

Single Umbilical Artery

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Case Scenario

A 23-year-old primigravida underwent a level-2 antenatal ultrasonography (USG) screening at 20 weeks of gestation. The USG revealed the presence of an isolated single umbilical artery (SUA) in the umbilical cord. No other fetal anomaly was identified on detailed evaluation.

Definition

SUA (Figure) is the commonest umbilical abnormality and one of the most common congenital anomalies, reported in 0.2% to 1.2% of live births.¹ The incidence is high in twins (4%–11%), abortuses, and autopsy series, with female co-twins being commonly affected.^{2–4} The incidence is even higher in twins born from assisted

reproductive technologies. The absence of left artery is more common than the absence of right artery, and 10% of SUAs have been reported to be associated with congenital anomalies.⁵

Etiology/Pathogenesis

The rudimentary umbilical cord forms during the 4th- to 8th-week of gestation and develops completely by the 12th week. The normal cord contains 2 arteries and a single vein embedded within the Wharton jelly.⁶ The development of SUA has 3 theories, namely, primary agenesis, secondary atresia of a previously normally developed vessel, and the persistence of original allantoic artery of the body stalk. In a majority of cases, the left artery is absent possibly because of secondary atresia, as suggested by histologically identifiable

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Figure. Ultrasound Doppler Image of a 2-Vessel Cord With Single Umbilical Artery

Courtesy: Dr Bheemashanker Nandyal, Shardha Scan Center, Bagalkot, Karnataka

remnants of a second artery.⁷ The right artery is usually larger than the left; this asymmetry may explain propensity of the right SUA.⁸ Pre-eclampsia, chronic hypertension, smoking, intake of alcohol, diabetes, obesity, epilepsy, polyhydramnios, oligohydramnios, twin or multiple gestations, and assisted reproductive technologies have all been shown to increase the risk of SUA.^{4,9}

Significant Associations

SUA has been found to be associated with monozygous twins, sirenomelia, vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, limb abnormalities (VACTERL) association, fetal hydantoin syndrome, Meckel–Gruber syndrome, Jarcho–Levin syndrome, multiple lentiginos syndrome, trisomy 13, trisomy 18, and Zellweger syndrome. Chromosomal abnormalities are reported in 8% to 11% of fetuses with SUA. Commonly reported chromosomal anomalies are trisomies 13 and 18 and rarely trisomy 21 and monosomy 45X.^{4,10} The intrauterine growth retardation (IUGR) has been reported to be more common in fetuses with SUA (the prevalence is 2 times higher even in isolated SUA); this increased prevalence may be because of placental insufficiency. Preterm labor, fetal and neonatal death, placental anomalies, abnormal umbilical cord insertions, and

other structural anomalies (genitourinary, cardiovascular, and musculoskeletal system abnormalities) are the other conditions associated with SUA. Overall, the perinatal mortality in SUA pregnancies may be as high as 20%.¹¹

Antenatal Diagnosis and Management

The guidelines by the International Society of Ultrasound in Obstetrics and Gynecology (2013) recommend that the number of cord vessels, cord insertion to the umbilicus, and cysts in the umbilical cord should be checked for during the first-trimester ultrasound. The guidelines also suggest to evaluate the fetal paravesical area briefly with power or color Doppler to identify the presence of 2 umbilical arteries.¹² However, this procedure is not followed regularly because the sensitivity and positive predictive value of the first-trimester sonography to detect SUA is poor.¹³ Hence, the American Institute of Ultrasound in Medicine recommends evaluation of the umbilical cord during routine prenatal examinations in the second and third trimesters.¹⁴

After the diagnosis of SUA, it is important to look for other structural malformations in the fetus (especially genitourinary, cardiovascular, and musculoskeletal system malformations). If SUA is detected, a thorough fetal heart scan (ie, 4-chamber view, outflow tracts, and 3-vessel view) on ultrasound is warranted. If ultrasound is unable to scan completely or associated malformations are noticed in the scan, an anatomy scan and fetal echocardiography should be done from a specialized center. However, if the second trimester anatomy scan is performed efficiently and only an isolated SUA is detected, a fetal echocardiography is not required.¹⁵ Invasive testing with chromosomal evaluation (microarray) is recommended only if associated malformations are detected and is not done routinely for isolated SUA.^{16,17} It is pertinent to mention that the maternal serum pregnancy-associated plasma protein A (PAPP-A) level in the first trimester and alpha-fetoprotein (AFP) level in the second trimester increase in isolated SUA pregnancies. This must be taken into account while

interpreting results of these tests in SUA pregnancies. Other components of quadruple test in the second trimester generally remain unaffected by isolated SUA.¹⁸

The risk of IUGR in isolated SUA increases by at least 2 times vis-à-vis a 3-vessel cord, while the risk of placental and amniotic fluid anomalies increases by 2 to 4 times.¹⁶ Because of the high risk of IUGR, preterm labor, and adverse fetal outcomes, even isolated SUA pregnancies should be frequently evaluated.⁹ A growth scan is recommended for fetuses with isolated SUA in 28 to 32 weeks and 32 to 36 weeks of gestation. If SGA/IUGR and/or other placental/cord anomalies are observed, the follow-up should be individualized and more frequent. For normal-weight fetuses with isolated SUA and normal cord insertion, delivery should be managed as per the standard obstetric indications. Neonates born with isolated SUA do not require any special neonatal care for the condition per se, and the long-term outcomes can be expected to be the same as in those neonates with 3-vessel cord.¹⁶ Although some studies suggest a screening renal ultrasound, with or without a micturating cystourethrogram, to rule out occult renal abnormalities and vesicoureteric reflux in isolated SUA, large prospective and retrospective series refute the clinical benefit of this practice.¹⁹⁻²¹ The odds of incidence of inguinal hernias may be high in later life in the isolated SUA population.²²

Conclusion

SUA affects 0.2% to 1.2% of all live births, with a high incidence in twins and abortuses. The second and third trimester ultrasound scans should routinely include umbilical cord evaluation. Once SUA is diagnosed, the fetus should be screened for other structural anomalies, including genitourinary, cardiovascular, and musculoskeletal defects. A detailed repeat scan, fetal echocardiography, and invasive tests are warranted to detect chromosomal abnormalities such as trisomies 13 and 18 only if an isolated abnormality is detected. The SUA may elevate maternal serum PAPP-A and AFP levels, hence the adjustments should be made for SUA before the results are interpreted. Even in the absence of other congenital anomalies, because of the high incidence of IUGR, preterm labor, and adverse fetal outcomes,

isolated SUA pregnancies should be kept under frequent surveillance.

Recommendations and Practice Points

1. All routine second and third trimester ultrasound scans should include umbilical cord evaluation.
2. Associated congenital anomalies are seen in 10% of fetuses with SUA. Hence, a detailed screening should always be done once SUA is detected. This should also include evaluating genitourinary, cardiovascular, and musculoskeletal defects.
3. A detailed repeat scan, fetal echocardiography, and invasive tests are warranted to detect chromosomal abnormalities such as trisomies 13 and 18 only after detecting an associated abnormality. Routine chromosomal analysis is not recommended for isolated SUA.
4. The fetal surveillance, including growth scans at 28 to 32 weeks and 32 to 36 weeks of gestation age, should be done repeatedly even in isolated SUA pregnancies.
5. Routine postnatal renal ultrasound and/or micturating cystourethrogram are not indicated for neonates born with antenatal isolated SUA.
6. Neonates born with isolated SUA should be provided routine neonatal care, and the long-term outcome can be expected to be the same as those neonates with 3-vessel cord.

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