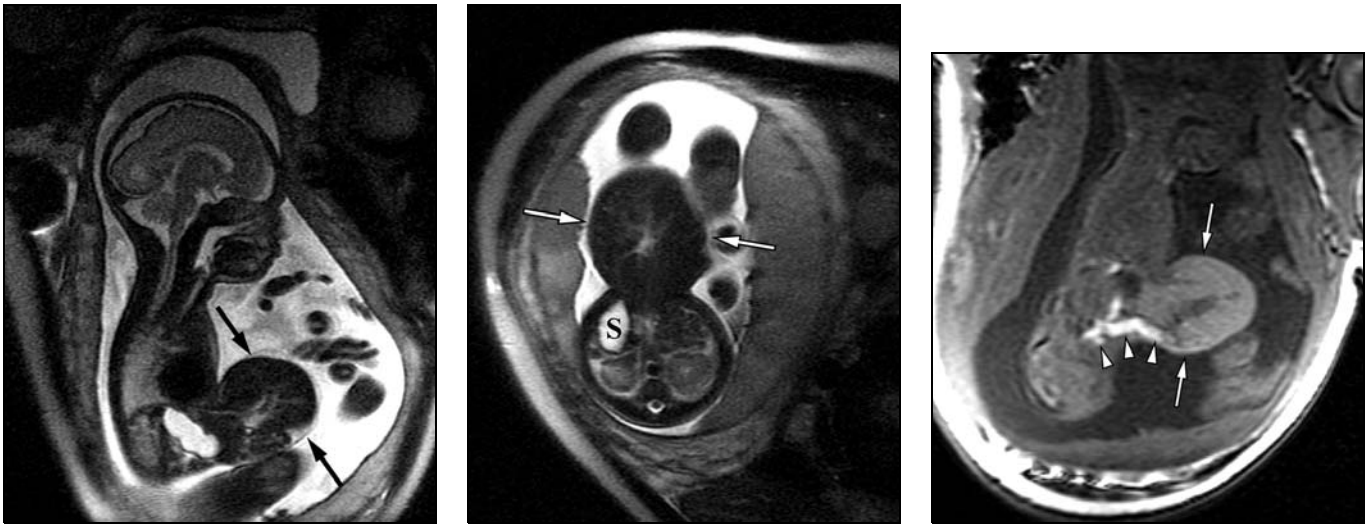


Fig. 2.—Omphalocele in 26.3-week fetus.
A and **B**, Sagittal MR images show herniation through anterior abdominal wall defect of liver and bowel and peritoneal covering (*arrows*). In **B,** stomach (S) is interior to abdominal wall.
C, Sagittal MR image shows omphalocele (*arrows*) with meconium-filled colon (*arrowheads*) extending through anterior abdominal wall defect.

Fetal MRI is performed with a single-shot rapid acquisition sequence with refocused echoes. This sequence minimizes effects from fetal motion by producing T2-weighted images in less than a second. Gradient-echo T1-weighted imaging is used as an adjunct to verify meconium in the bowel, liver anatomy, or intracranial hemorrhage.

Omphalocele

An omphalocele is an anterior abdominal wall defect encased by parietal peritoneum that results in the herniation of abdominal contents into the base of the umbilical cord [2, 3]. Omphalocele occurs in one of every 4,000 live births [2, 4]. Omphaloceles that contain liver parenchyma are thought to result from the failure of the lateral mesodermal body fold to migrate centrally and close the body wall [2, 3]. Omphaloceles consisting of mostly bowel with no evidence of liver parenchyma may result from the persistence of the primitive body stalk beyond 12 weeks' gestation [2].

Omphaloceles are commonly (54.2%) found to be associated with other anomalies, which determines prognosis [2, 3]. Chromosomal abnormalities, especially trisomy 13 and 18 syndromes, occur in 30–40% of patients [2, 3]. Beckwith-Wiedemann syndrome, which includes macrosomia, macroglossia, and organomegaly, is found in 5–10% of patients born with omphalocele [2, 3]. Other associated defects include cardiac, genitourinary, gas-

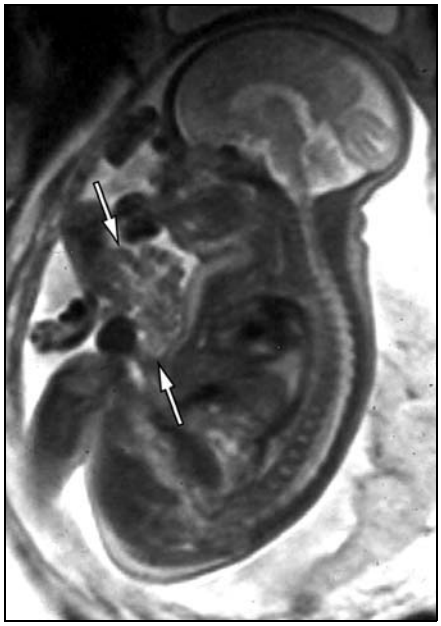


Fig. 3.—Gastroschisis in 29-week fetus. Sagittal MR image shows bowel (*arrows*) protruding through anterior abdominal wall defect. (Reprinted with permission from [9])

trointestinal, musculoskeletal, neural tube, and head and neck anomalies [2, 3].

Diagnosing omphalocele on prenatal sonography in the second or third trimester has varying rates of success (66–93%) [5, 9]. Additional anatomic detail can be obtained on MRI. MRI evaluation of the fetus with omphalocele shows a central abdominal wall defect, with abdominal viscera herniating into a thin-walled sac separate from the surrounding amniotic fluid (Fig. 1). The liver can be seen on MRI as a solid organ of low signal intensity on T2-weighted images (Fig. 2). The signal of the liver on prenatal MRI is often lower than that seen in the liver on similar sequences in children and adults.

The mortality rate is 80% when any associated defect is present and increases to near 100% when chromosomal or cardiovascular anomalies exist [2]. However, if an omphalocele is found in isolation, then the mortality rate decreases to 10% [2]. Therefore, detection of associated abnormalities on fetal imaging has great prognostic significance. In uncomplicated cases, the size of an omphalocele does not affect prognosis [4]. Accurate diagnosis is essential for parental counseling to avoid misclassification between omphalocele and gastroschisis, which may influence elective termination rates [3].

Gastroschisis

Gastroschisis is the herniation of fetal bowel loops into the amniotic cavity through a typically right-sided paraumbilical abdominal wall defect [2, 6]. This anomaly does not have a surrounding membrane and has an incidence of 1–6 per 10,000 live births [6]. The cause of gastroschisis is unclear but likely is heterogenous [2, 3, 6, 10]. One theory states that premature atrophy or abnormal persistence of the right umbilical vein results in rupture of the anterior abdominal wall at a point of weakness [2, 6]. Another speculates the defect is caused by ischemic damage to the anterior abdominal wall by premature interruption of the omphalomesenteric artery [2, 6].

Associated anomalies are rare in gastroschisis except related bowel abnormalities, usually intestinal atresia or stenosis from vascular compromise [2, 5, 6]. Other bowel complications include obstruction, perforation, peritonitis, motility dysfunction, necrotizing enterocolitis, short-gut syndrome, and fistulas [6]. The intrauterine mortality rate is 10–15%, and the condition of the bowel at birth is the single most important prognostic factor [6].

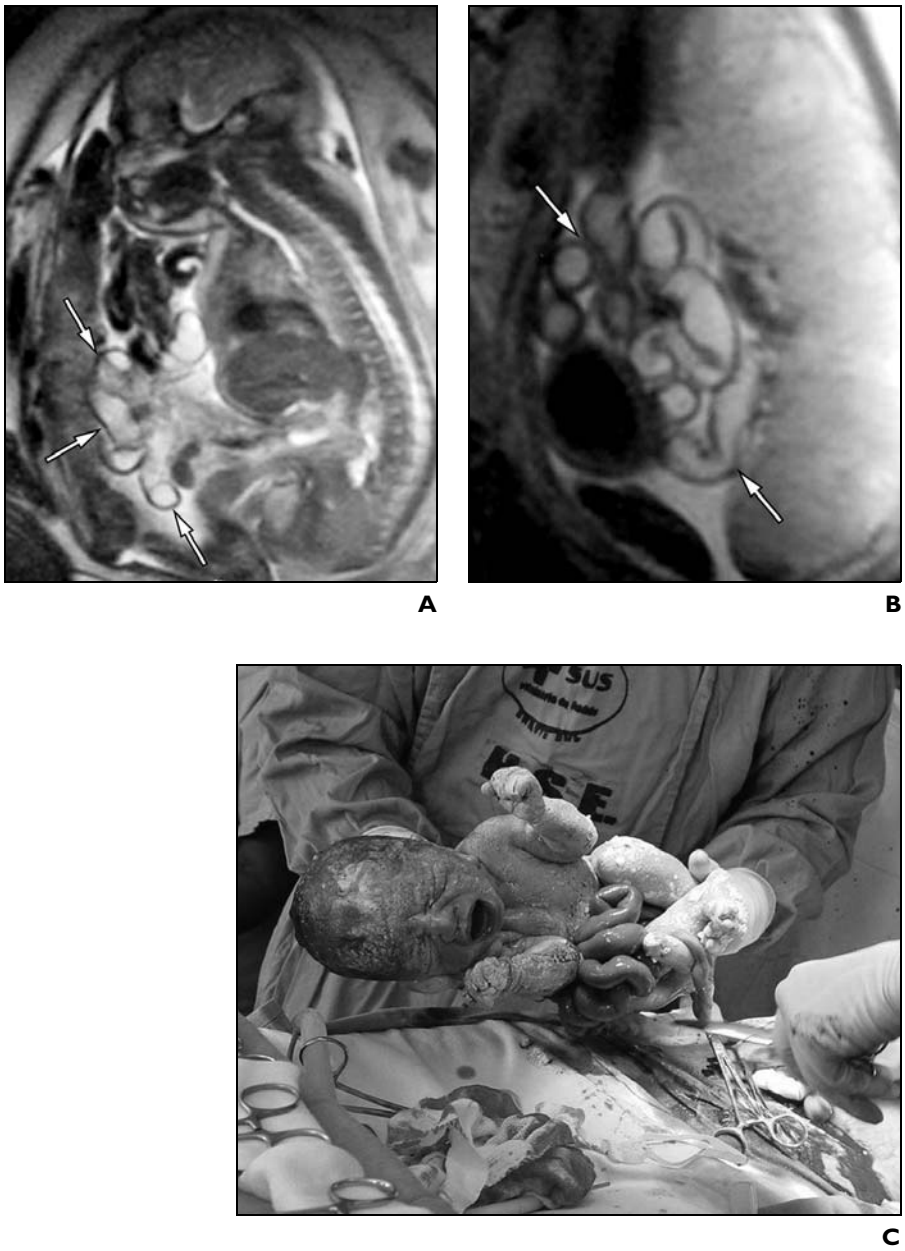
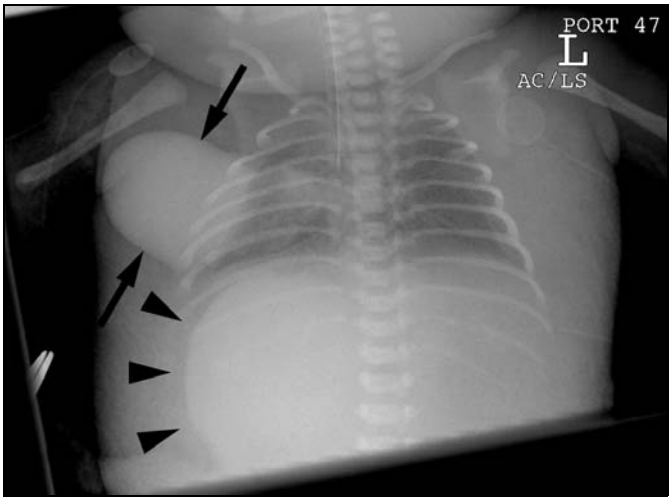
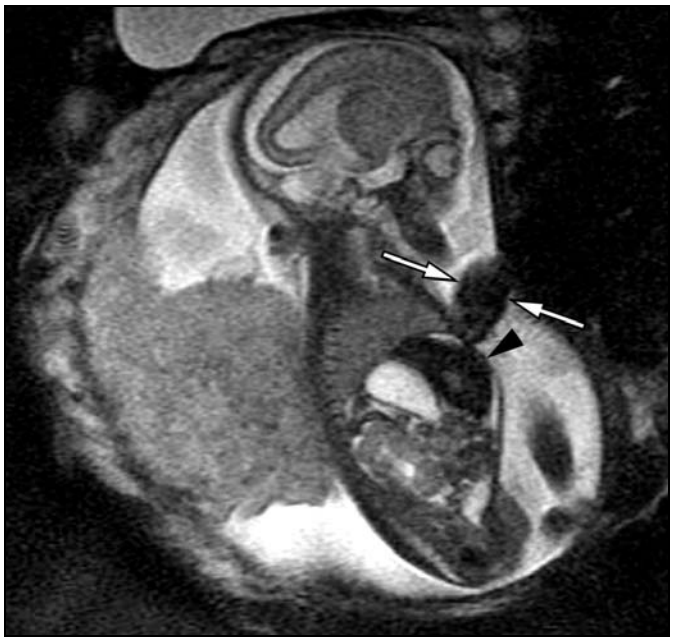


Fig. 4.—Gastroschisis in 30-week fetus.
A, Sagittal MR image shows bowel loops (*arrows*) outside abdominal cavity. Bowel is dilated and fluid-filled, indicative of bowel abnormality. No peritoneal covering is seen.
B, Axial MR image shows bowel (*arrows*) floating freely in amniotic fluid. Bowel is dilated and fluid-filled, with thickened wall.
C, Photograph of cesarean delivery shows bowel protruding through anterior abdominal wall defect.

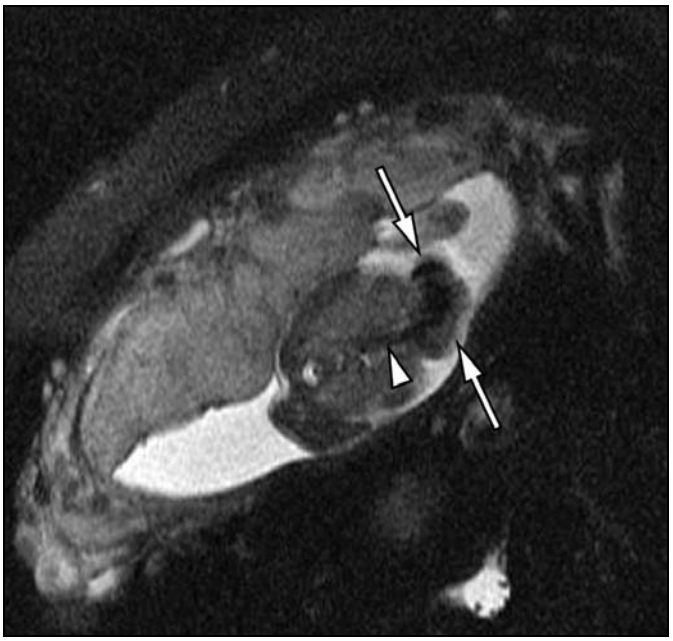
MRI evaluation shows bowel loops herniating through an abdominal wall defect adjacent to an intact umbilical cord. The bowel loops are not surrounded by a membrane and can be seen freely floating in the amniotic fluid (Fig. 3). Bowel loops may be dilated and fluid-filled with a thickened bowel wall, indicative of bowel abnormalities [2, 5, 6] (Fig. 4). The presence of free-floating extracorporeal solid organs may indicate a ruptured omphalocele is present rather than gastroschisis [5]. Overall survival rates are good (85–97%) [6], but a mortality rate of up to 28% has been reported, usually from prematurity, sepsis, and associated bowel abnormalities [4]. Antenatal diagnosis of gastroschisis may fa-



A



B



C

Fig. 5.—Pentalogy of Cantrell in 32-week fetus.
A, Frontal chest radiograph on day 1 of life (after 32 weeks' gestation) shows heart (*arrows*) outside thoracic cavity. Rim of increased radiolucency (*arrowheads*) is seen around liver, suggesting liver is outside abdominal cavity.
B, Sagittal MR image obtained at 20 weeks' gestation shows heart (*arrows*) outside thoracic cavity. Liver (*arrowhead*) is seen protruding through anterior abdominal wall defect.
C, Axial MR image shows heart (*arrows*) external to chest wall, with aorta (*arrowhead*) traversing chest wall defect.

cilitate planned delivery in a specialized unit, parental counseling, and surgical planning.

Pentalogy of Cantrell

In 1958, Cantrell described a syndrome consisting of defects of the midline supraumbilicus, the lower sternum, the anterior diaphragm, the diaphragmatic pericardium, and the heart [2, 4, 7, 9]. Pentalogy of Cantrell is rare, with fewer than 60 cases reported in the literature [4]. The cause is thought to result from incomplete ventral migration of sternal anlage and myotomes at 14–19 days of gestation [7]. The sternal and abdominal wall defects are thought to arise from this abnormal mesodermal migration [4]. Failure of the transverse septum to develop and abnormal development of the myocardium result in diaphragmatic and cardiac defects, respectively [4].

The abdominal wall defect is commonly an omphalocele, but diastasis recti abdominis, epigastric hernia, and umbilical hernia have also been reported [7]. An inferior sternal cleft usually constitutes the sternal defect, with the heart external to the defect [7]. The anterior diaphragmatic defect is present in 91% of cases, and continuity between the pericardial and peritoneal cavities is not rare [7]. Ventricular septal defect is almost universal, with pulmonic stenosis, atrial septal defect, and tetralogy of Fallot occurring less commonly [7]. Patients may also have diverticula of either ventricle, with left diverticula occurring more frequently [7]. This syndrome may be associated with chromosomal anomalies, including trisomy 13 and 18 syndromes and Turner's syndrome [4]. Other associated anomalies include ascites, a two-vessel cord, scoliosis, and craniofacial abnormalities [4].

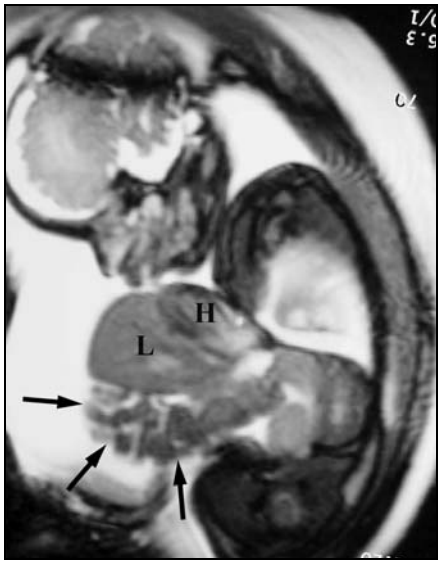
Imaging findings usually show omphalocele, ectopic heart, and pericardial or pleural effusions [4] (Figs. 5 and 6). The diaphragmatic defects may be difficult to characterize on MRI [4]. The prognosis of pentalogy of Cantrell depends on the severity of associated abnormalities but is generally poor, with a high infant mortality rate [2, 4, 7]. Cardiac diverticula should be repaired early to prevent rupture, and staged repair may help alleviate high intraabdominal and intrathoracic postoperative pressures [7].

Limb–Body Wall Complex

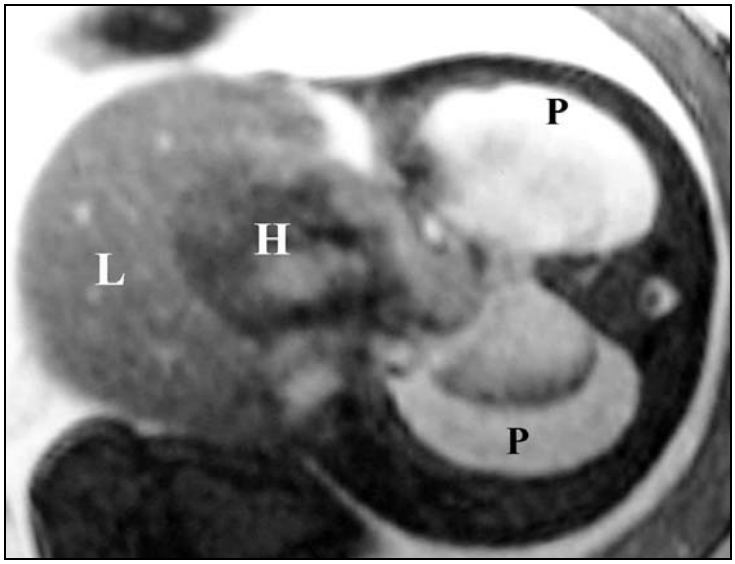
Limb–body wall complex consists of a variable group of congenital limb and body wall defects of the chest and abdomen [4]. Reported defects include a large, usually left-sided, abdominoschisis; a thoracic wall defect; rotational anomalies of the lower limbs; clubfoot; brachydactyly; polydactyly; syndactyly, absent limbs; and scoliosis [4, 8]. This syndrome is rare, with a reported incidence of one in more than 14,000 live births [4]. The etiology is controversial; the proposed mechanisms are amnion rupture, early vascular disruption, and embryologic malformation with abnormal development of the body folds [4, 8].

Associated craniofacial anomalies include exencephaly, encephalocele, and facial defects [4, 8]. Myelomeningocele may be present, leading to Arnold-Chiari malformation and hydrocephalus [8]. In addition, the internal organs are frequently affected with a high incidence of cardiac and diaphragmatic defects, bowel atresia, renal agenesis, and hydronephrosis [4, 8]. The umbilical cord is typically short or absent, with the placenta attached to the fetal trunk and viscera herniated [4] (Fig. 7). Amniotic bands are present in up to 40% of cases [4].

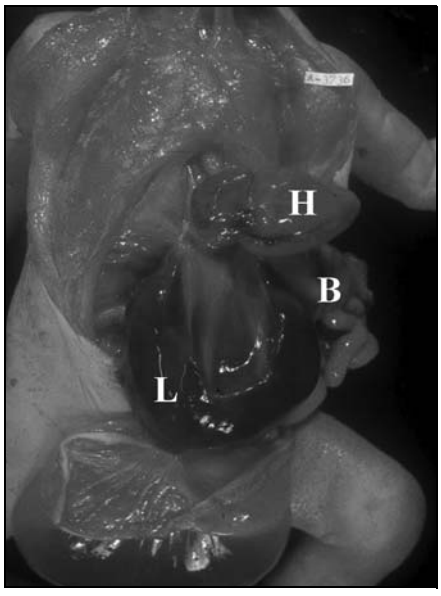
Imaging findings may show an abnormally located placenta without evidence of an umbilicus [8]. The abdominal, thoracic, limb, craniofacial, and internal organ anomalies are



A



B



C

Fig. 6.—Pentalogy of Cantrell in 27-week fetus.
A, Sagittal MR image shows heart (H), liver (L), and bowel (*arrows*) protruding through anterior abdominal and chest wall defect.
B, Axial MR image shows heart (H) and liver (L) protruding from defect in anterior body wall of fetus. Bilateral pleural effusions (P) are evident. (Reprinted with permission from [9])
C, Photograph of gross abortus shows heart (H), liver (L), and bowel (**B**).

variably displayed on imaging evaluation [4]. Often, herniated abdominal organs are seen entangled in membranes and forming a complex mass [4] (Fig. 7). Amniotic bands, when present, may be seen as linear structures but can be nonvisualized as a result of volume averaging [4]. Limb–body wall complex is universally fatal [4]. Thus, the prenatal diagnosis and differentiation of limb–body wall complex from other abdominal wall defects are crucial for parental counseling [8].

Bladder Exstrophy

Bladder exstrophy is the herniation of the urinary bladder through an infraumbilical anterior abdominal wall defect [4]. The incidence of bladder exstrophy is one in every 33,000 live births [2, 4]. This defect occurs during cloacal development, with abnormal retraction of the cloacal membrane resulting in eversion of the bladder [2, 4].

Associated genitourinary findings include extension of the bladder defect into the urethra, incomplete testicular descent, and bilateral inguinal hernias [2]. Bladder exstrophy is also associated with OEIS complex (omphalocele, extrophy [bladder], imperforate anus,

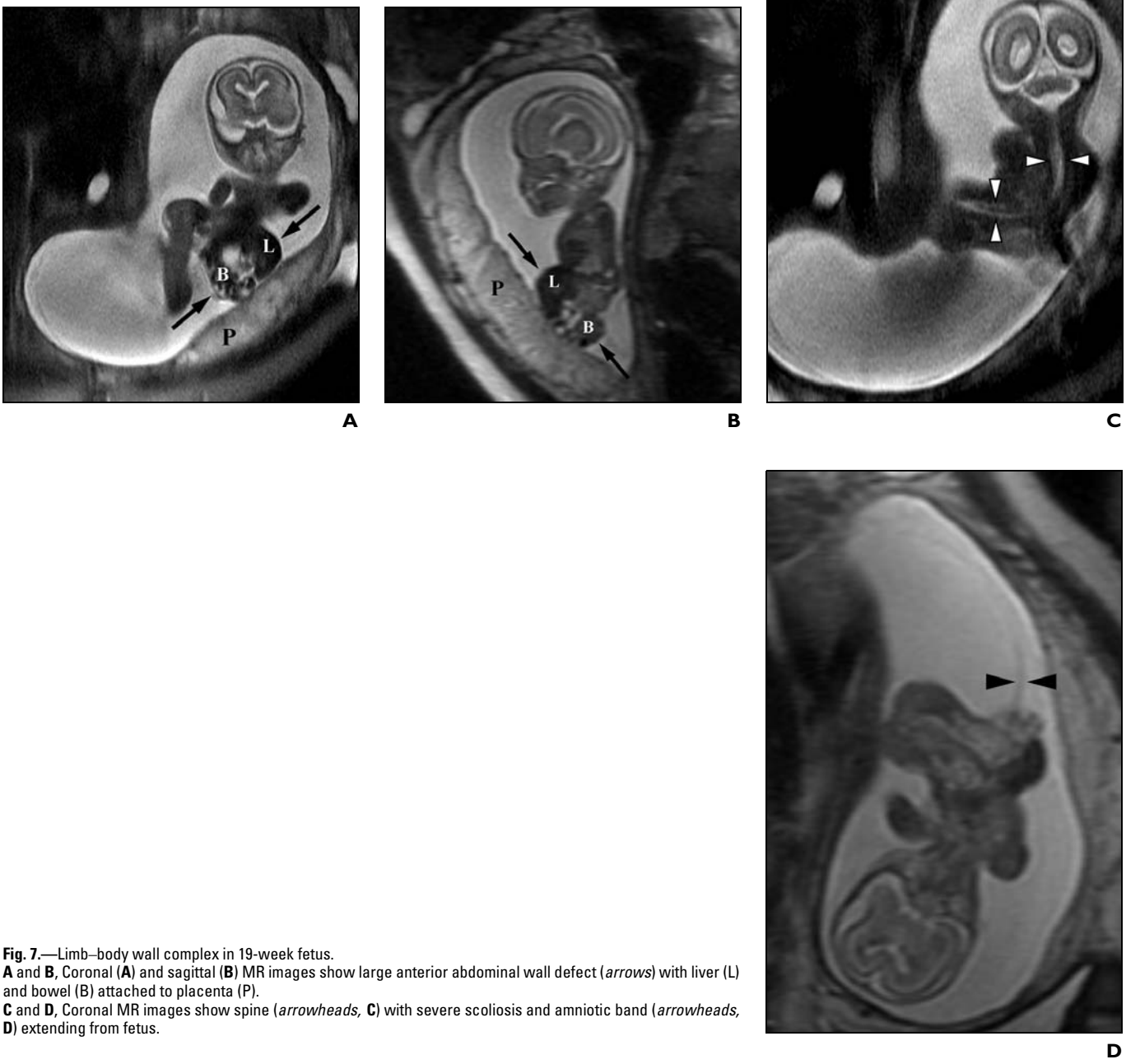


Fig. 7.—Limb–body wall complex in 19-week fetus.
A and **B**, Coronal (**A**) and sagittal (**B**) MR images show large anterior abdominal wall defect (*arrows*) with liver (L) and bowel (B) attached to placenta (P).
C and **D**, Coronal MR images show spine (*arrowheads,* **C**) with severe scoliosis and amniotic band (*arrowheads,* **D**) extending from fetus.

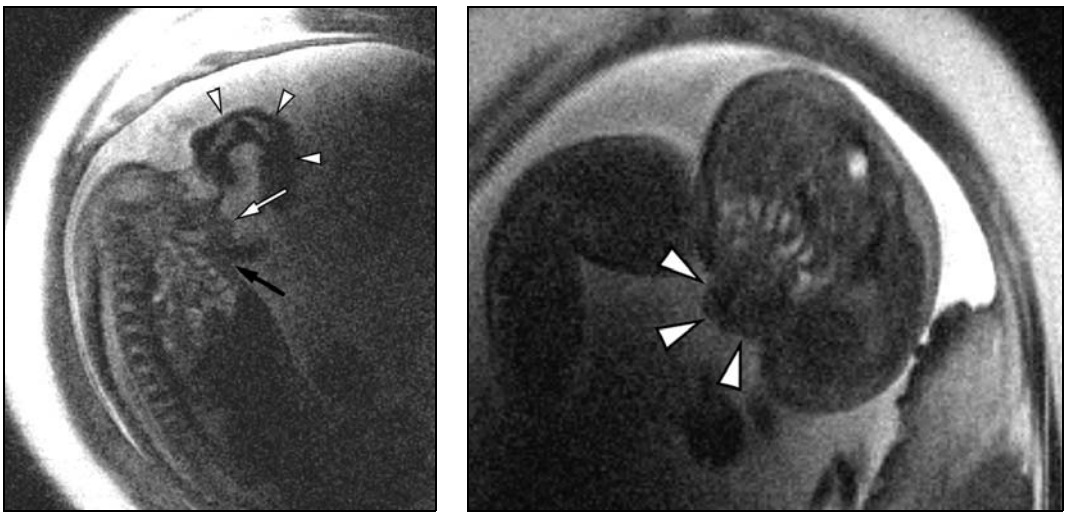


Fig. 8.—Bladder exstrophy in 32-week fetus.
A, Sagittal MR image shows bladder (*white arrow*) outside anterior body wall. Umbilical cord (*arrowheads*) with low-lying insertion (*black arrow*) is also seen.
B, Axial MR image shows bladder (*arrowheads*) extending through anterior abdominal wall defect.

spinal defects) [4]. This complex can be differentiated from limb–body wall complex by the placental involvement and scoliosis seen with limb–body wall complex in OEIS complex [4]. Complications of bladder exstrophy include urinary incontinence, infertility, and pyelonephritis [2].

Imaging findings of bladder exstrophy include a soft-tissue mass extending from a large infraumbilical anterior wall defect [2, 4] (Fig. 8). The absence of a normal urinary bladder and a low-lying umbilical cord insertion may also indicate the diagnosis [4]. Prognosis is good for bladder exstrophy, with surgical intervention required for primary closure or excision with urinary diversion [2, 4].

References

1. Levine D. Magnetic resonance imaging in prenatal diagnosis. *Curr Opin Pediatr* 2001;20:1233–1240
2. Emanuel PG, Garcia GI, Angtuaco TL. Prenatal detection of anterior abdominal wall defects with US. *RadioGraphics* 1995;15:517–530
3. Calzolari E, Bianchi F, Dolk H, Milan M. Omphalocele and gastroschisis in Europe: a survey of 3 million births 1980-1990—EUROCCAT Working Group. *Am J Med Genet* 1995;58:187–194
4. Fogata ML, Collins HB 2nd, Wagner CW, Angtuaco TL. Prenatal diagnosis of complicated abdominal wall defects. *Curr Probl Diagn Radio* 1999;28:101–128
5. Chen CP, Liu FF, Jan SW, Sheu JC, Huang SH, Lan CC. Prenatal diagnosis and perinatal aspects of abdominal wall defects. *Am J Perinatol* 1996;13:355–361
6. Durfee SM, Downard CD, Benson CB, Wilson JM. Postnatal outcome of fetuses with the prenatal diagnosis of gastroschisis. *J Ultrasound Med* 2002;21:269–274
7. Herman TE, Siegel MJ. Special imaging casebook: pentalogy of Cantrell. *J Perinatol* 2001;21:147–149
8. Casken H, Atas B, Tuncer O, Odabas D, Dal H. A case of limb-body wall complex diagnosed in utero. *J Emerg Med* 2003;25:104–106
9. Werner H Jr, Brandao A, Daltro P. *Ressonancia magnética em obstetricia e ginecologia.* Rio de Janeiro, RJ, Brazil: Revinter, 2003