18.3 Abdominal Masses

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FETAL ABDOMINAL CYSTIC MASSES

Cystic abdominal masses in the fetus are common. Ozyuncu et al.[1] found a positive predictive value of 75% for identifying the body system of origin for prenatal cysts. Since the diagnosis rarely affects prenatal intervention or delivery, it is important to recognize the abnormality, give a differential diagnosis, and obtain postnatal follow-up to document resolution or make a definitive diagnosis for possible postnatal therapy.

Ovarian Cyst

Incidence: Fetal ovarian cysts are the most common abdominal cysts reported prenatally. The incidence, however, is not documented. The presence of small ovarian cysts in the perinatal period is normal. Cohen et al.[2] evaluated postnatal ultrasounds (US) and found that 82% of normal infants in the first 3 months of life had ovarian cysts measuring 0.1 to 1.4 cm.

Embryology and Pathology: Neonatal cysts are of germinal or Graafian epithelial origin and consist of follicular, theca lutein, corpus luteum, or simple cysts and result from enlargement of an otherwise normal follicle.[3]

A follicle may enlarge secondary to hormonal stimulation by follicle-stimulating hormone (FSH) the fetal pituitary, maternal estrogen, and placental human chorionic gonadotropin (HCG). There is increased incidence of fetal ovarian cysts in cases of maternal diabetes, rhesus sensitization, and preeclampsia, all of which are associated with increased serum chorionic gonadotropins. There is also a reported association with fetal hypothyroidism.[4]

After birth, HCG and estrogen drop, while FSH and luteinizing hormone (LH) increase until 3 months and then fall as the hypothalamic–pituitary axis matures. Cysts regress with time, likely because of hormonal changes.

Diagnosis

Ultrasound: Fetal ovarian cysts are most often identified after 28 weeks' gestation, though they are also noted as early as 19 weeks.[4,5] They are typically unilateral, but may be bilateral.

Ovarian cysts may be simple (Fig. 18.3-1) or complicated (Fig. 18.3-2). Simple cysts are anechoic and smooth with thin or imperceptible walls on US. Complicated cysts demonstrate hemorrhage as a fluid–debris level, retracting clot, multiple internal septations, or a solid mass. The wall may be thick and echogenic from dystrophic calcification.[3]

MRI: Simple cysts are hyperintense on T2w and hypointense on T1w following fluid signal. On T2w, the blood products

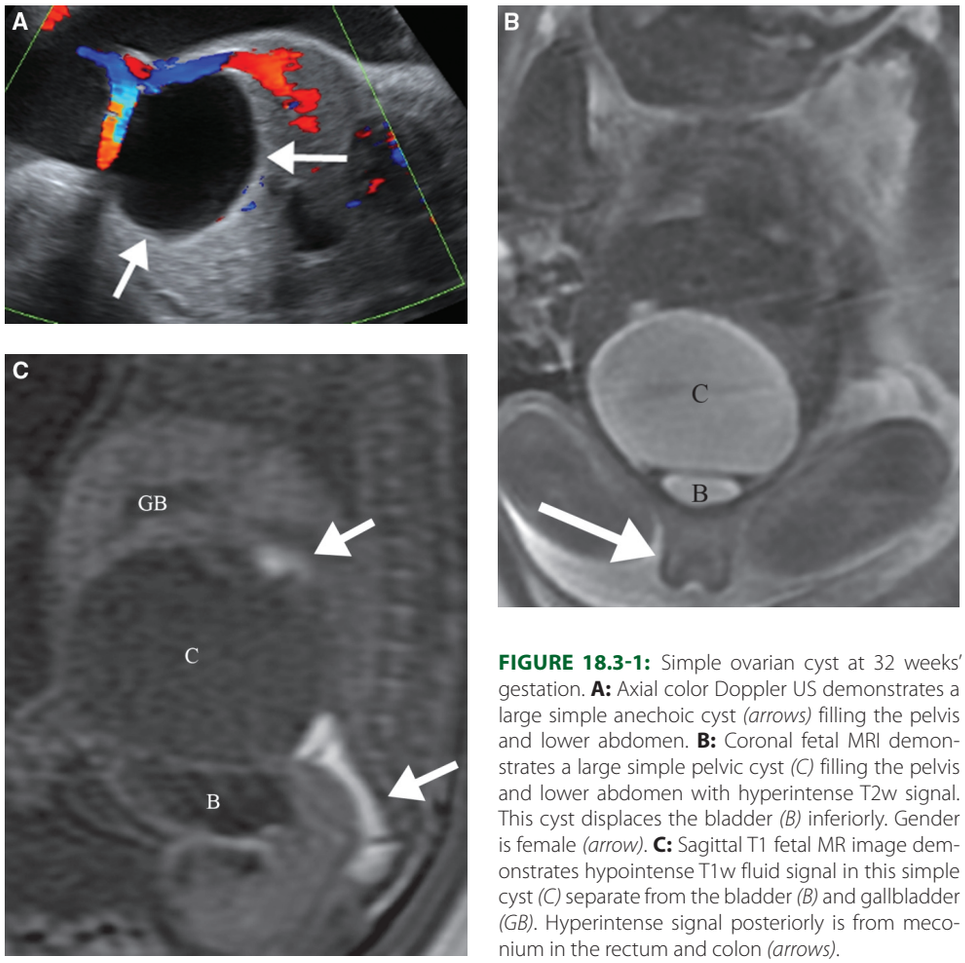

FIGURE 18.3-1: Simple ovarian cyst at 32 weeks' gestation. **A:** Axial color Doppler US demonstrates a large simple anechoic cyst *(arrows)* filling the pelvis and lower abdomen. **B:** Coronal fetal MRI demonstrates a large simple pelvic cyst *(C)* filling the pelvis and lower abdomen with hyperintense T2w signal. This cyst displaces the bladder *(B)* inferiorly. Gender is female *(arrow)*. **C:** Sagittal T1 fetal MR image demonstrates hypointense T1w fluid signal in this simple cyst *(C)* separate from the bladder *(B)* and gallbladder *(GB)*. Hyperintense signal posteriorly is from meconium in the rectum and colon *(arrows)*.

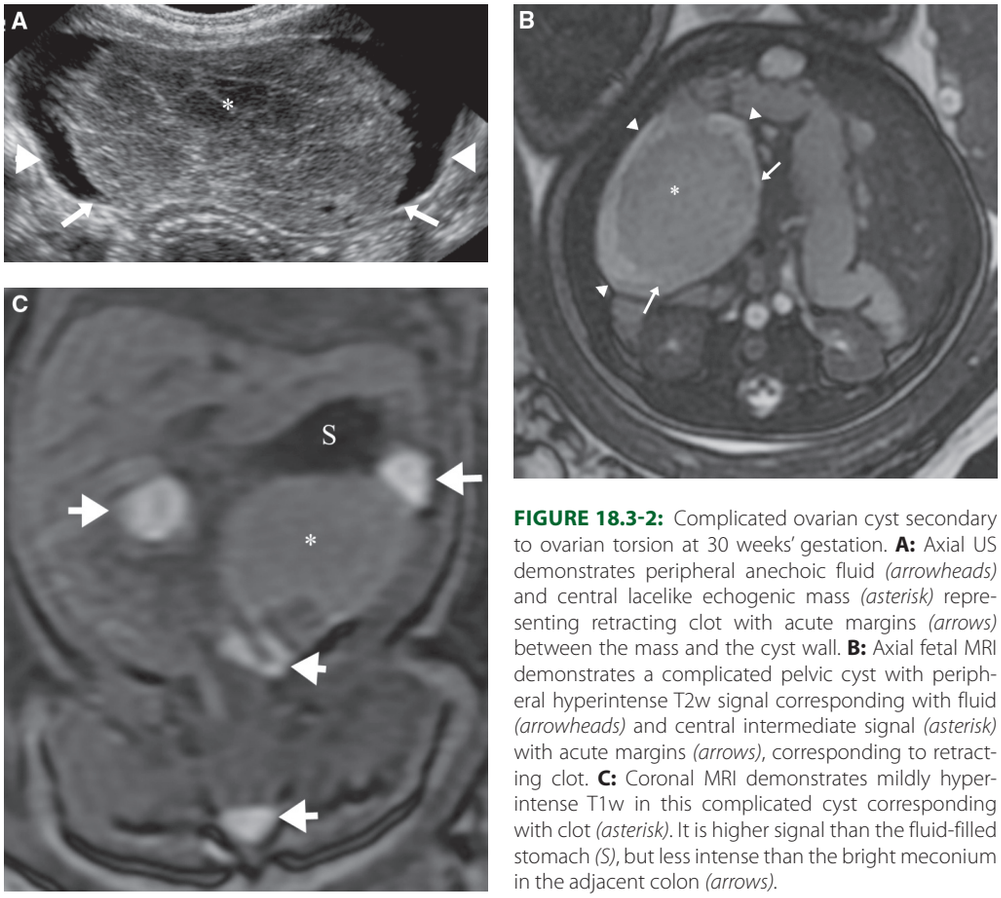

FIGURE 18.3-2: Complicated ovarian cyst secondary to ovarian torsion at 30 weeks' gestation. **A:** Axial US demonstrates peripheral anechoic fluid *(arrowheads)* and central lacelike echogenic mass *(asterisk)* representing retracting clot with acute margins *(arrows)* between the mass and the cyst wall. **B:** Axial fetal MRI demonstrates a complicated pelvic cyst with peripheral hyperintense T2w signal corresponding with fluid *(arrowheads)* and central intermediate signal *(asterisk)* with acute margins *(arrows)*, corresponding to retracting clot. **C:** Coronal MRI demonstrates mildly hyperintense T1w in this complicated cyst corresponding with clot *(asterisk)*. It is higher signal than the fluid-filled stomach *(S)*, but less intense than the bright meconium in the adjacent colon *(arrows)*.

in complicated cysts are lower signal than fluid. Hemorrhage causes hyperintense T1w. Marked hypointensity on gradient echo or echoplanar imaging indicates hemosiderin deposition. Nemac et al.[6] describes restricted diffusion and hyperintense FLAIR signal with intracystic hemorrhage. MRI can be useful to evaluate soft tissue components, but for the vast majority of cases, US diagnosis and follow-up is the imaging standard.[6]

Suggested criteria for fetal ovarian cyst include (1) female gender, (2) unilateral or bilateral pelvic or lower abdominal location, and (3) normal kidneys, bladder, and intestines. Cysts may be located anywhere in the abdomen or pelvis because of ligamentous laxity and a relatively long mesosalpynx, or because of torsion with autoamputation.[4,5] Cysts are considered pathologic if they measure 2 cm or greater. When very large, a cyst may fill the abdomen and pelvis, displacing normal structures and making it difficult or impossible to determine the source.

Cyst hemorrhage, with or without torsion, is the most common complication and is suspected when the cyst appears complicated. Ovarian cyst hemorrhage is highly associated with torsion. Ascites may occur with cyst rupture or torsion.[5] Anemia has been reported. Other complications are due to mass effect. Polyhydramnios occurs in up to 18% of cases,[7] and is likely secondary to extrinsic bowel obstruction by large cysts >6 cm.[8] Severe mass effect may rarely result in elevation of the diaphragm and pulmonary hypoplasia.[5]

Differential Diagnosis: The differential diagnosis for a fetal pelvic cyst includes lymphangioma, gastrointestinal (GI) duplication cyst, cystic teratoma, hydrocolpos, urachal cysts, and persistent cloaca. Identifying a normal urinary system excludes bladder outlet obstruction, megacystis microcolon intestinal hypoperistalsis syndrome, prune belly syndrome, and hydroureteronephrosis.

When a cyst is very large, it may not be possible to determine whether it arises from the abdomen or pelvis, and whether abdominal cysts should also be considered (Tables 18.3-1 and 18.3-2). Extremely rare fetal ovarian tumors such as benign cystic teratomas, mucinous and serous cystadenomas, and a single case of carcinoma have been reported.[5]

Prognosis: In a meta-analysis including 420 fetuses with ovarian cysts from 1984 to 2005, cysts spontaneously regress in 50%.[9] Spontaneous regression may occur either in utero or postnatally, and typically occurs within 1 to 12 months.[3,5] Both simple and complicated cysts may resolve, although there is a slightly lower regression rate of complicated cysts.[10]

Ovarian cysts are complicated by hemorrhage and/or torsion in 35%.[9] Regarding cyst size and prognosis, 98% of cysts <5 cm regressed spontaneously, whereas 93% of cysts greater than 5 cm resulted in complications.[9] However, there are studies indicating that size does not predict torsion and the complicated appearance, rate of growth, and length of the pedicle are more important.[3,7]

Management: Most authors recommended serial US follow-up for cysts less than 5 cm to document size and monitor for increasing complexity indicating hemorrhage/torsion. In utero needle

Table 18.3-1 Differential Diagnosis of Fetal Abdominal Cysts Based on Location

- Right upper quadrant
  - Choledochal
  - Hepatic cyst or cystic tumor
  - Duodenal duplication
  - Duodenal atresia
  - Pancreatic (head)
- Left upper quadrant
  - Splenic
  - Gastric duplication
  - Pancreatic
- Midabdomen
  - Lymphatic (mesenteric, omental)
  - Meconium pseudocyst
  - Umbilical vein varix
- Retroperitoneal
  - Common
    - Hydronephrosis
    - Renal cystic disease
  - Uncommon
    - Urinoma
    - Adrenal cyst
    - Cystic neuroblastoma
    - Lymphatic (retroperitoneal)
- Lower abdomen and pelvis
  - Common
    - Hydrometrocolpos
    - Ovarian
    - Ureterocele
    - Urachal
    - Megacystis
  - Uncommon
    - Persistent cloaca
    - Anterior sacral meningocele
    - Sacrococcygeal teratoma
- Any location
  - Alimentary duplication
  - Dilated bowel (obstruction, atresia)
  - Cystic teratoma

aspiration appears to be safe and effective, but is used rarely, with an 89% regression rate after aspiration. Persistent complicated or persistent large simple cysts (>5 cm) can be treated surgically to decrease the possibility of torsion or exclude an alternative diagnosis.[9] Since torsion often occurs in utero, postnatal surgery is primarily for the purpose of preventing complications such as hemorrhage, rupture with peritonitis, and intestinal obstruction from adhesions, rather than for ovarian salvage.

Recurrence Risk: Since the hormonal stimulation is removed in the neonatal period, they are not expected to recur after they spontaneously resolve or are surgically treated.

Hepatic Cyst

Incidence: Low but not precisely known.[11] There is a female predominance.

Embryology and Pathology: Simple hepatic cysts are presumed secondary to interruption of the intrahepatic biliary system with growth arrest and dilation, representing cysts of biliary origin.

Most antenatally diagnosed hepatic cysts are simple.[11] They most commonly have a cuboidal epithelial lining with positive CK-7 staining and a fibrous wall with occasional smooth muscle fibers, suggesting bile origin and supporting a biliary growth arrest etiology.[12] However, no epithelial lining[13] and simple squamous lining has also been reported.[11] Avni et al.[13] hypothesize that some hepatic cysts may represent areas of necrosis from an ischemic event, particularly those without an epithelial lining.

Diagnosis

Ultrasound: A hepatic cyst is most commonly seen in the second trimester (median gestational age 22 weeks), but has been reported in the third and first trimesters.[11,12] Hepatic cysts may be unilocular or multilocular. They may be clearly intrahepatic or subhepatic and attached to the liver capsule. Cysts as large as 20 cm have been reported.[14] Antenatally detected liver cysts are more often present in the left lobe.[11,14] On US, hepatic cysts are round, well defined, anechoic structures with posterior enhancement.

A large cyst may compress the umbilical vein, resulting in hydrops and fetal demise, may elevate the diaphragm and cause pulmonary hypoplasia, or may compress the bowel and cause polyhydramnios.[11,14]

MRI: Hepatic cysts follow fluid signal on MRI with hyperintense T2-weighted signal and hypointense T1-weighted signal surrounded by higher signal liver parenchyma (Fig. 18.3-3).

Differential Diagnosis: Simple hepatic cysts, intrahepatic choledochal cysts (Caroli disease), and mesenchymal hamartomas may look identical on prenatal imaging,[11] although simple hepatic cysts are statistically more common.[11,12] Consider rare entities like cystic hepatoblastoma, cystic teratoma, and hemangioendothelioma, particularly if there is question of a solid component or multilocularity. If the cyst is large and/or subhepatic, it may be difficult to differentiate from extrahepatic fetal abdominal cysts (Tables 18.3-1 and 18.3-2).

Prognosis: Most simple hepatic cysts are asymptomatic and remain unchanged or regress over time.[11] Complications are rare and secondary to large size and mass effect. Infection, hemorrhage, and torsion of the cyst are theoretical complications.

Management: Hepatic cysts are typically managed with serial prenatal US examinations to document size and mass effect on adjacent structures. A very large cyst resulting in mass effect on the chest, umbilical cord, or bowel may require in utero aspiration to allow normal fetal development.[15]

Postnatally, cholangiography and/or hepatobiliary scan may help differentiate a choledochal cyst from a simple hepatic cyst, which is managed differently. Postnatal surgery for simple cysts, including aspiration, fenestration, cystectomy, or hepatic lobectomy, is typically reserved for patients with symptoms because of cyst size or uncertainty about the diagnosis.[11,12]

Recurrence Risk: Hepatic cysts tend to stay the same or regress on follow-up imaging.[11]Surgically resected cysts have not demonstrated recurrence.[11,12]

Table 18.3-2 Comparison of Fetal Abdominal Cysts

Cyst	Locularity	Trimester	Gender	Location
Ovarian	U > M	3rd >> 2nd	F	Pelvis ± abdomen
Hepatic	U >> M	2nd > 3rd > 1st	F >> M	RUQ
Choledochal	U	2nd > 3rd	F >> M	RUQ
Gallbladder duplication	U	2nd or 3rd	—	RUQ
Enteric	U >> M	2nd or 3rd	M > F	Any
Lymphatic	M or U	2nd > 3rd	M ≥ F	Any
Splenic	U	3rd	—	LUQ
Pancreatic-isolated	M or U	3rd > 2nd	F > M	Upper abdomen
Pancreatic-with anomalies	M or U	2nd > 3rd	F ≥ M	Upper abdomen

U, unilocular; M, multilocular; F, female; M, male; Trimester, trimester at diagnosis; RUQ, right upper quadrant; LUQ, left upper quadrant.

Choledochal Cyst

Incidence: Variable depending on geography with two-thirds of reported cases involving the Japanese population. Females are affected more often than males by a ratio of 3:1 in the Japanese literature and 9:1 in the Western literature.[16,17]

Embryology and Pathology: The bile duct elongates and begins to recanalize at the end of the 5th week. By the 12th week, bile is secreted from the liver and transported to the duodenum by the extrahepatic biliary system.[18] The pancreas secretes enzymes in the 5th month.

The etiology is not proven. The most widely cited theory states that pancreaticobiliary reflux secondary to an anomalous long common pancreaticobiliary channel results in increased pressure, inflammation, and weakening resulting in cystic dilation. This does not explain cysts seen before pancreatic enzyme production. Alternatively, choledochal cysts may be due to distal obstruction by a web, stricture, biliary atresia, or sphincter of Oddi dysfunction.[19] Some hypothesize that prenatal infectious cholangitis causes biliary duct scarring resulting in a spectrum of obstructive cholangiopathy that includes choledochal cyst, biliary atresia, and neonatal hepatitis.[20] In utero accident has been suggested as well.

Choledochal cysts are classified by the Todani modified Alonso–Lej Classification (Fig. 18.3-4).[21–23]

There is evidence that 22% of patients in the United States with choledochal cysts have associated congenital cardiac anomalies including ASD, VSD, PDA, and persistent fetal circulation.[24] Choledochal cysts may also be associated with concurrent biliary atresia or be confused with the rare cystic biliary atresia.[25]

Diagnosis

Ultrasound: A choledochal cyst is a cystic mass at the liver hilum, subhepatic, or in the liver parenchyma (Type V), that is separate from the gallbladder. Choledochal cysts have been detected during second-or-third trimester US. It can be differentiated from other cystic masses if one can demonstrate a fusiform shape with tapered ends in continuity with the biliary system. On US, it is typically anechoic with posterior enhancement, separate from the stomach, duodenum, and gallbladder, and

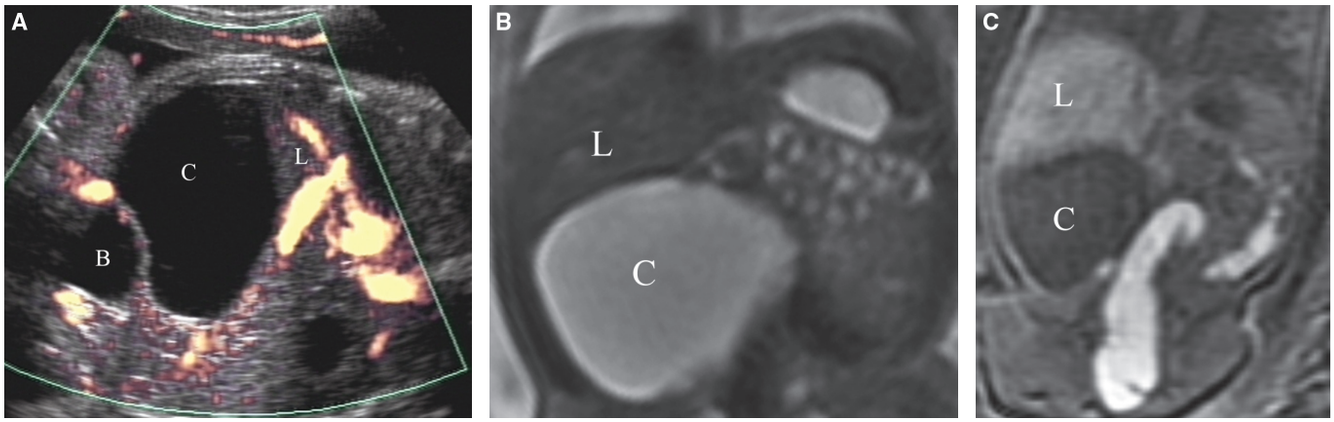

FIGURE 18.3-3: Hepatic cyst at 27 weeks' gestation. **A :** Coronal color Doppler US demonstrates a large anechoic cyst *(C)* along the inferior margin of the right lobe of the liver *(L)*, separate from the bladder *(B)*. There is no internal Doppler flow. **B:** Coronal MR image demonstrates hyperintense T2 fluid signal in the cyst *(C)* at the inferior margin of the right hepatic lobe *(L)*. **C:** Coronal T1w MR demonstrates hypointense fluid signal of the cyst *(C)* at the inferior margin of the right hepatic lobe *(L)*.

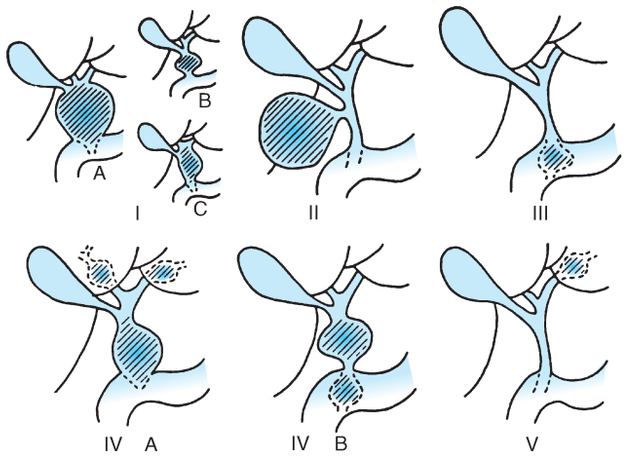

FIGURE 18.3-4: Classification of choledochal cysts as proposed by Alonso-Lej et al.[21] and modified by Todani et al.[22] Type IA, cystic extrahepatic biliary dilation; type IB, segmental extrahepatic biliary dilation; type IC, diffuse extrahepatic biliary dilation; type II, saccular diverticulum of extrahepatic bile duct; type III, choledochocele; IVA, multiple cysts of the intrahepatic and extrahepatic biliary tree; type IVB, multiple cysts of the extrahepatic biliary tree only; type V, intrahepatic cysts only (Caroli disease). (From Novak DA, Suchy FJ, Balistreri WF. Disorders of the liver and biliary system. In: McMillan JA, Feigin RD, DeAngelis C, et al, eds. *Oski's Solution*. Philadelphia, PA: Lippincott Williams & Wilkins; 2006:2035.)

without peristalsis (Fig. 18.3-5). Choledochal cysts with internal echoes have been reported.[26] Doppler US demonstrates lack of internal flow and intimate relationship with the portal vein and hepatic artery.[27]

MRI: Choledochal cysts follow fluid signal on MRI with hypointense T1w signal and hyperintense T2w signal. In some cases, MRI may be more useful than US in demonstrating the relationship to the biliary tree (Fig. 18.3-5).

Differential Diagnosis: If a cyst is in the porta hepatis and demonstrates intimate association with the portal vein and hepatic artery, the differential diagnosis includes choledochal cyst, cystic biliary atresia, duodenal duplication cyst, and pancreatic head cyst. Biliary atresia with cystic dilation can look identical to a choledochal cyst on prenatal imaging. Amniotic fluid analysis for digestive enzymes may help differentiate these various cysts in utero; biliary atresia demonstrates decreased γ-glutamyl transferase.[19] Figure 18.3-6 schematically differentiates the types of prenatally diagnosed biliary anomalies. Some authors suggest that (1) choledochal cysts increase in size over time, whereas cystic biliary atresia does not, (2) cysts of biliary atresia tend to be <2.5 cm, whereas choledochal cysts tend to be >4 cm or have intrahepatic biliary dilation, or (3) anechoic cysts correspond to atresia whereas large, echoic, or enlarging cysts correspond with choledochal cysts; however, the number of cases is small and exceptions are reported, so these criteria should not be used for definitive prenatal diagnosis.[26,28]

Other abdominal or pelvic cysts, including hepatic cysts, mesenchymal hamartomas, ovarian cysts, and adrenal cysts, should be considered in the differential as well.

Prognosis: Untreated choledochal cysts may be complicated by rupture with peritonitis, biliary obstruction, recurrent cholangitis, pancreatitis, cirrhosis, portal hypertension, liver failure, and malignant degeneration. With modern early surgical treatment, symptoms resolve and prognosis is good, with increasing morbidity and mortality with delayed surgery.[28,29]

There is evidence that choledochal cysts are associated with congenital cardiac anomalies, so echocardiographic screening may be warranted before surgery.[24]

Management: Postnatal US, MRI and/or hepatobiliary scan confirm hepatobiliary origin and further differentiate a choledochal cyst from the rare cystic biliary atresia. Intraoperative cholangiography and/or pathology may be necessary to definitively exclude biliary atresia.[30]

Complete surgical excision of the choledochal cyst with bilioenteric reconstruction, typically Roux-en-Y hepaticojejunostomy, is the preferred treatment. Early surgery in the first month of life may be warranted since even asymptomatic (not jaundiced) patients tend to develop liver fibrosis immediately after birth.[29]

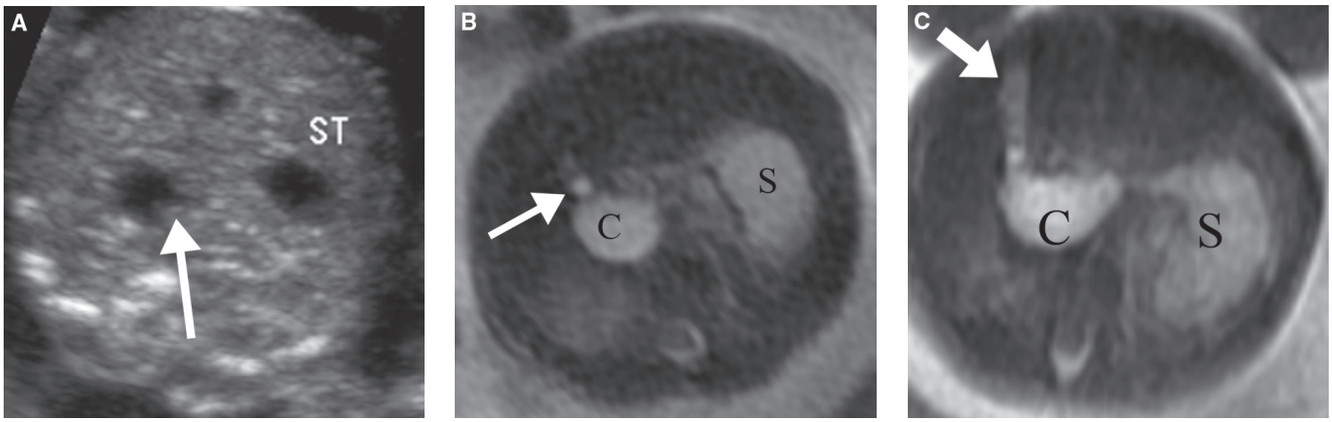

FIGURE 18.3-5: Choledochal cyst at 20 weeks' gestation. **A:** Transverse US through the fetal abdomen demonstrates a cyst in the liver hilum that tapers medially *(arrow)*. ST, stomach. **B:** Axial MRI demonstrates T2 hyperintense fluid signal of the cyst *(C)* at the hepatic hilum, separate from the stomach *(S)* and communicating with the bile duct *(arrow)* at the liver hilum. **C:** The next slice demonstrates the proximity of the cyst *(C)* to the gallbladder *(arrow)*, but separate from it. S, stomach.

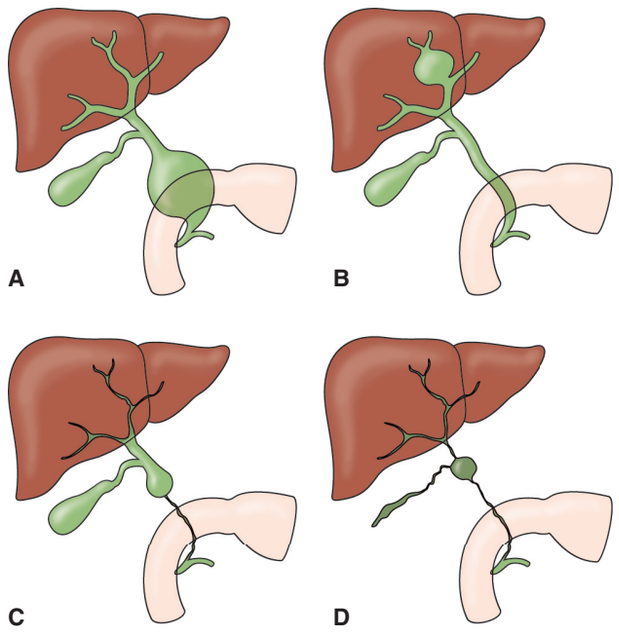

FIGURE 18.3-6 Schematic diagram of congenital biliary anomalies diagnosed antenatally. **A:** Type I-c choledochal cyst. **B:** Type V choledochal cyst. **C:** Type 1 biliary atresia. **D:** Type 3 biliary atresia with segmental cystic dilation. (Reprinted from Redkar RM, Howard ER. Antenatal diagnosis of congenital anomalies of the biliary tract. *J Pediatr Surg*. 1998;33:700–704, copyright © 1998, with permission from Elsevier.)

Recurrence Risk: Postsurgical follow-up of antenatally diagnosed, postnatally resected choledochal cysts has not demonstrated recurrence with follow-up ranging from 10 months to 7 years.[25–29]

Gallbladder Duplication

Incidence: One in 3,800 incidence is based on Boyden's 1926 work citing only 5 cases out of 19,000 autopsy and patient cases.[31] Bronshtein et al.[32] found 2 gallbladders described as septated, bilobed, or duplicated in 10,016 prenatal ultrasounds, for an estimated incidence of 1 in 5,008 screening ultrasounds.

Embryology and Pathology: The gallbladder arises from the caudal aspect of the hepatic diverticulum of the foregut in the 4th week and canalizes by the 12th week.[32]

In true duplication, there are two gallbladders, each with its own cystic duct.[33] It may result from continued growth, rather than the expected regression of diverticula that arise from the cystic or common bile duct in the 5th and 6th weeks.[31] Alternatively, two cystic primordia may arise separately from the common bile duct, resulting in an accessory gallbladder.[33]

A disturbance of cell division of the cystic primordium in the 5th or 6th week may result in splitting with two gallbladders in a septate, V-shaped, or Y-shaped duplication, but only one cystic duct entering the common duct.[33]

Diagnosis

Ultrasound: A duplicated gallbladder appears as parallel elongated cysts in the gallbladder fossa. Duplicated gallbladder has been reported on prenatal imaging between 20 and 32 weeks gestation.[34–36] On US, fluid in the gallbladder is typically anechoic (Fig. 18.3-7).

MRI: On MRI, a duplicated gallbladder follows fluid signal with hyperintense T2-weighted signal and hypointense T1-weighted signal (Fig. 18.3-7C). In the third trimester, gallbladder content signal is more variable.

Differential Diagnosis: The primary differential consideration is a gallbladder fold, but this is oriented along the short axis of the gallbladder. Other considerations include choledochal cyst, hepatic cyst, enteric duplication cyst, or lymphatic cyst. Its parallel configuration in the gallbladder fossa is characteristic.[35]

Prognosis: Duplicated gallbladder is an incidental finding with a good prognosis. The greatest importance lies in knowledge of the anatomy prior to cholecystectomy for symptomatic disease

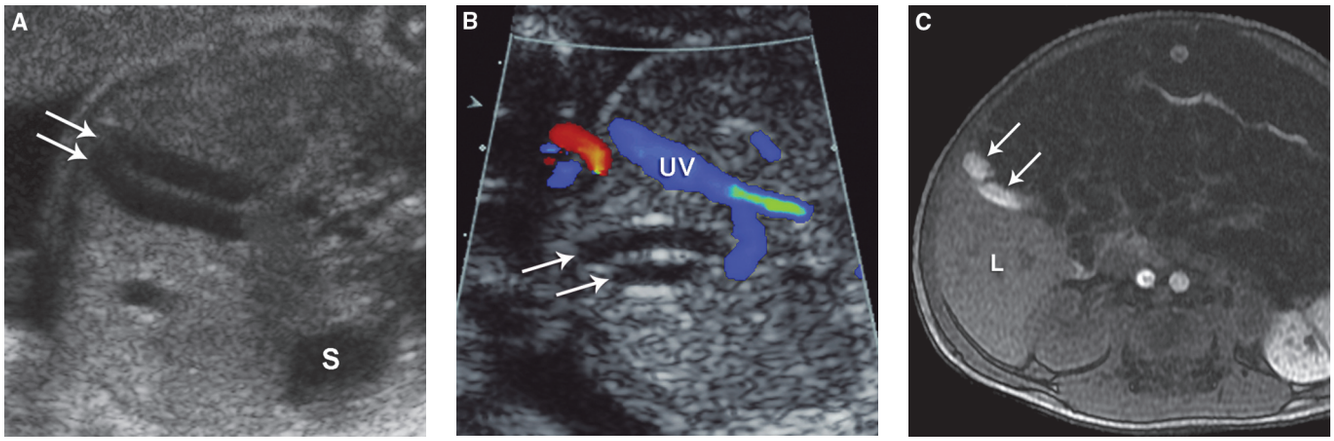

FIGURE 18.3-7: Gallbladder duplication at 30 weeks' gestation. **A:** Transverse US demonstrates two parallel elongated anechoic structures *(arrows)* in the gallbladder fossa. S, stomach. **B:** Color Doppler US demonstrates that the cystic structures *(arrows)* are to the right of the umbilical vein *(UV)* and lack internal flow. **C:** Postnatal MRI demonstrates hyperintense signal of bile in the parallel structures *(arrows)* in the gallbladder fossa of the liver *(L)*. (Reprinted from Gerscovich EO, Towner D, Sanchez T, et al. Fetal gallbladder duplication. *J Ultrasound Med*. 2011;30:1310–1312, with permission from American Institute of Ultrasound in Medicine.)

later in life, and for differentiating it from prenatal cysts that may require postnatal surgery.

Management: If the diagnosis is uncertain prenatally, postnatal US or MR may be useful for diagnosis. No intervention is required.

Recurrence Risk: None.

Enteric Duplication Cyst

Incidence: Prenatal detection is reported to be 1 in 18,333 live births, compared to 1 in 4,500 detected by autopsy, suggesting that prenatal US detects approximately 25% of duplication cysts.[37] There is a 2:1 male predominance.[38]

Embryology and Pathology: Between the 6th and 8th weeks of gestation, the solid GI tract canalizes via coalescence of multiple vacuoles. Duplications of the esophagus and intestines may be related to an error of canalization according to the "aberrant recanalization theory." The split notochord theory states that an adhesion between the endoderm and the ectoderm during the fifth gestational week causes the notochord to split, resulting in vertebral abnormality and an intraspinal or extraspinal neurenteric cyst, mostly applying to foregut duplications. Other theories include incomplete twinning, persistent embryologic diverticula, and intrauterine vascular accident theories.[39]

Duplication cysts are spherical (74% to 82%) or tubular (18% to 26%), typically unilocular structures. They are often single but may be multiple in 5% to 7%[38,39] (Fig. 18.3-8). Duplication cysts occur anywhere along the alimentary tract from the mouth/tongue to the rectum, but most commonly involve the ileum. The spherical type usually does not communicate with the lumen, whereas the tubular type frequently communicates.[39] Intestinal duplications usually occur along the mesenteric border, and gastric duplications typically occur along the greater curvature. Rarely, a duplication cyst may have its own separate mesentery. By definition, they should be associated with the GI tract, but there are reports of cysts separate from the GI tract with the same histology as duplication cysts, often called isolated duplication cysts.

Diagnosis

Ultrasound: On US, duplication cysts are typically anechoic. Intracystic hemorrhage results in internal echoes. Rarely, duplication cysts communicate with the bowel lumen, and enteric content/meconium is present in the cyst, causing internal echoes.[40] Enteric duplication cysts may demonstrate peristalsis and gut signature with echogenic mucosal and echolucent muscular layers. However, gut signature is easier to see on postnatal imaging and may not be detectable on prenatal US (Fig. 18.3-8).

Polyhydramnios may be present if the cyst results in GI obstruction or if there is concurrent bowel atresia, but this is uncommon. There is an association of bowel atresia with

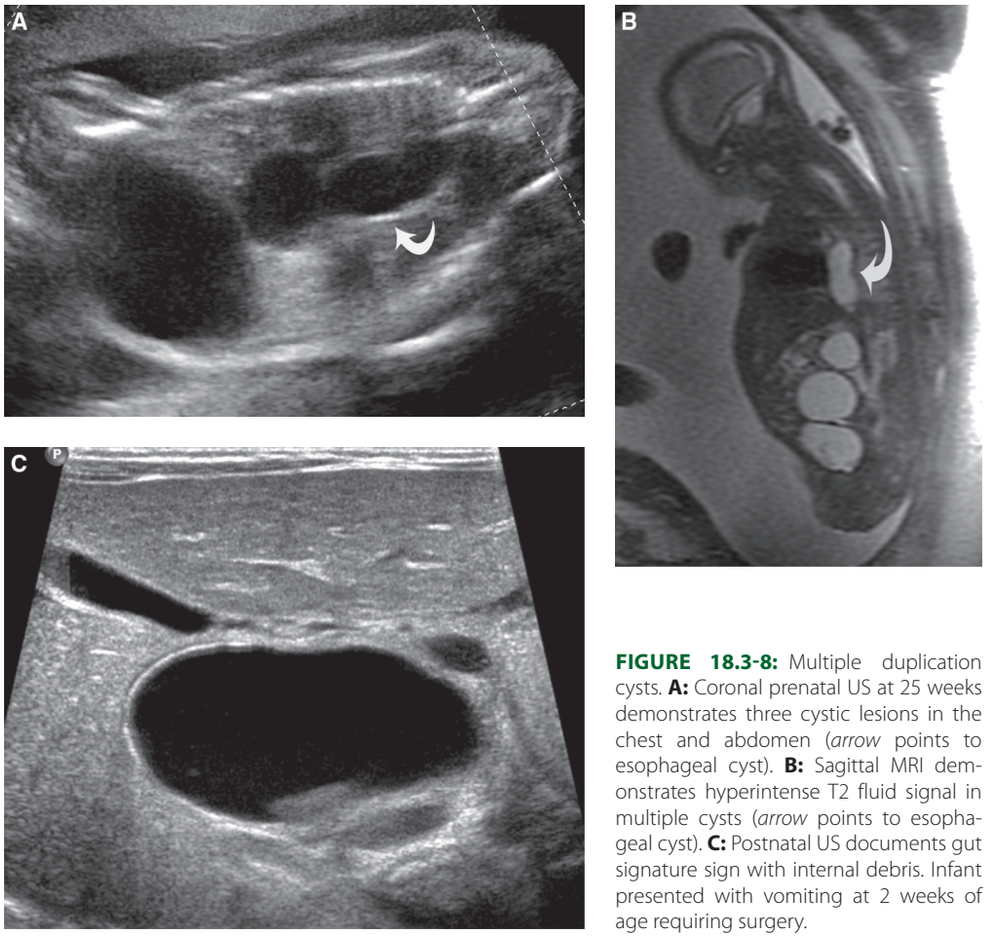

FIGURE 18.3-8: Multiple duplication cysts. **A:** Coronal prenatal US at 25 weeks demonstrates three cystic lesions in the chest and abdomen (*arrow* points to esophageal cyst). **B:** Sagittal MRI demonstrates hyperintense T2 fluid signal in multiple cysts (*arrow* points to esophageal cyst). **C:** Postnatal US documents gut signature sign with internal debris. Infant presented with vomiting at 2 weeks of age requiring surgery.

ileojejunal duplications in about 11%.[38] Half of foregut duplication cysts are associated with skeletal malformations and one-third of midgut and hindgut duplications are associated with GI or GU anomalies.[41]

MRI: On MRI, duplication cysts follow fluid signal, unless hemorrhage or meconium is present, resulting in hyperintense T1-weighted signal and hypointense T2-weighted signal (Fig. 18.3-8). Large field of view may help identify cyst origin and identify additional cysts. Associated GI or GU anomalies may be further assessed by MRI.

Differential Diagnosis: The differential diagnosis depends on location (Tables 18.3-1 and 18.3-2). Features that make a duplication cyst more likely include a thick wall with gut signature and peristalsis. Identification in the second trimester favors duplication cyst over ovarian cyst or bowel obstruction. Lymphatic cyst may be indistinguishable unless there is peristalsis.

Prognosis: Duplication cysts may be complicated by ulceration and hemorrhage if acid secreting gastric mucosa is present. Ectopic pancreatic tissue is most commonly found in gastric duplications and may cause pancreatitis. Duplications may result in mass effect on adjacent structures and intestinal obstruction. They may undergo torsion, become a lead point for intussusception or intestinal volvulus, or become infected or inflamed and perforate. Foley et al. found that of 12 prenatally detected duplications, 4 required surgery in the first 2 months for symptoms of bowel obstruction. Of the asymptomatic patients, 5/8 duplications demonstrated ulcerated acid secreting mucosa.[37] Prognosis is good with surgical resection.

Management: Since most enteric duplications become symptomatic in the first 2 years of life, they are typically completely excised, with or without segmental bowel resection.

Recurrence Risk: They do not recur after resection.

Lymphatic Cyst

Lymphatic cysts (including mesenteric, omental, and retroperitoneal cysts) and lymphangiomas are often used as interchangeable terms, although some authors separate them based on histology and clinical course. Lymphangiomas tend to be more aggressive and infiltrative. For this discussion of prenatal imaging, both will be grouped together, with an emphasis on imaging follow-up to assess behavior.

Incidence: Lymphangioma of any location is common, occurring in 1 in 6,000 pregnancies, but abdominal lymphangiomas account for less than 5% of these.[42] Localized lymphatic cysts are often asymptomatic, and the incidence is not known.

Embryology and Pathology: Abdominal lymphatic cysts are hypothesized to arise from abnormal development of the lymphatic system in the 6th week of gestation with lack of lymphatic drainage resulting in cystic collections.[43]

Obstructed lymphatic drainage results in a benign cystic mass of lymphatic channels that most commonly occur in the small bowel mesentery, but may occur in the large bowel mesentery, omentum, or retroperitoneum.[44] Some authors describe them as obstructed lymphatics and others as hamartomatous lymphatic tumors.

Diagnosis

Ultrasound: Review of case reports indicates that lymphatic cysts are commonly discovered on prenatal imaging in the second and early third trimesters (19 to 31 weeks), they increase in size over time, are more often left sided, and they may change from unilocular to multilocular over time. Small localized cysts are most often centrally located in the abdomen but may be anywhere in the abdomen or pelvis and may change position because of attachment to the mobile mesentery or omentum. Lymphangiomas may be isolated to the abdomen, or may infiltrate surrounding structures and may extend to the abdominal wall, chest, or legs.[43,45–47]

On US, lymphatic cysts are typically thin-walled anechoic unilocular or multilocular structures. Hemorrhage or chylous fluid may cause internal echoes. There may be associated skin edema, polyhydramnios, and hydrops.

MRI: MRI is useful for large field of view assessment. Lymphangiomas follow fluid signal with hypointense T1-weighted signal and hyperintense T2-weighted signal. If there is hemorrhage, T1 signal is hyperintense, T2 signal is more hypointense, and there may be marked hypointense signal on gradient echo imaging with hemosiderin deposition (Fig. 18.3-9).

Differential Diagnosis: Enteric duplication cysts and ovarian cysts may look identical to unilocular lymphatic cysts. Other cyst differential considerations depend on location (Tables 18.3-1 and 18.3-2). Pedunculated mesenchymal hamartoma and cystic teratoma are also considerations.

Prognosis: The prognosis depends on the location, rate of growth, extent, and effect on adjacent structures. The prognosis is good for small mesenteric cysts localized to the abdomen where there is often complete postnatal resection without recurrence. Poor prognosis is associated with early diagnosis, rapid growth, large size, hydrops/polyhydramnios, and extra-abdominal extension. Postnatally, localized cysts result in abdominal pain and/or distension from mass effect, torsion, hemorrhage, rupture, intussusception, or volvulus.

Management: Prenatally, follow-up USs are recommended to assess growth. Fetal karyotype is suggested because of the association with chromosomal abnormalities with lymphangiomas of the neck, but thus far the few reported cases of abdominal lymphangiomas have not demonstrated abnormal karyotype or other congenital abnormalities.

Cases have either been postnatally resected,[48–50] monitored clinically,[46] or, rarely, pregnancy was terminated because of the extent and fast growth of the lesion.[42,43,45,47]

Postnatally diagnosed symptomatic cysts have been treated surgically, preferably with complete enucleation, and bowel resection may be necessary. Marsupialization is the least preferable choice since incomplete resection may result in recurrence.[44]

Recurrence Risk: In children less than 10 years old, there is a 2% recurrence rate after resection. Recurrence is more likely for retroperitoneal cysts than mesenteric cysts.[44]

Splenic Cyst

Incidence: Dankovcik et al.[51] found 3 fetal splenic cysts in 7,897 screening USs for an incidence of 1 in 2,632.

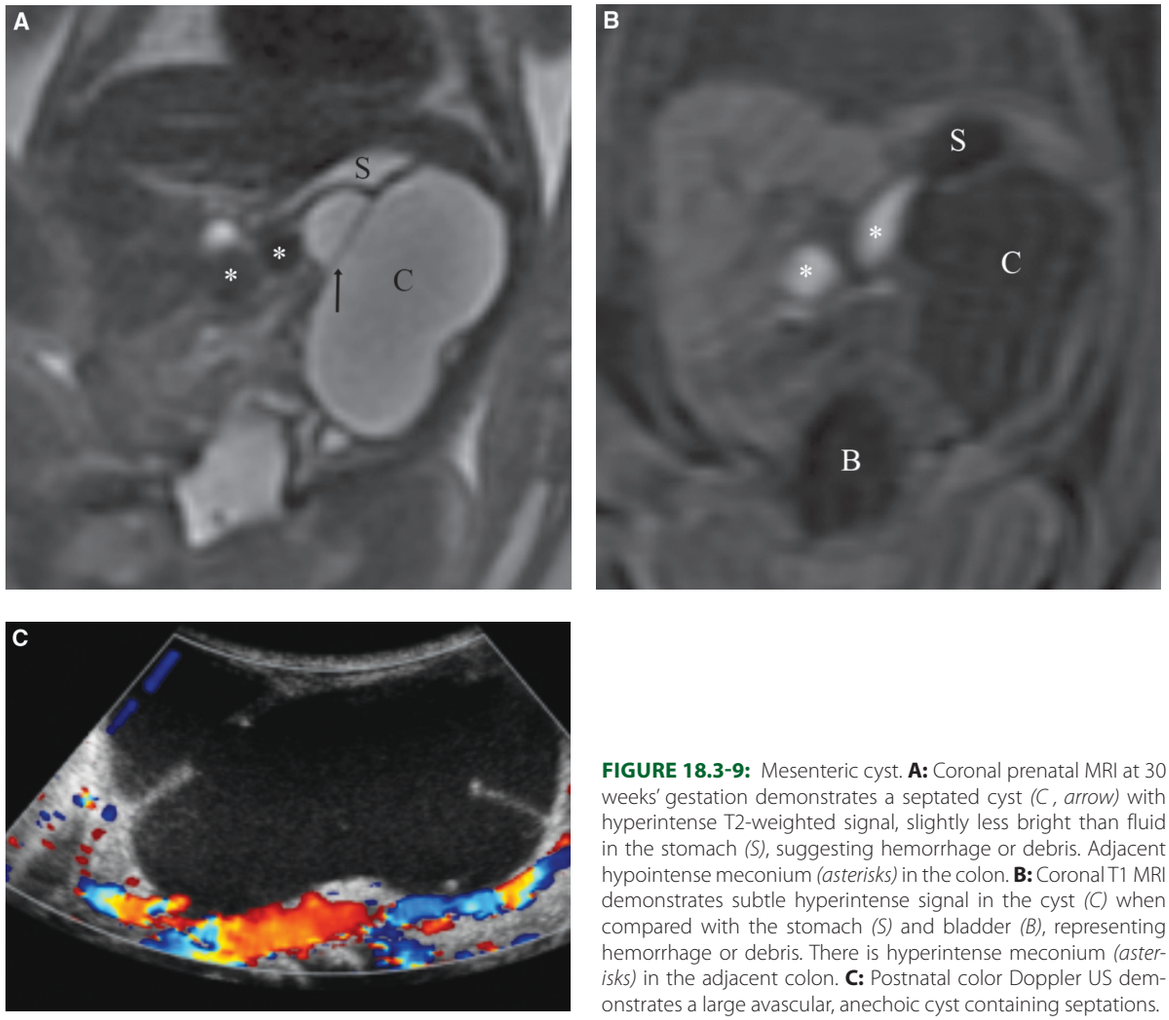

FIGURE 18.3-9: Mesenteric cyst. **A:** Coronal prenatal MRI at 30 weeks' gestation demonstrates a septated cyst *(C , arrow)* with hyperintense T2-weighted signal, slightly less bright than fluid in the stomach *(S)*, suggesting hemorrhage or debris. Adjacent hypointense meconium *(asterisks)* in the colon. **B:** Coronal T1 MRI demonstrates subtle hyperintense signal in the cyst *(C)* when compared with the stomach *(S)* and bladder *(B)*, representing hemorrhage or debris. There is hyperintense meconium *(asterisks)* in the adjacent colon. **C:** Postnatal color Doppler US demonstrates a large avascular, anechoic cyst containing septations.

Embryology and Pathology: The spleen begins development in the 6th to 7th week.

Prenatally diagnosed splenic cysts most likely represent lymphangiomas, epidermoids, or pseudocysts.[52] The pathogenesis is not proven, and theories include (1) pluripotent cell invasion of the spleen with subsequent metaplasia, (2) inclusion of coelomic mesothelial cells with squamous metaplasia, (3) invagination of peritoneal endothelial cells, and (4) lymphatic dilation.[51]

Diagnosis

Ultrasound: A splenic cyst is unilocular with a thin or imperceptible smooth wall most often diagnosed in the third trimester. Published cases of splenic cysts in the English literature measure 6 to 31 mm.[51,53] On US, it is an anechoic left upper quadrant mass.

MRI: On MRI, it follows fluid signal with hyperintense T2-weighted signal and hypointense T1-weighted signal.

Differential Diagnosis: Differential considerations include pancreatic cyst, hepatic cyst, lymphatic cyst, duplication cyst, renal cyst, urinoma, urinary collecting system dilation, and adrenal cyst.

Prognosis: Prenatally detected splenic cysts tend to be small and most often decrease in size or resolve in utero or in the first 2 years of life without symptoms.[51,52] Reports of splenic cyst complications of torsion, hemorrhage, rupture, and infection described in children and adults have not been described in the fetal/perinatal period.

Management: US monitoring in utero and postnatally is recommended to document cyst regression or stability and exclude growth, subsequent complications, or alternative diagnoses.

Recurrence Risk: Case reports of antenatally diagnosed splenic cysts indicate stability or regression without recurrence.

Pancreatic Cyst

Incidence: Fetal pancreatic cysts are rare, with only seven reported cases of isolated congenital cysts[54–56] and rare cases associated with other congenital anomalies.[56–58]

Embryology and Pathogenesis: True pancreatic cysts are epithelium-lined cysts hypothesized to result from anomalous development of the pancreatic ducts.

It is suggested that they represent anomalous development of the pancreatic ductal system, where sequestered secretory cells give rise to cysts.[59] There is a predilection for female gender and body/tail location for isolated cysts. Prenatal pancreatic cysts have been prenatally associated with Beckwith–Weidemann syndrome, asphyxiating thoracic dysplasia, renal-hepatic-pancreatic dysplasia, and narrow thorax with short limb dwarfism.[56,58] Small cysts were reported with short rib polydactyly syndrome Type 1.[57] Postnatally, there is an association with polycystic renal disease, anorectal malformation, and Von Hippel Lindau disease.[59]

Diagnosis

Ultrasound: Reported cases without congenital anomalies were diagnosed between 21 and 38 weeks' gestation, and cyst size at diagnosis was 21 to 58 mm. They may stay the same or increase in size.[54–56] Those with other congenital anomalies were diagnosed between 15 and 32 weeks with varied size, number, and appearance of cysts.[56–58]

Pancreatic cysts may be unilocular, multilocular, or transition from a unilocular to a septated cyst. They may be single or multiple. They are anechoic on US.

MRI: Pancreatic cysts follow fluid signal on MRI with hyperintense T2 signal and hypointense T1 signal.

Differential Diagnosis: Consider renal, liver, biliary, adrenal, or alimentary duplication cyst.

Prognosis: The prognosis is good for fetal pancreatic cysts without other congenital abnormalities where postnatal resection is successful (however, no long-term follow-up is documented in the literature). Potential serious postnatal complications of pancreatic cysts include infection, cholangitis, pancreatitis, cyst rupture, and peritonitis if not resected.

Prognosis is worse with concurrent congenital anomalies.

Management: Pancreatic cysts located in the body and tail are treated with surgical excision. Cysts in the pancreatic head are treated with internal drainage procedures.[56]

Recurrence Risk: There is one case report of recurrence in an incompletely resected antenatally diagnosed pancreatic cyst.[60]

Hepatosplenomegaly

Fetal hepatosplenomegaly (HSM) may be secondary to immune or nonimmune hydrops, congenital infection, cardiac failure, hemolytic anemia, metabolic disease, overgrowth disorders, myeloproliferative disorders, or tumors (Table 18.3-3).[61–67] Detection of hepatic and/or splenic enlargement should prompt careful evaluation of the entire anatomy for other anomalies and clinical screening for congenital infection.

A myeloproliferative disorder may result in hepatomegaly or hepatosplenomegaly, with or without hydrops. It may represent either transient abnormal myelopoieses or acute leukemia; definitive diagnosis requires cordocentesis and analysis of blast forms. It has been mostly seen in infants with trisomy 21, but has also been described in infants with normal karyotype. Patients require intensive care after birth, so prenatal assessment of severity may be helpful. Ogawa et al.[68] suggest that the degree of HSM may correlate with severity of postnatal disease, but there are no large studies to prove it.

Table 18.3-3 Causes of Fetal Hepatosplenomegaly (HSM)

- Immune hydrops
- Nonimmune hydrops
- Cardiac failure
- Congenital infection
  - Toxoplasmosis
  - CMV
  - Syphilis
  - Rubella
  - Tuberculosis
- Metabolic
  - Hyperthyroidism
  - Storage disease (Hunter, Gaucher, Niemann Pick, Wolman, Glycogen storage disease type IV have been described with prenatal HSM)
  - Congenital disorders of glycosylation
  - Mitochondrial hepatopathy
  - Smith–Lemli–Opitz syndrome (defect in cholesterol synthesis)
  - Enzyme deficiencies resulting in hemolytic anemia
    - Congenital erythropoietic porphyria
    - Pyruvate kinase deficiency
    - Glucose–phosphate isomerase deficiency
    - Transaldolase deficiency
- Overgrowth disorders (Beckwith–Weidemann syndrome)
- Zellweger syndrome
- Myeloproliferative disorder/leukemia (usually associated with trisomy 21)
- Primary tumor
- Metastases

Studies have demonstrated that splenic circumference is an excellent predictor of severe fetal anemia secondary to Rh alloimmunization in nonhydropic fetuses not treated with transfusion, and is more useful late in pregnancy (after 30 weeks).[69] It was less sensitive and specific, but still useful, in the second-trimester evaluation of hemoglobin Bart (homozygous alpha thalassemia1).[63]

SOLID LIVER MASSES

Solid hepatic masses of the fetus and neonate are rare and account for approximately 5% of total neoplasms in this age group. Primary hepatic tumors include infantile hemangioma/hemangioendothelioma, mesenchymal hamartoma, and hepatoblastoma. Liver metastases may be secondary to neuroblastoma, leukemia, or renal tumors. Less common liver metastases may be secondary to yolk sac tumor, rhabdomyosarcoma, or rhabdoid tumor.[70] Rare cases of fetal focal nodular hyperplasia and hepatic adenomas have been reported.[71,72]

Hemangioendothelioma and Hepatic Vascular Lesions

Incidence: Hemangioendothelioma (HAE) accounts for 60% of perinatal liver tumors.[70]

Embryology and Pathology: North's placental theory suggests that placental mesodermal progenitor cells or placental

angioblasts are responsible, as evidenced by the striking similarity of glucose transporter protein (GLUT)-1 positive infantile hemangioma tissue to placental tissue and supported by increased incidence after chorionic villous sampling. Alternatively, a vascular precursor cell may undergo somatic mutation or may be influenced by local inductive influences to proliferate.[73]

Hemangioendothelioma is a term applied to a benign vascular lesion of the liver. In the literature, several terms have been used, sometimes considered synonymous and sometimes discussed as separate entities (Table 18.3-4). These vascular liver lesions have more recently been grouped into solitary, multiple, and diffuse lesions, and into GLUT 1 reactive and nonreactive, which better describes their behavior.[74,75] *Focal lesions* are histologically GLUT-1 negative and behave similar to the rapidly involuting congenital hemangiomas (RICH) of the skin, which are fully grown at birth and regress over 12 to 14 months. These are more likely to be seen on prenatal imaging. *Multifocal lesions* are GLUT-1 positive and behave similar to infantile hemangiomas (IH) of the skin. They tend to present clinically in early infancy with a proliferative phase during infancy followed by an involutional phase over several years, and demonstrate female predominance. *Diffuse lesions* are multiple GLUT-1 positive lesions that are too numerous to count and may cause massive hepatomegaly with compartment syndrome, respiratory compromise, and impaired venous return.[74,75]

Multifocal and diffuse forms express type 3 iodothyronine-deiodinase, which degrades thyroid hormone and causes hypothyroidism. It is theorized that multifocal lesions may coalesce and become diffuse.[75] High-flow shunting may result in heart failure, more common in the diffuse form where more than half of patients are affected (Fig. 18.3-10F). Intralesional thrombosis may result in transient anemia and thrombocytopenia.[75]

Diagnosis

Ultrasound: On ultrasound, the *solitary* GLUT-1(−) lesions are typically large, well-circumscribed lesions with heterogeneous echogenicity from central necrosis and hemorrhage, and may have echogenic shadowing dystrophic calcification. They sometimes demonstrate high-flow arteriovenous or portovenous shunts with large tubular anechoic vessels (Fig. 18.3-10). *Multifocal* GLUT-1(+) lesions are homogeneous small round masses that are usually hypoechoic. There may be large vessels in or adjacent to the masses with shunting. *Diffuse* tumors may be difficult to discern because of near complete replacement of the liver parenchyma with hepatomegaly and heterogeneous echotexture.[74]

Doppler may demonstrate hypervascularity with high-velocity arterial waveforms and arterialized venous waveforms. Decreased aortic caliber below the celiac axis with hepatic artery, hepatic vein, and cardiac dilation suggest shunting.

Table 18.3-4 Comparison of Infantile Hepatic Hemangioma Subtypes[74,75]

Subtype	Focal	Multifocal	Diffuse
Synonyms	Hemangioendothelioma, cavernous hemangioma, arteriovenous malformation, solitary hemangioma, RICH of the liver, hepatic vascular malformation with associated capillary proliferation (HVMCP)	Hemangioendothelioma, capillary hemangioma, cellular hemangioma, infantile hepatic hemangioma	Hemangioendothelioma, capillary hemangioma, cellular hemangioma, infantile hepatic hemangioma, hemangiomatosis
GLUT 1 reactivity	GLUT 1 negative (–)	GLUT 1 positive (+)	GLUT 1 positive (+)
Time of clinical presentation	Prenatal imaging or first few weeks of life	First weeks–months of life	First weeks–months of life
Natural history	Fully grown at birth, involute in infancy	Proliferate in infancy, then involute	Proliferate in infancy, then involute
% Female	48.5	66.2	70
Prenatal appearance	Large, spherical, often heterogeneous with central necrosis/fibrosis, calcification	Multiple small spherical homogeneous masses, hepatomegaly	Innumerable masses or heterogeneous enlarged liver
Postnatal enhancement	Peripheral rim enhancement with no central enhancement of necrotic areas on delay	Centripetal enhancement with delayed homogeneous enhancement	Centripetal enhancement with delayed homogeneous enhancement
Cutaneous	15.3%	77.4%	53.3%
Complications	High-flow shunts → CHF, mild anemia and thrombocytopenia	Occasional high-flow shunts → CHF, anemia, thrombocytopenia	Mass effect on IVC, renal and hepatic veins and chest with respiratory compromise, multiorgan failure
Hypothyroidism	0%	21.4%	100%, profound hypothyroidism
Response to medical treatment	Unclear	Good	Poor

MRI: All types are predominantly T2 hyperintense and T1 hypointense, but large solitary tumors may be heterogeneous secondary to necrosis, hemorrhage, thrombus, fibrosis, and calcification (Fig. 18.3-10). Dilated vessels are tubular flow voids on T2-weighted MRI. Shunting is suspected when there are large flow voids with aortic caliber change. Small nodules of the multifocal or diffuse subtypes may be more apparent on MRI rather than US because of the soft tissue contrast and tend to be homogeneous in signal.[74]

There is an association with cutaneous hemangiomas (Table 18.3-4). When HAE is diffuse, there may be extrahepatic involvement of many organs.[70] Hemangioendothelioma is rarely associated with other congenital anomalies.

Differential Diagnosis: Other hepatic tumors, including mesenchymal hamartoma, hepatoblastoma, and metastases, are the primary differential considerations. Urine catecholamines help differentiate neuroblastoma metastases in the case of multifocal disease. Alpha-fetoprotein (AFP) does not differentiate it from hepatoblastoma since AFP may be elevated in HAE and not elevated in half of congenital hepatoblastomas.

Prognosis: Asymptomatic patients have an excellent prognosis. Prognosis is poorer for patients with the diffuse form, hydrops, cardiac failure, jaundice, or thrombocytopenia.[75–77] Disseminated intravascular coagulation (DIC) rarely occurs.

Management: Prenatal fetal blood sampling to diagnose DIC with subsequent in utero platelet transfusion,[78] prenatal treatment with maternal oral steroids,[79] and intrauterine steroids via the umbilical vein and amniotic fluid[80] have been described. Rarely, tumor rupture during delivery may result in massive hemorrhage, so cesarean section should be considered.[70] Postnatally, asymptomatic patients are monitored with serial imaging. Symptomatic patients are treated medically. Corticosteroids are first-line treatment to hasten involution. Propranolol, vincristine, or interferon may be used in addition to steroids or separately. Medical therapy is often effective for GLUT-1(+) tumors, but effectiveness is unknown for GLUT-1(−) tumors.[75] Approximately one-third of patients with shunts fail pharmacologic therapy and require embolization. If medical management is ineffective, surgical options include hepatic artery ligation or embolization of shunts, rarely resection, and as

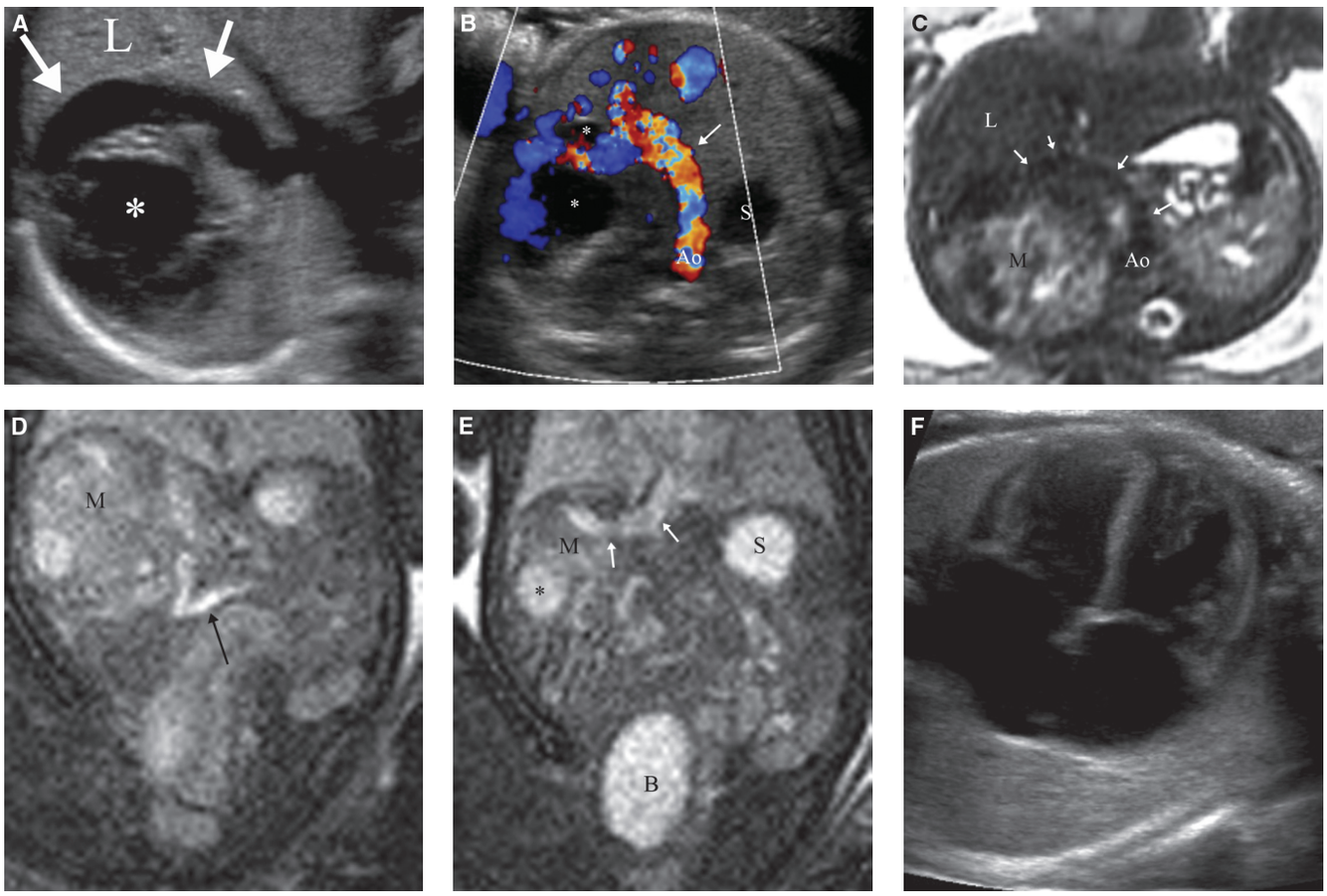

FIGURE 18.3-10: Hepatic hemangioma. **A:** Axial US image of the right lobe of the liver *(L)* demonstrates a heterogeneous, predominantly hypoechoic mass with a large round anechoic area centrally *(asterisk)* and a tubular anechoic structure anteriorly *(arrows)*. **B:** Doppler confirms that the tubular structure is a large celiac and hepatic artery *(arrow)* arising from the aorta *(Ao)*. There is no Doppler flow in anechoic cystic areas *(asterisks)* of degeneration. S, stomach. **C:** Axial MR T2w image demonstrates the heterogeneous, predominantly hyperintense mass *(M)*. The tubular hypointense flow void of the large artery *(arrows)* is displaced by the mass. Coronal SSFP images demonstrates **(D)** the large feeding artery *(arrow)* supplying the heterogeneous hepatic mass *(M)* and **(E)** large hepatic vein drains into the IVC *(arrows)*. S, stomach; B, bladder; asterisk, cystic degeneration. **F:** Four-chamber heart view demonstrates cardiomegaly secondary to high-flow shunt. (Courtesy of Chris Cassady, MD.)

a last resort, liver transplant. Symptomatic patients with diffuse disease should be quickly referred for transplant consideration while attempting medical therapy since their course is often short with high mortality. Thyroid function screening is recommended, since consumptive hypothyroidism associated with the multifocal and diffuse forms requires high-dose supplementation.[74]

Recurrence Risk: There are reports of hepatic IH regressing, and then recurring between 2½ and 5 years of age with malignant histology, and often metastatic disease, considered synonymous with angiosarcoma.[81]

Mesenchymal Hamartoma

Incidence: Mesenchymal hamartomas (MH) account for 23% of liver tumors diagnosed in the perinatal period.[70]

Embryology and Pathology: Several chromosomal translocations have been described, favoring the hypothesis that MH may actually represent a neoplasia.[82]

There are four hypotheses (1) developmental, arising from a ductal plate malformation, (2) the result of a local vascular insult/ischemia, (3) a response to a toxic insult, or (4) a neoplasia rather than a hamartoma.[82]

MH is associated with placental mesenchymal stem villous hyperplasia (vascular malformation), and is not typically associated with other congenital anomalies.[82]

Diagnosis

Ultrasound: MH is more common in the right lobe. Twenty percent are pedunculated and may not appear intrahepatic.[82] It has been diagnosed as early as 19 weeks gestation. Typically, it is a multilocular cystic structure with a variable soft tissue component (Fig. 18.3-11). There can be multiple thin mobile septations and hyperechoic intracystic nodules. Rare appearances include intracystic hemorrhage or debris, hypervascular angiomatous component or mixed MH-HAE, cysts so small that the mass appears solid on imaging, or echogenic calcifications.

Look for complications related to mass effect such as polyhydramnios, hydrops, and diaphragm elevation with pulmonary hypoplasia.

MRI: Large field of view is useful in assessing the origin of these masses by MRI. MH is T1-weighted hypointense, but variable signal intensity on T2-weighted imaging (Fig. 18.3-11).[82] Lung hypoplasia can be assessed.

Differential Diagnosis: The differential diagnosis for lesions in the liver includes hepatic cyst, hemangioendothelioma, and cystic hepatoblastoma. Pedunculated tumors may mimic intra-abdominal cysts, particularly lymphatic cysts or cystic teratoma because of the septations.[82]

Prognosis: The prognosis is poorer for those diagnosed in utero than in childhood, with 29% mortality, predominantly because of mass effect.[83] There are reports of partial spontaneous regression, particularly in cases with a prominent angiomatous component. There are also rare reports of undifferentiated embryonal cell carcinoma of the liver arising within MH or after incomplete resection.[82]

Management: Prenatal follow-up with US is important to monitor size. They often enlarge quickly and can cause significant mass effect. Prenatal cyst decompression may alleviate mass effect and allow more normal fetal development, but fluid typically reaccumulates quickly. If the cyst is large, cesarean section may be performed for potential dystocia. Postnatally, symptomatic cysts are usually treated with surgical resection. Imaging and clinical follow-up is recommended for 5 years.[82]

Recurrence Risk: Recurrence rate is not documented, but recurrence of MH is reported after incomplete resection, and there are also cases of recurrence from small unrecognized satellite lesions.[82]

Hepatoblastoma

Incidence: The incidence of hepatoblastoma in U.S. children under age 1 is 10.5/1,000,000, and has been increasing.[84] Less than

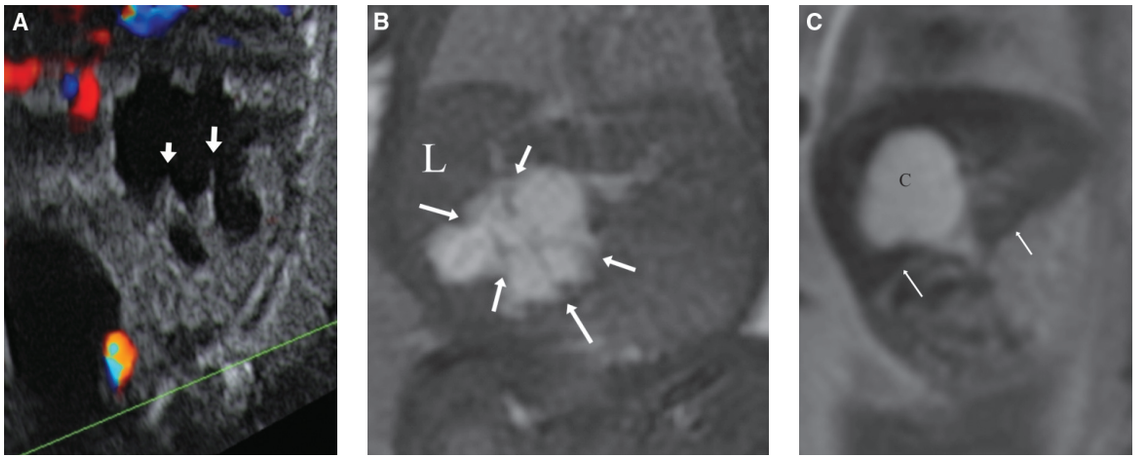

FIGURE 18.3-11: Mesenchymal hamartoma. **A:** Parasagittal US demonstrates an anechoic lobulated right upper quadrant abdominal mass with septations *(arrows)* and lack of internal Doppler signal. **B:** Coronal SSFP image demonstrates hyperintense signal in the lobulated mass in the liver *(L)* with several thin hypointense septations *(arrows)*. **C:** A sagittal T2w image best depicts its location within the liver (inferior margin of the liver is denoted by *arrows*). C, cyst.

10% of all hepatoblastomas occur in the perinatal period. It accounts for 17% of liver tumors diagnosed in the perinatal period.[70]

Embryology and Pathology: Multiple cytogenetic abnormalities have been described in association with childhood hepatoblastoma, including gain of chromosome 20, 2, or 8. There is also an association with trisomy 18.[84] An association with familial adenomatous polyposis has led to the discovery of adenomatous polyposis coli (APC)/β-catenin gene mutations found even in some sporadic hepatoblastoma cases. Changes in expression of *H19* and *IGF2* have been described, and are also commonly seen in Beckwith–Weidemann syndrome, with which hepatoblastoma is associated.[84,85]

It is hypothesized that oxygen free radicals may impede hepatocyte differentiation. There is also a high incidence of hepatoblastoma in very lowbirth weight infants, but whether the tumor is initiated or promoted by environmental factors of the NICU or owing to a common etiology is controversial and not proven.[84]

Hepatoblastoma is derived from undifferentiated embryonal tissue and may be classified as epithelial (subclassified as fetal, embryonal, or small cell undifferentiated) or mixed epithelial/mesenchymal subtypes. Congenital hepatoblastomas are more often the pure fetal histology. In childhood, metastases occur in the lung. In prenatal cases the lungs are spared, presumably because of fetal circulation, and metastases occur in the brain, bone, and placenta.[86]

Diagnosis

Ultrasound: Review of the antenatally diagnosed case reports indicates that fetal hepatoblastomas are typically diagnosed by US late in the third trimester as a large mass measuring 6 to 10 cm, although it has been seen as early as 30 weeks, and as small as 2.5 cm.[87] They are typically single, but may be multiple, and are more common in the right lobe.[70] On US, it is usually hyperechoic but may be heterogeneous with areas of degeneration demonstrating necrosis, hemorrhage, and calcification. It is a vascular, predominantly solid tumor. It may appear polylobular with a "spoke-wheel appearance."[87]

Mass effect may result in compression of vessels (umbilical vein, portal vein, and IVC), hydrops and compression of the lungs resulting in respiratory distress.

MRI: Hepatoblastomas are typically T2-weighted hyperintense, and T1-weighted hypointense, but there may be heterogeneity in large tumors if there is associated degeneration (Fig. 18.3-12). MRI is useful for lung volume calculation and metastasis detection.

Differential Diagnosis: The differential diagnosis includes hemangioendothelioma, metastases, and mesenchymal hamartoma. The serum AFP is not a good discriminator. It is only elevated in half of congenital cases, and may rarely be elevated in hemangioendothelioma and mesenchymal hamartoma.[70]

Prognosis: The prognosis is poor. Mass effect may compress vessels, resulting in hydrops and stillbirth. Diaphragmatic elevation may compress the lungs, resulting in respiratory distress. Tumor hemorrhage, spontaneous or related to vaginal delivery, may result in anemia or death. Rarely, metastases contribute to mortality.[89] The survival rate for treated congenital hepatoblastoma patients is 40%, for untreated patients is 0%, and the overall survival is 25%. There is a difference in survival based on histology with 50% survival for pure fetal histology and 30% for fetal + embryonal.[70] There have been improved mortality rates with treatment of childhood hepatoblastomas, but it is not known whether modern treatment has improved mortality for those diagnosed in the perinatal period.

Management: Prenatally, hepatic masses are followed with imaging to assess growth, mass effect on adjacent structures, and to monitor for hydrops. Cesarean section should be considered since these tumors are known to bleed and rupture during delivery and are typically large and may cause dystocia.[70] Postnatally, they are treated with surgical resection. Chemotherapy may be used preoperatively to shrink the tumor, and/or postnatally to control microscopic disease.

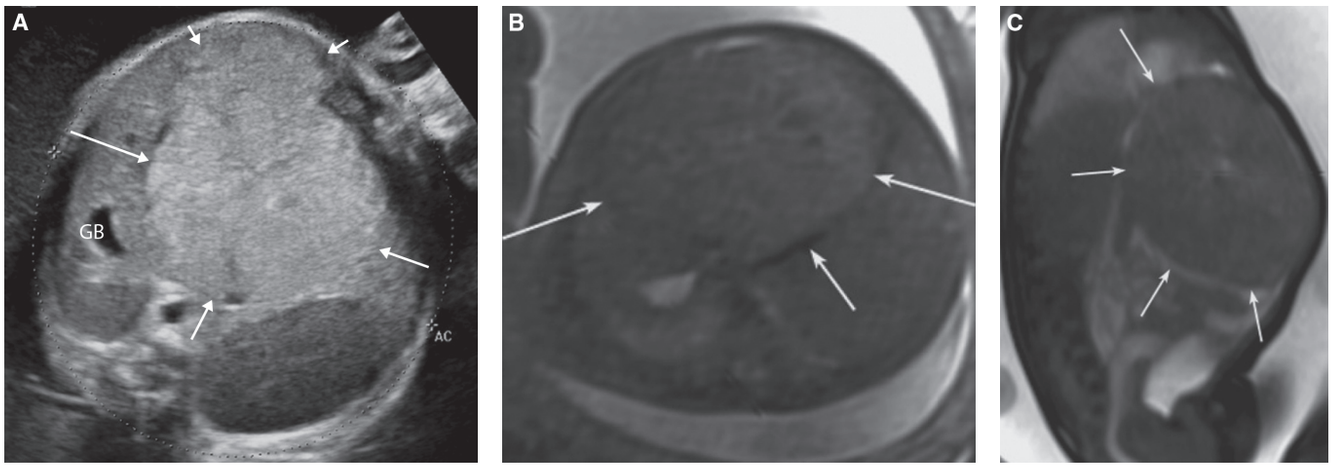

FIGURE 18.3-12: Hepatoblastoma. **A:** Transverse US demonstrates a large echogenic liver mass *(arrows)* displacing the gallbladder *(GB)*. There are subtle linear hypoechoic structures in a spoke-wheel orientation. **B:** Axial T2-weighted prenatal MR image demonstrates a large liver mass *(arrows)* with mild hyperintense T2-weighted signal relative to liver. **C:** Sagittal SSFP image demonstrates mild heterogeneous signal centrally within the mass *(arrows)*. (Reprinted from Al-Hussein H, Graham E, Tekes A, et al. Pre- and postnatal imaging of a congenital hepatoblastoma. *Fetal Diagn Ther*. 2011;30(2):157–159. Copyright © 2011, with permission from S. Karger AG, Basel, Switzerland.)[88]

Recurrence Risk: Hepatoblastoma diagnosed in childhood recurs, so presumably congenital hepatoblastoma may recur, but there are no data to cite a recurrence rate.

OTHER ABDOMINAL MASSES

Teratoma and fetus in fetu both demonstrate heterogeneous appearance on US and MR, often with soft tissue, bone, and fat elements, and with varying degrees of organization. Fetus in fetu is a rare diagnosis in which a monochorionic, monozygotic twin is incorporated into the host twin. Some authors believe that presence of four limbs appropriately arranged relative to a vertebral column differentiates it from a teratoma, and others believe that a vertebral column is not required for the diagnosis.[90,91] Both fetus in fetu and teratoma are completely surgically excised with their surrounding membranes to reduce the risk of recurrence.[91]

Gastric pseudomass is a rounded echogenic area in the lumen of the stomach, possibly as a result of swallowed cells or hemorrhage in the amniotic fluid, which aggregates because of poor peristalsis in the second trimester. They resolve on follow-up imaging.[92]

Meconium pseudocyst is a well-defined hypoechoic mass with a hyperechoic-calcified wall and indicates bowel perforation[92] (see Chapter 18.1).

Extralobar subdiaphragmatic pulmonary sequestration occurs more commonly on the left (4:1) as an echogenic subdiaphragmatic mass, and the diagnosis is supported by identifying a thoracic aortic feeding vessel with Doppler. On MRI, it is homogeneously T2 hyperintense, and a feeding artery may be seen as a hypointense tubular flow void on T2-weighted imaging. They are typically seen in the second trimester and more often on the left, as opposed to neuroblastomas which are more hypoechoic and more often seen in the third trimester on the right[92] (see Chapter 17).

The Gallbladder

The gallbladder is first visible by US at 13 to 14 weeks gestation as a fluid-filled, anechoic structure in the right upper quadrant, subhepatic or intrahepatic in location, and to the right of the umbilical vein. Size increases linearly with gestational age and plateaus at 32 to 35 weeks. It has a sinusoidal contractility pattern over a 3-hour interval that is independent of maternal meals. While the percent contractility increases through the third trimester, the minimum volume is relatively constant, so the gallbladder should be visible on imaging throughout the cycle.[93] The MRI signal of the gallbladder contents is variable and is age dependent. Before 27 weeks, gallbladder contents are T1 hypointense and T2 hyperintense. After 30 weeks, T1 and T2 signal are variable. This is likely due to sludge or accumulation of paramagnetic substances in the gallbladder mucous.[94]

Nonvisualization of the Fetal Gallbladder

This is typically defined as not seeing the gallbladder on two USs within 7 to 15 days. With modern equipment and transvaginal technique at 14 to 16 weeks, the gallbladder is absent in only 0.1% of pregnancies.[95] The most recent data for transabdominal detection are from 1996 when nonvisualization was 68.6% at 12 to 16 weeks and decreased with gestational age to 7% at 20 to 24 weeks.[96] Blazer found that absent gallbladder was isolated in 59% (incidence 1 in 6,000 pregnancies) with normal outcome. Associated structural malformations were present in 41%, and 36% of those also had abnormal karyotype. The gallbladder was detected later in pregnancy or postnatally in 4/5 structurally abnormal babies that were not terminated, and in 13/20 normal babies.[95] Nonvisualization of the gallbladder is associated with cystic fibrosis, biliary atresia, and aneuploidy, so amniocentesis with karyotype and evaluation of amniotic digestive enzymes may be useful.

Cholecystomegaly

This is defined as gallbladder area more than 2 standard deviations above the mean for gestational age. Despite small retrospective studies suggesting an association with aneuploidy or biliary abnormalities, a large prospective study found cholecystomegaly in 43 of 775 (5.5%) fetuses and no such association.[97]

Gallbladder Stones and Sludge

Echoes in the gallbladder with posterior acoustic shadowing suggest stones, and those without shadowing suggest sludge. There are few reported cases with an estimated incidence of 1% after 28 weeks' gestation.[98] They often resolve in utero or in the first year of life without symptoms. Management includes follow-up US and possibly postnatal ursodeoxycholic acid.[99]

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