INTRODUCTION

Structural abnormalities of the umbilical cord may be identified during a prenatal ultrasound examination. This topic will describe several such abnormalities, as well as their potential clinical significance and management of affected pregnancies. Findings on postnatal examination of the umbilical cord and their clinical significance are reviewed separately.

●See "Gross examination of the placenta", section on 'Umbilical cord.

●See "The placental pathology report", section on 'Umbilical cord.

NORMAL UMBILICAL CORD ANATOMY — The umbilical cord contains two arteries and one vein surrounded by a gelatinous stroma (ie, Wharton's jelly) and covered by a single layer of amnion. The arteries carry deoxygenated blood from the fetus to the placenta, and the vein carries oxygenated blood from the placenta to the fetus. Arterial blood flow is pulsatile, with a smaller pulse in the umbilical vein [1]. The physical dimensions of the cord correlate with fetus size.

●Arteries – The left and right umbilical arteries are branches of the left and right internal iliac arteries, respectively. In the pelvis, the two arteries are separated by the bladder but then lie adjacent to each other at the umbilicus, where they exit the fetus and enter the umbilical cord (figure 1). The arteries follow a helical course around the umbilical vein until reaching the placenta, where they separate again and form the chorionic arteries on the surface of the placenta, with perforating branches to the underlying villi (figure 2).

●Vein – The confluence of chorionic veins at the chorionic plate form a single ("left") umbilical vein, which courses through the cord to the umbilicus. Upon entering the fetal abdomen, it runs cephalad until it enters the liver, where it anastomoses with the portal sinus (ie, confluence of portal veins) and the ductus venosus (figure 1)

A "right" umbilical vein is present very early in embryogenesis and then usually degenerates. Rarely, it persists as the only umbilical vein [3,4] or as a fourth umbilical vessel. (See 'Supernumerary vessels' below.

After birth, the intraabdominal portions of the umbilical vessels degenerate; the umbilical arteries become the lateral ligaments of the bladder, and the umbilical vein develops into the round ligament of the liver.

VASCULAR ABNORMALITIES

Single umbilical artery

Single umbilical artery (SUA) refers to a variation of umbilical cord anatomy in which there is only one umbilical artery, rather than the normal two umbilical arteries.

Prenatal diagnosis, clinical significance, and management

Prenatal diagnosis of SUA is based on the finding of a two-vessel cord on transverse and longitudinal views of a free loop of cord (image 3). Visualization of one artery going around the fetal bladder instead of one artery on each side of the bladder supports the diagnosis (image 4).

SUA may be an isolated finding or associated with aneuploidy or other congenital anomalies. Prenatal diagnosis, clinical significance, and pregnancy management are reviewed in detail separately. (See "Single umbilical artery".)

Hypoplasia of one umbilical artery

Hypoplasia of one umbilical artery is a variant of SUA in which both arteries are present but with a gross disparity in their size. The hypoplastic but functional artery can occasionally be detected prenatally by ultrasound.

Prenatal diagnosis

The prenatal diagnosis of a hypoplastic umbilical artery is based on identification of all of the following :

- Two patent umbilical arteries
- One artery that is smaller than the other (at least 50 percent smaller)
- Discordant umbilical artery flow velocity waveforms between the arteries

Size discordancy is best appreciated in a magnified cross-sectional view of the umbilical cord [19]. There is no universal standard definition of discordancy; differences in arterial diameter of 1 to 3 mm between arteries have been described and occur in up to 2 percent of pregnancies [18,21,22]. The prevalence of discordancy is much lower if it is defined as >50 percent difference between arterial diameters. Using this definition, only 12 cases of hypoplastic umbilical artery were identified among 31,000 consecutive second- and third-trimester ultrasound scans [20]. The low prevalence (0.04 percent) in this series was probably the result of using the very strict definition of hypoplastic umbilical artery and the inclusion of many early second-trimester pregnancies. The detection of marked discordancy between umbilical arteries is difficult to document before 20 weeks due to the small size of the arteries early in gestation.

Color Doppler ultrasound is used to confirm flow within the hypoplastic artery (image 5), since absence of flow suggests an atrophic nonfunctional artery. Doppler is also used to document arterial flow velocity waveforms, which are discordant when one artery is hypoplastic [18,19]. The resistance index is almost always higher in the smaller artery, and there may be absent end-diastolic flow.

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Clinical significance

The clinical significance of hypoplastic umbilical artery has not been clearly established. An increased risk of adverse pregnancy outcome has been reported, as well as associations with maternal diabetes mellitus, polyhydramnios, fetal growth restriction, congenital anomalies, abnormal insertion of the umbilical cord, and placental abnormalities [18-23]. Among 39 prenatally diagnosed cases of hypoplastic umbilical artery, seven fetuses (18 percent) had associated anomalies, including three cases of trisomy 18 and one case each of biliary atresia, hydronephrosis and duplex collecting system, agenesis of the corpus callosum and kyphoscoliosis, and tetralogy of Fallot [18-23]. The high prevalence of associated structural anomalies in these reports should be interpreted with caution because most cases of hypoplastic umbilical artery were diagnosed in a high-risk population.

Management

A thorough examination of fetal anatomy and both cord insertion sites should be performed to look for additional abnormalities. The author suggests ultrasound examination for fetal growth and assessment of amniotic fluid at 28 and 34 weeks. This approach is based on the limited clinical experience obtained from the few case reports and small series reported in the literature.

At any follow-up ultrasound examination, he suggests measuring Doppler flow in both umbilical arteries. Clinical management is based on the Doppler findings of the normal diameter artery.

Postnatal confirmation

Constriction of the umbilical arteries after birth makes the

CLASSIFICATION

SUA has been classified into four types based upon the likely developmental etiology:

●Type 1 is the most common form of SUA, comprising 98 percent of cases. The umbilical cord contains two patent vessels, an artery of allantoic origin and a vein derived from the left umbilical vein. This type of SUA has been associated with genitourinary anomalies.

●Type 2 accounts for 1.5 percent of cases. The umbilical cord contains two patent vessels, an umbilical artery of vitelline origin arising from the superior mesenteric artery and a vein arising from the left umbilical vein. Severe fetal anomalies, such as caudal regression syndrome and sirenomelia, have been associated with Type 2 SUA.

●Type 3 is rare. The umbilical cord contains three patent vessels, one artery of allantoic origin and two veins. The veins arise from the left umbilical vein and a persistent anomalous right umbilical vein. This form of SUA has been associated with major congenital anomalies, resulting in a poor fetal prognosis.

●Type 4 is extremely rare. It consists of one artery of either vitelline or allantoic origin and a vein derived from the right umbilical vein. The risk of embryonic loss may be increased in these cases.