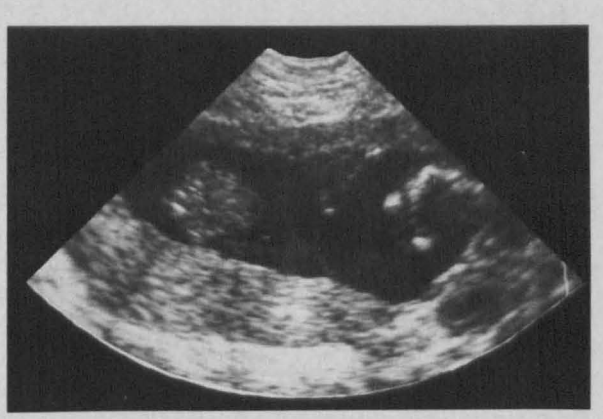

Figure 1. Gestational age at 13 weeks, with two visible sacs.

In order to assure continuity of care for this patient, a team consisting of the family physician and an obstetrician/gynecologist was established, and biweekly office visits were alternated between them. Weekly coagulation profiles, which included a complete blood count, platelet count, prothrombin time, partial thromboplastin time, fibrinogen and fibrin degradation products, were obtained, and all of these studies remained within normal limits throughout the pregnancy. There was no fever, cramping, abdominal pain, or further spotting on clinical follow-up. Fundal height grew consistently throughout gestation and reached a maximum 36 cm at 39 weeks. Monthly sonograms showed normal fetal growth of the live twin and gradual degeneration of the dead one until approximately 27 weeks' gestation (Figure 2). Thereafter, the remnant of the dead twin became virtually imperceptible.

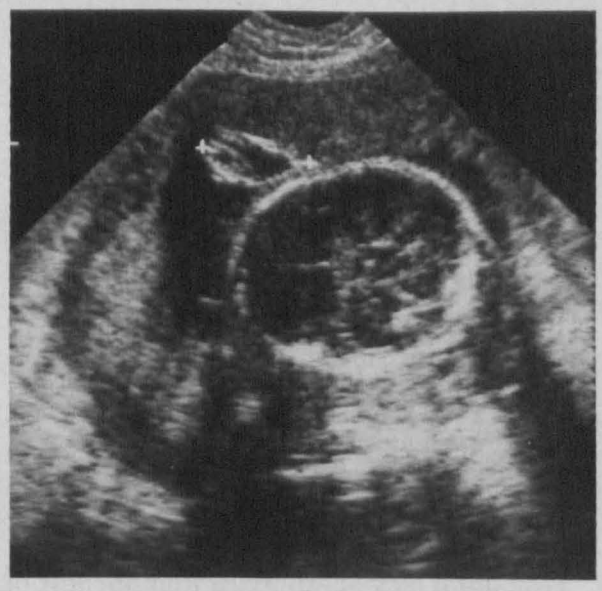

Figure 2. Twenty-seven weeks with a small disappearing fetal remnant as marked + +.

On September 15, 1984, spontaneous labor with intact membranes began and progressed normally. A 3000 g, vigorous female infant was delivered in a birthing chair over a midline episiotomy under local anesthesia. Apgar scores were 8 for 1 minute and 9 for 5 minutes. A single placenta followed, which measured 19 × 2 cm and had a central umbilical cord insertion. A second 8 cm infarcted placenta was adherent to the liveborn's placenta (Figure 3). This might have been interpreted as an accessory lobe; however, because of the previous sonographic identification of twins, a second placenta was expected. It showed extensive retroplacental clotting with membranous necrosis, supporting the suspicion of fetal death due to placental infarction. The uterine cavity was explored to ascertain complete delivery of the intrauterine contents. A second fetus was not identified in the uterine cavity.

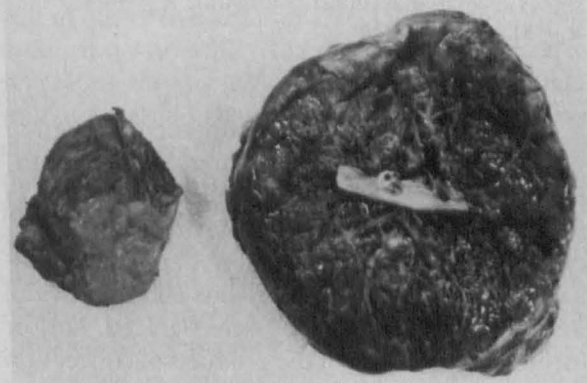

Figure 3. The small infarcted placenta at delivery was adherent to the term liveborn's placenta.

Both placentas were fixed in formalin, and a compressed fetus papyraceus was later recognized (Figure 4). Radiographs further defined the fetal structure as an apparently normal fetus, having died at approximately 13 weeks, with a femoral length of 11 mm (Figure 5). Figure 6 shows dissection of the fetus papyraceus.

Discussion

Landy, et al. in 1982 reported a well-organized investigation of the "vanishing twin syndrome."[1] Their summary of the literature and review of correspondence with members of the International Society of Twins Studies provided new insight about the potential frequency of twinning and fetal death.

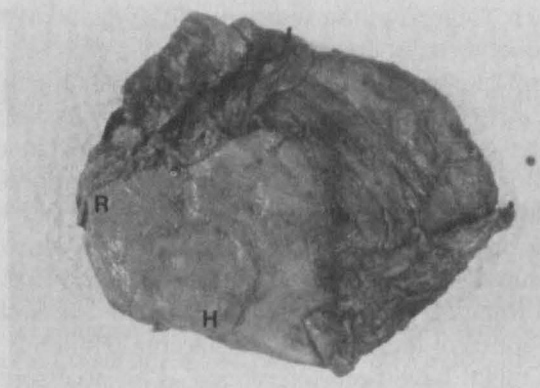

Figure 4. Visual examination of the small placenta shows several anatomical landmarks. H = Head, R = Ribs.

Twinning frequency and spontaneous abortion statistics are presumably inaccurate due to the unrecognized loss of some multiple pregnancies in utero, resulting in a single, surviving twin. Moreover, such twin gestations do not appear in ordinary birth statistics. Spontaneous abortion before 6 weeks may be clinically unrecognized. Lost or incomplete recovery of the products of conception in a spontaneous abortion at home, complete or partial expulsion of a second sac during spotting episodes with a surviving single gestation, therapeutic abortions without sonographic recognition of singleton versus multiple gestations, and unrecognized placental evidence of multiple gestation contribute to inaccurate twinning and fetal death statistics. Indeed, this case could have gone unrecognized if prenatal sonographic studies had not been done or the placenta had not been scrutinized closely.

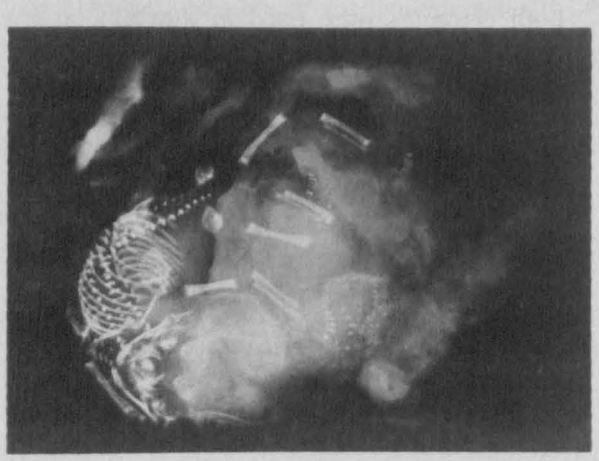

Figure 5. Radiographic evidence of fetal bony structure compressed in the smaller infarcted placenta.

The relative frequency of twins increases with maternal age and parity.[2,3] The dizygotic twinning rate rises to a maximum at age 37 years and falls abruptly near menopause. Martin's study supported the hypothesis that mothers of dizygotic twins have higher serum gonadotropin levels in the early follicular phase of the menstrual cycle than mothers of singletons.[4] This increase in twinning has been postulated to be due to the increase of gonadotropins with increasing age.[5,6] The sudden fall to zero near menopause is presumably related to aging and declining ovarian function.

Physiological explanations for the vanishing twin include reabsorption of an embryo or fetus papyraceus. Fetus papyraceus occurs as the result of a variety of lethal circumstances affecting one of the twins in the middle stages of pregnancy. Posner and Klein postulated that fetal deaths

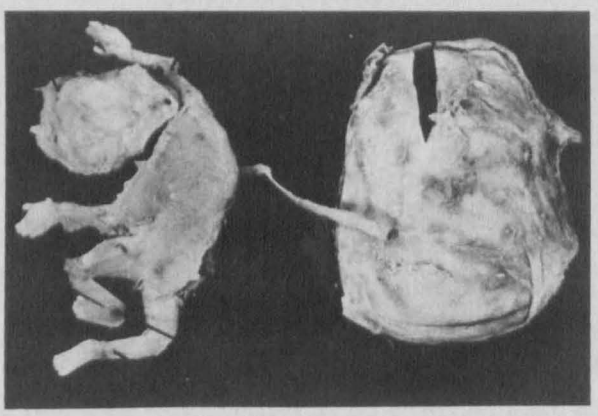

Figure 6. Dissection of the fetus papyraceus.

earlier than 3 months are resorbed, and later deaths fail to mummify in time.[7] Fetal deaths of twins may also be secondary to the transfusion syndrome, hemolytic disease, decidual vascular disease, velamentous cord insertion, cord entanglement or infarct, anomalies, trisomies, uteroplacental compromise, placental abruptio or infarct, or chronic maternal disease. Close examination of the placenta, cord, and membranes is essential to discern causes of death and also the presence of multiple gestation. Documentation of data is important if the rate of multiple gestation and the frequency of fetus papyraceus are to be identified.

Fetal death may be suspected from the loss of fetal heart tones or fetal movement and vaginal bleeding and confirmed by amniocentesis, sonog-

raphy, or examination of the placenta following delivery. Bleeding in the first trimester is common with fetal death and should signal attempts to identify a multiple gestation pregnancy. When prenatal diagnosis has been made, sonographic study should be used throughout the pregnancy to detect possible complications of the live fetus, to assess intrauterine growth, and to follow the fetal remnant.

There is a lack of accurate information about the natural history of coagulopathy associated with the death of one twin in utero. Maternal and fetal consumptive coagulopathy may occur with the absorption of thromboplastins released by the dead fetus. There are no case reports of maternal hypofibrinogenemia when fetal death of one twin occurred before week 5.[8] Reversal of consumptive coagulopathy has been reported with the use of heparin.[9] Typically, the clotting defect is characterized by decreasing plasma fibrinogen, increasing fibrin degradation products, and decreased platelet count, often followed by vaginal bleeding.[10,11]

Of more serious concern are the reported frequencies of morbidity and mortality in the surviving twin.[8-22] This, presumably, is related to the presence of vascular connections and coagulopathy in the live fetus.[17] Maternal chronic illness and placental insufficiency may also lead to fetal death, papyraceus formation, and the subsequent death of the second twin because of a hostile intrauterine environment.[12]

Yet, the presence of fetal death does not consistently cause a negative outcome in the live fetus. Landy, et al., 1982, found that the prognosis for the surviving twin is good.[1] Of 28 patients suspected of having a vanishing fetus, four pregnancies led to the death of the second fetus, leaving an 86.7 percent survival rate.[23] As Enbom noted, this is overly optimistic and does not take into account the potential rate of morbidity in the surviving twin.[24] Gindoff, et al. identified resorption of first trimester empty sacs and found that one empty sac does not adversely affect the development of a coexisting normal fetus, but multiple empty sacs seem to be associated with pregnancy loss.[25]

Neuman believed that the frequency of intrauterine fetal death in monochorionic twins is three times more likely to occur than in dichorionic twins.[26] Johnson's large-scale review of 34,677 deliveries showed a 64 percent survival rate for the co-twin associated with a single intrauterine death. Monochorionicity of the intrauterine death was present in 20 percent, supporting Neuman's claim.[22] Vascular anastomosis and cord accidents, as well as velamentous cord insertions, are seen more frequently in monochorionic placentas. The number of placentas in a twin pregnancy depends in part on zygosity. Dizygotic twins develop two separate placentas, whereas the majority of monozygous twins share a single placenta through which their circulation communicates. With two separate placentas there are no shared vascular beds, and the risk of exchange transfusion does not exist. One must assess the usefulness of sonographic identification of two separate placentas. In dichorionic pregnancies that implant side by side, which makes it difficult to detect two separate circulations, the physician may be able to identify membranes separating the two twins. Sonographic identification of a male and female twin pregnancy can also clarify the issue of separate placentas. With two placentas, one can reason that the death of a fetus followed by papyraceus formation would not be directly responsible for morbidity or mortality in a dizygous second twin and that a favorable outcome is likely. In our case, the sonographic studies revealed two separate placentas and sacs.

Elevated maternal serum and amniotic fluid alpha-fetoprotein, as well as the presence of a positive acetylcholinesterase band in fluid from the sac of a healthy fetus after the death of a co-twin have been reported.[27,28]. Early gestational sonographic examination with attention to the placenta may allow identification of a second gestational sac or fetus, and therapeutic abortion of a normal fetus because of persistently elevated alpha-fetoprotein secondary to the death of a co-twin could be avoided.

Multiple pregnancy can be diagnosed by sonography before 10 weeks.[29] Nakano, in his study of multiple gestations and fetal loss using ultrasonic tomography during 7–12 weeks' gestation, observed that misinterpretation may occur.[30] Technical ultrasonic artifact, distortion, and changing conditions in pregnancy may account for inaccurate diagnosis. Loss of an early pregnancy may result in plaque formation and appear sonographically similar to a fetus papyraceus. Skeletal formation can clarify this finding, except in fetal death under 10 weeks of age. Moreover, if one fully examines a placenta, earlier sacs may be identified. Accurate evaluation of multiple gestations with fetal loss will require precise sonographic technique followed by thorough exami-

nation of the placenta and uterine contents after delivery to confirm previous findings.

Landy, et al., 1986, attempted to establish a correct rate of disappearance of human fetuses based on sonography in the first trimester in 1000 pregnancies.[23] Their conclusion was that the frequency of multiple gestation is 3.29 percent to 5.39 percent greater than previously believed. This disparity was based in part on sonographic documentation of the vanishing twin, with a frequency of 21.2 percent. Two factors continue to prevent exact diagnosis of the multiple gestations in early pregnancy. Identifying gestational sacs before the 12th week of pregnancy is technically more difficult, and very early first trimester sonographic examinations are not standard practice. One could postulate that the true rate of twin conception is greater than is currently recognized.

Conclusion

We believe that the vanishing twin phenomenon constitutes a definite clinical problem and have reported here a patient in whom this occurred. Furthermore, we believe that this condition occurs more frequently than has been known in the past. It occurs at the end of the first and the beginning of the second trimester of pregnancy and is more likely to occur in older women who are multiparous. It should be suspected whenever there is first trimester bleeding, and early sonography should be used to make the diagnosis. Such pregnancies should be considered high risk and managed optimally with attention to repeated sonography and tests for coagulopathy that might affect the surviving twin and the mother.

The prognosis for the surviving twin is good and is improved in dizygotic pregnancies. A dizygotic twin pregnancy may be identified with chromosomal or sonographic studies looking for separate placentas or membranes and gender differences. It is worth noticing that a dead twin can cause increases in alpha-fetoprotein and acetylcholinesterase in the amniotic fluid of the survivor.

Careful examination of placentas after delivery can confirm the existence of a fetus that did not survive, especially when a fetus papyraceus is identified.

We gratefully acknowledge the assistance of Katherine Camacho Carr, R.N, C.M.N., M.S.; David Luthy, M.D.; and Robert Osborne, M.D., in the team management of this patient, and Chris Sherman for providing photographic detail.

References

1. Landy HJ, Keith L, Keith D. The vanishing twin. Acta Genet Med Gemellol (Roma) 1982; 31:179-94.
2. Weinberg W. Beitrge zur physiologie und pathologie der mehrlingsgeburten beim menschen. Arch Fuer Die Gesamte Physiologie Des Menschen und Der Tiere 1901; 88:346-430.
3. Bulmer MG. The biology of twinning in man. Oxford: Clarendon Press, 1970:78.
4. Martin NG, Olsen ME, Theile H, El Beaini JL, Handelsman D, Bhatnagar AS. Pituitary-ovarian function in mothers who have had two sets of dizygotic twins. Fertil Steril 1984; 41:878-80.
5. Milhan S Jr. Pituitary gonadotrophin and dizygotic twinning. Lancet 1964; 2:566.
6. Albert A, Randall RV, Smith RA, et al. The urinary excretion of gonadotrophin as a function of age. In: Engle ET, Pincus G, eds. Hormones and the aging process. New York: Academic Press, 1956:49-62.
7. Posner AC, Klein MA. Fetus papyraceus. Obstet Gynecol 1954; 3:106-10.
8. Hanna JH, Hill JM. Single intrauterine fetal demise in multiple gestation. Obstet Gynecol 1984; 63:126-30.
9. Romero R, Duffy TP, Berkowitz R, Chang E, Hobbins JC. Prolongation of a preterm pregnancy complicated by death of a single twin in utero and disseminated intravascular coagulation. N Engl J Med 1984; 310:772-4.
10. Skelly H, Marivate M, Norman R, Kenoyer G, Martin R. Consumptive coagulopathy following fetal death in a triplet pregnancy. Am J Obstet Gynecol 1982; 142:595-6.
11. Pritchard JA, MacDonald PC, eds. Williams Obstetrics. 16th ed. New York: Appleton-Century-Crofts, 1980:652.
12. Kindred JE. Twin pregnancies with one twin blighted. Am J Obstet Gynecol 1944; 48:642-82.
13. Camiel MR. Fetus papyraceus with intrauterine sibling death. JAMA 1967; 202:247.
14. Livnat EJ, Burd L, Cadkin A, Keh P, Ward AB. Fetus papyraceus in twin pregnancy. Obstet Gynecol 1978; 51:41s-45s.
15. Moore CM, McAdams AJ, Sutherland J. Intrauterine disseminated intravascular coagulation: a syndrome of multiple pregnancy with a dead twin fetus. J Pediatr 1969; 74:523-8.
16. Yoshioka H, Kadomoto Y, Mino M, Morikawa Y, Kasubuchi Y, Kusunoki T. Multicystic encephalomalacia in liveborn twin with a stillborn macerated co-twin. J Pediatr 1979; 95:798-800.
17. Benirschke K. Twin placenta in perinatal mortality. New York State J Med 1961; 61:1499-508.
18. Durkin MV, Kaveggia EG, Pendleton E, Neuhauser G, Opitz JM. Analysis of etiologic factors in cerebral palsy with severe mental retardation. I. Analysis of gestational, parturitional and neonatal data. Eur J Pediatr 1976; 81:67-81.
19. Melnick M. Brain damage in survivor after in-utero death of monozygous co-twin. Lancet 1977; 2:1287.

20. Hoyme HE, Higginbottom MC, Jones KL. Vascular etiology of disruptive structural defects in monozygotic twins. Pediatrics 1981; 67:288-91.
21. Schinzel AA, Smith DW, Miller JR. Monozygotic twinning and structural defects. J Pediatr 1979; 95:921-30.
22. Johnson S, Barss V, Driscoll S. Incidence and impact of single intrauterine death in multiple gestation. Lab Invest 1986; 54:29A.
23. Landy HJ, Weiner S, Corson SL, Batzer FR, Bolognese RJ. The vanishing twin: ultrasonographic assessment of fetal disappearance in the first trimester. Am J Obstet Gynecol 1986; 155:14-9
24. Enbom JA. Twin pregnancy with intrauterine death of one twin. Am J Obstet Gynecol 1985; 152:424-9.
25. Gindoff PR, Yeh MN, Jewelewicz R. The vanishing sac syndrome. J Reprod Med 1985; 31:322-5.
26. Newman HH. The physiology of twinning. Chicago: The University of Chicago Press, 1923: 147
27. Winsor EJ, Brown BS, Luther ER, Heifetz SA, Welch JP. Deceased co-twin as a cause of false positive amniotic fluid AFP and ACHE. Prenat Diagn 1987; 7:485-9.
28. Bass HN, Oliver JB, Srinivasan M, et al. Persistently elevated AFP and ACHE in amniotic fluid from a normal fetus following demise of its twin. Prenat Diagn 1986; 6:33-5.
29. Levi S. Ultrasonic assessment of the high rate of human multiple pregnancy in the first trimester. JCU 1976; 4:3-5.
30. Nakano H, Kubota S, Koyanagi T, Taki I. The prognosis of multiple pregnancy assessed by ultrasonic tomography. Acta Obstet Gynaecol Jpn 1981; 33:839-43.