Single umbilical artery

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Literature review current through: May 2017. | **This topic last updated:** Dec 01, 2016.

INTRODUCTION — The umbilical cord typically contains two arteries and one vein. Single umbilical artery (SUA) refers to a variation of umbilical cord anatomy in which there is only one umbilical artery. It may be an isolated finding, or associated with aneuploidy or other congenital anomalies.

EMBRYOGENESIS — Three mechanisms have been proposed to explain the development of SUA [1]:

- Secondary atresia or atrophy of a previously normal umbilical artery
- Primary agenesis of one of the umbilical arteries
- Persistence of the original single allantoic artery of the body stalk

Approximately 40 percent of umbilical cords with SUA undergoing microscopic examination have muscular remnants, which supports secondary atresia or atrophy of a previously normal umbilical artery as the likely mechanism of SUA in these cases [2].

CLASSIFICATION — SUA has been classified into four types based upon the likely developmental etiology [3,4]:

- 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. There may be an increased risk of embryonic loss.

PREVALENCE AND EPIDEMIOLOGY — The prevalence of SUA depends on characteristics of the population studied:

- At 11 to 14 weeks of gestation before chorionic villus sampling – 5.9 percent [5]
- At 16 to 23 weeks of gestation on prenatal ultrasound in an unselected population – 0.48 percent [6]
- Among fetal deaths, autopsies, or aborted fetuses – 2.13 percent [7]
- Among live born infants – 0.55 percent [7]

SUA has been reported to occur more frequently in pregnancies at the extremes of maternal age [8] and in Eastern Europeans, and less frequently in Japanese and Africans [3]. There are inconsistent data that male fetuses are more often affected than females [9] and are more likely to have additional malformations [8].

SUA is more common in twin pregnancies (3.9 to 8.8 percent) [8,10-13], with similar prevalence in monochorionic and dichorionic twins [14]. Isolated SUA has been reported on prenatal ultrasound in 1.7 percent of twins at 17 to 22 weeks of gestation [15]. In twins discordant for SUA, the smaller twin is more likely to have the single artery [8,14,16]. SUA is invariably present in acardiac twins [3].

RISK FACTORS — SUA has been associated with maternal smoking, diabetes, hypertension, and seizure disorders [3,11].

CLINICAL PRESENTATION — SUA is a finding on obstetrical ultrasound examination. In the second and third trimesters, the American Institute of Ultrasound in Medicine recommends imaging the umbilical cord as part of a standard prenatal ultrasound examination and evaluating the number of vessels in the cord, when possible [17]. SUA can be detected in the first trimester,

but sensitivity and specificity are higher in the second trimester (sensitivity: 57.1 versus 86.6 percent, specificity: 98.9 versus 99.9 percent in one study [18]). Associated fetal and/or placental abnormalities are common. (See 'Assessment for associated abnormalities' below.)

Findings — Ultrasound examination shows only two vessels in the umbilical cord. Larger studies report the left artery is absent more often than the right (59 versus 41 percent [19]) [1,19,20]. The side (left or right) of the missing artery does not appear to have clinical significance [19-22], with the exception of one study that reported complex fetal anomalies and chromosomal abnormalities occurred exclusively when SUA resulted from the absence of the left umbilical artery [1].

Most umbilical cords have a normal diameter despite SUA, but the cord may be velamentous or have a small diameter [23,24]. In the longitudinal view, the two-vessel cord frequently appears straight and non-coiled, although occasionally the single artery may loop around the vein.

Absence of one umbilical artery in the cord necessarily implies the absence of the corresponding intraabdominal portion of the umbilical artery.

The single umbilical artery is larger than normal. In the normal three-vessel cord, blood flow to the placenta is approximately equally distributed through both arteries. In the two-vessel cord, the entire fetoplacental circulation is transported through only one artery, resulting in a compensatory increase of the arterial diameter. The diameter of the single artery usually measures more than 50 percent of the diameter of the vein, resulting in an umbilical vein-to-artery ratio <2 (figure 1) [25,26]. In contrast, the arteries in a three-vessel cord usually measure less than 50 percent of the diameter of the vein, resulting in an umbilical vein-to-artery ratio >2. The compensatory increase in the single artery diameter can be seen as early as the early second trimester [26,27].

High-resolution color Doppler ultrasound may demonstrate differences in the impedance indices in the pelvic circulation of fetuses with SUA [28,29]. The discordant intraluminal blood flow between common iliac arteries is more marked as pregnancy progresses, which may be due to both the decrease in resistance in the placental circulation and the increase in resistance in femoral arteries [28,29].

DIAGNOSIS

Normal three-vessel cord — Examination of the number of umbilical vessels in the umbilical cord can be difficult in the first trimester due to the small size of the vessels. In the second and third trimesters, the three umbilical vessels in a normal umbilical cord (image 1) can be easily visualized in a cross-sectional image of the cord by two-dimensional ultrasound examination. Color flow Doppler imaging of the intra-abdominal portions of the umbilical arteries running alongside the fetal bladder confirms the diagnosis.

Single umbilical artery — Prenatal diagnosis of SUA is based on the following:

- Ultrasound examination showing only two vessels in the umbilical cord (cross section or longitudinally) (image 2A) [30]. The larger vessel is the vein and the smaller vessel is the artery [3]. In the transverse plane, the two-vessel cord resembles a “soda can tab” when the amount of Wharton's jelly is adequate.
- Color flow Doppler in the region of the bifurcation of the fetal aorta shows intra-abdominal umbilical vessels on only one side of the fetal bladder, which is pathognomonic of SUA (image 3A-B). This is particularly helpful in the first trimester and can facilitate early diagnosis of SUA (image 4).

Counting the number of cord vessels in a free loop of cord is difficult because multiple loops of cord are in close proximity to each other. Counting the number of cord vessels in a hypercoiled cord is also difficult (image 5). Color Doppler of intra-abdominal vessels can be more informative in these settings than standard real-time imaging.

Differential diagnosis

Fused arteries at the insertion site — A potential diagnostic pitfall is only imaging the cord close to the placental insertion because the umbilical arteries may fuse along a short (<3 cm) segment of cord at this site [31]. This results in an umbilical cord with a short distal (placental) segment with two vessels and a long proximal (fetal) segment with three vessels (image 6) [32,33]. Imaging intra-abdominal vessels on both sides of the bladder excludes the diagnosis of SUA in these cases.

Hypoplasia of one umbilical artery — Hypoplasia of one umbilical artery is a variant of SUA in which both arteries are present, but there is a gross disparity in their size and blood flow. The hypoplastic but functional artery can occasionally be detected prenatally by ultrasound [34-40]. Constriction of the umbilical arteries after delivery makes the diagnosis difficult to document postnatally in spite of routine examination of the umbilical cord. (See "Prenatal diagnosis and management of umbilical cord abnormalities", section on 'Hypoplasia of one umbilical artery'.)

If one artery is atrophic rather than hypoplastic, the muscular remnants of the atrophic artery can only be detected by histological examination of the umbilical cord [2].

POSTDIAGNOSTIC EVALUATION

Assessment for associated abnormalities — A comprehensive sonographic evaluation of the fetus, cord, and placenta should be performed to look for fetal anomalies, especially genitourinary, cardiac, gastrointestinal, musculoskeletal, and central nervous system anomalies [7,8,41-45]. Specific but uncommon anomalies that have been reported with SUA include diaphragmatic hernia, exstrophy of cloaca sequence, sirenomelia sequence, and VATER (VACTERL) association (Vertebral anomalies, Anal atresia, Cardiac defects, Tracheoesophageal fistula and/or Esophageal atresia, Renal & Radial anomalies, and Limb defects) [3,43,46].

In a meta-analysis of 37 studies that evaluated pregnancy complications associated with SUA, the mean incidence of associated anomalies was 66.3 percent in series of fetal deaths, autopsies, or aborted fetuses and 27 percent in series of live born infants [7]. In one of the largest studies published after this analysis, both cardiac anomalies and renal anomalies were significantly more prevalent in infants with SUA than in those without SUA (cardiac anomalies 7.1 versus 0.4 percent, renal anomalies 4.8 versus 1.7 percent) [42].

The sonographic examination should include:

- A detailed fetal anatomic survey. (See "Ultrasound examination in obstetrics and gynecology", section on 'Detailed examination'.)

  A standard four-chamber view of the heart and views of the great arteries is performed; fetal echocardiography is not necessary if these standard cardiac assessments are normal and the patient has no other indications for fetal echocardiography [41,47]. (See "Fetal cardiac abnormalities: Screening, evaluation, and pregnancy management".)

  The number, location, and appearance of the kidneys should be documented as renal anomalies and are common, but not usually clinically significant [7,48].

- Assessment of the placenta and umbilical cord. Velamentous insertion, abnormally short cord, placenta previa, true knot, circumvallate placenta, and placental infarcts are more common with SUA [8,11].

Fetal genetic studies — The rate of aneuploidy with isolated SUA does not appear to be increased. In a series of 643 fetuses with SUA, 424 fetuses had isolated SUA and none of these fetuses had a chromosomal abnormality [43]. The same authors' review of the literature found 809 cases of isolated SUA of which six (0.7 percent) had an abnormal karyotype (three trisomy 18 and three trisomy 21).

Most experts do not consider isolated SUA a high-risk factor for offering invasive fetal genetic studies [1,40,43,49-51]. However, offering noninvasive conventional serum screening or a cell-free DNA screening test for trisomies 21, 18, and 13 is reasonable. Invasive diagnostic testing (karyotype with microarray) should be offered to women in whom additional fetal abnormalities are observed (eg, anatomic abnormalities, symmetric growth restriction) or the aneuploidy screening test is positive [52], given the increased risk of fetal genetic abnormalities in each of these settings. Karyotype abnormalities have been reported in as many as 45 percent of fetuses with nonisolated SUA [6,19,43]; autosomal trisomies, particularly trisomy 18, trisomy 13, or triploidy, appear to be the most common abnormalities [43]. This approach minimizes the number of women who will require an invasive test while potentially detecting aneuploid fetuses misdiagnosed as isolated SUA prenatally [3,53,54].

PREGNANCY MANAGEMENT

Monitoring for growth restriction — The authors perform ultrasound examination at 28 and 35 weeks to screen for growth delay, given the increased risk of this complication in SUA [52] (see 'Small for gestational age and birth weight' below). However, there is no evidence that routine sonographic screening for growth restriction improves perinatal outcome in these patients [55].

The Society of Obstetricians and Gynaecologists of Canada recommends monitoring fetal growth by physical examination, with follow-up ultrasound if clinically warranted (eg, size less than dates, development of additional risk factors for growth delay) [49], while other authors suggest routine ultrasound follow-up for fetal growth assessment [42,50] or a single 32 week screening examination.

Antepartum fetal monitoring — With isolated SUA and normal fetal growth, routine fetal testing is generally not recommended [56].

In the presence of recognized fetal growth restriction, regular fetal testing is employed. In general, this is comprised of modified biophysical profile and umbilical artery Doppler. After 32 weeks of gestation, nonstress test is added to the testing and the frequency of testing may be increased to twice weekly, depending upon the individual circumstances. (See "Overview of antepartum fetal surveillance", section on 'Indications for fetal surveillance'.)

Delivery — SUA alone does not affect the timing or route of delivery. The pediatric provider should be notified of the prenatal findings, and the neonate should be closely examined at delivery. Opinions differ on the value of postnatal renal screening of infants with isolated SUA prenatally. (See "Care of the umbilicus and management of umbilical disorders", section on 'Single umbilical artery'.)

Special issues

Cordocentesis — The safety of cordocentesis, when indicated, in fetuses with SUA has been addressed in two studies involving a total of 43 fetuses with SUA and associated structural anomalies undergoing fetal blood sampling for karyotyping [57,58]. No procedure-related complications were reported and no fetal deaths occurred in the two weeks after the procedure. Vasospasm or tamponade of the cord vessels, which can occur during fetal blood sampling, could have more serious consequences in a fetus with SUA [57,58].

First-trimester diagnosis — There is only limited information on the management of SUA diagnosed in the first trimester. If detected at this early gestational age, nuchal translucency thickness and fetal anatomy should be assessed. Increased nuchal translucency thickness or fetal anomalies are markers of an increased risk for aneuploidy [5], and warrant offering genetic testing. (See "Cystic hygroma and increased nuchal translucency" and "Prenatal genetic evaluation of the anomalous fetus".)

PREGNANCY OUTCOME — The type and frequency of adverse outcomes in fetuses with SUA depend largely on whether it is isolated or nonisolated [53,59,60]. Studies evaluating the outcome of SUA are often small, retrospective, and subject to several types of bias, thus limiting the reliability of their findings.

Preterm delivery — An increased risk of preterm delivery has been reported by several authors [42,54,60-63]. In a population-based study (194,809 deliveries), infants with isolated SUA were born on average one week earlier than infants without SUA (38.3±3.0 versus 39.3±2.1 weeks, p <0.001) [61]. Earlier delivery has been related to an increased frequency of a variety of pregnancy complications, including premature rupture of membranes, placenta previa, abnormalities of amniotic fluid, placental abruption, pregnancy-induced hypertension, and maternal medical disorders [13,61,62].

Small for gestational age and birth weight — Isolated SUA may have a small effect on fetal growth and birth weight, but data are inadequate to make a clear conclusion. In a 2013 systematic review and meta-analysis that pooled data from three cohort studies and four case-control studies (n = 928 pregnancies) comparing pregnancy outcomes with isolated SUA versus a three-vessel cord, isolated SUA was associated with trends toward small for gestational age (SGA) (odds ratio [OR] 1.6, 95% CI 0.97-2.6) and a small reduction in mean birth weight (51 grams, 95% CI -154.7 to 52.6 g) that were not statistically significant [64]. Studies were eligible for the analysis if they described at least 30 cases of isolated SUA as identified by ultrasound before an average gestational age of 24 completed weeks. A subsequent study of 219 consecutive women carrying a fetus with an isolated SUA diagnosed in the second trimester reported a significant association with SGA, pregnancy-induced hypertension, and medically indicated preterm birth [62].

In nonisolated SUA, chromosomal and anatomic abnormalities likely have a greater impact on fetal growth than the number of arteries in the umbilical cord.

Perinatal outcome — Increases in fetal, neonatal, and infant death have been reported with isolated SUA, but findings have been inconsistent and inconclusive. In the 2013 systemic review and meta-analysis described above, isolated SUA was associated with a trend toward increased perinatal mortality that was not statistically significant (OR 2.0, 95% CI 0.9-4.2) [64].

Uncontrolled case series have noted an increased frequency of various adverse outcomes in fetuses with isolated SUA (eg, neonatal intensive care unit admission, nonreassuring fetal heart rate leading to cesarean delivery, SGA, perinatal death) [54,61,65]. A reduction of Wharton's jelly and abnormal cord twist have been observed in cords with SUA, which might account, in part, for the adverse outcomes observed in some studies [66,67].

In nonisolated SUA, perinatal morbidity and mortality are increased and depend on the underlying chromosomal and anatomic anomalies.

Long-term outcome in offspring — Studies have generally reported that long-term physical and neurologic development of children with isolated SUA is similar to that in unaffected children [53,59,68].

In nonisolated SUA, long-term outcomes depend on the underlying chromosomal and anatomic anomalies.

SUMMARY AND RECOMMENDATIONS

- Single umbilical artery (SUA) refers to a variation of umbilical cord anatomy in which there is only one umbilical artery. It may be an isolated finding or associated with fetal abnormalities. The left artery is absent more often than the right; however, the side (left or right) of the missing artery does not appear to have clinical significance. (See 'Embryogenesis' above and 'Findings' above.)

- SUA occurs in approximately 0.5 percent of unselected pregnancies undergoing second-trimester ultrasound examination, but is more common in twins and in fetal deaths, autopsies, and aborted fetuses. (See 'Prevalence and epidemiology' above.)

- Prenatal diagnosis of SUA is based on the following (see 'Diagnosis' above):

    - Ultrasound examination showing only two vessels in the umbilical cord (cross section or longitudinally) (image 2A-B).

- Color flow Doppler in the region of the bifurcation of the fetal aorta showing intra-abdominal umbilical arteries on only one side of the fetal bladder (image 3A and image 3B).

- A potential diagnostic pitfall is only imaging the cord close to the placental insertion, where a short segment may contain only two vessels while the remainder of the cord contains three vessels. (See 'Fused arteries at the insertion site' above.)

- A comprehensive sonographic evaluation of the fetus, cord, and placenta should be performed to look for other anomalies, especially genitourinary, cardiac, gastrointestinal, and central nervous system anomalies. Fetal echocardiography is not necessary if a standard four-chamber view of the heart and views of the great arteries are normal and the patient has no other indications for fetal echocardiography. (See 'Assessment for associated abnormalities' above.)

- For pregnancies with isolated SUA, screening for trisomy 21, 13, or 18 by conventional serum screening or cell-free fetal DNA screening is a reasonable first-line approach. Invasive testing with microarray should be offered to women who have pregnancies with nonisolated SUA or who screen positive. (See 'Fetal genetic studies' above.)

- Isolated SUA may have a small effect on fetal growth and birth weight, but data are inadequate to make a clear conclusion. In nonisolated SUA, chromosomal and anatomic abnormalities likely have a greater impact on fetal growth than the number of arteries in the umbilical cord. (See 'Small for gestational age and birth weight' above.)

- Increases in fetal, neonatal, and infant death have been reported with isolated SUA, but findings have been inconsistent and inconclusive. In nonisolated SUA, perinatal morbidity and mortality are increased and depend on the underlying chromosomal and anatomic anomalies. (See 'Perinatal outcome' above.)

- The authors perform ultrasound examinations every four to six weeks to screen for growth delay, given the possibly increased risk of this complication in SUA. However, there is no evidence that routine sonographic screening for growth restriction improves perinatal outcome in isolated SUA, and not performing serial ultrasound examinations or performing a single screening examination at 32 weeks is also reasonable. (See 'Small for gestational age and birth weight' above.)

- Because an increase in perinatal mortality has been reported for isolated SUA without comorbidities, the author orders weekly fetal assessment by biophysical profile or nonstress test from 32 weeks to delivery, even in the absence of growth restriction or other standard indications for antepartum fetal surveillance. However, it is also reasonable to restrict this testing to pregnancies with standard clinical indications. (See 'Antepartum fetal monitoring' above.)

- SUA alone does not affect the timing or route of delivery. The pediatric provider should be notified of the prenatal findings. (See 'Delivery' above.)

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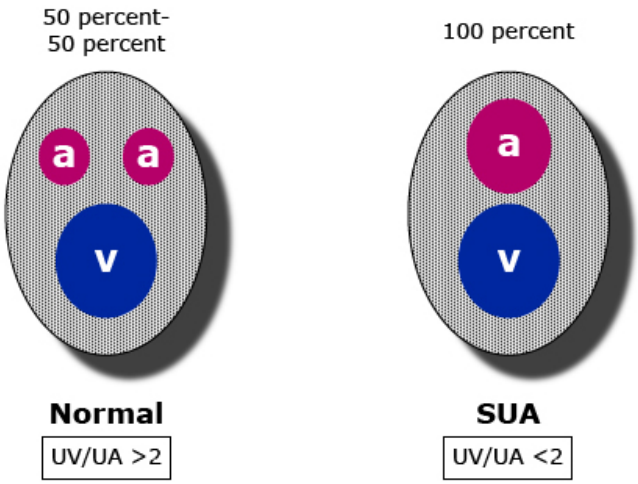
Topic 15245 Version 19.0

GRAPHICS

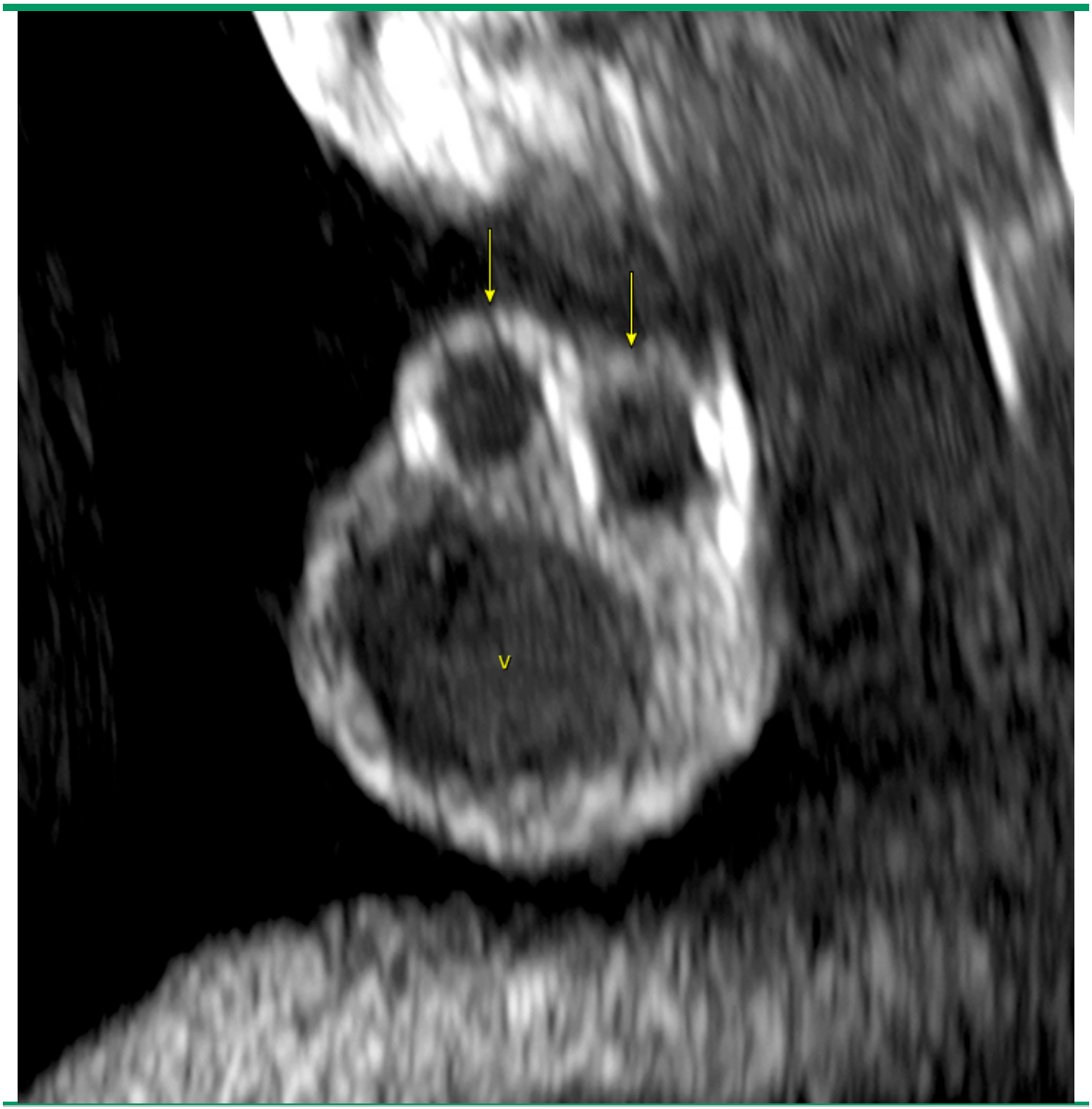
Umbilical vein to umbilical artery ratio with single umbilical artery



Schematic drawing shows the compensatory increase in size of the single umbilical artery. In the normal three-vessel umbilical cord the circulation is theoretically distributed equally in two arteries (left), resulting in an umbilical vein to umbilical artery ratio (UV/UA) greater than 2. In fetuses with single umbilical artery (SUA) the size increase of the artery results in a UV/UA ratio lower than 2.

Graphic 63821 Version 3.0

Normal three-vessel umbilical cord

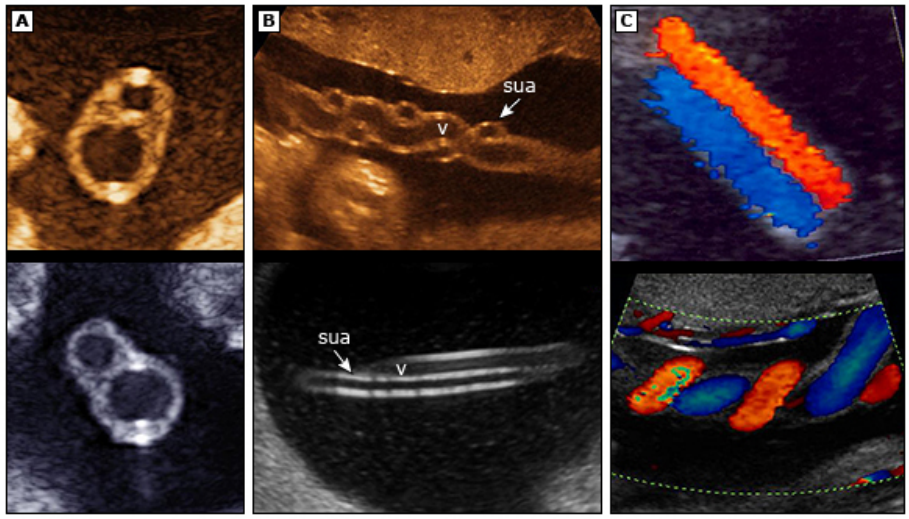


Cross-sectional ultrasound image of a normal three-vessel umbilical cord. The arrows point to the two umbilical arteries.

v: single umbilical vein.

Graphic 109920 Version 1.0

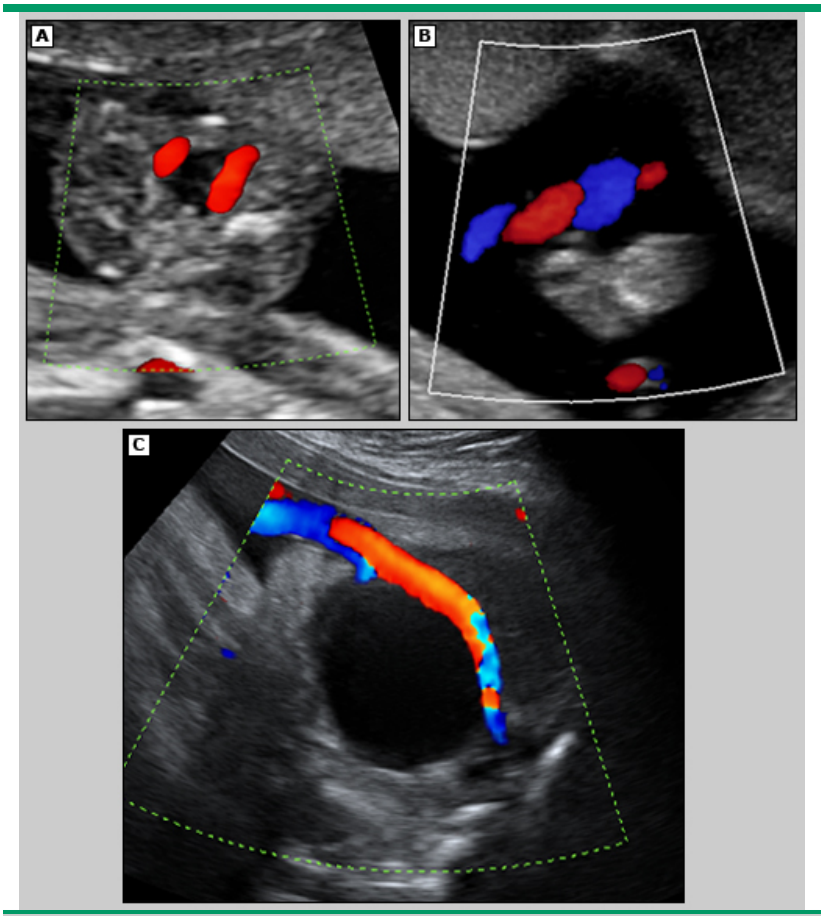
Prenatal ultrasound images of single umbilical artery



(Panel A) Single umbilical artery. Transverse section of a two-vessel umbilical cord in two different fetuses shows the typical "soda can tab" sign.
(Panel B) Single umbilical artery. Longitudinal views of a two-vessel umbilical cord as depicted by two-dimensional ultrasound show the single umbilical artery (sua) and the vein (v). Upper panel, fetus with a coiled umbilical cord. Lower panel, fetus with a straight, non-coiled umbilical cord.
(Panel C) Single umbilical artery. Color Doppler ultrasound shows a straight, non-coiled two-vessel umbilical cord (upper panel) and a coiled two-vessel umbilical cord (lower panel).

Graphic 73747 Version 3.0

Color Doppler images of two-vessel umbilical cord

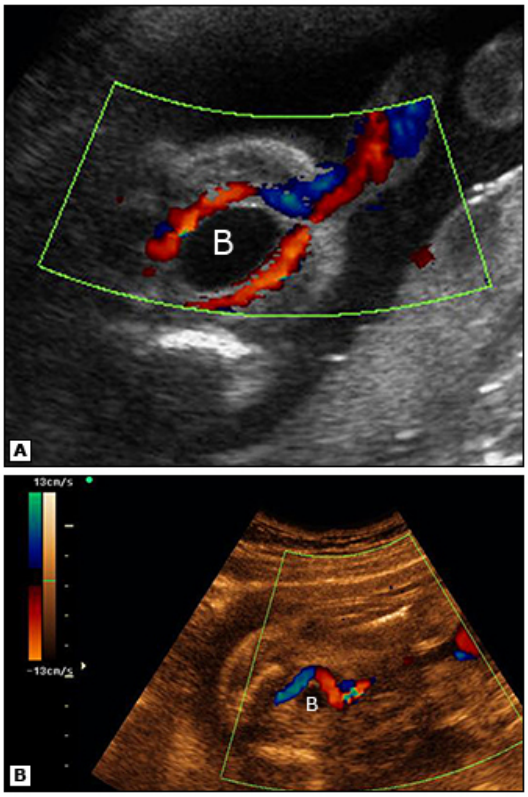


(A) Normal three-vessel umbilical cord. Color Doppler shows presence of two umbilical arteries, one on each side of the bladder.
(B) Two-vessel umbilical cord. Color Doppler images show alternating single artery and vein.
(C) Two-vessel umbilical cord. Transverse view at the level of the fetal bladder shows an artery on only one side of the fetal bladder.

Courtesy of Deborah Levine, MD.

Graphic 56497 Version 9.0

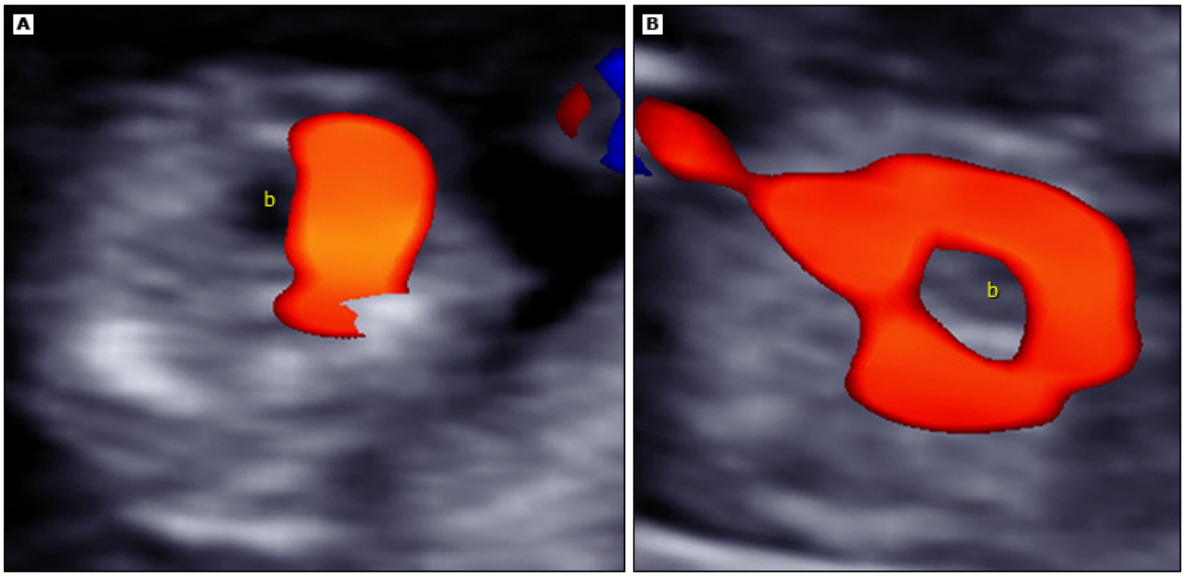
Color Doppler of intraabdominal single umbilical artery



Color Doppler ultrasound depicting the intra-abdominal segment of the umbilical arteries running along side of the fetal bladder (B).
(Panel A) Normal two umbilical arteries in a fetus with a three-vessel cord.
(Panel B) Only a single artery is seen in a fetus with a two-vessel cord.

Graphic 72664 Version 2.0

First-trimester color Doppler image of one versus two (normal) umbilical arteries

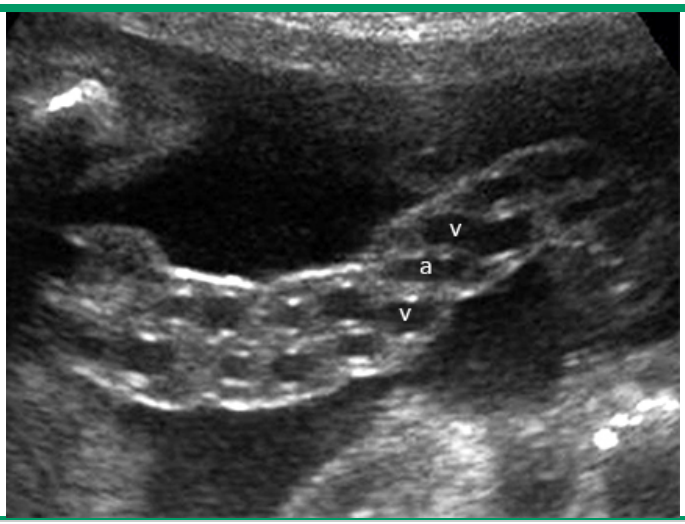


First-trimester color flow Doppler image showing (A) a single umbilical artery alongside the fetal bladder and (B) two umbilical arteries with the bladder between them.

b: bladder.

Graphic 109922 Version 1.0

Prenatal ultrasound image of a single umbilical artery in a hypercoiled umbilical cord

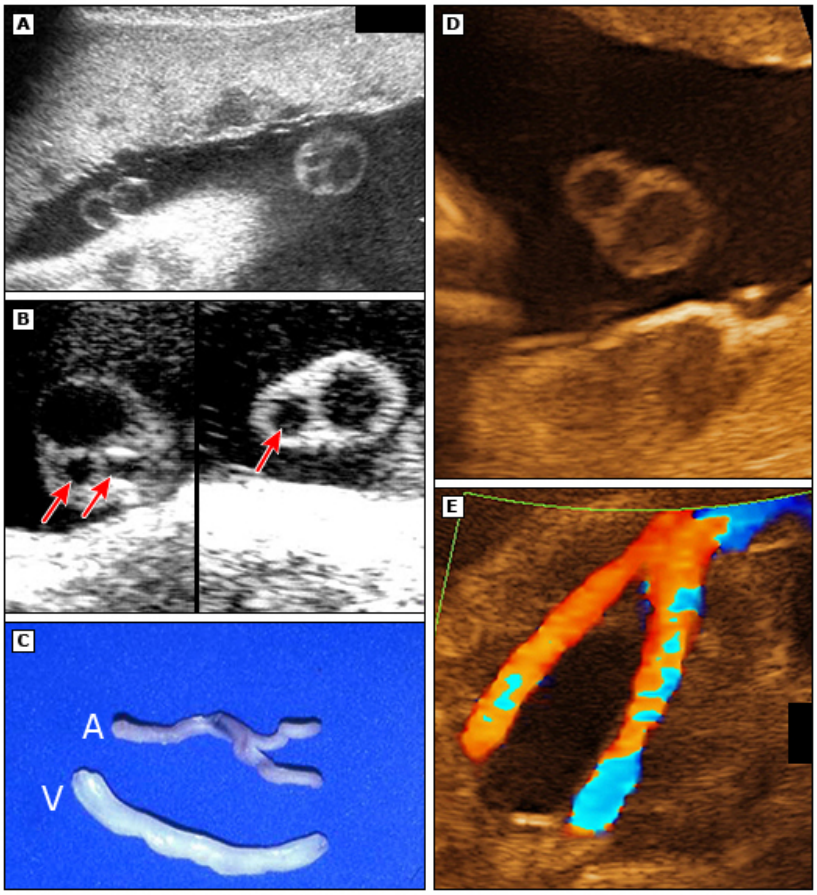


Conventional two-dimensional ultrasound shows a potential pitfall in the diagnosis of single umbilical artery. The diagnosis can be easily missed in hypercoiled umbilical cords.

v: vein; a: single umbilical artery.

Graphic 79742 Version 3.0

Fused umbilical arteries

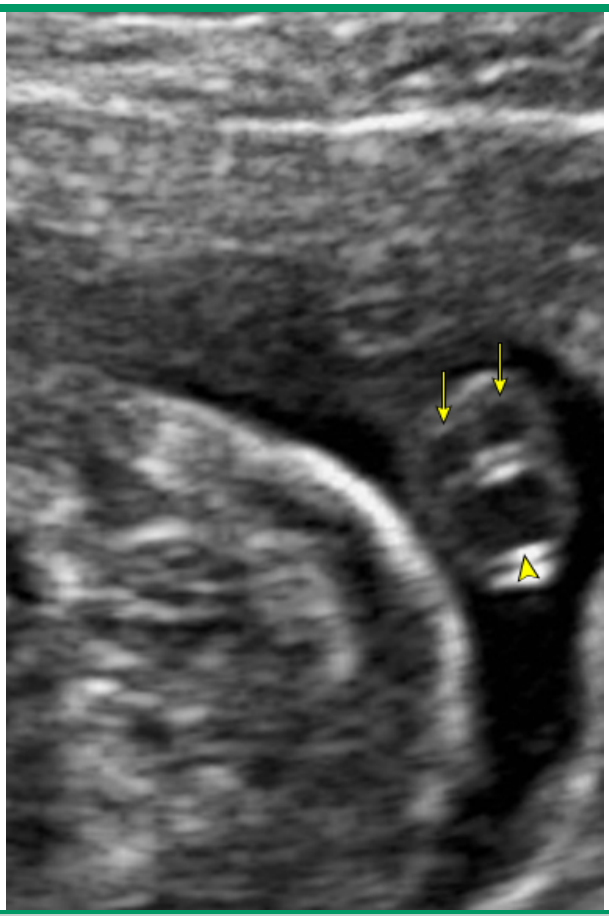


(Panels A, B, and C) Fused umbilical arteries.
(Panel A) Simultaneous view of an umbilical cord with a fused umbilical artery. The segment closest to the placenta shows a two-vessel cord, while the segment closest to the fetus shows the normal three-vessel cord.
(Panel B) Dual view of the umbilical cord in a fetus with fused umbilical arteries shows a segment with three vessels (left) and two vessels (right). At delivery, the exact site at which the arteries fused is shown. The Wharton's jelly was removed after fixation with formalin.
(Panels D and E) Dual view of the umbilical cord from a fetus with fused umbilical cord. The umbilical cord has two vessels. However, the normal two intra-abdominal arteries are depicted with Color Doppler ultrasound.

A: umbilical artery; V: umbilical vein.

Graphic 51055 Version 3.0

Ultrasound image of a normal three-vessel umbilical cord



Cross-sectional image of the umbilical cord at 18 weeks of gestation shows two arteries (arrows) and one vein (arrowhead).

Courtesy of Deborah Levine, MD.

Graphic 78465 Version 6.0

Contributor Disclosures

Martin L Gimovsky, MD Nothing to disclose **Eva Tejero Rosa, MS, CGC** Nothing to disclose **Waldo Sepulveda, MD** Nothing to disclose **Deborah Levine, MD** Nothing to disclose **Louise Wilkins-Haug, MD, PhD** Nothing to disclose **Vanessa A Barss, MD, FACOG** Nothing to disclose

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