# Fetuses with single umbilical artery: analysis of 45 cases

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**Summary**

*Objective:* The aim of this study was to analyze cases and determine the clinical significance of a prenatally detected single umbilical artery (SUA) in our population. *Materials and Methods:* All second and third trimester sonographic examinations carried out between January 2004 and September 2007 in our perinatology unit were reviewed. The postnatal results of the fetuses with SUA were obtained from the medical records and the patients. *Results:* From a total of 5,620 pregnant patients who were examined by ultrasound (US) scan between 15-36 weeks, a single umbilical artery was found in 45 cases, representing an incidence of 0.8%. Of these, 20 (45%) also presented with other malformations. There were six neonatal deaths, one fetal demise, and six terminations of pregnancy due to severe malformations in this group. Three cases with associated anomalies underwent surgery and one case required intensive care in the neonatal period. The only cytogenetic abnormality was trisomy 18 in one case. Six of 45 fetuses (13%) with single umbilical arteries had abnormal echocardiographic findings. In two of the fetuses associated anomalies (cleft palate and esophageal atresia) were detected after birth. In pregnancies without associated anomalies no aneuploidy was found and they were completely normal at birth and during the neonatal period. *Conclusions:* Scanning the umbilical cord is one of the essential parts of US examination. As the rate of cardiac malformations seen with single umbilical arteries is high, fetal echocardiography should be performed in suspected cases. The newborn should be reexamined immediately after birth due to the possibility of undetected anomalies.

*Key words*: Single umbilical artery; Prenatal diagnosis; Ultrasound; Congenital malformation; Fetal echocardiography.

## Introduction

The umbilical cord normally contains three vessels; two arteries and one vein. The reported incidence of single umbilical artery (SUA) is 1.5% in spontaneous abortuses, and 0.5 to 2.5% of uncomplicated neonates [1]. This is one of the most common congenital malformations with an incidence of approximately 1% of all deliveries [2].

These fetuses have been shown to have a high rate of structural abnormalities, ranging from 18 to 68% [3, 4]. Chromosomal abnormalities are reported in 8-11% of fetuses with SUA, particularly with trisomies 13 and 18, whereas trisomy 21 does not appear to be associated with this anomaly [4, 5]. In a recent study all chromosomally abnormal fetuses with SUA were found to have associated malformations detected by ultrasound (US) [6]. SUA was also reported to be associated with increased risk of fetal growth retardation, prematurity, and increased perinatal mortality rate [7].

We evaluated the associated anomalies and perinatal outcome in fetuses with a SUA detected on US scanning in our clinic.

## Materials and Methods

All of the records from detailed sonographic examinations at second and third trimester in fetuses of low- and high-risk pregnant women between January 2004 and July 2007 were reviewed.

We followed by scanning the fetus a standard protocol that included images of the central nervous system, spine, heart, diaphragm, stomach, kidneys, bladder, umbilical cord and cord insertion, and extremities. Cardiac outflow tracts were also routinely imaged and suspected cases were referred to echocardiography. Patients are counseled concerning the findings and offered amniocentesis or cordocentesis to assess fetal karyotype.

The presence of a single umbilical artery was suspected when a cross-sectional image of the umbilical cord demonstrated only two vessels. We used color Doppler US to confirm the diagnosis of a single umbilical artery, with a transverse view of the fetal pelvis showing only one umbilical artery around the fetal bladder.

Fetuses were classified into groups: the first group with an isolated SUA, the second one with a SUA and congenital malformations. Maternal and neonatal data were obtained by a review of the medical records and from the patients. Umbilical artery and associated abnormalities were confirmed after delivery in all cases.

## Results

During the study period 5,620 women were examined by US scan between 15-36 weeks of gestation; 45 were found to have a single umbilical artery (0.8%). The mean gestational age at the time of the first examination was 22 weeks/3 days.

In 25 (55%) cases we observed SUA as an isolated finding. One fetus in this group was lost in follow-up and one of the pregnancies is continuing. In the group of 23 fetuses without additional anomalies the mean birthweight was 3,031 ± 477 g and all children were phenotypically healthy at birth. Two cases were under 2,500 g at term.

Twenty of the fetuses were found to have associated anomalies at pre and postnatal examinations. One of these pregnancies is still continuing. Cytogenetic abnor-

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maly (trisomy 18) was diagnosed in one of the 13 (7.7%) fetuses in the group with anomalies. There were six neonatal deaths and six terminations of pregnancy (one fetus with trisomy 18; five fetuses with severe non-chromosomal malformations) in this group. Two cases were operated due to cardiac defects, and one of them died after one year. One case with undetected esophageal atresia was also operated during the neonatal period. Two cases with minor anomalies required intensive care after birth.

Six (42%) of these cases with SUA had abnormal echocardiographic findings. Cleft palate and esophageal atresia were detected after birth in two of the fetuses. Type of anomalies, karyotype and outcomes of the fetuses with associated anomalies are listed in Table 1.

Table 1. — *Outcomes and karyotypes of the cases with associated anomalies.*

| GW | Types of anomalies | Outcome | Karyotype |
|---|---|---|---|
| 18 | VSD, CPC | TOP | Trisomy |
| 24 | VSD | Live birth, operated | Normal |
| 34 | VSD, Hydrocephaly | Live birth, NND | Normal |
| 20 | Atrio ventricular septal defect | Operated, death after 1 year | Normal |
| 33 | Single ventricle | Live birth, NND | Normal |
| 19 | VSD, Acrania, Spina bifida | TOP | – |
| 22 | CPC | Live birth, NICU | Normal |
| 20 | CPC, EIF, PE | TOP | Normal |
| 24 | CPC | 32 weeks of gestation | – |
| 23 | DWM, Ventriculomegaly | Live birth, NND | Normal |
| 28 | Anencephaly | TOP | – |
| 20 | Anencephaly-spina bifida | TOP | – |
| 20 | Hydrocephaly | Live birth, NND | – |
| 17 | Exstrophy of the bladder | TOP | Normal |
| 20 | Mild renal pelviectasy | Live birth | Normal |
| 26 | Omphalocele | Live birth, NND | Normal |
| 32 | Gastroschisis | Live birth, NND | – |
| 21 | Small bowel atresia | Fetal demise | – |
| 24 | Esophageal atresia*, short femur | Live birth, operated | Normal |
| 20 | Cleft palate* | Live birth, NICU | Normal |

GW: gestational week; CPC: Choroid plexus cyst; EIF: Ecogenic intracardiac focus; PE: Pes equinovarus; VSD: Ventricular septal defect; DWM: Dandy Walker Malformation; TOP: Termination of the pregnancy; NND: Neonatal death, NICU: Neonatal intensive care unit; * This anomaly was not detected prenatally.

## Discussion

In some pregnancies one of the umbilical arteries is absent due to either primary agenesis, atrophy of one of the arteries or the persistence of the original allantoic artery in the body stalk of the embryo [8]. With the use of US, the presence of a SUA should be detectable in most pregnancies, even as early as 12 weeks of gestation [9]. Despite the methods available for the detection of a SUA [10, 11] and its relatively common occurrence, the antenatal detection rate is reported to be poor with only one-third of the cases identified in previous studies [12]. The availability of color Doppler in routine scanning has improved the detection rate. In a recent study the missing rate of a SUA between cases having sonographic examination was 38% [13].

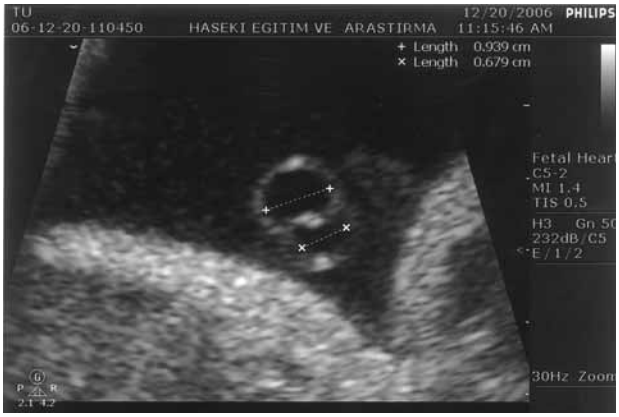


Figure 1. — Umbilical vein/artery < 2.

The reported rate at second trimester scanning is 0.7% [14]. In this study the incidence of a SUA was found to be 0.8% among second and third trimester fetuses at sonografic examination. However the real rate should be higher due to the fact that examination of the cord becomes lower in priority, if the anomalies are noted with US before the presence of a SUA. Therefore the reported rate of associated anomalies in previous studies has varied between 15.4 and 67% [14, 15].

The increase in the diameter of the umbilical artery relative to the umbilical vein has been reported by some authors [16]. They noticed that the diameter of the umbilical artery was larger then 50% of that of the umbilical vein, resulting in a vein to artery (V/A) ratio of < 2. In all of our cases with SUA the V/A ratio was < 2 (Figure 1). We confirmed the presence of SUA by using color Doppler.

According to the reports of several investigators, a variety of congenital anomalies have been associated with SUA, including cardiovascular malformations, central nervous system defects, gastrointestinal or urogenital defects, and musculoskeletal malformations [2, 17, 18].

Some authors have proposed that there should be an association between a SUA and other fetal anomalies. Their findings suggested a possible common underlying vascular pathogenetic factor, which explains the frequent concurrence of a SUA formation, limb reduction defects, atresias, and organ aplasias [19]. The cases with small bowel and esophageal atresia can be examples of this phenomenon.

In a series by Gornall *et al.* there was a preponderance of urogenital anomalies [20]. In our series only two of the fetuses (4%) were found to have anomalies of this system (exstrophy of the bladder and mild renal pelviectasy). In a recent study it was also concluded that it is not necessary to screen for renal anomalies in infants with a SUA without other anomalies seen at physical examination [21].

In our study population, the incidence of cardiac abnormalities among fetuses with a SUA was 13%. In three of these cases the cardiac defect was the only detected asso-

ciated anomaly. The incidence was 10.7% (3 of 28 cases) among an apparently isolated SUA. Budorick *et al.* reported an incidence of 5% and advised fetal echocardiography in all of these cases [22].

However as a result of another study with an incidence of 27.6% of cardiac anomalies among all of the cases with a SUA the authors concluded that fetal echocardiography may not be indicated as a routine part of evaluation of the fetus with an isolated SUA, unless the four-chamber view and outflow tracts are abnormal or cannot be obtained [23]. We also did not perform detailed cardiac examination in cases with normal findings at first scan.

SUAs had an incidence of 3.3% among cytogenetically abnormal pregnancies and was found in 77.8% of trisomy 18 cases [24]. Our single case was also trisomy 18; choroid plexus cysts and ventricular septal defects (VSDs) were additional anomalies. In this case all of these findings were missed at the first trimester scan and nuchal translucency was in normal range.

By second trimester scanning the presence of SUA implied a search for other anomalies. VSD was confirmed with echocardiography at 19 weeks of gestation.

In a review by Pierce *et al.* the rate of aneuploidy for fetuses with an apparent isolated SUA (n: 367) was 0.54%, and this rate increased to 19.9% for fetuses with other identifiable abnormalities (n: 161) [25]. The aneuploidy rate in our series in the group with associated anomalies was 7.7%. In cases with an isolated SUA, most authors do not recommend routine karyotype, as we did in our series.

Although the significantly higher perinatal mortality rate of fetuses with a SUA and no other obvious abnormality has been recognized by some authors [26, 27], prenatal sonography is found to be reliable in the identification of major concurrent anomalies, and no alteration in pregnancy management is recommended whatsoever if no other abnormalities are detected at sonography by others [28]. In two of our cases which required intensive care after birth, cleft palate and esophageal atresia were not detected prenatally and a choroid plexus cyst in the second trimester was the only finding. In two of the cases ventriculomegaly and hydrocephaly were late second and third trimester findings. The cases with an isolated SUA should be reexamined at late gestation and after birth due to the possibility of new occurring anomalies.

In a previous study fetuses with an isolated SUA were found to be at similar risk for being small for gestational age (SGA) compared with fetuses with 3-vessel umbilical cords. It was concluded that antepartum serial US examination does not provide more information for interval fetal growth assessment [29]. In our study the mean birthweight of these fetuses was 3,031 ± 477 g, and only two were SGA (8.6%).

## Conclusion

Examination of the umbilical arteries should be a part of first trimester scanning. If a SUA is prenatally detected, a detailed sonographic evaluation is necessary. Fetal echocardiography should also be performed in suspected cases. Although the routine protocols for follow-up should not be altered, all babies with SUA should be examined immediately after birth due to the possibility of undetected life-treating anomalies.

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