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A variety of neoplasms can develop in each tetal organ. Most fetal neoplasms can be detected by careful prenatal ultrasonographic examination. Some neoplosms show specific ultrasonographic findings suggesting the differential diagnosis, but others do not. Knowledge of the presence of a neoplasm in the fetus may alter the prenatal management of a pregnancy and the mode of delivery, and facilitates immediate postnatal treatment. During the last five years, we experienced 32 cases of fetal neoplasms in a variety of organs. We describe their typical ultrasonographic findings with correlating postnatal CT, MRI, and pathologic findings.

lthough various neoplasms may occur in any organ system during fetal life, a fetal neoplasm is very rare and descriptions of the related US findings have been sporadic and inconclusive. During the last five years, we encountered 32 cases of fetal neoplasms in a variety of organs (Table 1), and in this article we describe their typical US findings, correlating these with the postnatal US, CT, MRI, and pathological findings.

Index terms: Ultrasound(US) Fetus Neoplasms

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Fetal Brain Tumors

Prenatal US can diagnose several kinds of congenital brain tumors occurring during fetal life. Such tumors can cause spontaneous intracranial hemorrhage during this period or dystocia during delivery. An awareness of their presence, as demonstrated by fetal US, may alter the mode of delivery and facilitates immediate postnatal treatment. The early diagnosis of a fetal brain tumor is therefore important.

Teratoma

Teratoma is the most common congenital neoplasm composed of tissues originating from all three germinal layers, and may occur in a variety of locations. Fetal brain teratoma usually appears as a large, solid and/or cystic tumor, often replacing normal brain tissue and sometimes eroding the skull (Fig. 1) (1). Associated congenital defects are frequently encountered, and if a fetal teratoma is suspected, a careful US survey should thus be performed (2). Brain teratomas usually cause intrauterine or early neonatal death, and require cesarean section for delivery of an enlarged fetal head (1).

Glioblastoma

Glioblastomas account for 2-9% of congenital brain tumors (3). At prenatal US, at which the mass effect and hydrocephalus are observed (Fig. 2), this tumor appears as a large homogeneous hyperechoic mass involving the supratentorial cerebral parenchy-

ma (4), and is thus distinct from a teratoma, which is multicystic. Both congenital glioblastoma and other congenital brain tumors may cause spontaneous intracranial hemorrhage in a fetus and an infant. In congenital tumors, the prevalence of hemorrhage has been reported as 18%, a rate much higher than that otherwise seen in children and adults (3). The rapid growth of congenital tumors, which explains the poor prognosis of glioblastomas, may be related to this higher incidence of bleeding. In a few reported cases, the immediate cause of death was heart failure, which may have been due to anemia resulting from the sequestration of a large amount of blood by the highly vascular tumor (5).

Tumors of the Fetal Face and Neck

Fetal tumors of the face and neck are a diverse group of

Table 1. Fetal Neoplasms Diagnosed by Prenatal Ultrasound

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Organ	Neoplasm	Number
Brain	Teratoma	3
	Glioblastoma	2
Face and neck	Lymphangioma	5
	Teratoma	1
Thorax	Lymphangioma	3
	Rhabdomyoma	1
Abdomen	Lymphangioma	3
	Omental cyst	1
Liver	Adenoma	1
	Hemangioma	1
Adrenal gland	Neuroblastoma	1
Kidney	Mesoblastic nephroma	1
Spine	Sacrococcygeal teratoma	7

lesions. Vascular lesions including lymphangioma and hemangioma are common, while teratomas and dermoid cysts, representing true congenital neoplasms, are relatively uncommon. The prenatal differential diagnosis of these lesions is often very difficult.

Lymphangioma

Lymphangiomas are common congenital lymphatic malformations that are frequently present at birth and are due to aberrations in normal lymphatic development. There are four histological types of lymphangioma, namely capillary lymphangioma (a small, ill-defined mass of thin-walled dilated lymphatics in a rich cellular connective tissue stroma), cavernous lymphangioma (a larger spongy compressible mass of dilated lymphatics with fibrous adventitial coat), cystic hygroma (a large macroscopic lymphatic space resulting from sequestration of the central lymphatics), and vasculolymphatic malformation (6). These types are considered to be a spectrum of manifestations of the same pathologic process. Most lymphangiomas occur in the neck, generally in the posterior triangle.

US demonstrates unicameral or multilocular cystic masses with thin or thick-walled septa (Fig. 3). Scattered low-level echoes, a solid component, or fluid-fluid levels, all due to bleeding and fibrin deposition may occur (7). The border of a lymphangiomas is indistinct and diffuse infiltration may occur there (8).

Cystic hygromas are often isolated malformations in which the remainder of the lymphatic system is normal. However, a fetal nuchal cystic hygroma has a poor prognosis when associated with chromosomal abnormalities and fetal hydrops.

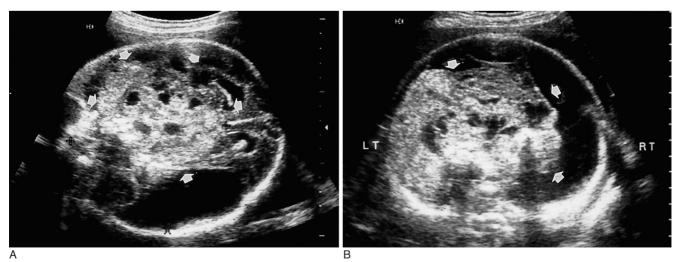


Fig. 1. Prenatal ultrasonographic findings of fetal brain teratoma in a 35-week fetus.

A, B. Axial and coronal ultrasonographic images of the fetal head show that a large mass of mixed echogenicity (arrows) has replaced normal brain structures.

Teratoma

About 5% of all teratomas occur in the face and neck region, most commonly in the anterior and lateral aspect of the latter. Cervical teratomas usually originate during early development of the tongue, when all three germ cell layers are in close proximity to each other. A fetal oropharyngeal teratoma is also known as an epignathus and is easily detected by prenatal US; findings of a pedunculated solid and cystic mass in the anterolateral aspect of the fetal face and neck suggest this diagnosis (Fig. 4) (9). Because of the difficulty in swallowing of amniotic fluid, one-third of these tumors are associated with polyhydramnios (10).

Neonatal survival depends on the size and extent of the involved tissues, respiratory compromise being the major cause of morbidity and mortality. Untreated cervical teratomas are associated with a high mortality rate, though after surgical removal the chances of survival are excellent (9).

Tumors of the Fetal Chest

Lymphangioma

Although lymphangiomas most commonly occur in the cervical region, they may also be found in the axillae, mediastinum, and extremities. The US findings are not different to those of cervical lymphangiomas (Fig. 5).

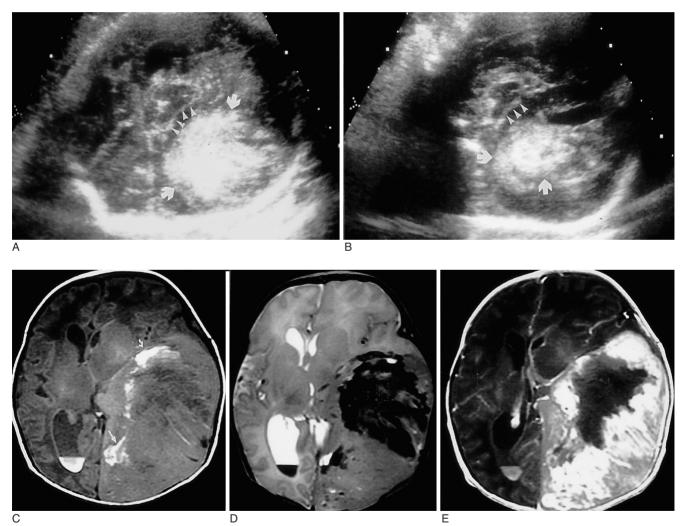


Fig. 2. Prenatal ultrasonographic and postnatal MR findings of fetal glioblastoma.

A, B. Axial and coronal ultrasonographic images of the fetal head depict a large mass of mixed echogenicity (arrows) at the left side of cerebral hemisphere. The margin of the mass is ill defined. The midline of the brain (arrowheads) has shifted to the right, and the right hemisphere is compressed by the mass.

- C. T1WI demonstrates a large ill-defined mass in the left cerebral hemisphere. The observed high signal intensity (small arrows) is due to intratumoral hemorrhage.
- D. T2WI reveals heterogeneous signal intensity due to hemorrhage and necrosis.
- E. Contrast-enhanced T1WI shows bright enhancement, with central necrosis.

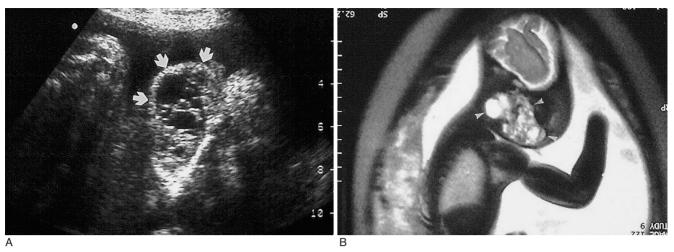


Fig. 3. Prenatal ultrasonographic and MRI findings of a 26-week fetus with bilateral cervical lymphangioma.

A. Coronal ultrasonographic image shows multiseptated cystic masses (arrows) at both sides of the fetal neck and chin.

B. Sagittal fetal MRI demonstrates a multiseptated cystic mass (arrowheads) at the left side of the fetal neck and chin.

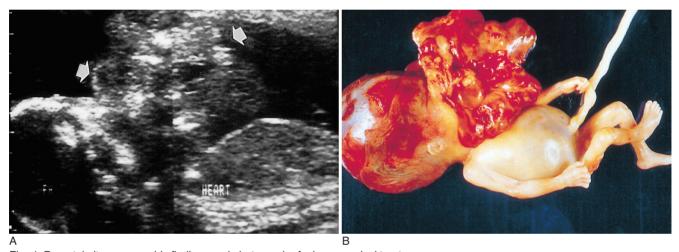


Fig. 4. Prenatal ultrasonographic findings and photograph of a large cervical teratoma.

A. Sagittal fetal image shows a large pedunculated mass of mixed echogenicity (arrows) at the anterior aspect of the fetal face and neck.

B. Photograph of the fetus shows a large pedunculated mass originating from the oral cavity.

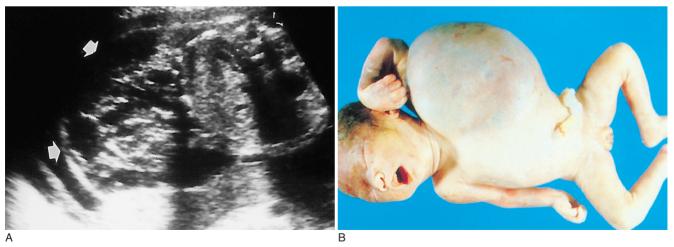


Fig. 5. Prenatal ultrasonographic finding and photograph of a 22-week fetus with a large chest-wall lymphangioma.

A. Axial image of the fetal upper abdomen demonstrates a large exophytic mass of mixed echogenicity (solid arrows) in the left chest wall.

B. The photograph depicts a large exophytic mass in the left chest wall.

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Tumors of the Fetal Heart

Congenital cardiac tumors are rare, with a prevalence of 0.01–0.05% (11). Rhabdomyomas are the most common cardiac tumors in infants and children, accounting for up to 60% of all tumors in this lesion. They may occur as single or multiple lesions within the ventricles and may be detected prenatally. In more than 50% of patients with cardiac rhabdomyomas, tuberous sclerosis is also present (12). The majority of such tumors are discovered incidentally at routine obstetrical US, whereas a few are discovered in patients with a family history of tuberous sclerosis or fetal cardiac arrhythmia, or in association with nonimmune hydrops. At US, a rhabdomyoma is most commonly seen as a homogeneous, echogenic, intracardiac mass, and is most commonly located in the ventricle, especially the septum

(Fig. 6). Cardiac rhabdomyomas are often multiple, occurring in more than one cardiac chamber and may produce arrhythmias, obstruct blood flow in the heart, or diminish contractility because of replacement of the myocardium (13). Although cardiac tumors can easily be detected by prenatal echocardiography at as early as 20 weeks of gestation, diagnosis requires that they are of sufficient size. During the fetal period they are known to either become larger or regress.

Tumors of the Fetal Abdomen

Abdominal Lymphangioma

Intra-abdominal lymphangiomas are rare but are usually located in the small bowel mesentery. This malformation

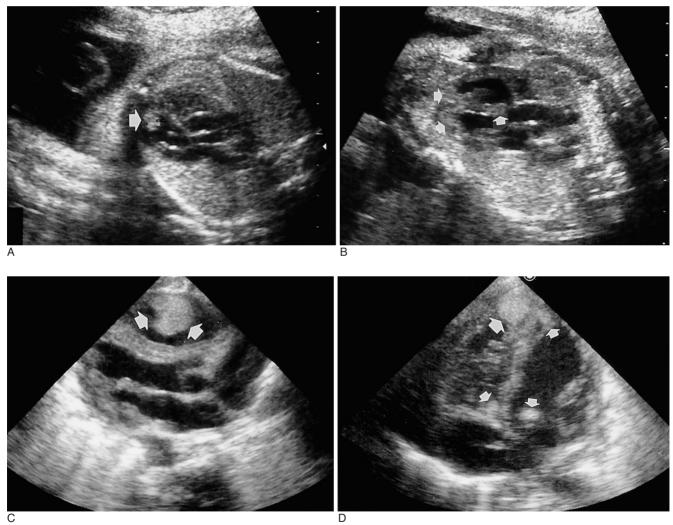


Fig. 6. Prenatal and postnatal ultrasonographic findings of cardiac rhabdomyomas.

- A. Prenatal ultrasonographic of a 27-week fetus reveals a round echogenic mass (arrow) in the lower portion of the right ventricle.
- B. Out-tract view of the fetal heart shows thickened interventricular septum and ventricular wall with multifocal increased echogenicities (arrows) suggesting multiple rhabdomyomas.
- C. Neonatal echocardiography depicts a round echogenic mass (arrows) in the right ventricle.
- D. Multiple echogenic nodules (arrows) are seen at the interventricular septum and ventricular wall.

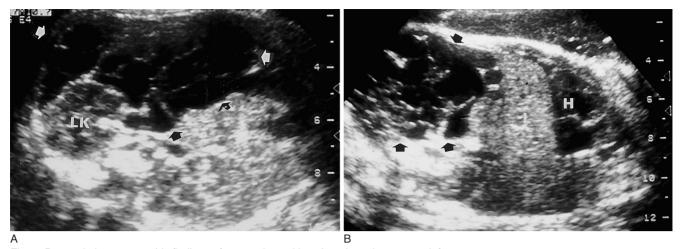


Fig. 7. Prenatal ultrasonographic findings of retroperitoneal lymphangioma in a 32-week fetus.

A, B. Axial and coronal images of the fetal abdomen depict a large multiseptated cystic lesion (arrows) in the left retroperitoneal space (LK: left kidney, LV: liver, H: heart).

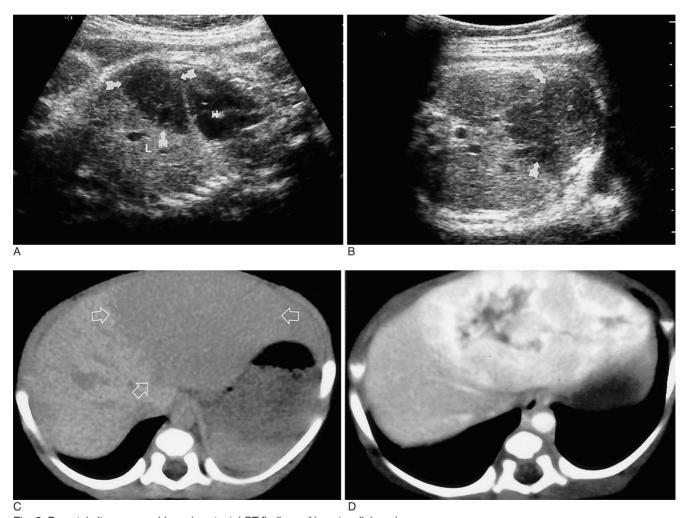


Fig. 8. Prenatal ultrasonographic and postnatal CT findings of hepatocellular adenoma.

- A, B. Coronal and axial ultrasonographic images of a 33-week fetus reveal a well-defined mass of low echogenicity (arrows) in the left lobe of the fetal liver (L: liver).
- C. Nonenhanced CT scan depicts a well-demarcated large mass (open arrows) in the left lobe of the liver.
- D. Contrast-enhanced CT scan demonstrates the early enhancement of the mass. Irregular unenhanced areas suggesting necrosis are apparent.

has been referred to a variety of names, such as mesenteric cyst, cystic lymphangioma of the mesentery, chylangioma or chylous cyst. Mesenteric cysts and abdominal lymphangiomas differ, however, in terms of their location, histology and potential for recurrence, and should be considered separate clinical entities (14). At US, lymphangiomas appear as cystic or multicystic masses, often with internal septations or scattered internal echoes (Fig. 7) (15). Clinical presentation depends on the site of involvement. A tumor may be very large, for example causing clinical problems early in life, but many patients are completely asymptomatic and the tumor is discovered during surgical exploration undertaken for other reasons. A tumor may occur in any portion of the small or large bowel mesentery, with symptoms arising due to the compression of adjacent organs (16).

Tumors of the Fetal Liver

1) Hepatocellular Adenoma

Hepatocellular adenoma in the fetus is very rare and its radiological findings have not been extensively described. These tumors are generally round masses with variable echogenicity, and are clearly distinguishable from normal liver parenchyma (Fig. 8). They are usually quite large, and infarction, hemorrhage and rupture are frequent. At postnatal CT and MRI, internal hemorrhage, fatty change, or necrosis may be observed (17, 18).

2) Hemangioma

Although hemangioma is the most common benign hepatic tumor, its antenatal diagnosis has rarely been reported in the literature (19). Because hemangiomas are often uncomplicated and resolve spontaneously during the first

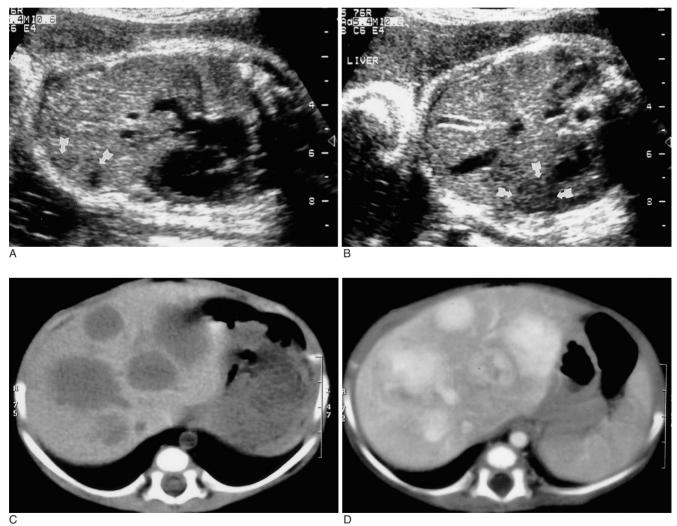


Fig. 9. Prenatal ultrasonographic and postnatal CT findings of multiple hepatic hemangiomas.

A, B. Coronal and axial ultrasomographic images of a 31-week fetus depict dilated hepatic veins, probably due to arteriovenous shunt. Ill-defined lesions of low echogenicity (arrows) are seen in the left lobe of the fetal liver, but were not by prenatal ultrasonographic.

- C. Nonenhanced CT scan shows multiple well-demarcated large masses in the liver.
- D. Contrast-enhanced CT scan demonstrates strong early enhancement.

two years of life, the incidence of this lesion certainly underestimated.

At antenatal US, a hepatic hemangioma is seen as a well-circumscribed abdominal mass with ranging in diameter from 1 to 10 cm (Fig. 9). It may be solitary or multiple, the latter sometimes forming part of generalized hemangiomatosis syndrome. Its internal structure is generally heterogeneous, with hypoechoic areas located at its center, and both hypo- and hyperechogenicity may be observed. Color Doppler ultrasound may show vascular flow with low resistive index (19, 20).

The mortality rate in infantile hepatic hemangioma has been reported as 12-90%. Complications include congestive cardiac failure, thrombocytopenia, and intra-abdominal hemorrhage as a consequence of rupture of the hemangioma (21).

Tumors of the Fetal Kidney (Mesoblastic nephroma)

Mesoblastic nephroma is a rare benign mesenchymal renal tumor, arising either from the kidney or the renal fossa. It occurs almost exclusively during in the neonatal period or early infancy, It is the most common fetal renal neoplasm. The prognosis after surgical excision is good. The echogenicity of the mass is varies: it homo- or heterogeneous, showing either necrotic or cystic change (Fig. 10) (22, 23). Rapid enlargement of the mass may lead to hemorrhaging. A mesoblastic nephroma is frequently associated with polyhydroamnios and consequently with premature labor (22, 24).

Tumor of the Adrenal Gland (Neuroblastoma)

Neuroblastoma is the most common malignant tumor in

neonates, accounting for 20% of all such malignancies (25). The presence of a solid or cystic fetal adrenal mass suggests fetal neuroblastoma, though fetal adrenal hemorrhage should also be considered. Fetal neuroblastoma is usually detected in the third trimester of pregnancy. The US findings vary, ranging from cystic to solid. Mixed foci of calcification are seen in the suprarenal area, though the cystic pattern is more frequent (Fig. 11) (26).

Overall prognosis for these patients is excellent, and this may be due in part to the detection of an otherwise clinically silent neuroblastoma in situ, which would undergo spontaneous regression (27). It may thus be appropriate to avoid overly aggressive therapeutic interventions, especially when many favorable prognostic indicators are present at diagnosis.

Sacrococcygeal Teratoma

Sacrococcygeal teratoma is the most common congenital neoplasm, usually presenting as a large midaxial exophytic mass in the sacrococcygeal region. It may be almost entirely external (type I), internal and external in equal parts (type II), mainly internal (type III), or entirely internal (type IV) (28). At prenatal US, the majority of those tumors are seen as a solid, or mixed cystic and solid, external caudal mass (Fig. 12). Only small proportions are entirely cystic (29). A sacrococcygeal teratoma may cause significant perinatal morbidity and mortality, and may lead to various complications such as severe dystocia, fetal hydrops, polyhydroamnios, or bleeding during parturition. Due to vascular steal from high metabolic demand and secondary high-output cardiac insufficiency, the prognosis for





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Fig. 10. Prenatal ultrasonographic findings of mesoblastic nephroma in a 34-week-old fetus.

A. Sagittal image reveals a markedly enlarged right kidney (arrows), with increased echogenicity throughout the entire renal area. The crescent-shaped lesion (\Leftrightarrow) occupying the perirenal space is probably due to the tumor extension and hematoma.

B. Power Doppler ultrasonographic image shows a vessel penetrating the renal capsule as far as the perirenal space.

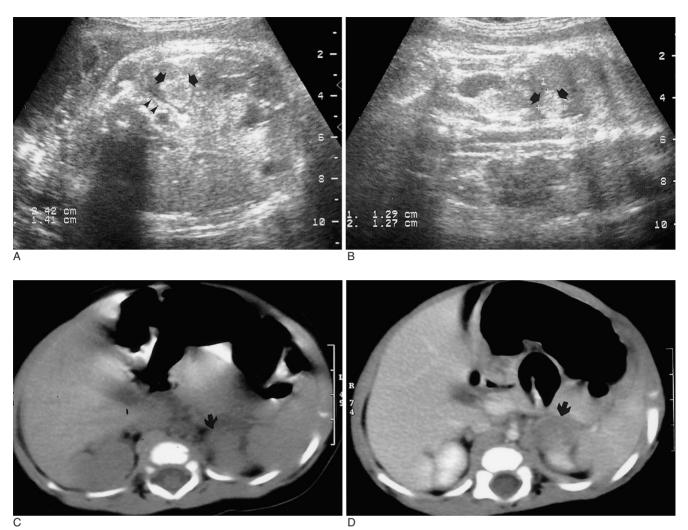


Fig. 11. Prenatal ultrasonographic and postnatal CT findings of neuroblastoma.

- A, B. Axial and coronal ultrasonographic images of a 32-week fetus depict a round echogenic mass (arrows) in the left suprarenal paraspinal area. The mass abuts to the right adrenal gland (arrowheads).
- C. Nonenhanced CT image demonstrates a small solid mass (arrow) in the supero-anterior portion of the left kidney.
- D. Contrast-enhanced CT image shows mild enhancement of the mass (arrow).

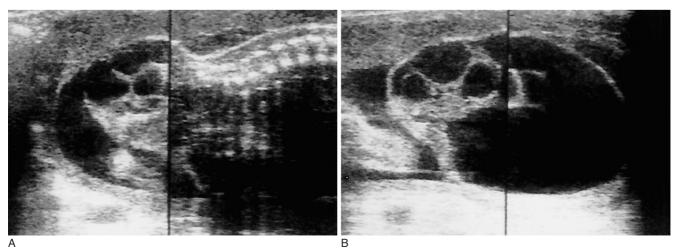


Fig. 12. Typical ultrasonographic findings of sacrococcygeal teratoma in a 33-week fetus.

- A. Sagittal image reveals an exophytic solid and cystic mass, the intrapelvic extent of which is not well defined, in the perineal area.
- B. Axial image of the same mass depict its cystic and solid components.

fetuses with solid tumor is usually poor (30).

SUMMARY

A variety of neoplasms can develop in each fetal organ. Teratomas and lymphangiomas may arise during early pregnancy, but other neoplasms usually develop during late pregnancy. Most fetal neoplasms can be detected by careful prenatal US until this time, but a few, such as hemangiomas of the liver, cannot be detected until the time of birth. Some show specific US findings suggesting the differential diagnosis, but others do not. Knowledge of the presence of a neoplasm in the fetus may alter the prenatal management of a pregnancy and the mode of delivery, and facilitates immediate postnatal treatment. The early detection of a fetal neoplasm and understanding of its US findings is therefore very important.

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