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Presence of an umbilical artery notch in monochorionic/monoamniotic twins

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Abstract

Objective—To examine the association between an umbilical artery notch and fetal deterioration in monochorionic/monoamniotic (MC/MA) twins.

Methods—Six MC/MA twin pregnancies were admitted at 24–28 weeks of gestation for close fetal surveillance until elective delivery at 32 weeks or earlier in the presence of signs of fetal deterioration. Ultrasound (US) examinations were performed twice weekly. The presence of cord entanglement, umbilical artery notch, abnormal Doppler parameters, a non-reassuring fetal heart rate pattern, or an abnormal fetal biophysical profile was evaluated.

Results—Umbilical cord entanglement was observed on US in all pregnancies. The presence of an umbilical artery notch was noted in four out of six pregnancies and in two of them an umbilical artery notch was seen in both twins. The umbilical artery pulsatility index was normal in all fetuses. Doppler parameters of the middle cerebral artery and ductus venosus, fetal biophysical profile and fetal heart rate monitoring remained normal until delivery in all pregnancies. All neonates experienced morbidity related to prematurity; however, all were discharged home in good condition.

Conclusion—The presence of an umbilical artery notch and cord entanglement, without other signs of fetal deterioration, is not indicative of an adverse perinatal outcome.

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Disclosure Statement

The authors report no conflicts of interest.

Keywords

cord entanglement; Doppler velocimetry; fetal hypoxia; fetal surveillance; umbilical cord knot

Introduction

Monochorionic/monoamniotic (MC/MA) twin gestations are infrequent, with a prevalence of 1 in 8,000 pregnancies [1, 2], and at risk of complications related to monochorionicity [3–5], such as twin-to-twin transfusion syndrome [6, 7], twin-reversed arterial perfusion [8,9], fetal growth restriction [10–12], congenital anomalies [13], and preterm delivery [14–16]. In addition, MC/MA pregnancies may be affected by unique complications, such as conjoined twins [17], and umbilical cord entanglement that can increase the risk of fetal death [18, 19]. Kofinas et al. [20] first reported the presence of an umbilical artery notch associated with cord entanglement in a triplet pregnancy with MC/MA twins. One of the twins died in utero and the co-twin had a non-reassuring fetal heart rate pattern requiring an emergency cesarean delivery [20]. Therefore, the presence of an umbilical artery notch in the setting of MC/MA twins has been considered a poor prognosis sign, but has seldom been reported [20–22].

In MC/MA twin gestations, it is important to identify the presence of cord entanglement and signs of fetal deterioration, such as abnormal Doppler velocimetry and non-reassuring fetal heart rate patterns [23–25]. The prevalence of cord entanglement can reach 70–80% in all MC/MA pregnancies [26], yet most will have a favorable outcome [27]. In addition, Hugon-Rodin et al. [22] and Abuhamad et al. [21] have reported favorable outcomes in MC/MA twin gestations when an umbilical artery notch is present; however, further observations are desirable. In this study, we aimed to examine the association between an umbilical artery notch and signs of fetal deterioration in MC/MA twin pregnancies.

Study Population

From 2007 to 2013, 53 MC twin pregnancies were evaluated; nine of them (17%) were MA: one was a conjoined twin pregnancy, two presented early deliveries, and six MC/MA twin pregnancies without evidence of congenital anomalies were followed longitudinally. All pregnant women were enrolled in research protocols approved by the Institutional Review Boards of the *Eunice Kennedy Shriver* National Institute of Child Health and Human Development and by the Human Investigation Committee of Wayne State University, and provided written informed consent for ultrasound (US) examination.

The pregnant women were admitted at 24–28 weeks of gestation for close fetal monitoring until delivery. US examinations were performed twice weekly in all pregnancies. Daily fetal heart rate monitoring and fetal biophysical profile evaluations were also conducted. All sets of twins were electively delivered by cesarean section at 32 weeks of gestation or earlier in presence of signs of fetal deterioration. Before delivery, all pregnant women received a course of steroids to accelerate lung maturity (12 mg betamethasone/2 doses, 24 h apart).

US Examination

Evaluation of fetal anatomy and weight was performed in all fetuses. In all pregnancies, signs of twin-to-twin transfusion syndrome, consisting of discrepancy in the size of the bladders and in the amount of amniotic fluid, and abnormal Doppler parameters of the umbilical artery and ductus venosus (absent diastolic flow and absent atrial flow, respectively) were investigated [28, 29]. The presence/absence of cord entanglement was documented by visualization of the umbilical cords from the fetal abdomen to the placental insertion site. If present, the location of the cord entanglement (either close to the placenta or to one of the fetuses) was registered. Doppler recordings of the umbilical artery were obtained for each twin at three locations: (1) close to the entrance into the fetal abdomen, (2) at a free loop of the umbilical cord, and (3) near the cord entanglement. In addition, a Doppler evaluation of the umbilical arteries within the cord entanglement was performed. An umbilical artery notch was defined as a discontinuous trace in the late systolic phase of the waveform (Figure 1A). The pulsatility index [PI, (peak systolic velocity – end-diastolic velocity)/mean maximum average velocity; normal: <95th centile for gestational age [30]] and characteristics of the diastolic flow in the umbilical artery, middle cerebral artery and ductus venosus were also evaluated in both twins.

Healthcare providers were informed of the presence of cord entanglement and/or umbilical artery notching. In one pregnancy, the decision to deliver at 28 weeks of gestation was based only on the presence of cord entanglement without other signs of fetal deterioration. In the other five pregnancies, there were no fetal Doppler abnormalities or abnormal fetal heart tracings, and the twins were delivered at 32 weeks of gestation. Neonatal outcomes along with pathology examination of the placenta and umbilical cord were registered.

Results

Descriptive characteristics of the six pregnancies are presented in Table 1. Intrauterine growth restriction was diagnosed in two pregnancies, with both twins affected in one of them. Cord entanglement was observed in all six pregnancies, and in four pregnancies it was located close to the placental insertion of the cords. An umbilical artery notch was visualized in four out of six pregnancies, and in two of them an umbilical notch was noted in both twins. The presence of a notch did not affect the PI or the diastolic flow velocities in the umbilical artery of any fetus. In all four pregnancies, the location of the notch within the umbilical cord changed from one examination to another and was more frequently observed close to the cord entanglement (Figure 1A, 1B). When the umbilical was examined within the cord entanglement, a dynamic change in the Doppler waveform was observed with intermittent presence of an umbilical artery notch, but with normal diastolic flow (Figure 1C). Doppler velocimetry of the middle cerebral artery and ductus venosus, fetal heart rate monitoring, and the fetal biophysical profile all remained within normal limits until delivery.

Characteristics of the neonates and pathology examinations of the placenta/umbilical cord are shown in Table 1. One pregnant woman had a cesarean delivery at 28 weeks and 5 days of gestation due to cord entanglement and an umbilical artery notch without other signs of fetal deterioration or the presence of additional pregnancy complications. All other sets of

twins were electively delivered at 32 weeks of gestation. Umbilical cord pH values were normal (7.20) in all newborns; three newborns had a 1-min Apgar score of 4, but recovered at 5 min; 10 newborns (83%) presented with mild to severe respiratory complications at birth, from transient tachypnea (n = 2) to respiratory distress syndrome (RDS; n = 9); 7 neonates (58%) had hyperbilirubinemia, and 4 (33%) developed an intraventricular hemorrhage grade I. There were no neonatal deaths, and all neonates were discharged home in good condition.

Cord entanglement was confirmed at the time of birth in all pregnancies (Figure 2A), an umbilical cord knot was observed in two pregnancies (Figure 2A, 2B), a forked placental insertion of the umbilical cords in one set of twins (Figure 2C), and a marginal placental insertion of the cords for the other set of twins. In three cases, the placental insertions of the umbilical cords were observed at approximately 1–2 cm to one another.

Discussion

The main findings of this study are: (1) umbilical cord entanglement was observed in all six studied pregnancies; (2) an umbilical artery notch was observed in four pregnancies and in two pregnancies a notch was noted in both twins; (3) umbilical artery notching was more commonly observed close to the cord entanglement; however, in repeated examinations the notch changed its location in the umbilical cord and its shape; (4) the PI and diastolic velocities in the umbilical artery remained normal; and (5) no signs of fetal deterioration were documented in any case.

Clinical Significance of an Umbilical Artery Notch

Notching of the umbilical artery usually affects the late systolic [21] or early diastolic phase [20] of the Doppler waveform and has been proposed to result from narrowing, distortion or compression of the umbilical vessels [31, 32]. In singleton pregnancies, an umbilical artery notch has been associated with an abnormal insertion of the umbilical cord [21, 32], a tight umbilical cord knot [33], fetal intra-abdominal umbilical vein dilatation [34], and gastroschisis [35]. Abuhamad et al. [36] proposed that the presence of an umbilical artery notch in singleton gestations should prompt targeted US examination to search for anomalies in the umbilical cord and in the placenta as well as increased fetal surveillance due to the risk of non-reassuring fetal heart rate pattern.

In MC/MA twins, an umbilical artery notch has been associated with cord entanglement and adverse perinatal outcome [20, 33]. Table 2 summarizes the findings and obstetrical outcome of previous studies reporting of notching in the umbilical artery waveform in MC/MA twin gestations [20–22]. All were reported in the third trimester and 3 out of 4 had a favorable perinatal outcome. In our cases, we were able to observe an umbilical artery notch in four pregnancies, with the location of the notch changing, but more apparent close to the cord entanglement. Mechanical factors that stretch the cord, such as entanglement, true knots and fetal movements, can increase the tension in the arterial wall of the umbilical artery, thus contributing to umbilical artery notching.

The only report on umbilical artery notching and adverse perinatal outcome in MC/MA twins was by Kofinas et al. [20]. In this report, when the umbilical artery notch was observed, there was already one fetal demise, and the surviving twin demonstrated an elevated systolic/diastolic ratio in the umbilical artery, pulsation of the umbilical vein and variable decelerations on the nonstress test. Subsequent studies, such as that from Hugon-Rodin et al. [22] and ours, have not found such an association. The presence of an umbilical artery notch in both fetuses was previously reported by Abuhamad et al. [36] with a favorable outcome for both twins. We observed an umbilical artery notch in both co-twins in two pregnancies, one of which had a true knot confirmed after delivery. It is possible that the presence of an umbilical artery notch might be more frequent than expected if the umbilical artery is evaluated in different parts of the umbilical cord. In general, Doppler interrogation of the umbilical artery in twins is done close to the fetal insertion of the cord where a notch is less frequently observed. It might be possible that, when evaluating the vessels involved in the cord entanglement, overlapping of the Doppler spectrum of the two fetuses can give the impression of an umbilical artery notch. Overton et al. [37] described this finding as the 'galloping' pattern. The galloping pattern is different from notching of the umbilical artery, as the systolic peaks of the two heart frequencies can be noted at any time during the cardiac cycle. The umbilical artery notch is always observed in the late systolic or early diastolic phase of the cardiac cycle. In addition, the galloping pattern is only observed within the region of cord entanglement, whereas an umbilical artery notch can be observed in a free loop of the umbilical cord.

Placenta and Umbilical Cord Pathology

Cord entanglement was confirmed in all pregnancies at delivery. The high prevalence of cord entanglement has been previously reported, suggesting that it does not necessarily lead to an abnormal perinatal outcome [2, 26, 37–44]. All other placental and umbilical cord findings, such as a true knot, forked placental insertion [45] and the proximity of cord placental insertions to each other, might contribute to the presence of an umbilical artery notch. Fortunately, no signs of fetal deterioration were documented in our cases.

Management of MC/MA Twins

Although intensive surveillance is recommended for MC/MA twins [46–49], controversy exists regarding the frequency and timing of the initiation of tests as well as the optimal gestational age for delivery [48, 50, 51]. Two retrospective cohort studies have demonstrated that early hospitalization between 24 and 28 weeks of gestation accompanied by intensive fetal monitoring and delivery at 32–34 weeks of gestation improved perinatal survival when compared to outpatient management [49, 52]. A close evaluation of these pregnancies might help to identify early signs of fetal deterioration [53]. It is likely that neither the isolated presence of cord entanglement nor the presence of an umbilical artery notch is an indication alone for an emergency delivery. However, the presence of additional abnormalities in the umbilical artery waveform, such as increased PI, reduced diastolic flow [20, 42] or umbilical vein pulsations [34], may suggest tight cord entanglement which could lead to fetal death. Our results suggest that cord entanglement and an umbilical artery notch may be frequent findings in MC/MA twins. However, a tight cord entanglement might lead to severe notching

and acute fetal deterioration. Close surveillance of these pregnancies is therefore recommended.

All newborns experienced morbidity related to gestational age at delivery. None of the neonates with intraventricular hemorrhage grade I had further progression. These results suggest that in the absence of signs of fetal deterioration, even when cord entanglement and umbilical artery notching are present, these pregnancies could be prolonged to at least 34 weeks, when the risk of neonatal morbidity is substantially reduced.

Conclusion

In MC/MA twin gestations, the presence of umbilical cord entanglement and umbilical artery notching in the absence of signs of fetal deterioration do not necessarily lead to an adverse perinatal outcome.

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Figure 1.

Changes in the shape of the umbilical artery notch recorded at different locations: **a** close to the fetal abdominal insertion of the umbilical cord, **b** close to the site of cord entanglement, **c** within the cord entanglement, and **d** high-definition color flow Doppler using three-dimensional US of the umbilical cord entanglement.

Figure 2.
Gross examination of the umbilical findings after birth: **a** entanglement of the umbilical cords with a true knot, **b** umbilical cord knot, and **c** forked placental insertion of the umbilical cords of both twins.

Table 1

Clinical characteristics of six MC/MA twin pregnancies with an umbilical artery notch

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6
Maternal age (years)	23	29	28	23	17	28
Gestational age at hospitalization (weeks)	24	26	24	26	28	27
Gestational age at delivery (weeks+days)	28+5	32+0	32+1	32+0	32+2	32+2
Birthweight (g) (Twin A, Twin B)	1190 / 1300	1745 / 1685	1950 / 1895	1485 / 1320	2000 / 1870	1675 / 1730
APGAR at 1 and 5 minutes (Twin A, Twin B)	6/8 7/8	8/9 8/8	7/8 6/8	3/8 6/7	3/6 4/8	6/8 5/7
Cord pH (Twin A, Twin B)	7.2/7.2	7.3/7.2	7.3/7.3	7.3/7.2	7.2/7.3	7.3/7.3
Ultrasound findings	Cord entanglement Umbilical artery notch in both twins	Cord entanglement No umbilical artery notch	Cord entanglement No umbilical artery notch	Cord entanglement Umbilical artery notch in one twin	Cord entanglement Umbilical artery notch in one twin	Cord entanglement Umbilical artery notch in both twins
Placenta and umbilical cord pathology examination	Cord entanglement Marginal placental insertion of the two cords	Cord entanglement Close placental insertion of the umbilical cords	Cord entanglement Close placental insertion of the umbilical cords	Cord entanglement Close placental insertion of the umbilical cords	Cord entanglement Forked placental cord insertion Cord knot	Cord entanglement Cord knot
Neonatal complications						
Twin 1	RDS Hyperbilirubinemia	IVH grade I Hypoglycemia Hyperbilirubinemia	IVH grade I RDS Hyperbilirubinemia	IVH grade I, RDS	RDS	RDS Hyperbilirubinemia
Twin 2	RDS Hyperbilirubinemia	TTN, Bradycardia Anemia,	TTN Hyperbilirubinemia	IVH Grade I, RDS Bradycardia Anemia	RDS	RDS Hyperbilirubinemia

RDS, respiratory distress syndrome; TTN, transient tachypnea of the newborn; IVH, intraventricular hemorrhage.

Table 2

Previous studies reporting an umbilical artery notch in monochorionic/monoamniotic twin gestations

Author	GA at detection (weeks)	Cord entanglement (US or pathology)	Clinical characteristics
Kofinas et al.(20)	32	Yes	Fetal demise of one twin at time of ultrasound, an umbilical artery notch was observed in the surviving twin Nonreactive non-stress test, abnormal fetal biophysical profile, abnormal umbilical artery Doppler. Delivery at 32 weeks. Tight cord entanglement
Abuhamad et al.(21)	33	Yes	Normal fetal heart rate monitoring, decreased amniotic fluid. Delivery at 34 weeks.
	31	Yes	Normal heart rate monitoring, umbilical artery notch in both twins. Delivery at 32 weeks
Hugon-Rodin et al.(22)	31	Yes	Normal fetal heart rate monitoring, Delivery at 32 weeks

GA: gestational age; UA: Umbilical artery; US ultrasound