Ultrasound Obstet Gynecol (2010)
Published online in Wiley Online Library (wileyonlinelibrary.com). **DOI:** 10.1002/uog.7711

Isolated single umbilical artery: need for specialist fetal echocardiography?

D. DEFIGUEIREDO, T. DAGKLIS, V. ZIDERE, L. ALLAN and K. H. NICOLAIDES
Harris Birthright Research Centre for Fetal Medicine, King's College Hospital Medical School, London, UK

KEYWORDS: cardiac defect; fetal echocardiography; prenatal diagnosis; single umbilical artery; ultrasound

ABSTRACT

Objective *To examine the association between single umbilical artery (SUA) and cardiac defects and to determine whether patients with SUA require specialist fetal echocardiography.*

Methods *Incidence and type of cardiac defects were determined in fetuses with SUA detected at routine second-trimester ultrasound examination.*

Results *A routine second-trimester scan was performed in 46 272 singleton pregnancies at a median gestation of 22 (range, 18–25) weeks and an SUA was diagnosed in 246 (0.5%). Cardiac defects were diagnosed in 16 (6.5%) of these cases, including 10 (4.3%) in a subgroup of 233 with no other defects and in six (46.2%) of the 13 with multiple defects. In 11 (68.8%) of the 16 cases with cardiac defects the condition was readily diagnosable by evaluating the standard four-chamber view and the views of the great arteries. In the remaining cases there was left persistent superior vena cava or small ventricular septal defect, where prenatal diagnosis may not be important because they are not associated with adverse outcome.*

Conclusion *Although SUA is associated with an increased incidence of cardiac defects it may not be necessary to refer such patients for specialist fetal echocardiography because the defects are detectable by evaluating standard cardiac views that should be part of the routine second-trimester scan.* *Copyright © 2010 ISUOG. Published by John Wiley & Sons, Ltd.*

INTRODUCTION

A single umbilical artery (SUA) is found in about 0.5% of pregnancies and is associated with chromosomal defects and malformations of all major organ systems[1,2]. In previous studies on a combined total of 1038 cases of SUA diagnosed prenatally the prevalence of fetal abnormalities was 33.6% (Table 1)[3–15]. Consequently, the prenatal diagnosis of SUA should motivate the sonographer to undertake a systematic and detailed examination of the fetal anatomy for the diagnosis or exclusion of associated defects. In the reported series of SUA, the prevalence of cardiac defects was 11.4%, but it is not stated whether these were isolated or whether they were associated with other, more easily detectable, defects (Table 1)[3–15].

In this study we examined the association between SUA and cardiac defects with the aim of determining whether patients with SUA require specialist fetal echocardiography.

METHODS

All pregnant women booked for antenatal care and delivery in our hospital are offered two ultrasound scans, one at 11–13 weeks' gestation as part of screening for chromosomal defects[16,17] and another at 20–23 weeks for detailed fetal examination according to a standard protocol. All scans are carried out by sonographers who had obtained The Fetal Medicine Foundation certificate of competence in the 20–23-week scan (www.fetalmedicine.com). The standard examination includes the use of color-flow mapping in the fetal pelvis to visualize the two umbilical arteries and the diagnosis of SUA. In all cases of SUA, the fetal heart is examined either by an experienced fetal medicine consultant or by a fetal cardiologist. This includes, as a minimum, examination of the four-chamber view, outflow tracts and transverse arches. Demographic characteristics and ultrasound findings are recorded in a fetal database at the time of the examination. Data on pregnancy outcome were obtained from the hospital records.

We searched the fetal database to identify all patients with an SUA among those singleton pregnancies undergoing a routine second-trimester scan between January 2000 and December 2008.

Correspondence to: Prof. K. H. Nicolaides, Harris Birthright Research Centre for Fetal Medicine, King's College Hospital Medical School, Denmark Hill, London SE5 8RX, UK (e-mail: fmf@fetalmedicine.com)

Accepted: 28 May 2010

Copyright © 2010 ISUOG. Published by John Wiley & Sons, Ltd.

Table 1 Prenatal sonographic studies reporting on the incidence of cardiac and other abnormalities in fetuses with single umbilical artery

Reference	*Gestational age at scan (weeks)*	*Total* (n)	*Fetal abnormalities* (n *(%))*	*Cardiac defects* (n *(%))*
Abuhamad *et al.* (1995)[3]	25 (10–40)	77	20 (26.0)	9 (11.7)
Catanzarite *et al.* (1995)[4]	16–39	82	41 (50.0)	15 (18.3)
Blazer *et al.* (1997)[5]	15 (14–16)	46	6 (13.0)	1 (2.2)
Ulm *et al.* (1997)[6]	21 (16–41)	103	58 (56.3)	10 (9.7)
Chow *et al.* (1998)[7]	29 (16–41)	118	37 (31.4)	19 (16.1)
Geipel *et al.* (2000)[8]	21 (13–39)	102	43 (42.2)	15 (14.7)
Budorick *et al.* (2001)[9]	2nd trimester	57	26 (45.6)	5 (8.8)
Gossett *et al.* (2002)[10]	22.8 (17–28)	103	29 (28.2)	18 (17.5)
Gornall *et al.* (2003)[11]	19 (19–20)	107	20 (18.7)	7 (6.5)
Martinez-Payo *et al.* (2005)[12]	20 ($\geq$ 13)	40	6 (15.0)	1 (2.5)
Volpe *et al.* (2005)[13]	20 (17–22)	40	16 (40.0)	9 (22.5)
Granese *et al.* (2007)[14]	16–23	61	22 (36.1)	3 (4.9)
Lubusky *et al.* (2007)[15]	16–22	102	25 (24.5)	6 (5.9)
Total		1038	349 (33.6)	118 (11.4)

RESULTS

During the study period a routine second-trimester scan was performed in 46 272 singleton pregnancies at a median gestational age of 22 (range, 18–25) weeks. SUA was diagnosed in 246 (0.5%) and in this group the median maternal age was 34 (range, 16–44) years. One of the 246 women had diabetes mellitus and another had a family history of congenital heart defect (CHD). None of the women was taking any medications associated with CHD, such as antiepileptic drugs or lithium.

The 246 cases with SUA were divided into three groups. In the first group there were 223 cases in which no cardiac or extracardiac defects were identified prenatally, but in two of the live births from this group a ventricular septal defect was detected postnatally. In the second group there were 10 cases with no extracardiac defects but with a cardiac abnormality that was diagnosed prenatally. In the third group there were 13 cases with extracardiac defects (multiple, $n = 5$; spina bifida, $n = 2$; ventriculomegaly, $n = 2$; encephalocele, $n = 1$; hydrops, $n = 1$; unilateral multicystic kidney, $n = 1$; pelvic kidney, $n = 1$) and in six of these a cardiac abnormality was diagnosed prenatally.

In total, cardiac defects were diagnosed in 16 (6.5%) cases, including 10 (4.3%) of 233 with no other defects (Groups 1 and 2) and in six (46.2%) of the 13 with at least one extracardiac defect (Table 2).

In the 223 cases of isolated SUA, there were 214 (96.0%) live births, three neonatal deaths (one due to birth asphyxia and two due to prematurity) and six intrauterine deaths (five due to fetal growth restriction and one unexplained). In the group of 23 cases of SUA with prenatally detected fetal defects, there were 12 (52.2%) live births, seven terminations of pregnancy at the request of the parents and four intrauterine deaths.

In 185 (75.2%) of the 246 cases of SUA the patients had first-trimester screening for chromosomal defects in addition to the second-trimester scan. The incidence of cardiac or extracardiac defects in the second-trimester scan was 4.3% (8 of 185) in those with first-trimester screening and 24.6% (15 of 61) in those without (chi^2 test $P < 0.001$).

Table 2 Cardiac defects in fetuses with single umbilical artery in the presence and absence of other defects classified according to the standard sonographic view necessary for prenatal diagnosis

Cardiac defect	*No other defects* (n = *233)*	*Other defects* (n = *13)*
Detectable in the four-chamber view		
Atrioventricular septal defect	1 (0.4)	—
Atrioventricular valve dysplasia	—	1 (7.7)
Coarctation of the aorta	3 (1.3)	1 (7.7)
Detectable in the great artery view		
Tetralogy of Fallot	1 (0.4)	2 (15.4)
Double outlet right ventricle and pulmonary atresia	1 (0.4)	—
Transposition of the great arteries	1 (0.4)	—
Requires specialist echocardiography		
Ventricular septal defect	2 (0.9)	2 (15.4)
Left superior vena cava	1 (0.4)	—
Total	10 (4.3)	6 (46.2)

Data are given as *n* (%).

DISCUSSION

The findings of this study confirm the previously reported association between SUA and cardiac defects[3–15]. The incidence of cardiac defects was much higher in the presence of extracardiac abnormalities than in cases without other abnormalities (46.2% vs. 4.3%).

The overall incidence of cardiac defects in fetuses with SUA inevitably depends on the design of the study and whether the patients included constitute a routinely screened unselected population, as in our study, or whether it includes patients referred to a specialist center from a routine service following the diagnosis of cardiac or other defects. Another important factor that influences both the incidence of SUA and the coincidence of cardiac as well as other defects in the second trimester is the proportion of pregnancies undergoing first-trimester screening. Early diagnosis of chromosomal and other major defects often results in termination of pregnancy and consequently a substantial reduction in the incidence

Copyright © 2010 ISUOG. Published by John Wiley & Sons, Ltd.

of such abnormalities in the second trimester. A study of pregnancies undergoing chorionic villus sampling at 11–14 weeks' gestation reported that the incidence of SUA was 5.9%, which is substantially higher than the incidence of 0.5% in our patients, and that half of the fetuses with SUA had chromosomal defects, mainly trisomy 18, which is strongly associated with cardiac and multiple other defects[18]. In our study the incidence of fetal defects in the second-trimester scan was approximately six times higher in those who had not had first-trimester screening than in those who had (24.6% vs. 4.3%).

The pattern of cardiac defects was similar in the groups with and without other abnormalities and was also similar to that reported in previous studies of fetuses with SUA[3–15]. In two-thirds of the cases the cardiac defect was readily diagnosable by evaluating the standard views of the four chambers and great arteries. In the case of left persistent superior vena cava and small ventricular septal defect diagnosis of the defect would require a more detailed scan. However, prenatal diagnosis of such defects may not be important because they are not associated with adverse outcome.

In all cases with SUA the fetal heart was examined either by an experienced fetal medicine consultant or by a fetal cardiologist. However, a limitation of the study, which may have resulted in underestimation of the incidence of cardiac defects, is that the neonates had routine clinical, rather than detailed cardiological, examination.

The main issue raised by our findings is whether patients with SUA detected at the routine second-trimester scan should be referred for specialist fetal echocardiography. Certainly the incidence of cardiac defects in such patients (4.3%) is substantially higher than in patients with a family history of cardiac defects and diabetes mellitus (about 2%), which are widely accepted as indications for fetal echocardiography. The alternative argument is that examination of the fetal heart should be an integral part of the routine second-trimester scan and that sonographers undertaking such an examination should be competent in obtaining and evaluating the four-chamber view and outflow tracts[19]. Consequently, detection of an SUA should alert the ultrasonographer to examine more carefully the standard cardiac views and refer for specialist echocardiography only those with a suspected abnormality. If the routine scan does not include appropriate examination of the heart, patients with SUA should be referred to a specialist in fetal echocardiography.

ACKNOWLEDGMENT

This study was supported by a grant from The Fetal Medicine Foundation (Charity No: 1037116) and is part of the PhD thesis of D. DeFigueiredo, Universidade de Brasilia, Brasilia, Brazil.

REFERENCES

1. Leung AK, Robson WL. Single umbilical artery. A report of 159 cases. *Am J Dis Child* 1989; **143**: 108–111.
2. Thummala MR, Raju TN, Langenberg P. Isolated single umbilical artery anomaly and the risk for congenital malformations: a meta-analysis. *J Pediatr Surg* 1998; **33**: 580–585.
3. Abuhamad AZ, Shaffer W, Mari G, Copel JA, Hobbins JC, Evans AT. Single umbilical artery: does it matter which artery is missing? *Am J Obstet Gynecol* 1995; **173**: 728–732.
4. Catanzarite VA, Hendricks SK, Maida C, Westbrook C, Cousins L, Schrimmer D. Prenatal diagnosis of the two-vessel cord: implications for patient counseling and obstetric management. *Ultrasound Obstet Gynecol* 1995; **5**: 98–105.
5. Blazer S, Sujov P, Escholi Z, Itai BH, Bronshtein M. Single umbilical artery – right or left? Does it matter? *Prenat Diagn* 1997; **17**: 5–8.
6. Ulm B, Ulm M, Deutinger J, Bernaschek G. Umbilical artery Doppler velocimetry in fetuses with a single umbilical artery. *Obstet Gynecol* 1997; **90**: 205–209.
7. Chow JS, Benson CB, Doubilet PM. Frequency and nature of structural anomalies in fetuses with single umbilical arteries. *J Ultrasound Med* 1998; **17**: 765–768.
8. Geipel A, Germer U, Welp T, Schwinger E, Gembruch U. Prenatal diagnosis of single umbilical artery: determination of the absent side, associated anomalies, Doppler findings and perinatal outcome. *Ultrasound Obstet Gynecol* 2000; **15**: 114–117.
9. Budorick NE, Kelly TF, Dunn JA, Scioscia AL. The single umbilical artery in a high-risk patient population: what should be offered? *J Ultrasound Med* 2001; **20**: 619–627.
10. Gossett DR, Lantz ME, Chisholm CA. Antenatal diagnosis of single umbilical artery: is fetal echocardiography warranted? *Obstet Gynecol* 2002; **100**: 903–908.
11. Gornall AS, Kurinczuk JJ, Konje JC. Antenatal detection of a single umbilical artery: does it matter? *Prenat Diagn* 2003; **23**: 117–123.
12. Martinez-Payo C, Gaitero A, Tamarit I, Garcia-Espantaleo M, Iglesias Goy E. Perinatal results following the prenatal ultrasound diagnosis of single umbilical artery. *Acta Obstet Gynecol Scand* 2005; **84**: 1068–1074.
13. Volpe G, Volpe P, Boscia FM, Volpe N, Buonadonna AL, Gentile M. ["Isolated" single umbilical artery: incidence, cytogenetic abnormalities, malformation and perinatal outcome] *Minerva Ginecol* 2005; **57**: 189–198.
14. Granese R, Coco C, Jeanty P. The value of single umbilical artery in the prediction of fetal aneuploidy: findings in 12,672 pregnant women. *Ultrasound Q* 2007; **23**: 117–121.
15. Lubusky M, Dhaifalah I, Prochazka M, Hyjanek J, Mickova I, Vomackova K, Santavy J. Single umbilical artery and its siding in the second trimester of pregnancy: relation to chromosomal defects. *Prenat Diagn* 2007; **27**: 327–331.
16. Snijders RJ, Noble P, Sebire N, Souka A, Nicolaides KH. UK multicentre project on assessment of risk of trisomy 21 by maternal age and fetal nuchal-translucency thickness at 10–14 weeks of gestation. Fetal Medicine Foundation First Trimester Screening Group. *Lancet* 1998; **352**: 343–346.
17. Kagan KO, Wright D, Baker A, Sahota D, Nicolaides KH. Screening for trisomy 21 by maternal age, fetal nuchal translucency thickness, free beta-human chorionic gonadotropin and pregnancy-associated plasma protein-A. *Ultrasound Obstet Gynecol* 2008; **31**: 618–624.
18. Rembouskos G, Cicero S, Longo D, Sacchini C, Nicolaides KH. Single umbilical artery at 11–14 weeks' gestation: relation to chromosomal defects. *Ultrasound Obstet Gynecol* 2003; **22**: 567–570.
19. CG62 Antenatal care: full guideline. National Institute for Health and Clinical Excellence. http://www.nice.org.uk/nicemedia/pdf/CG62FullGuidelineCorrectedJune2008July2009.pdf.

Copyright © 2010 ISUOG. Published by John Wiley & Sons, Ltd.