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Outcome of the Surviving Cotwin of a Fetus Papyraceus or of a Dead Fetus

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Abstract. Serial ultrasound examinations have demonstrated that one of two gestational sacs in a twin pregnancy may often disappear. When it disappears at an early stage of gestation, the pregnancy may advance without any disturbance and the cotwin can be delivered well developed and lively. When the intrauterine death occurs in the second trimester, the dead fetus usually results in a fetus papyraceus and the cotwin continues to be alive near term. However, when death occurs in the last trimester, the viable twin may be spontaneously delivered soon and be premature. In some cases of late fetal death, the dead fetus may induce intravascular thromboses in many organs of the surviving cotwin, so that the living infant may develop cerebral palsy later after birth.

Key words: Vanishing twin, Surviving cotwin, Blighted ovum, Fetus papyraceus, Twin transfusion syndrome, Intrauterine coagulation

INTRODUCTION

Serial ultrasound examination in the course of a multiple pregnancy not unfrequently show one of the two or more gestational sacs to disappear during early pregnancy. The rate of disappearance ranges widely, depending on the timing of discovery by ultrasound, the ultrasonar diagnostic techniques, or other factors. The phenomenon is still unexplained, although some morphologic evidence has also been described following the birth of a surviving cotwin [3,6,8]. Aiming at finding traces of a disappearing twin, we undertook a careful examination of delivered placentas of twins delivered at our hospital.

MATERIALS AND METHODS

A total of 189 placentas of twins delivered at Tokyo Medical College Hospital from February 1961 to May 1985 were examined. According to the method of Benirschke [1], twin placentation was classified into monochorionic (MC) and dichorionic (DC), and the presence of intraplacental vascular anastomoses in MC placentas was assessed by injecting air or a colored dye into surface vessels on the chorionic plate.

Of the 189 twin pairs, 115 had a MC placenta. In 21 of these, one fetus died in utero during pregnancy. These were subdivided into three groups according to the trimester when fetal death was likely to have occurred.

CASE REPORTS

Of the 115 pair of MC twins, 9 pairs, who were admitted to the hospital with episodes of vaginal bleeding associated or not with abdominal tension in the first trimester, had a chorionic cyst-like sac including a stunted embryo on the fetal surface of the placenta delivered at term. All except one (Case 9) delivered a normal baby, at term and following an uneventful course of pregnancy except for slight vaginal bleeding. Case 9, a male infant weighing 2934 g, was affected by tetralogy of Fallot and died at 36 days of age (Table 1).

Case 4 involved a 29-year-old primigravida admitted to our hospital complaining of vaginal bleeding in the first trimester. Twin pregnancy was diagnosed through an ultrasound examination at 6 weeks' gestation. She recovered from vaginal bleeding without any treatment 2 weeks later. On the ultrasound examination at 10 weeks' gestation, an intact gestational sac, containing a viable fetus, was visible in the center of the uterus, but in addition, a deformed gestational sac, including an amorphous mass, was recognized close to the cervix. At 40 weeks' gestation, the woman delivered a normal female baby weighing 2748 g.

The placenta contained a 7 × 7 cm chorionic cyst-like sac on the fetal surface. On opening the sac, a strand of umbilical cord-like tissue and a highly compressed embryo were recognized.

Table 2 shows 7 cases of fetus papyraceus coexisting with a viable fetus in a MC twin pregnancy. The early death of one of monozygotic twins occurred in utero in the 2nd trimester, apparently resulted in a fetus papyraceus with the cotwin sometimes growing to near term without serious adverse effects. In 4 of these cases, the viable twins developed well and were born near term uneventfully. In 1 case, however, the male infant weighed 1148 g, was born together with a male fetus papyraceus of 290 g at 29 weeks' gestation, and died of severe respiratory distress shortly after birth (Case 13). The remaining 2 viable twins suffered from congenital anomalies; one male infant, weighing 3322 g, was born at 40 weeks' gestation with meningocele and spina bifida (Case 12), while the other male infant, weighing 3280 g, was born at 41 weeks' gestation complicated by skin defects on both flanks through waist (Case 16).

Case 16 (Table 2) involved a 26-year-old mother, who had a miscarriage in her first pregnancy, but then had a normal pregnancy and delivered a male infant the following year. The infant suffered from skin defects, but is now 8 years old, regularly

Table 1. Cystic Form in Monochorionic Twins

Case	Gestational weeks; Placental weight	Sex; fetal weight; Size of cyst	Remarks
1. S.H.	40w	F: 3250g	stunted embryo (5g)
	435g	4 × 3 cm	1.8 × 1.5 cm
2. H.K.	41w	F: 3300g	stunted embryo
	435g	4 × 3.3 cm	1.5 × 1.2 cm
3. T.S.	41w	F: 3730g	stunted embryo
	580g	5 × 3 cm	
4. Y.I.	40w	F: 2748g	stunted embryo
	420g	7 × 7 cm	3.2 × 1.7 cm
5. S.S.	42w	F: 3258g	
	510g	3 × 2 cm	blighted umb. cord
6. J.A.	40w	F: 3510g	blighted umb. cord
	525g	6 × 4 cm	
7. I.Y.	42w	M: 3803g	
	495g	5 × 4.6 cm	blighted umb. cord
8. S.N.	40w	F: 3017g	stunted embryo
	380g	2.8 × 2 cm	
9. R.T.	39w	M: 2934g	A: tetralogy of Fallot
	400g	5 × 5 cm	stunted embryo

developed, and he can exercise without any limitation, even though old scars on his waist are found. The placenta was found to have a fetus papyraceus associated with marginal insertion of the umbilical cord and a marked infarct of 11.0 × 4.0 × 1.4 cm in size. The skin lesions of the liveborn twin might be due to a thrombotic cause derived from a dead fetus, but fortunately the damages were localized only in the skin. The mother also had another healthy boy two years later.

In Table 3, 5 cases of monozygotic twins associated with later fetal death are presented. When a twin dies in utero in the last trimester, delivery of the cotwin may be spontaneously induced in a few days. Accordingly, the most viable twins were prematurely delivered. In such cases, the dead fetus may cause extensive vascular thromboses in many organs of the surviving cotwin by release of thromboplastin-like substances, so that a living infant may sometimes develop cerebral palsy later after birth. In Case 19, the living baby suffered from severe cerebral palsy, and in Cases 20 and 21, the surviving infants suffered from hyperbilirubinemia during the neonatal period.

In Case 19 (Table 3), a 28-year-old primigravida complained of spontaneous labor pains when admitted to our hospital at 35 weeks' gestation. She soon delivered a female

Table 2. Fetus Papyraceus in Monochorionic Twins

Case	Gestational weeks; Placental weight	Fetal sex and weight	Placental vascular anastomoses	Remarks
10. H.O.	39w	F: 2750g	unknown	A: marginal cord insert.
	345g	F.pap.: 18g		B: velamentous cord insert.
11. N.I.	44w ?	M: 3365 g	A – A	B: amnion nodosum, velamentous cord insert.
	670g	F.pap.: 25g	A – V	
12. C.U.	40w	M: 3322g	unknown	A: spina bifida, meningocele,
	450g	F.pap.: 35g		B: velamentous cord insert.
13. Y.E.	29w ?	F.pap.(M): 290g	A – A	A: area of placenta: thin,
	450g	M: 1148g	V – V	B: died soon, B cord: edematous
14. M.K.	37w	F: 2444g	unknown	Both: velament. cord insert.
	340g	F.pap.: 5g		B: squamous metaplasia
15. T.S.	42w	M: 3484g	unknown	B: monster, amnion nodosum, velamentous cord insert.
	325g	F.pap.: 25g		
16. K.W.	41w	M: 3280g	unknown	A: cong. skin defect,
	395g	F.pap.: 8g		B: whole infarct of placenta, Both: marginal cord insert.

Table 3. Late Intrauterine Fetal Death of One Fetus in Monochorionic Twins

Case	Gestational weeks; Placental weight	Fetal sex and weight (dead fetus)	Placental vascular anastomoses	Remarks
17. Y.S.	32w	F: 1625g	A – A	A: Infarct
	620w	(F: 871g)		B: Infarct, Intervillous thrombosis Velament. cord insert.
18. N.S.	39w	M: 2555g	A – A	B: Infarct Marg. cord insert.
	730g	(M: 2565g)		
19. A.M.	35w	(F: 1709g)	A – A	A: Marg. cord insert.
	710g	F: 2210g	V – A	B: Edema of cord Developed cerebral palsy
20. S.K.	39w	F: 2281g	A – A	A: Intervillons thrombosis Marg. cord insert.
	775g	(F: 1287g)	V – V	B: Infarct, Short cord, Marg. cord insert.
21. K.K.	30w	M: 1756g	A – A	A: Marg. hemorrhage
	360g	(M: 1410g)	V – V	B: Short cord Velament. cord insert.

Table 4. Classification According to Estimated Timing of Intrauterine Death of One Twin

Estimated timing of fetal death	No. of cases	Deceased twins	Weight of twins	Gestational weeks at labor
1st trimester	9	Blighted	3283 ± 334g	40.6 ± 1.0
2nd trimester	7	Fetus papyraceus	2828 ± 77g	38.9 ± 4.5
3rd trimester	5	Late IUDF	2085 ± 345g	35.0 ± 3.6

still-born infant weighing 1709 g; probably, the infant had died in utero a few days earlier. Eight minutes later, another female infant, weighing 2210 g, was delivered by breech extraction, with Apgar scores of 6 and 9 at 1 and 5 minutes after birth. Although the living infant had an uneventful neonatal period, she later developed cerebral palsy. As the placenta was diamniotic monochorionic with artery-to-artery and vein-to-vein anastomoses, that might be the result of intravascular coagulation induced by the dead fetus.

The 21 cases of monochorionic twins with one died during pregnancy were subdivided into three groups according to the time of the fetal death. As shown in Table 4, when death occurs in the first trimester, the living cotwin usually continue to grow without any adverse effect to term. A survived twin is often mistaken for a singleton at delivery. Therefore, careful examination of the placenta is required. A blighted ovum can be sometimes recognized as a chorionic cyst-like sac on the fetal surface of the placenta when a fetus died in a very early stage of pregnancy.

When death occurs in the second trimester, the dead fetus becomes mummified and usually remains in utero until the cotwin's delivery. Most surviving cotwins continue to develop until near term without any trouble.

Finally, if death occurs early in the third trimester, the viable cotwin is frequently spontaneously delivered in a few days. Accordingly, the living twins associated with later intrauterine death of a cotwin will be premature.

DISCUSSION

Although the etiology of intrauterine death of one of monozygotic twins is yet to be known, a major cause is likely to be the twin transfusion syndrome, which can occur even in an early stage of pregnancy. If a severe type of the twin transfusion syndrome occurs in a very early state, fetal death may be induced. As the dead fetus is unable to produce coagulative factors at this time, the surviving fetus can continue to grow without any trouble.

The fetal death of a twin in the second trimester might be closely related to complications such as marginal or velamentous insertions of the umbilical cord, as are frequently found in twin placentas, especially monochorionic ones (Table 5). These complications frequently result in fetal morbidity [5,9] on account of circulatory disturban-

Table 5. Fetal Outcome in Velamentous and Marginal Insertions of the Umbilical Cord

	Cord insertion	N(%)	IUFD	Neonatal	Fetal
			Stillbirth N (%)	death N (%)	anomaly N (%)
Monochorionic twins (N=230)	Velamentous	56(24.3)	13(23.2)	4(7.1)	8(14.3)
	Marginal	25(10.9)	10(40.0)	5(20.0)	8(32.0)
Dichorionic twins (N=148)	Velamentous	14(9.5)	5(35.7)	0	3(21.4)
	Marginal	13(8.8)	0	0	0
Singletons (N=3458)	Velamentous	51(1.5)	5(9.8)	3(5.9)	7(13.7)
	Marginal	171(4.9)	4(2.3)	4(2.3)	7(4.1)
	Central or eccentric	3236(93.6)	44(1.4)	9(0.3)	84(2.6)

Table 6. Anastomoses in Twin Transfusione Syndrome and Fetal Outcome

Anastomoses	N	Survivors			Cases of anomaly (No. of fetuses)
		both	one	none	
Artery-artery	2	1	1	0	0
Artery-artery & artery-vein	4	3	1	0	1 (1)
Artery-artery & vein-vein	11	4	5	2	6 (7)
Vein-vein & artery-vein	0	0	0	0	0
Vein-vein	1	0	1	0	1 (1)
Artery-vein	3	2	0	1	1 (1)
Unknown or none	14	0	13	1	14 (17)
Total	35	10	21	4	23 (27)

ces induced by constriction on the cord vessels. In the cases of fetus papyraceus or late intrauterine fetal death, all dead fetuses, except one, had marginal or velamentous insertion of the cord, but such cord complications were also found in 4 of 12 survived twins. Therefore, other factors than cord complications should be considered as possible causes of fetal death during twin pregnancies.

Intraplacentar vascular anastomoses in monozygotic twin placentas develop into a twin transfusion syndrome in 15-36% of cases [4,10,11]. The severe type of this syndrome leads not only to marked dissimilarity in size between twins, but also to

death of the retarded twin in utero. The most severe type of the condition in our study [12] was the coexistence of vein-to-vein and artery-to-artery anastomoses (Table 6).

Of 29 cases with undetermined anastomosis, 14 were associated with a blighted ovum or a fetus papyraceus. In such cases, some types of vascular anastomosis might have existed previously and become obliterated after fetal death. In fact, we succeeded in finding the traces of vascular anastomosis in some cases.

Table 7 shows the outcome of the liveborn twins. Twelve of 21 cases were born alive without any complication and 2 were born prematurely but uneventfully. Seven of the 21 cases (33.3%) presented some complications. In 3 cases, one premature baby died of respiratory distress shortly after birth and 2 mature infants died later of severe anomalies. Two premature liveborn twins had transient hyperbilirubinemia, but they recovered soon by phototherapy with or without blood exchange transfusion.

Since Benirschke reported in 1961 [1] that a liveborn infant with a macerated still-born twin suffered from multiple thrombi in many organs, possible coagulation troubles in the surviving cotwin of a dead fetus have frequently been investigated [2,7,13]. In our series, these might have occurred only in two cases. Possibly, coagulation troubles are related to some type of intraplacental anastomoses between the twins, while the early onset of labor pains following the late intrauterine death of one of twins may result from circulatory disturbances in the placenta.

Table 7. Outcome of Liveborn Cotwins of a Dead Fetus

Fetal outcome	No. of cases	%
Mature and well	12	57.1
Premature and well	2	9.5
Neonatal or infant death	3	14.3
Hyperbilirubinemia	2	9.5
Congenital skin defect	1	4.8
Cerebral palsy	1	4.8

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