Pseudo-monoamniotic Pregnancy: Case Report and Review of Etiologic Considerations

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ABSTRACT

Pseudo-monoamniotic gestations are increasingly recognized through sonographic surveillance of monochorionic twins, though etiologic factors remain undefined. We present a case of spontaneous pseudo-monoamniotic twins and propose umbilical cord insertion proximity as a sonographic marker. Systematic review of the literature was performed and additional cases with similar findings were noted. Approximately 75% of reported cases (28/37) were deemed spontaneous and several included short inter-cord distances. Shunting of blood away from the membranes in the region between the cord insertions may be responsible for membrane rupture. Further investigation is needed into short inter-cord distance as a marker for monochorionic twins at risk to become a pseudo-monoamniotic gestation.

KEYWORDS: monochorionic, twins, pseudo-monoamniotic,

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INTRODUCTION

Twin gestations occur in 2% of all pregnancies, yet account for 10% of all perinatal morbidity and mortality. This discrepancy is attributed to an increased risk of prematurity in all twin gestations, as well as additional risks based on chorionicity. Monochorionic twin gestations are susceptible to additional morbidity due to the potential for anastomotic placental vessels and resultant Twin-Twin Transfusion Syndrome (TTTS). Monochorionic-monoamniotic gestations lack a dividing membrane, resulting in cord entanglement in up to 70% of twins and increased rates of intrauterine fetal demise (IUFD).[1,2]

Chorionicity is set at the earliest stages of development and often characterized during the initial sonographic exam. In rare cases, the dividing membrane in a monochorionic-diamniotic twin gestation can rupture and create a functional monochorionic-monoamniotic, or "pseudo-monoamniotic", gestation. Septostomy may be spontaneous, though iatrogenic events are more commonly encountered. Intentional septostomy is used as a treatment for TTTS, while unintentional septostomy has occurred after invasive diagnostic procedures such as amniocentesis, fetoscopy, or laser photocoagulation. The occurrence of partial monochorionic or partial monoamniotic twinning has also been documented, though these terms refer to incomplete septum formation secondary to the timing of zygotic cleavage.[3]

Increased use of sonographic imaging for antenatal surveillance of monochorionic gestations has led to better recognition of pseudo-monoamniotic twins. However, the etiology of spontaneous rupture of the dividing membrane remains unknown due to the scarcity of this population. We present a case of pseudo-monoamniotic twins diagnosed in the antenatal period within the context of the existing literature. Additionally, we discuss a hypothesis for the etiology of spontaneous membrane rupture and propose a sonographic marker common to several reported cases.

CASE

A 35 year old G3P2002 at 18 weeks, 6 days gestation was referred to Maternal-Fetal Medicine for advanced maternal age and twin gestation. During the initial sonogram, chorionicity was determined to be monochorionic-diamniotic due to the presence of a thin dividing membrane, absence of a twin peak sign, and concordant gender. Fetal growth was appropriate for gestational age. The patient was advised to return bi-weekly for TTTS surveillance and monthly for growth assessments. Fetal echocardiograms were obtained and noted to be normal.

The subsequent sonographic evaluation one month later (22 weeks, 6 days) did not clearly visualize the dividing membrane. The possibility of rupture of the dividing membrane was discussed. At 24 weeks and 6 days, the dividing membrane was again difficult to see in its entirety. The likelihood of rupture was determined to be high and the diagnosis of a pseudo-monoamniotic twin gestation was made. The patient was counseled on the potential for cord entanglement and increased risk of intrauterine demise. Antepartum management was adjusted to parallel monochorionic-monoamniotic pregnancies, including weekly evaluation for hydrops until 28 weeks, hospitalization at 28 weeks, and delivery at 32-34 weeks.

Subsequent imaging studies demonstrated a progression of events linked to rupture of the dividing membrane. At 26 weeks gestation, imaging demonstrated both fetuses in the same compartment with remnants of the dividing membrane appearing to sequester the bulk of the umbilical cords. At 29 weeks gestation, the umbilical cords were no longer compartmentalized by the membrane and were visualized intermingling without evidence of compression. Cord entanglement was suspected and supported by additional sonographic imaging.

Upon admission to the antepartum unit at 28 weeks gestation, the patient underwent intermittent monitoring for fetal well-being assessment. Fetal monitoring remained overall reassuring. A biophysical profile (BPP) was obtained for fetus B following a repeated episode of tachycardia at 30 weeks, 5 days gestation; the BPP was 8 out of 8 with spontaneous resolution of fetal tachycardia. Tocometry infrequently demonstrated uterine irritability and irregular contractions. Weekly sonographic assessments demonstrated concordant growth of the fetuses with no signs of TTTS through the final assessment at 32 weeks. Betamethasone was administered 48 hours prior to delivery to promote fetal lung maturity (12mg IM q24 hours x 2 doses).

A scheduled primary cesarean section was performed at 32 weeks, 6 days gestation. Twin A was a female infant weighing 1,640gm with APGAR scores of 8 and 9 at 1 and 5 minutes, respectively. The uterus clamped down following delivery of Twin A, necessitating nitroglycerin (200mcg) for rapid uterine relaxation. Twin B, a female infant weighing 1,700gm with APGAR scores of 5 and 8 at 1 and 5 minutes, was subsequently delivered. The newborns were taken to the neonatal intensive care nursery for further care. Twin A was noted to have a small amniotic band over the right forearm that was removed after delivery, but no additional deficits.

The placenta was carefully examined after delivery. Knotting and looping of the umbilical cords were noted at two distinct sites. The sites of umbilical cord insertion into the placenta were found to be closely approximated with only 1 cm separation. A single placental disc with an amniotic plica was observed, confirming the monochorionic gestation with remnants of a diamniotic dividing fetal membrane (Figure 1). The placenta was sent to pathology for formal evaluation. The pathology examination revealed no significant histological changes or evidence of chorioamnionitis. The point of rupture was indeterminate. The placental cord insertion site for both Twin A and Twin B were paracentric at 3.5cm and 4.5cm from the disc margin, respectively. Per discussion with pathology, superficial anastomosing vessels were suspected between the adjacent cord insertion sites.

COMMENT

Pseudo-monoamniotic twins are an uncommon variant of monochorionic-diamniotic gestations with an unknown incidence. The increased use of sonographic imaging has improved antenatal detection of this population, resulting in an increased reporting of cases over the past decade. Early reports diagnosed pseudo-monoamniotic twins at the time of delivery due to the absence of a dividing membrane on examination and cord entanglement. More recent reports have documented this finding during the course of routine antenatal surveillance of monochorionic-diamniotic twin gestations. One group of monochorionic twins found spontaneous septostomy rates to be as high as 1.8%, however this is likely an overestimate in the general twin population.[4]

A search of the English-language literature on PubMed and Ovid with review of relevant references yielded 37 reported cases of pseudo-monoamniotic twins resulting from initially diagnosed monochorionic-diamniotic gestations.[1-2,4-26] Notably, recognition of pseudo-monoamniotic twins has grown with 47% (18) of all publications occurring within the past 5 years (2009-2014). Table 1 lists all the published cases of pseudo-monoamniotic twins. Of the reported cases, 76% (28) were attributed to spontaneous rupture, 19% (7) occurred after invasive procedures in the antepartum period, and 2% (1) was suggested to be the result of chorioamnionitis.[15]

Multiple etiologies have been associated with rupture of the dividing membrane in monochorionic twin gestations. Traumatic disruption of the membrane may occur from maternal or iatrogenic causes, such as abdominal trauma or unintentional septostomy during invasive procedures. Fetal movement has also been cited as a cause of rupture of the dividing membrane.[8] Additional findings associated with rupture of the dividing membrane include developmental abnormalities of the membranes such as amniotic plica, polyhydramnios, and infection.[4,15]

Previous investigators have reported pseudo-monoamniotic twins in the absence of preceding external triggers such as maternal trauma, infection, or invasive antenatal procedures. Lee, et al. proposed that the proximity of the umbilical cord insertion sites in the placenta may be related to spontaneous rupture of the dividing membrane. They hypothesized that anastomosis of the placental vessels between the closely approximated umbilical cords could lead to diversion of blood flow and reduced perfusion of the dividing membrane. While vascular anastomoses are more commonly seen in TTTS, only a minority of the reported cases of pseudo-monoamniotic twins were associated with TTTS. Our patient demonstrated cord insertion sites that were closely approximated, which has also been noted in multiple case reports of spontaneous septostomy.[7,15,17,18,22,26] Two reported cases included pictures suggestive of a short distance between cord insertion sites.[15,17] Oh and colleagues noted nearly adherent cord insertion sites visible across the amniotic membrane after cesarean delivery.[22] Chadha et al. also described a 1 cm distance between umbilical cord insertion sites with vascular anastomosis, but no cord entanglement at the time of delivery.[7] Likewise, Yoshimura et al. also noted contiguous cord insertion sites after delivery.[26]

Abnormalities in the umbilical cord insertion in monochorionic twins has been associated with discordant growth, TTTS, and increased perinatal mortality. A study of 150 monochorionic-diamniotic twin placentas demonstrated vascular anastomoses in 97% and a median intercord distance of 15cm.[27] In monochorionic twin gestations, the pattern of the cord insertions into the placenta has been demonstrated to influence the vasculature at the chorionic plate. De Paepe, et al. has shown that the umbilical cord insertion site influences the chorionic vascular distribution of the contralateral twin in monochorionic-diamniotic pregnancies.[28] Their study demonstrated the lowest density of superficial perforating chorionic arteries when both placenta demonstrated a paracentral umbilical cord insertion.[28] In a subsequent study, the same group demonstrated a non-significant decrease in chorionic vascular density of intermediate villi and an increased density of terminal villi in MD twins with concordant paracentral/paracentral cord insertions.[29]

The principles for management of pseudo-monoamniotic twin gestations centers on minimizing risk to the fetuses. The functional monoamniotic gestational sac incurs similar risks of cord entanglement and perinatal morbidity as monochorionic-monoamniotic gestations. Antepartum management may include weekly-biweekly sonographic assessment of hydrops/TTTS, monthly assessment of fetal growth, and inpatient hospitalization after 28 weeks with daily fetal surveillance. Van Mieghan, et al. has recently demonstrated a low risk of perinatal morbidity from cord entanglement in monoamniotic gestations less than 33 weeks, and that elective delivery should be pursued at that time.[30] Sonographic determination of the distance between umbilical cord insertion sites may be a potential marker for screening monochorionic-diamniotic gestations at risk for spontaneous septostomy. Systematic evaluation of the intercord distance in monochorionic gestations is needed to confirm the true value of this marker.

REFERENCES

1 Abraham, R. J. (2013). The protective effect of oligohydramnios in a case of missed diagnosis of spontaneous septostomy in monochorionic diamniotic twins. *J Obstet Gynaecol, 33*(2), 205-207.

2 Sherer, D. M., Bitton, C., Stimphil, R., Dalloul, M., Khoury-Collado, F., & Abulafia, O. (2005). Cord entanglement of monochorionic diamniotic twins following spontaneous antepartum septostomy sonographically simulating a true knot of the umbilical cord. *Ultrasound Obstet Gynecol, 26*(6), 676-678.

3 Galjaard, S., Moerman, P., Corveleyn, A., Devlieger, R., Lewi, L. (2014). Partial monochorionic and mono-amniotic twin pregnancies. *Ultrasound Obstet Gynecol, 44(6), 722-4.*

4 Chmait, R. H., Aghajanian, P., Kontopoulos, E. V., & Quintero, R. A. (2009). Prenatal diagnosis of spontaneous septostomy of the dividing membranes in complicated monochorionic diamniotic multiple gestations. *J Ultrasound Med, 28*(5), 663-668.

5 Aisenbrey, G. A., Catanzarite, V. A., Hurley, T. J., Spiegel, J. H., Schrimmer, D. B., & Mendoza, A. (1995). Monoamniotic and pseudomonoamniotic twins: sonographic diagnosis, detection of cord entanglement, and obstetric management. *Obstet Gynecol, 86*(2), 218-222.

6 Beasley E., Megerian G., Gerson A., Roberts NS. (1999). Monoamniotic twins: case series and proposal for antenatal management. *Obstet Gynecol*, 93(1): 130-4.

7 Chadha, R., Lange, I. R., Bratz, L., Cooper, S. L., Roggensack, A., & Johnson, J. A. (2012). A rare case of antepartum spontaneous septostomy in a monochorionic diamniotic twin pregnancy. *Case Rep Obstet Gynecol, 2012*, 748614.

8 Chen, S. E., Trupin, L., & Trupin, S. (1994). Antepartum rupture of diamniotic membranes separating monozygotic twins. A case report. *J Reprod Med, 39*(1), 67-70.

9 Cook, T. L., & O'Shaughnessy, R. (1997). Iatrogenic creation of a monoamniotic twin gestation in severe twin-twin transfusion syndrome. *J Ultrasound Med, 16*(12), 853-855.

10 De Lia, J. E., & Worthington, D. (2000). Intrauterine sling with umbilical cord entanglement in diamniotic twins. *Ultrasound Obstet Gynecol, 15*(5), 447.

11 Dunnihoo, D. R., & Harris, R. E. (1966). The diagnosis of monoamniotic twinning by aminography. *Am J Obstet Gynecol, 96*(6), 894-895.

12 Eddib, A., Rodgers, B., Lawler, J., & Yeh, J. (2006). Monochorionic pseudomonoamniotic twin pregnancy with fetal demise of one twin and development of maternal consumptive coagulopathy. *Ultrasound Obstet Gynecol, 28*(5), 736-737.

13 Feldman, D.M., Odibo, A., Campbell, W.A., Rodis, J.F. (1998) Iatrogenic monoamniotic twins as a complication of therapeutic amniocentesis. *Obstet Gynecol,* 91(5Pt2), 815-16.

14 Fleming, T. a. M., T. (2012). Spontaneous septostomy in mono-chorionic diamniotic twins resulting in cord entanglement and fetal demise. *Australasian Society for Ultrasound in Medicine*.

15 Gilbert, W. M., Davis, S. E., Kaplan, C., Pretorius, D., Merritt, T. A., & Benirschke, K. (1991). Morbidity associated with prenatal disruption of the dividing membrane in twin gestations. *Obstet Gynecol, 78*(4), 623-630.

16 Hackney, D. N., Khalek, N., Moldenhauer, J., & Ozcan, T. (2013). Twin-twin transfusion syndrome presenting as polyhydramnios in both fetuses secondary to spontaneous microseptostomy. *AJP Rep, 3*(2), 83-86.

17 Krause, H. G., & Goh, J. T. (1998). Cord entanglement in monochorionic diamniotic twins. *Aust N Z J Obstet Gynaecol, 38*(3), 341-342.

18 Lee, K. J., Kim, M. K., Lee, S. Y., Lee, W. S., & Lee, Y. H. (2012). Spontaneous rupture of the dividing membrane in a monochorionic pregnancy resulting in a pseudo-monoamniotic pregnancy with cord entanglement. *J Obstet Gynaecol Res, 38*(5), 863-866.

19 Megory, E., Weiner, E., Shalev, E., & Ohel, G. (1991). Pseudomonoamniotic twins with cord entanglement following genetic funipuncture. *Obstet Gynecol, 78*(5 Pt 2), 915-917.

20 Miura, N., & Suzuki, S. (2008). Fetal asphyxia due to cord entanglement in a monochorionic diamniotic twin pregnancy complicated by 2nd-trimester single intrauterine demise. *Fetal Diagn Ther, 23*(1), 69-71.

21 Nasrallah, F. K., & Faden, Y. A. (2005). Antepartum rupture of the intertwin-dividing membrane in monochorionic diamniotic twins: a case report and review of the literature. *Prenat Diagn, 25*(9), 856-860.

22 Oh, J. E., Bae, J. Y., Hwang, I. A., Kim, M. J., & Seong, W. J. (2011). Antepartum spontaneous rupture of the intertwin-dividing membrane in monochorionic diamniotic twins; Nearly adhered cords. *Korean Journal of Obstetrics, 54*(6), 304.

23 Suzuki, S. (2013). Case series of monoamniotic and pseudomonoamniotic twin gestations. *ISRN Obstet Gynecol, 2013*, 369419.

24 Suzuki, S., Ishikawa, G., Sawa, R., Yoneyama, Y., Otsubo, Y., & Araki, T. (1999). Iatrogenic monoamniotic twin gestation with progressive twin-twin transfusion syndrome. *Fetal Diagn Ther, 14*(2), 98-101.

25 Tamura, T., Miura, A., & Suzuki, S. (2011). Spontaneous disruption of the dividing membrane in monochorionic diamniotic twin pregnancy. *Fetal Diagn Ther, 30*(3), 241-242.

26 Yoshimura, K., Aiko, Y., Inagaki, H., Nakata, M., & Hachisuga, T. (2009). Prenatal spontaneous disruption of the dividing membrane in monochorionic diamniotic twins detected at the time of fetoscopic laser photocoagulation. *J Obstet Gynaecol Res, 35*(6), 1129-1131.

27 Hack KE, Nikkels PG, Koopman-Esseboom C, Derks JB, Elias SG, van Gemert MJ, Visser GH. Placental characteristics of monochorionic diamniotic twin pregnancies in relation to perinatal outcome. Placenta. 2008 Nov;29(11):976-81.

28 De Paepe ME, Shapiro S, Hanley LC, Chu S, Luks FI. Correlation between cord insertion type and superficial choriovasculature in diamniotic-monochorionic twin placentas. Placenta. 2011 Nov;32(11):901-5.

29 Chu, S., Mao, Q., Shapiro, S., Luks, SL., De Paepe, M.E. (2013). Correlation between cord insertion type and chorionic villus vascularization of the co-twin in diamniotic-monochorionic twin pregnancies. *Early Human Development,* 89(2013), 243-247.

30 Van Mieghan, T.M., Heus, R.D., et al. (2014). Prenatal Management of Monoamniotic twin pregnancies. *Obstet Gynecol,* 124(3), 498-506.

FIGURE LEGEND

Figure 1: Image of the placenta and umbilical cords from the pseudo-monoamniotic gestation. Note the close proximity of the placental cord insertions and presence of an amniotic plica. Knotting of the umbilical cord is present, which is characteristic of monoamniotic gestations.

Table 1. Published reports of pseudo-monoamniotic twin gestations with corresponding sonographic and pathologic findings.

Reference	Subject Age (years)	Diagnostic Timing	Antenatal Intervention	Sonographic Findings	Gestational Age at Delivery (weeks)	Pathologic Findings
Abraham, 2013	22y	Postnatal		Dividing membrane not visualized after 13 weeks	34+2	Cord entanglement
Aisenberry, 1995		Postnatal		Unremarkable 26 week scan	30+2	Cord entanglement
Chadha, 2012		Prenatal		Dividng membrane not visualized after 26 weeks	29+5	Umbilical cord insertions into placenta 1cm apart
Eddib, 2006	36y	Postnatal		IUFD diagnosed at 29 weeks; cord entanglement noted and dividing membrane not visualized	34+0	Cord entanglement; dividing membrane between insertion sites
Beasley, 1999	26y	Prenatal		Dividing membrane not visualized at 21 week scan	32+0	Cord entanglement
Gilbert, 1991	36y	Prenatal	Amniocentesis	Dividing membrane present at 22 weeks; membrane rupture noted prior to delivery of IUFD		Cord entanglement; cord insertion sites approximated
	36y	Postnatal		Dividing membrane present at 23 week scan	25+0	Umbilical cord insertion sites approximated; amnion remnant
	40y	Postnatal		Dividing membrane present at 25 week scan	27+1	Twin B with velamentous cord insertion
	31y	Postnatal			34+0	Amniotic plica between insertion sites
	15y	Postnatal		TTTS	22+0	Amniotic plica with evidence of degeneration
	29y	Postnatal		Dividing membrane present at 20 week scan	30+3	Cord entanglement; amniotic plica; velamentous cord insertions
	28y	Prenatal	Laser	TTTS and amnion interuption noted	34+0	Cord entanglement; amnion disruption
Chen, 1994	22y	Prenatal		Dividing membrane not visualized after 29 weeks	37+0	Cord entanglement
Nasrallah, 2005	29y	Prenatal		Dividing membrane not visualized after 36 weeks	36+3	Cord entanglement; cords with eccentric insertion sites
Krause, 1998	40y	Postnatal		Dividing membrane visualized at 38 week scan	39+0	Cord entanglement; umbilical cords closely approximated
De Lia, 2000		Postnatal	Amnioreduction	Tangled cords visualized at 26 week scan; TTTS	27+0	Cord entanglement; central cord insertion x2
Oh, 2011	31y	Postnatal		Dividing membrane indeterminate for rupture at 30 week scan	35+3	Cord entanglement; umbilical cords were adherent

Yoshimura, 2009	28y	Prenatal	Fetoscopy	Dividing membrane rupture and cord entanglement noted at 24 weeks by fetoscopy for TTTS	32+0	Cord entanglement; umbilical cord insertions closely approximated
Lee, 2012	37y	Prenatal		Dividing membrane not visualized after 27 weeks	35+1	Cord entanglement; cord insertion sites approximated; amniotic plica
Sherer, 2005	17y	Postnatal		Dividing membrane rupture noted at 29 week scan	34+0	Cord entanglement
Chmait, 2009	27y	Prenatal	Laser	Dividing membrane detachment and rupture seen on fetoscopy for TTTS at 20 weeks	27+0	
	30y	Prenatal	Yes	Dividing membrane rupture suspected at 18 week scan; TTTS	27+0	
	30y	Prenatal	Fetoscopy; Laser	Cord entanglement noted at 20 week scan; TTTS	34+0	
	24y	Prenatal	Yes	Dividing membrane separation and umbilical cord entanglement noted on 19 week scan; TTTS	38+0	
Feldman, 1998	28y	Prenatal	Amniocentesis	Dividing membrane visualized at 28 week scan; TTTS	32+0	Cord entanglement
Hackney, 2013	30y	Prenatal	Amniocentesis, Reduction	Microseptostomies noted at 20 weeks; TTTS	23+6	Membrane intact on exam; vascular anastomoses
Dunihoo, 1966	18y	Prenatal	Amniocentesis, Amniography	26 week u/s with possible membrane rupture	38+0	Amniotic plica between cord insertion sites
Fleming, 2012	28y	Postnatal		Dividing membrane visualized at 35 week scan	35+6	Cord entanglement; Twin A IUFD
[a]Suzuki, 2013		Prenatal	Amnioreduction	Dividing membrane not visualized at 24 week scan; TTTS	27+0	Twin A IUFD; Twin B IUFD
		Postnatal		Dividing membrane visualized at 21 week scan	28+0	Twin B IUFD; cord entanglement
			Laser	TTTS	31+0	
			Laser	TTTS	33+0	
					35+0	
		Postnatal		Dividing membrane appeared normal on biweekly scans	36+0	Cord entanglement; Twin A cord attached to dividing membrane
					37+0	Cord entanglement

[a]Case series with summary of all pseudo-monoamniotic twin gestations from the Suzuki group.

Abbreviations: IUFD, intrauterine fetal demise; TTTS, twin-to-twin-transfusion syndrome