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MFM Consult: Single umbilical artery: What you need to know

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Q: A 23-year-old woman (gravida 1,para 0) is found to have a fetus with a single umbilical artery at the time of initial ultrasound, which was performed at 18 weeks' gestation. What other abnormalities are associated with a single umbilical artery?

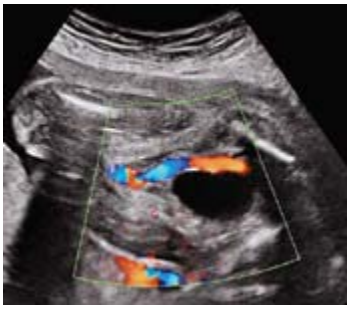


Figure Color Doppler image of single umbilical artery around the fetal bladder.

A: The finding of a single umbilical artery (SUA) can be a source of concern for both patients and providers. The normal umbilical cord contains 2 arteries and 1 vein; SUA is the result of atrophy or agenesis of 1 of the arteries. An SUA can usually be detected on cross-section of the umbilical cord during an otherwise routine gray-scale, second-trimester ultrasound exam, usually after 18 weeks' gestation. An SUA can also be detected using color-flow Doppler to examine the umbilical arteries in the pelvis (Figure) even at an earlier gestational age (eg, 14 weeks). [1] The incidence of SUA is 0.25% to 1% of all singleton pregnancies and up to 4.6% of twin gestations. [2]

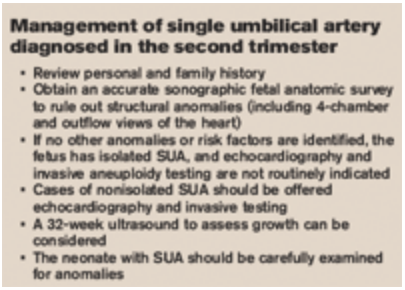
An isolated SUA with no other structural or chromosomal abnormalities should be distinguished from an SUA that is present with other abnormalities. The rate of associated fetal structural anomalies when an SUA is detected has been reported to range from 13% to 56%. [3] The most common associated anomalies have been noted to occur in the renal, cardiovascular, gastrointestinal, and central nervous systems. Genetic syndromes that may feature an SUA include VATER complex (a group of congenital anomalies consisting of vertebral defects, imperforate anus, tracheoesophageal fistula, and radial and renal dysplasia), Meckel-Gruber, and Zellweger. Teratogenic exposures such as maternal hyperglycemia and phenytoin have also been associated with SUA.

When other abnormalities are present, there is an increased likelihood of chromosomal abnormalities as well as other syndromes. Aneuploidy occurs in about 9% (range, 0%-26%) of fetuses with SUA when it is associated with other anomalies. [4] Trisomies 21 and 18 have been reported in prenatal case series of SUA, but Turner syndrome, triploidy, and chromosomal deletions and rearrangements have also been reported in babies with SUA.

If there are no other evident fetal abnormalities, how should this patient be further evaluated? Is additional testing indicated if additional fetal abnormalities are identified?

Once the diagnosis of a SUA has been made, a complete evaluation should be performed. The patient's personal and family histories should be reviewed, including any previous pregnancy complications, medication or teratogen exposure, other children with congenital anomalies, and any genetic disorders in the maternal and paternal families. Any aneuploidy screening (such as first-trimester screen, quadruple

screen) that the patient has undergone during the pregnancy should be reviewed. If the patient has not undergone screening and the timing is appropriate, it should be offered.



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A targeted ultrasound anatomic survey should be performed to identify any other anomalies that may be present. Special attention should be paid to the cardiac, genitourinary, gastrointestinal, and central nervous systems. Four chambers and outflow cardiac views should be obtained in all cases of SUA. If the ultrasound reveals only SUA with no other anomalies, the fetus has an isolated SUA. The extent of further testing when there is an apparently isolated SUA is controversial. If there is clinical confidence that the SUA is isolated, it appears that routine fetal echocardiogram is of little added benefit. [3] Similarly, because the incidence of chromosomal abnormalities is not increased in fetuses with an isolated SUA (<1%), invasive genetic testing is not routinely indicated. [3,4-6] Physical exam may detect anomalies: up to 7% of fetuses diagnosed with a suspected isolated SUA in one series had anomalies detected postnatally. [7]

In cases when SUA is not isolated (ie, associated with anomalies on ultrasound), further prenatal diagnosis should be considered. Fetal echocardiogram is indicated if the SUA is not isolated, if any abnormality is suspected on initial cardiac imaging, or if there are other indications. Fetal karyotype assessment should be offered to patients with nonisolated SUA, those with abnormal genetic screening, or with intrauterine growth restriction (IUGR).

If initial evaluation after identification of the single umbilical artery is reassuring, how should the patient's pregnancy be managed?

Perinatal management of an isolated SUA is still debated. An isolated SUA has been suggested to increase the risk of perinatal morbidity, such as intrauterine growth restriction and possibly perinatal mortality. [5-10] Despite all of these associations, the finding of an SUA is nonspecific and is most commonly seen in a normal fetus.

Although most fetuses with an SUA have an uncomplicated antepartum course, some studies have reported an increased risk of IUGR when an isolated SUA is present. However, this association remains controversial. Although some recent studies suggest that an isolated SUA does not actually place the fetus at increased risk for IUGR, [8,9] other studies have shown an increased risk for IUGR. [5-10] Wiegand et al reviewed serial growth ultrasounds on 138 fetuses with an SUA and reported 4 fetuses (2.9%) with IUGR, which was within the expected range for an unselected population. [8] Bombrys et al reviewed 255 fetuses with an SUA and 289 fetuses without an SUA, and noted similar rates of small-for-gestational-age infants between the groups. [9] Both studies concluded that fetuses with an SUA were not at increased risk for IUGR. Burshtein et al compared 243 fetuses with an SUA with more than 194,500 fetuses having normal umbilical vessels. They noted that fetuses with an SUA had an increased risk of IUGR, polyhydramnios, oligohydramnios, placental abruption, cord prolapse, and perinatal mortality after controlling for other confounders. [10] Most (15 of 18) of the fetal deaths in a large (n=354) series of isolated SUAs with karyotyping occurred in IUGR SUA fetuses. [4] These data suggest that if IUGR (especially <5% of estimated fetal weight) is detected, perinatal morbidity and mortality may increase. [4,10]

Given the inconsistency of the data, it seems reasonable that a single scan at approximately 32 weeks for assessment of fetal growth be considered for fetuses with an isolated SUA. A process for managing SUA diagnosed in the second trimester accompanies this article.

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member of the Publications Committee (see list of 2010 members on www.smfm.org) have a conflict of interest, financial or otherwise, to disclose with regard to the content of this article.

Disclaimer: The practice of medicine continues to evolve and individual circumstances will vary. This opinion reflects information available at the time of acceptance for publication and is not designed nor intended to establish an exclusive standard of perinatal care. This publication is not expected to reflect the opinions of all members of the Society for Maternal-Fetal Medicine.

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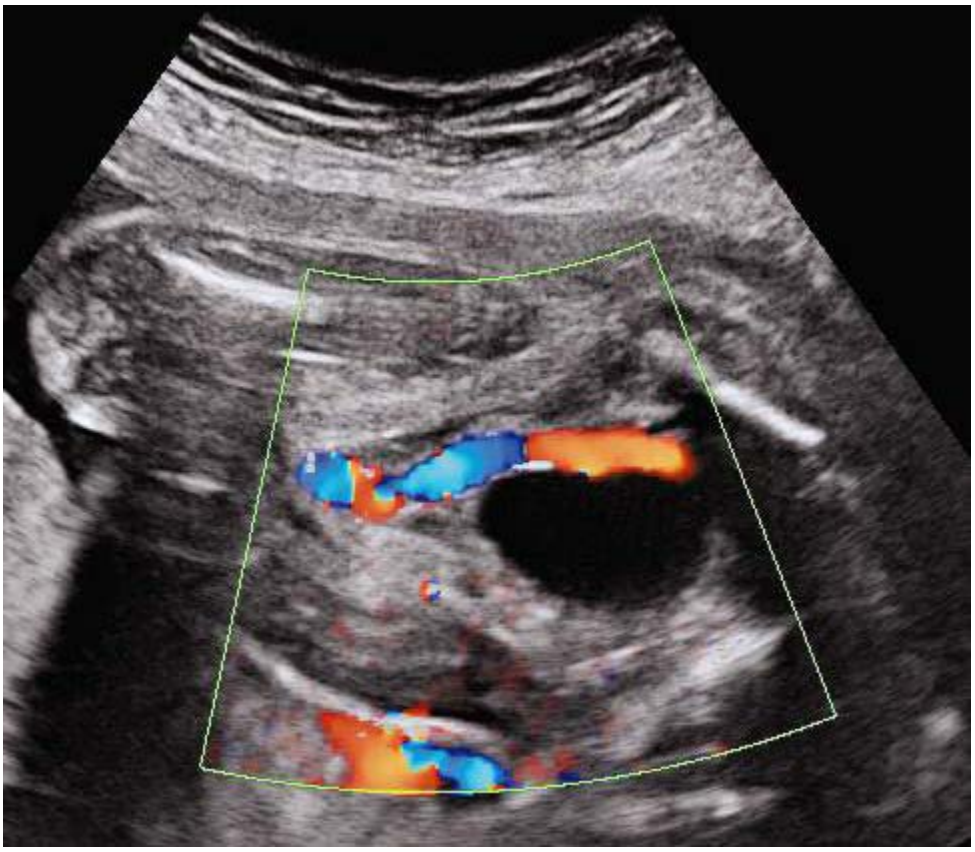

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- Review personal and family history
- Obtain an accurate sonographic fetal anatomic survey to rule out structural anomalies (including 4-chamber and outflow views of the heart)
- If no other anomalies or risk factors are identified, the fetus has isolated SUA, and echocardiography and invasive aneuploidy testing are not routinely indicated
- Cases of nonisolated SUA should be offered echocardiography and invasive testing
- A 32-week ultrasound to assess growth can be considered
- The neonate with SUA should be carefully examined for anomalies

Figure Color Doppler image of single umbilical artery around the fetal bladder.
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