tissue structures external to the face or cranium should alert the sonographer to the possibility of nevus sebaceous, and a careful search for other associated abnormalities should be done.

Cystic adenomatoid malformation of the lung is also hamartomatous in nature and is characterized by failure of the terminal bronchioles to canalize, resulting in the formation of isolated cystic lesions. These malformations may be classified into three subtypes according to the size of the cysts. Type I has large cysts usually involving a single lobe; type II has multiple small cysts less than 1.2 cm in diameter distributed diffusely throughout one or both lungs; type III consists of noncystic lesions. Larger cysts generally have been associated with a better prognosis because they are more resectable and appear less likely to be associated with hydramnios and hydrops. Hydramnios is thought to occur secondary to esophageal compression and decreased gastrointestinal amniotic fluid absorption. The presence of nonimmune hydrops portends a dismal outcome, although some progress has been made in the prenatal treatment of these patients with placement of transabdominal cystoamniotic shunts or with fetal surgery. In a recent series reported by the Fetal Treatment Program at the University of California, San Francisco, four of six fetuses that underwent fetal surgery (hysterotomy and resection of chest mass) survived and are currently doing well.[6]

The present case illustrates the importance of a complete ultrasound examination once a single malformation has been detected. To our knowledge, this is the first report describing the ultrasound appearance of a nevus sebaceous and the first case in which a hamartomatous lesion of the lung was associated with a nevus sebaceous. This is somewhat surprising in view of the many different hamartomatous lesions described previously in association with this syndrome. By itself, the nevus sebaceous is not life-threatening; however, the presence of associated abnormalities can make effective treatment very difficult. Serial ultrasound examinations are recommended, and unless there are other abnormalities such as fetal hydrops or hydramnios, routine obstetric care should probably not be altered.

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Received July 26, 1993.
Received in revised form September 7, 1993.
Accepted September 8, 1993.

Umbilical artery steal syndrome and distal gangrene in a case of twin-twin transfusion syndrome

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***Background*: Vascular accidents in the survivor from pregnancies with twin-twin transfusion syndrome have been attributed to the release of thromboplastin from the dead fetus.**

***Case*: In a woman presenting at 22 weeks' gestation with acute polyhydramnios due to twin-twin transfusion syndrome, endoscopic laser coagulation of the communicating vessels was performed. At endoscopy, the toes of the left foot of the recipient fetus were noted to be gangrenous. This fetus had only a left umbilical artery, which was markedly dilated, and Doppler studies demonstrated impaired perfusion of the left leg. In four control fetuses with a single umbilical artery, Doppler studies demonstrated normal waveforms in both legs.**

***Conclusion*: In twin-twin transfusion syndrome, fetal peripheral gangrene may occur before the death of one twin. The ischemic necrosis of the toes probably resulted from the combined effect of polycythemia and umbilical artery steal syndrome. Interruption of twin-twin transfusion syndrome caused resolution of the polycythemia and prevented further**

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damage by removing one of the components of the peripheral ischemia. ***(Obstet Gynecol 1994;83:862–5)***

We recently observed a case suggestive of the umbilical artery steal syndrome, by which a major part of the blood volume from the common iliac artery may have been diverted into a single umbilical artery, at the expense of flow into the ipsilateral external iliac artery and leg.

Case Report

A 40-year-old woman presented at 22 weeks' gestation with acute polyhydramnios. Ultrasonographic examination demonstrated a monochorionic diamniotic twin pregnancy; both fetuses were morphologically normal but they were discordant in size. The larger twin had a normal biophysical profile, but was surrounded by polyhydramnios and had a single umbilical artery, which was markedly dilated and appeared as a cystic tortuous structure on the left side of the bladder (Figure 1). The smaller twin appeared adherent to the placenta because of the oligohydramnios, and no bladder could be demonstrated.

We diagnosed severe twin-twin transfusion syndrome and counseled the parents as to the available options of conservative management, repeated amniocenteses, termination, or laser coagulation of the communicating vessels.[1,2] The parents chose coagulation of the communicating vessels. After administration of local anesthesia, a 2.2-mm fetoscope (KeyMed, Southend, UK) was introduced into the amniotic sac of the recipient twin.[2] Examination of the fetus revealed a left foot with gangrene of the middle and distal phalanges of the second, third, and fourth toes (Figure 1). There were no amniotic bands. The right foot, both hands, and the face were normal. A fetal blood sample, taken by umbilical venous puncture, had a hemoglobin concentration of 17 g/dL (normal range for this gestation 11–14 g/dL). Subsequently, the fetoscope was directed toward the inter-twin membranes, and the six crossing vessels were coagulated by administering a total of 6000 J through a laser fiber (MBB, Munich, Germany) adjacent to the fetoscope, within the same cannula.[2]

Color flow mapping and pulsed Doppler velocimetry (Acuson 128, Mountain View, CA) of the fetal circulation were performed before and at regular intervals after laser coagulation of the communicating vessels. Before the procedure, the pulsatility index (PI) in the umbilical artery of the recipient twin was above the 95th percentile (1.99) and the mean blood velocity in the descending thoracic aorta was below the fifth percentile (12 cm/second) of our reference ranges for gestation. One week later, the values returned to within normal ranges, and both remained normal throughout pregnancy. The flow velocity waveforms from the femoral and tibial arteries were different in the two legs, and the PIs and maximum velocities were higher on the right side, both during pregnancy and postnatally (Table 1).

Doppler measurements at 22–27 weeks' gestation in four

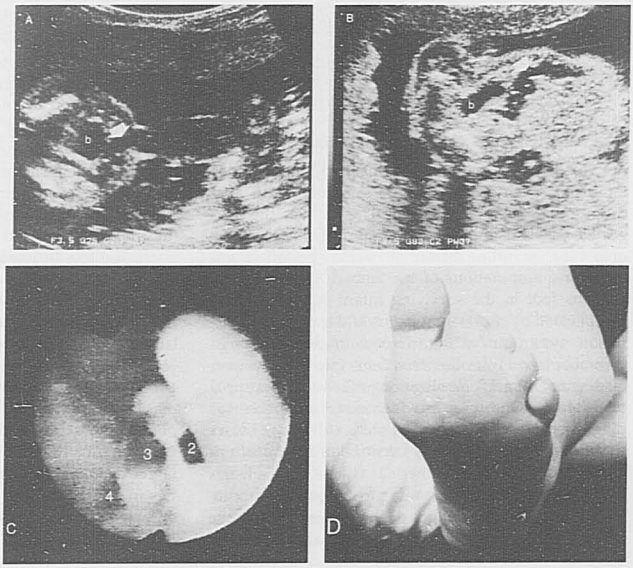

Figure 1. Ultrasound pictures demonstrating a single dilated left umbilical artery (*arrow*) in cross-sectional (A) and longitudinal view (B) of the abdomen (b = bladder). The fetoscopic dorsal view of the left foot (C) demonstrates necrotic (black) areas of toes 2–4. The postnatal plantar view (D) demonstrates absence of the distal phalanges of toes 2–4.

Table 1. Pulsatility Index and Maximum Velocity in the Femoral and Posterior Tibial Arteries at Various Gestations

GA (wk)	Left femoral		Right femoral		Left tibial		Right tibial	
	PI	Vmax	PI	Vmax	PI	Vmax	PI	Vmax
22	2.15	9	3.12	15	2.51	3	2.48	13
25					2.37	6	3.40	15
28	2.27	13	3.26	19	3.13	4	2.82	13
32	2.35	13	3.48	23	3.02	5	4.01	14
36	2.50	13	3.55	26				
Postnatal	3.36	51	5.78	80				

GA = gestational age; PI = pulsatility index; Vmax = maximum velocity (cm/s).
In the antenatal studies, the time average maximum velocity was measured; in the postnatal study, the systolic maximum velocity was used because there was reversed diastolic flow in the right femoral artery.

control fetuses with single umbilical arteries demonstrated blood flow waveforms from the femoral arteries similar to those in the right leg of our fetus. The mean PI was 3.65 for the left and 3.40 for the right, and the corresponding values of mean time average maximum velocity were 15 and 16 cm/second. In each fetus, the difference in values comparing the two legs was always less than 15%.

The day after fetoscopy, the donor twin was dead but the recipient had a normal biophysical profile. Subsequent scans demonstrated normal growth, and a healthy female infant was delivered vaginally after spontaneous labor at 39 weeks. The birth weight was 2500 g, the Apgar scores at 1 and 5 minutes were 9 and 10, respectively, and the hemoglobin concentration was 14.5 g/dL. The distal phalanges of the second, third, and fourth toes were absent in the left foot (Figure 1). The infant, now 8 months old, is developing normally. The placenta was monochorionic, and the dead fetus was papyraceous. The mother did not demonstrate features of disseminated intravascular coagulation (DIC).

Discussion

Congenital amputations of the extremities have been described in association with maternal ingestion of teratogens such as thalidomide, amniotic band syndrome, or ischemic accidents.[3] The latter is the suggested mechanism for the association between chorionic villus sampling before 10 weeks' gestation and transverse amputations of the limbs.[4] A case of gangrenous foot in the surviving infant of a pregnancy complicated by chorioangiopagus and twin-twin transfusion syndrome was attributed to the release of thromboplastin from the dead fetus into the survivor, leading to DIC and embolization of the vascular supply to the foot.[5] Our case, in which distal gangrene was diagnosed when both fetuses were still alive, shows that this phenomenon may occur before the death of one twin.

The ischemic necrosis leading to congenital amputation of the distal phalanges of the foot seems to have been the consequence of impaired perfusion due to the combined effect of polycythemia and umbilical artery steal syndrome. The main determinant of blood viscosity is hematocrit, and when polycythemia is present, hyperviscosity in the microcirculation could cause ischemic tissue damage. In two polycythemic neonates, distal gangrene in the hand developed within 48 hours of birth.[6,7] Margono et al[5] also reported polycythemia in a case of gangrenous foot in the survivor of a pregnancy with twin-twin transfusion syndrome. Nevertheless, gangrene of the extremities, unlike polycythemia, is very rare.

Single umbilical artery is found in 1% of singleton and 3–4% of multiple pregnancies, and in 20–50% of infants with associated anomalies.[8] However, ischemic digital defects are not common. We have studied four control cases with single umbilical artery, and Doppler velocimetry of the lower limbs demonstrated normal flow in both legs. In contrast, the case with gangrenous toes demonstrated features compatible with impaired perfusion of one leg, which persisted postnatally. This may be due to the marked dilatation of the single umbilical artery, which caused a steal phenomenon from the ipsilateral external iliac artery.

A steal of blood from the cerebral arteries has been described in premature neonates with persistent ductus arteriosus, and this phenomenon may be involved in the genesis of ischemic and hemorrhagic cerebral injury.[9] The steal phenomenon occurs during diastole, when there is shunting of blood from the aorta through the ductus arteriosus to the pulmonary circulation. Therefore, during diastole, a decrease in cerebral blood flow and a consequent increase in PI occur. Unlike the ductal steal syndrome, which occurs during diastole, the umbilical artery steal syndrome is a phenomenon observed mainly during systole, when blood is preferentially diverted toward the vessel with the larger diameter and lower resistance. Diastolic velocities in arterial vessels of the extremities are normally very low or even absent or reversed. Therefore, a decrease in pressure due to umbilical artery steal syndrome would affect mainly the systolic peak veloc-

ities, so the PI is decreased in the vessels of the leg with impaired perfusion.

The postnatal persistence of a difference in blood flow to the two legs may have reflected a reduction in the diameter of the left external iliac artery. Reduction in the rate of blood flow through an artery causes a structural alteration of the arterial wall and a reduction in vessel diameter within 2 weeks.[10]

When the ischemia is severe enough to cause gangrene, the process is unlikely to reverse spontaneously if the causal agent persists. In our case, it is possible that laser coagulation of the communicating vessels was associated with improved perfusion to the extremities because interruption of twin-twin transfusion syndrome caused resolution of the polycythemia, one of the two components of the peripheral ischemia.

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Reprints are not available.

Received July 29, 1993.
Received in revised form September 17, 1993.
Accepted October 18, 1993.

Etiology of prune belly syndrome: Evidence of megalocystic origin in an early fetus

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***Background*: Prune belly syndrome is a rare and complicated condition affecting the genitourinary organs and abdominal wall. The etiology of abdominal musculature deficiency in prune belly syndrome is controversial. We present a case that should elucidate the etiology of this syndrome.**

***Case*: A spontaneously aborted fetus at 12 weeks' gestation with an early stage of prune belly syndrome was investigated by necropsy and light and electron microscopy. Megalocystis resulting from urethral atresia was diagnosed. There was no hydroureter or hydronephrosis, and both light and electron microscopy demonstrated evidence of development of the abdominal musculature. Both testes were elevated as a result of the megalocystis.**

Conclusion*: These findings suggest that hypoplasia of the abdominal musculature and cryptorchidism might develop secondary to the presence of chronic megalocystis in this syndrome. *(Obstet Gynecol 1994;83:865–8)

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Prune belly syndrome is a rare disease[1,2] characterized by defective abdominal-wall musculature, cryptorchidism, and urinary tract anomalies.[3] The incidence is higher in males. Its pathogenesis has not been established,[4–12] partly because the syndrome is rarely detected in early fetal life[13–15] and because of the paucity of pathologic specimens. The present study investigated a male fetus that aborted spontaneously at week 12 and that had an early stage of prune belly syndrome. We present an hypothesis regarding the pathogenesis of this syndrome based on our findings and a review of the relevant literature.