Diagnosis and Management of Obstetrical Complications Unique to Multiple Gestations
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Obstetrical complications unique to multiple gestations pose a number of unique challenges. The presence of more than one fetus complicates the diagnosis and management of a pregnancy when one fetus has a structural or chromosomal abnormality, intrauterine demise, preterm premature rupture of the membranes, or delivers prematurely. Similarly, the diagnosis and management of monoamniotic twins and conjoined twins is challenging. These obstetrical complications that are unique to multiple gestations require thorough counseling of the expectant parents, as well as care by physicians with expertise in the management of multiple gestations.

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Common obstetric complications, such as preeclampsia, gestational diabetes, preterm labor, and intrauterine fetal growth restriction, occur more frequently in multiple gestations, yet the diagnosis and management of these conditions are similar as for singleton pregnancies. In contrast, unique challenges arise when one fetus in a multiple gestation has a structural or chromosomal abnormality, intrauterine demise, preterm premature rupture of the membranes, or delivers prematurely. The presence of more than one fetus in utero confounds the diagnosis and management of these serious obstetric complications. Furthermore, multifetal gestations are at increased risk for complications that are unique to monochorionic gestations, such as twin–twin transfusion syndrome, monoamnionicity, twin reversed arterial perfusion syndrome, and conjoined twins. This chapter addresses the management of some of these complications that are unique to multiple gestations.

Discordant Fetal Abnormalities
Diagnosis of an Abnormal Fetus

A single fetus in multiple gestation may be affected by both chromosomal and structural abnormalities. In a dizygotic pregnancy, each fetus has its own independent risk for a chromosomal or structural abnormality, so the overall risk of an abnormality increases as the number of fetuses increases. The risk of a chromosomal abnormality in a monozygotic multiple gestation is similar to the risk in a singleton. However, monozygotic gestations are at an increased risk of complications that are unique to monochorionic gestations, such as twin–twin transfusion syndrome, twin reversed arterial perfusion syndrome, and conjoined twins.

The goals of prenatal diagnosis in a multiple gestation are the same as a singleton: to identify fetal abnormalities that could lead to a couple’s decision to terminate a pregnancy or alter the management of the pregnancy and delivery, and to identify fetuses that might benefit from fetal or early neonatal therapy. However, prenatal diagnosis in multiple gestations poses a number of unique challenges. Maternal serum screening is not as reliable in twins and may not be interpretable in higher-order multiples. Invasive diagnostic procedures, such as chorionic villus sampling and amniocentesis, are technically more challenging in multifetal gestations. Finally, the presence of multiple fetuses makes the assessment of fetal anatomy at the time of prenatal ultrasound more difficult. For a more thorough review of prenatal diagnosis in multiple gestations, the reader is referred to the chapter by Drs. Cleary-Goldman, D’Alton, and Berkowitz.

Management of Discordant Fetal Abnormalities

When fetal abnormalities are detected in one fetus of a preivable multiple gestation, couples can choose between termi-
nation of the entire pregnancy, expectant management, or selective termination of the abnormal fetus. When the abnormality is diagnosed after viability, the options are limited depending on the jurisdiction and the extent of the abnormality. For example, as in a singleton pregnancy, the third trimester termination of an anencephalic fetus in a multiple gestation may be considered. Termination of the entire pregnancy in a multifetal gestation, as in a singleton gestation, can be performed surgically or by prostaglandin induction.

Expectant management should be pursued only after a thorough discussion and documentation of the risks of continuing the pregnancy. It is important that the patient be informed of how the anomalous fetus might affect the prognosis for the entire pregnancy. In a monochorionic pregnancy, counseling should include the risks to the normal cotwin in the event of a demise of the abnormal fetus. Regardless of chorionicity, an abnormal fetus may increase the risk of miscarriage or preterm delivery through complications such as polyhydramnios. The management of labor, in particular intervention for nonreassuring fetal status or malpresentation of the anomalous fetus, should be defined before delivery.

Selective termination should be offered when a fetal anomaly is detected before viability in a multifetal gestation. The benefits of selective termination need to be weighed against the procedure-related risk of miscarriage or premature delivery. Selective termination in multichorionic pregnancies is most commonly performed by fetal intracardiac injection of potassium chloride. In a multicenter study of 402 selective reductions by intracardiac or intrafunic injection of potassium chloride, the overall rate of pregnancy loss before 24 weeks was 7.5%. The pregnancy loss rates based on the gestational age at the time of the procedure were 5.4% when the procedure was performed between 9 and 12 weeks, 8.7% between 13 and 18 weeks, 6.8% between 19 and 24 weeks, and 9.1% when the procedure was performed beyond 25 weeks. These differences were not statistically significant, suggesting that the risk of pregnancy loss following selective termination is similar in all trimesters. Further support for the safety of second trimester selective reduction comes from a study that reported similar loss rates following multifetal reduction in the first trimester and selective termination in the second trimester. In monochorionic pregnancies, selective reduction using an intracardiac injection of postassium chloride can result in demise of the cotwin either by transplacental passage of potassium chloride or acute hemodynamic changes in the untreated fetus. For this reason, a number of methods involving cord ligation and coagulation have been developed for performing selective termination in monochorionic pregnancies. For a more thorough review of the methods of selective reduction in monochorionic multifetal gestations, the reader is referred to the chapter by Drs. Spadola and Simpson.

The diagnosis of an abnormal fetus in a multiple gestation creates a complex set of circumstances for a family that must decide between termination of the entire pregnancy, expectant management, or selective reduction of the abnormal fetus. Furthermore, these cases are challenging for physicians involved in the counseling and management of such patients. There are ethical challenges that require an appreciation for beneficence and respect for autonomy of both the mother and the fetuses. Before viability, a fetus can only be considered a patient if the mother confers that status on the viable fetus, including the fetus with an anomaly. As such, respect for the autonomy of the mother is the guiding principle. When a woman chooses selective termination, she is withholding the status of becoming a patient from the fetus with the abnormality, but not the other normal fetuses. Because the risk of selective termination to the remaining fetuses is minimal, the procedure does not violate the beneficence-based obligation to the remaining fetuses. After viability, the ethical standard of care is to optimize perinatal outcome through aggressive antepartum and intrapartum management. However, other options that could be considered include selective termination and nonaggressive management of the pregnancy. Selective termination of a viable abnormal fetus is only ethical when there is certainty of the diagnosis, and certainty of death or absence of cognitive development as an outcome of the anomaly. When the diagnosis and/or prognosis are less certain, both the physician and the mother have a beneficence-based obligation to the viable fetus. Nonaggressive management should be reserved for cases in which there is a high probability of death or absence of cognitive function, but not certainty of the diagnosis and prognosis. However, nonaggressive management may not be appropriate when an abnormal fetus shares a placenta with a normal fetus. In such a case, nonaggressive management might place the normal fetus at risk and, for this reason, challenges the beneficence-based obligation to the normal fetus.

**Intrauterine Demise of One Fetus in a Multiple Gestation**

The increasing use of ultrasonography in early pregnancy has revealed that the incidence of a spontaneous multiple gestation is greater than previously believed. In fact, twin gestations may occur in as many as 12% of all spontaneous conceptions. However, it is estimated that only 50% of twin pregnancies identified in the first trimester will result in two live born infants. In some cases the entire pregnancy will be lost, and in other cases only one embryo will be lost and the pregnancy will continue as a singleton gestation. When one embryo in a multiple gestation is lost, this is known as a "vanishing twin." The prognosis for the surviving fetus(es) is excellent.

**Risks to the Surviving Fetus(es) Following the Demise of One Fetus**

Although the loss of one embryo in a multiple gestation is common in the first trimester, most patients are asymptomatic or have only slight vaginal spotting following the loss. Fortunately, when the demise occurs early in pregnancy, it appears that the prognosis for the surviving fetus(es) is excellent.
On the other hand, second and third trimester loss of a fetus in a multiple gestation has been associated with adverse outcomes. The demise of one fetus in a multiple gestation at this time is relatively rare, complicating anywhere from 2% to 5% of twin pregnancies.\textsuperscript{13-15} Late demise of both fetuses in a twin pregnancy occurs infrequently.\textsuperscript{16} It is estimated that there is a threefold to fourfold increase in intrapartum death with monochorionic twins compared with dichorionic twins.\textsuperscript{13,17} Twins with structural malformations are more likely to die in utero than their anatomically normal cotwins.\textsuperscript{15} The intrapartum demise of a single fetus is more common in higher-order multiple gestations, complicating 14% to 17% of triplet pregnancies.\textsuperscript{18,19} Intrauterine demise of one fetus in a multifetal gestation increases the risk for adverse events in the surviving fetus(es).\textsuperscript{20,21}

The greatest risk to the surviving fetuses regardless of chorionicity is preterm delivery and the associated complications of prematurity. Overall 50% to 80% of surviving cotwins are delivered before term, and of these premature deliveries, 86% are due to preterm labor.\textsuperscript{12-14} However, for monochorionic twins, there is the additional risk of multiorgan damage in the surviving cotwin. Ischemic injury has been documented in the spleen, kidney, gastrointestinal tract, skin, and brain of surviving cotwins.\textsuperscript{22-26} It is estimated that up to 20% of the surviving fetuses in monochorionic twin pregnancies may experience neurologic injury, such as multicystic encephalomalacia. Multicystic encephalomalacia is the development of cystic lesions in the cerebral white matter in areas supplied by the anterior and middle cerebral arteries. While the exact mechanism of neurologic injury is uncertain, it is thought to be related to the shared vasculature in monochorionic pregnancies. The extent of shared placental vasculature likely influences the degree to which a surviving twin will be affected by demise of its cotwin. There are two main theories regarding the pathophysiology of neurologic injury in surviving monochorionic twins. An early theory was that the retained fetus creates thromboplastic materials that embolize through placental anastomoses to the surviving twin.\textsuperscript{17}

A more recent and widely accepted theory is that the sudden decrease in blood pressure that occurs at the time of fetal death results in “back-bleeding” from the live twin to the dead twin’s vasculature, causing transient but severe hypotension and hypoxemia in the surviving fetus.\textsuperscript{13} This theory has been supported by the development of anemia with normal coagulation studies in the cotwin following an intrauterine fetal demise of a single fetus in a monochorionic pregnancy.\textsuperscript{27,28} Given that the changes in pressure are likely instantaneous with fetal death, it is not surprising that immediate delivery does not prevent the development of multicystic encephalomalacia in the surviving twin. In fact, immediate delivery may increase the risk to the surviving cotwin through the complications of prematurity.

In dichorionic twin pregnancies, the risk of major perinatal morbidity or mortality to the surviving twin appears to be negligible apart from that related to preterm delivery.\textsuperscript{13} Dichorionic twins complicated by a single intrauterine demise do not seem to be at increased risk of neurologic injury, other than the risks related to prematurity.\textsuperscript{13} The surviving twin in a dichorionic pregnancy is likely protected from ischemic injury because of the separate vascular systems.

In addition to the potential risks to the surviving fetus(es) following the demise of a single fetus in a multiple gestation, there is the potential for maternal complications as well. In multiple gestations, similar to singletons, retention of a dead fetus has been estimated to result in a 25% incidence of maternal disseminated intravascular coagulation.\textsuperscript{19} However, only a few cases of maternal coagulopathy have been reported under these circumstances, so the 25% incidence is likely an overestimation. In a review of 16 pregnancies complicated by intrauterine fetal death of 1 twin, there were no cases of maternal disseminated intravascular coagulation.\textsuperscript{13} While transient fibrin-split products and hypofibrinogenemia have been observed in some twin pregnancies complicated by demise of one fetus, medical therapy or intervention was not necessary.\textsuperscript{14} Although the maternal risk appears to be low, periodic coagulation profiles are recommended to detect the early development of this complication. Following the demise of a cotwin, mothers do not appear to be at increased risk of infection as a result of the retained fetus; however, there is an increased risk of delivery by cesarean section secondary to nonreassuring fetal status.\textsuperscript{29} In addition, there are psychological considerations for the parents following the death of one fetus in a multiple gestation. Although the psychological impact of an intrauterine demise in a singleton pregnancy has been described, less is known about the effects of losing a fetus in a multiple gestation.\textsuperscript{30} Feelings of loss could be complicated by concerns over well-being for the surviving fetus(es), as well as relief that the entire pregnancy was not lost. Providers should be aware that the parents’ response to the loss of a single fetus in a multiple gestation is complex and that these families may benefit from counseling by a therapist with experience in high-risk pregnancies.

**Management of a Multifetal Gestation Complicated by a Single Fetal Demise**

After the demise of one fetus in a multifetal pregnancy, the clinical management is challenging and depends on chorionicity, gestational age, fetal lung maturity, and the development of fetal or maternal complications. The optimal timing of delivery and the frequency with which antenatal testing should be performed are controversial. Management strategies can be based on the chorionicity and whether the demise occurs before or after fetal viability. Regardless of the gestational age and chorionicity, multifetal gestations that are complicated by an intrauterine demise of one fetus need to be followed closely for the development of preterm labor.

When the demise occurs before viability, the management of a dichorionic twin pregnancy consists of expectant management until 37 weeks’ gestation. Steroids for fetal lung maturity and surveillance for preterm labor, fetal well-being, and fetal growth should be considered until viability is reached. The management of preivable monochorionic twin pregnancies after the demise of one fetus is more complicated. Parents should be counseled about the risks of multiorgan injury, including multicystic encephalomalacia, and
our inability to predict with certainty whether a surviving cotwin will be affected. Serial ultrasounds and magnetic resonance imaging (MRI) may be useful to detect multicystic encephalomalacia antenatally. When demise occurs in a monochorionic previable twin pregnancy, termination of the pregnancy should be offered.

Intrauterine demise of one fetus after viability, regardless of chorionicity, is not an indication for delivery. Administration of corticosteroids to enhance fetal lung maturity between 24 and 34 weeks is reasonable given the increased risk of preterm delivery. In dichorionic pregnancies, weekly assessment of fetal well being is reasonable. Increased fetal surveillance should be considered if other complications, such as growth restriction or oligohydramnios, develop. Again, the management of neonatal complications is not an indication for delivery. In cases of multiple gestations, delivery by cesarean section is recommended. Serial ultrasounds and MRI, performed approximately 2 to 3 weeks after the demise, may be useful to detect multicystic encephalomalacia antenatally. However, it is important for parents to understand that close fetal surveillance cannot guarantee an intact fetus. In general, elective delivery should be considered at 37 weeks’ gestation or once fetal lung maturity is demonstrated. Delivery by cesarean section should be performed for routine obstetric indications, as vaginal delivery is not contraindicated in this situation.11 In monochorionic pregnancies with impending demise of one twin, preterm delivery may be indicated to prevent neurologic injury to the survivor or to potentially salvage both twins.

Autopsy of the stillborn fetus should be offered and pathologic examination of the placenta(s) should be performed to confirm chorionicity. Monochorionic twin survivors should be followed closely by their pediatrician for any evidence of neurologic injury or organ damage.21

Preterm Premature Rupture of the Membranes in One Fetus of a Multiple Gestation

Incidence and Diagnosis of Membrane Rupture in a Multiple Gestation

Preterm premature rupture of membranes (PPROM) is a complication in one-quarter to one-third of all preterm births, making it a leading cause of infant mortality and morbidity. PPROM affects 10% of all pregnancies, including multiple gestations. Whereas there is extensive literature on the clinical course of singleton pregnancies complicated by PPROM, there are limited data on PPROM in multiple gestations. As a result, the optimal management of this complication in multiple gestations is not clear and clinical decisions are compounded by the presence of one or more fetuses with intact membranes. The approach to PPROM in a multiple gestation involves a balancing of risks to both fetuses in the ruptured and unruptured sacs, as well as the mother.

The incidence of PPROM in multiple gestations is twice that of singleton pregnancies.31 In a retrospective review of multiples between 19 and 36 weeks’ gestation, PPROM complicated 7.4% of twin pregnancies compared with 3.7% of singleton pregnancies.31 The incidence of PPROM in triplets was similar to that in twins at 7.4%.31 PPROM in the midtrimester of pregnancy also occurs more frequently in twins (1.4%) compared with singletons (0.5%).31 In the majority of cases of PPROM in multiple gestations, rupture occurs in the presenting amniotic sac, and the diagnosis is easily confirmed on clinical examination of vaginal pooling, ferning, nitrazine testing, and ultrasound assessment of amniotic fluid volume. The diagnosis can be more difficult when PPROM of the nonpresenting sac occurs, as leakage of fluid from the vagina may be intermittent. The exact incidence of PPROM in a nonpresenting sac is unknown, but it is thought to be limited to complications related to invasive procedures, such as amniocentesis or fetal blood sampling. Information on the incidence of PPROM in multiples based on chorionicity is also unknown. A complication that is unique to monochorionic, diamniotic twin pregnancies is rupture of the dividing membrane.32 This can result in a twin pair sharing an amniotic sac, with all the risks associated with monoamniotic twins. Usually this complication follows an invasive procedure, such as an amniocentesis. However, in some cases, there is no precipitating event to explain the finding of a monoamniotic gestation that was previously diamniotic. Although the exact incidence of diamniotic gestations becoming monoamniotic is unknown, spontaneous rupture of the separating membrane is likely a rare event. In addition to invasive procedures, rupture of the separating membrane in a multiple gestation may be the result of infection or disturbances in membrane development.32 The resultant monoamniotic pregnancies have been complicated by preterm delivery, with an average gestational age at delivery of 29 weeks, and an increased risk of perinatal death with an overall perinatal mortality of 44%.32

Latency and Risks Following Premature Rupture of the Membranes

Overall, the latency period between rupture of membranes and delivery of expectantly managed twins and singletons is similar. For twin gestations with PPROM between 19 and 36 weeks, Mercer and coworkers reported a median latency of 1.1 days.32 Of 91 twin pairs, 64% delivered within 48 hours, and 91% delivered within 7 days of membrane rupture.31 These observations were not significantly different from those observed in singleton gestations, which had a median latency of 1.7 days, with 90% delivered at 7 days.31 In a comparison of 116 twins with PPROM to matched singleton controls, the median latency period in twins was 11.4 hours compared with 19.5 hours in singletons.33 The shorter latency in twins was not clinically significant, as the proportion...
of twins and singletons delivered at 48 hours and 7 days was similar.  

As in singleton pregnancies, the duration of the latency period in multiple gestations is related to the gestational age at the time of membrane rupture. Latency is significantly prolonged when PPROM occurs before 30 weeks' gestation for both singletons and twins. In a comparison of 48 twins to 131 singletons, the latency periods were similar when PPROM occurred before 30 weeks' gestation (4.8 days versus 5.6 days).  

When PPROM occurred after 30 weeks, latency for twins was significantly shorter than that of singletons (2.5 days versus 3.7 days). Another study found that twins had a significantly shorter latency period, regardless of gestational age. In a matched comparison of 116 twins and 116 singletons with PPROM, a significantly shorter latency period was found in twins overall (11.4 hours versus 19.5 hours). In addition, twins had a significantly shorter latency than singletons when PPROM occurred before 30 weeks (27.6 hours versus 75.1 hours). Despite the shorter latency for twins compared with singletons, there were no significant differences in neonatal outcomes between the two groups.  

Both singleton and twin pregnancies complicated by PPROM are at increased risk for infection, cord accidents, and placental abruption. Mercer and coworkers found no significant difference in the incidence of amnionitis (15% versus 23%), cord prolapse (4% versus 2%), or placental abruption (1% versus 6%) between singleton and twin pregnancies. Overall, there are no differences in perinatal morbidity or mortality between singletons and twins with PPROM. However, significant differences in neonatal morbidity have been reported between the presenting and nonpresenting fetuses. In twin pregnancies complicated by PPROM, the nonpresenting twin is at increased risk for respiratory distress syndrome (20.9% versus 7.1%) and prolonged oxygen therapy (43.6% versus 22.6%) when compared with the presenting twin. It is unknown whether this reflects accelerated fetal lung maturity after PPROM in the presenting twin or a relative immaturity in the nonpresenting twin. The finding of increased respiratory morbidity and lower Apgar scores in the nonpresenting twins does not appear to be related to the route or mode of delivery. Despite PPROM of the presenting sac, there were no statistical differences in infectious morbidity between the presenting and nonpresenting twins. Based on these findings, patients should be counseled that the nonpresenting fetus appears to be at higher risk for respiratory complications following PPROM of the presenting fetus.  

The most critical aspect to consider in the management of PPROM in multiple gestations is the gestational age at the time of membrane rupture. It is important to weigh the risks of prematurity versus the risks of infection and the potential adverse effects of oligohydramnios. Infectious complications from PPROM may affect the mother, the fetus in the ruptured sac, as well as the coetus(es) with intact membranes. The risks of oligohydramnios pertain to the fetus in the ruptured sac, and these are dependent on gestational age and the amount of remaining amniotic fluid. Oligohydramnios early in gestation may be associated with the development of pulmonary hypoplasia, skeletal deformities, and amniotic band syndrome.  

**Management of PPROM in a Multiple Gestation**  

PPROM can be classified as previable (before 23 completed weeks' gestation), remote from term (23 to 31 weeks' gestation), and near term (32 to 36 weeks' gestation). The contemporary management of singletons with PPROM is well established and can be applied to multiple gestations. Regardless of gestational age, once the diagnosis of PPROM is confirmed, digital examinations of the cervix should be avoided to increase latency and decrease infectious morbidity. Visualization of the cervix by sterile speculum should be performed, at which time appropriate cultures can be obtained. Patients should then be carefully evaluated for any evidence of advanced labor, infection, placental abruption, and nonreassuring fetal status. Apart from nonreassuring fetal status at a previable gestational age, the development of these complications warrants expeditious delivery.  

The management of multiple gestations with PPROM near term is similar to that of singletons. When PPROM occurs after 34 weeks, the perinatal complications related to prematurity are outweighed by the risks of chorioamnionitis and fetal loss due to cord compression, and therefore delivery is recommended. Antibiotic prophylaxis for group B streptococcus should be given if culture results are not available, or broad-spectrum antibiotics if there is any evidence of intrauterine infection. PPROM in multiple gestations between 32 and 34 weeks warrants delivery if fetal lung maturity is documented on amniotic fluid from vaginal pooling or from amniocentesis from the fetus with the ruptured sac, and by amniocentesis from the sac of the other fetus(es). If fetal lung maturity studies are negative, discordant, or if fluid is not available from all fetuses, then conservative management with close fetal surveillance and maternal monitoring as an inpatient are recommended. Consideration should be given to adjunctive antibiotic therapy to treat or prevent ascending infection to prolong the pregnancy. The effect of steroids on fetal lung maturity in multiple gestations has not been studied. Antenatal corticosteroids may be administered to gravidas with PROM up to 34 weeks of gestation. Tocolysis is not generally used at this gestational age because the risk of significant perinatal morbidity is low. In the absence of labor, placental abruption, nonreassuring fetal testing, or chorioamnionitis, delivery should be considered once 34 weeks of gestation is reached.  

Similarly, the management of multiples with PPROM remote from term parallels that of singletons. When PPROM occurs between 23 and 31 completed weeks, conservative management is recommended to prolong the pregnancy since delivery before 32 weeks is associated with significant risks of neonatal morbidity and mortality. Inpatient fetal and maternal surveillance, a single course of antenatal corticosteroids, and antibiotics to prolong latency are all recommended. Ideally, conservative management of multiple gestations complicated by PPROM should be performed at
centers that are capable of performing immediate cesarean deliveries and providing appropriate neonatal care. If necessary, patients should be transferred early in the course of management to avoid emergent transfer in the event that complications develop. Although data regarding the role of tocolytics in multiple gestations with PPROM are lacking, the use of prophylactic tocolytics to prolong pregnancy and allow time for the benefits of corticosteroids and antibiotics is reasonable. Again, in the absence of labor, placental abruption, nonreassuring fetal testing, or chorioamnionitis, delivery should be considered once 34 weeks of gestation is reached or between 32 and 34 weeks if fetal lung maturity is documented for each fetus.

In contrast to PPROM remote from term or near term, strategies for managing PPROM in multiple gestations at previable gestational ages are less uniform than those for singleton pregnancies. Currently, there are no data with which to make recommendations regarding initial conservative management in these cases. Options for previable PPROM range from expectant management to termination of the entire pregnancy. Patients should be counseled about the maternal and fetal risks and benefits of expectant management versus termination, and consent for any management plan should be obtained. If termination is declined, outpatient management after initial observation in the hospital to exclude infection or abruption is a reasonable option. As in singleton pregnancies with PPROM, infection must be excluded and prompt delivery for any evidence of intrauterine infection is recommended. Weekly visits to assess maternal and fetal status should be performed until fetal viability is reached. Once viability has been obtained, patients are usually readmitted to the hospital for closer maternal and fetal surveillance. Psychological support should be made available for patients and their families when faced with the difficult management choices and decisions dealing with PPROM in multiple gestations at previable gestational ages.

Investigational strategies, including selective reduction of the fetus in the ruptured sac or preterm delivery of this fetus followed by aggressive attempts to retain the remaining fetuses, have been reported when PPROM complicates a previable multiple gestation. Selective reduction of a previable fetus following rupture of membranes in a multiple gestation was first described by Dorfman and coworkers. This approach was based on the assumption that the risk of ascending infection would be reduced if the continuous leakage of amniotic fluid from the ruptured fetus was eliminated, and that a lower risk of infection would lead to an improved chance of the pregnancy progressing to viability. De Catte and coworkers reported on the outcome of 12 pregnancies in a comparison of expectant management versus selective reduction in multichorionic multiple pregnancies with PPROM between 13 and 20 weeks’ gestation. In 9 cases, patients were managed expectantly without antibiotics or tocolytics. Selective termination was performed in 3 cases within 3 days of membrane rupture, followed by a 5-day course of antibiotics. Weekly steroids were administered beginning at 24 weeks’ gestation. The mean interval between membrane rupture and delivery in the expectantly managed group was 7.4 weeks with only 3 survivors. In the selective reduction group, 2 of the 3 pregnancies progressed beyond 33 weeks. Although the numbers are small, selective reduction of a previable fetus in a ruptured sac of a multichorionic gestation may be a reasonable option in carefully selected cases; however, further study is needed before this strategy can be recommended.

Aggressive approaches to prolong pregnancy in cases of previable PPROM include the use of tocolytic therapy, antibiotics, and cervical cerclage. In a series of 8 multifetal gestations complicated by PPROM before 24 weeks’ gestation, Arias and coworkers reported on efforts to delay delivery of the retained fetuses after delivery of the ruptured fetus. Broad-spectrum antibiotics were administered, and, after delivery of the presenting fetus, either spontaneously or by induction, the umbilical cord was ligated as high as possible within the cervical canal. Tocolytics were given for uterine contractions and a cerclage was placed once contractions ceased. Prophylactic tocolysis with indomethacin was then administered for 48 to 72 hours and antibiotics were continued for 1 week, after which time patients were discharged to home. The authors reported a mean latency of 48 days. Although this aggressive approach successfully delayed delivery of the retained fetuses, subsequent reports by this same group suggested less favorable results.

The optimal management of previable PPROM in multiple gestations remains unclear. Expectant management results in an extremely high pregnancy loss rate and exposes the mother to the risks of infection and the complications of prolonged bed rest. In addition, pregnancies that progress to fetal viability are still subject to high neonatal morbidity and mortality from oligohydramnios, infection, and prematurity. Given the complexities of previable PPROM, an individualized approach is recommended. In certain circumstances, termination of the entire pregnancy may be the best approach. Although aggressive interventions, such as tocolysis and cerclage after delivery of a previable fetus with PPROM, may be considered, the benefits of these techniques need to be adequately demonstrated before they can be recommended. Similarly, selective reduction, which appears to have a higher success rate than expectant management, requires further study.
limited to case reports and, as such, was subject to bias of only reporting favorable outcomes. More recently, larger case series have been published. The largest series from a single practice is a review of 24 consecutive interval deliveries involving twin and triplet gestations. In this series, a uniform approach of tocolysis, antibiotics, and cerclage was used. These authors reported a mean interval delivery time of 36 days, with a range of 3 to 123 days. Although there were no survivors in the 16 first-born fetuses that delivered before 24 weeks’ gestation, 8 of the 18 retained fetuses in this group survived. In the 9 pregnancies in which the first-born delivered after 24 weeks, 4 infants survived. Of the 9 retained fetuses in this group, all survived. Overall, the perinatal mortality rate of first-born infants, regardless of gestational age, was 84%. The perinatal mortality rate of the infants born after delayed delivery was significantly less at 37%. In contrast to this favorable outcome, a series of 14 cases of attempted delayed interval delivery over a 12-year period reported less encouraging results. In this retrospective study, the median latency was only 2 days, with a range of less than 1 to 70 days. There was only 1 survivor among the first-born fetuses. Of the 19 retained fetuses, 7 survived until hospital discharge; however, only 1 of these neonates was discharged without major neurologic sequelae. Maternal morbidity included 2 cases of placental abruption and 8 cases of infectious morbidity, 1 of which resulted in septic shock and multiorgan failure. This study raises serious concerns about the fetal outcome and maternal risks associated with a delayed interval delivery; however, both studies are limited by a small number of cases. A second limitation of these studies is that they both include cases that were collected since 1991 and, therefore, may not represent contemporary outcomes because of improved perinatal care.

Two larger retrospective cohort studies using the “matched multiple birth file” prepared by the Centers for Disease Control and Prevention’s National Center of Health Statistics were recently published. This database includes information on all twin live births, still births, and infant deaths in the United States during the period of 1995 to 1998. One study identified 200 delayed twin deliveries in which the first twin was delivered between 17 and 29 weeks’ gestation. In this series, there was a significant improvement in the survival of the second-born twin when the latency period exceeded 2 days. When the outcomes were compared with matched twin pregnancies in which the second twin was delivered on the same or next calendar day, 56% of the delayed second twins survived to 1 year of age compared with only 24% of the nondelayed second twins. When the first twin delivered between 17 and 23 weeks, 32% of the delayed second twins survived to 1 year of age, compared with only 7% of the nondelayed second twins. Similarly, when the first twin delivered between 24 and 29 weeks, 89% of the delayed second twins survived to 1 year of age, compared with 66% of the nondelayed second twins. This study suggests that delayed interval delivery results in a significant improvement in fetal outcome when the latency exceeds 2 days, which might be attributable to the beneficial effects of steroids for fetal lung maturity. Using the same database, a second group identified 258 sets of twins in which the first twin was delivered vaginally between 22 and 28 weeks and the second twin was delivered at least 1 week later. In this series, there was only a decrease in perinatal and infant mortality for those second twins in which the first twin was delivered at 22 to 23 weeks and when the delivery interval was between 1 and 3 weeks. There was no benefit in outcome when the interval exceeded 3 weeks for any gestational age, or when the first twin was delivered at 24 to 28 weeks and the interval exceeded 1 week. It should be noted that, unlike the series that showed a benefit when the latency exceeded 2 days, this study defined a delayed delivery as only those deliveries in which the interval exceeded 1 week. For the cases in which the first twin delivered between 22 and 23 weeks, a delay of at least 1 week likely resulted in the second twin reaching viability and possibly receiving steroids for fetal lung maturity. After 24 weeks, a delayed delivery less than 1 week includes those pregnancies in which the second twin was retained for a period that would allow for the beneficial effects of steroids. Thus, it cannot be concluded that attempts at delayed interval delivery between 24 and 28 weeks for the administration of steroids do not benefit the second twin. This study also showed that the rates of small-for-gestational-age births in the second twin increased with increasing delivery interval. Although an increasing interval may be associated with impaired growth, other factors, such as multiple courses of steroids for fetal lung maturity, may have contributed to decreased fetal growth. Unfortunately, neither of these studies was able to analyze the different management strategies or the maternal risks associated with a delayed interval delivery.

Table 1: Unfavorable Conditions for Delayed Interval Delivery

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<td>Gestational age &lt;28 weeks</td>
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<td>Chorioamnionitis</td>
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<td>Placental abruption</td>
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<td>PPROM of retained fetus(es)</td>
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<td>Advanced cervical dilation</td>
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<td>Nonreassuring fetal testing</td>
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Candidates for Delayed Interval Delivery

Attempts to delay delivery of the remaining fetuses following the birth of the presenting fetus in a multiple gestation should only be undertaken in the absence of any maternal or fetal indications for delivery (Table 1). Consideration should be given to the number of fetuses, the gestational age at the time of delivery of the first fetus, placental chorionicity, evidence of intrauterine infection, rupture of membranes of the retained fetus(es), placental abruption, and maternal and fetal well-being.

Delayed delivery has been reported primarily in twin gestations; however, delayed delivery in higher-order multiples has been attempted. In a 1999 review of interval deliveries in multiple gestations, delayed delivery was attempted in 48 cases of twins, 12 cases of triplets, 2 cases of quadruplets, and a single case of quintuplets. Despite the limited data on higher-order multiples, attempts to delay delivery are reason-
able in the absence of contraindications irrespective of the starting number of fetuses.

The gestational age at the time of delivery of the first fetus should be at least 16 to 18 weeks before prolonged interval delivery is considered.\(^{39,40}\) In general, delayed delivery is not recommended beyond 28 weeks, as the risks of prolonging the pregnancy may not outweigh the potential benefits.\(^{40,45}\) Therefore, candidates for interval delivery are those who deliver the first fetus late in the second trimester or early in the third trimester.

It has also been recommended that delayed delivery be attempted only in multiples where each fetus has its own placenta.\(^{39,40}\) The concern is that the shared placental circulation in a monochorionic twin gestation might predispose the retained twin to neurological injury due to hypotensive ischemia at the time of delivery.\(^{27}\) However, normal neurologic development has been reported in the surviving cotwin of a monochorionic–diamniotic after delayed delivery.\(^{39}\) It is possible that umbilical cord ligation at the time of delivery of the first twin of a monochorionic pair might prevent the hemodynamic changes that are believed to cause neurological injury in the cotwin. Even with immediate umbilical cord ligation, patients and their families should be counseled about the potential risk of neurological injury to the retained twin in a monochorionic pregnancy. Although delayed delivery is reasonable to offer patients with previable delivery of a monochorionic twin, the benefits of delayed delivery beyond 24 weeks are uncertain.

One of the most common reasons for discontinuing attempts at delayed delivery in multiple gestations is chorioamnionitis, which complicates approximately 36% of cases.\(^{47}\) Although there have been successful outcomes in rare cases in which chorioamnionitis was treated with antibiotics, maternal sepsis complicates approximately 5% of cases of attempted delayed delivery.\(^{47}\) Despite isolated case reports of successful interval deliveries when chorioamnionitis is suspected, such a strategy cannot be recommended given the risks of both fetal and maternal morbidity and mortality.

Delayed delivery following PROM in the retained fetus(es) is controversial. In a series of 8 cases complicated by PROM before 24 weeks' gestation, aggressive management to prolong the pregnancies resulted in a mean interval time of 48 days, with a range of 8 to 114 days.\(^{39}\) Management included the use of broad-spectrum antibiotics, high cord ligation after birth of the first fetus, both prophylactic and therapeutic tocolysis, and cerclage placement. With this approach, 6 fetuses in 5 of the 8 pregnancies survived. An addendum to this series included 3 additional patients who were enrolled before publication who had less favorable results. No significant maternal morbidity was reported. Because of the mixed results in this series, further evaluation of aggressive attempts to delay delivery in cases of PROM in the retained fetus(es) is needed before such a strategy can be recommended.

**Approach to Delayed Interval Delivery**

If a delayed delivery is attempted, it is important to obtain detailed informed consent from the parents. Counseling should include a discussion of the benefits of a successfully prolonged pregnancy, specifically that even modest intervals during critical gestational ages can lead to a significant decrease in neonatal morbidity and mortality. However, patients should also be informed of the risk of delivering a perivable baby after a relatively short interval with severe morbidity related to prematurity, instead of delivering a previable fetus that would not survive. Once informed consent is obtained, consideration should be given to the use of steroids for fetal lung maturity, prophylactic antibiotics, tocolysis, cerclage, and inpatient management. If not documented previously, a detailed anatomic survey of the remaining fetus(es) is warranted as this may influence a family's decision to pursue aggressive management. In addition, aggressive attempts to prolong pregnancy should not be undertaken in cases of placental abruption, preeclampsia, chorioamnionitis, or nonreassuring fetal testing.

Ideally, candidates for delayed interval delivery should be identified before the first fetus is born to allow appropriate time for counseling and preparing a management strategy.\(^{39}\) After delivery of the presenting fetus, the umbilical cord should be ligated with an absorbable suture placed as high as possible inside the cervix. In women with a pregnancy in each horn of a didelphys uterus, the placenta can be successfully removed after delivery of one twin, leaving the remaining twin and placenta undisturbed. Although the benefits of steroids for fetal lung maturity have not been specifically studied in multiple gestations, they are recommended for patients at risk of preterm delivery between 24 and 34 weeks of gestation with intact membranes, or between 24 and 32 weeks of gestation for patients with ruptured membranes.\(^{48}\) Similar guidelines should be applied to cases of delayed interval delivery in multiple gestations. Patients having had a previable delivery should receive a single course of steroids once the retained fetus(es) reaches 24 weeks of gestation. The use of prophylactic antibiotics in these cases is controversial. The rationale is to prevent ascending infection, thereby prolonging the pregnancy for the remaining fetus(es). Before antibiotic prophylaxis is initiated, it is important to exclude chorioamnionitis in the remaining fetus(es). If there is any uncertainty about intrauterine infection, an amniocentesis should be considered. Although the data are limited, routine cultures of the vagina and cervix should be obtained. If prophylactic antibiotics are used, they should be broad-spectrum, such as ampicillin-sulbactam and metronidazole, to cover the wide variety of bacteria that have been isolated from the cervix, placenta, and amniotic fluid in cases of delayed interval delivery. Since the benefits of prophylactic antibiotics in cases of delayed interval delivery are uncertain, a short course of broad-spectrum antibiotics in the absence of infection is reasonable. Similarly, the role of tocolysis in delayed interval deliveries is uncertain. There is enormous variation in the literature regarding the choice of tocolytic, when tocolysis was started, and the length of time patients were treated. Indomethacin may mask the clinical signs of chorioamnionitis and, therefore, is not the ideal choice when tocolysis is used in cases of delayed interval delivery. Although no single strategy has proven to be beneficial, it is reasonable to use...
short-term tocolysis after delivery of the first fetus and avoid the use of prolonged tocolytic therapy in most cases.

The benefit of cerclage placement in multifetal pregnancies after delivery of one fetus is controversial. It has been suggested that a cerclage will close the cervix and decrease the risk of an ascending infection. Another potential benefit of a cerclage is that it adds stability to the cervix and support for the remaining fetus(es), particularly if the cervix remains dilated after delivery of the first fetus. In contrast, others have argued that placement of a cerclage may increase the risk of initiating labor and chorioamnionitis. In a review of the literature, immediate cervical cerclage placement after the first delivery was associated with a significantly longer interdelivery interval, 25 versus 8 days, without increasing the risk of intrauterine infection.47 If a cerclage is considered, it is recommended that it be done soon after delivery of the first fetus, as more favorable outcomes have been observed when the cerclage is placed within 2 hours.46 While most cerclages are performed using the McDonald technique, a two-cerclage technique using both a Shirodkar and McDonald cerclage may be employed to support the entire length of the cervix.49 Regardless of the elapsed time from delivery of the first fetus or the technique used, it is important that uterine contractions have ceased, placental abruption and infection have been excluded, and the well-being of the retained fetus(es) and mother have been confirmed before a cerclage is attempted.

Whether patients attempting a delayed delivery require hospitalization until the remaining fetus(es) has been delivered is uncertain. Advantages of outpatient management are a more familiar and comfortable environment for the patient and an overall reduction in hospital costs. All patients who have delivered one fetus of a multiple gestation should be hospitalized for an initial period of observation to monitor for any evidence of preterm labor, chorioamnionitis, nonreassuring fetal status, or placental abruption. Fetal assessment of the remaining fetus(es) will vary depending on gestational age, but may include intermittent heart rate auscultation, nonstress tests, or biophysical profiles. Fetal growth should be followed closely by ultrasound given the risk of a small-for-gestational-age birth in the retained twin.43 An individualized plan based on gestational age, degree of cervical dilation, compliance with modified bed rest, and social support should be considered before outpatient management is pursued.

Patients who are candidates for an attempt at delayed interval delivery should be carefully selected and counseled about the risks and benefits of this approach. Based on the current literature, it seems reasonable to counsel them that in the absence of any maternal or fetal indications for delivery, attempts to delay delivery of a preivable fetus might result in a more favorable outcome for the retained fetus(es). And, that delaying delivery between 24 to 28 weeks’ gestation for at least 2 days to complete a course of steroids for fetal lung maturity might also result in an improved outcome for the retained fetus(es). However, the advantage of extending the interval beyond 1 week at this gestational age is less certain. Patients need to be aware that there are risks of maternal infection and delivering a baby with serious neurological morbidity when a delayed interval delivery is attempted. At the present time, the optimal management of a delayed interval delivery is not known, but broad-spectrum antibiotics, tocolysis, and immediate cerclage is a reasonable strategy. Importantly, the management of multiple gestations following the preterm delivery of one fetus must be individualized according to the condition of the mother, the remaining fetus(es), and the wishes of the parents.

**Monoamniotic Twins**

Monoamniotic twins arise when the blastocyst splits between day 8 and day 13 postfertilization, after the establishment of the chorionic plate and amniotic sac. Monoamniotic twins occur in approximately 1% of all monzygotic twin pregnancies. The literature points to a predominance of female infants. In addition to the other complications that can occur in monochorionic gestations, monoamniotic twins are at elevated risk for fetal death due to cord entanglement. Because monoamniotic twins are rare, reports in the literature are limited to case reports, case series, and single-institution experiences over several decades, with more recent articles focusing on multicenter collection of cases or summation of cases from the literature during the most recent era of advanced neonatal care.

**Diagnosis of Monoamniotic Twins**

Correct diagnosis is crucial with respect to appropriate counseling and proposed management. Before imaging modalities, the diagnosis was made postnatally, and occasionally during the intrapartum period. Rodis and colleagues listed several ultrasound criteria that should be satisfied to diagnose monoamnionicity in utero50 (Table 2). Although ultrasound technology and operator experience have improved to the point that chorionicity and amnionicity can be correctly predicted in virtually all cases during the first trimester, the positive predictive rates are less later in pregnancy.31-35 The presence of a single yolk sac can significantly aid in early first trimester determination of amnionicity, and raises the suspicion for monoamniotic twins, as this structure can usually be seen before visualization of a dividing amnion.58,59 The detection of cord entanglement, especially with color Doppler and three-dimensional imaging also improves the accuracy of antenatal diagnosis.60-67 Other methods described in the literature to determine amnionicity include sequential amniocentesis with injection of dye near one fetus, and repeat am-

<table>
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<th>One placenta</th>
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<tr>
<td>Both fetuses with same sex</td>
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<tr>
<td>No dividing amniotic membrane should be visualized</td>
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<tr>
<td>Each fetus must have adequate amniotic fluid surrounding itself</td>
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<td>Both fetuses must be able to move freely within the uterine cavity</td>
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**Table 2 Ultrasound Criteria for Confirming Monoamniotic Twins**
niocentesis at the opposite quadrant of the uterus to assess for diffusion of dye, intraamniotic injection of air, creating microbubbles which act as an ultrasound contrast medium, and amniography, utilizing radiopaque dyes followed by flat plate x-rays or CT to assess for diffusion of the dye, or ingestion of dye by both fetuses. Although MRI and fetoscopy have been utilized for diagnostic purposes in other fetal conditions, there are no articles describing their use for making the diagnosis of monoamniotic twins.66-72 It is interesting to note that twin–twin transfusion syndrome is reported as occurring rarely in monoamniotic gestations. This may be due to an increase in numbers of all types of vascular connections present in the monoamniotic placenta when compared with the monochorionic diamniotic placenta.73-75 For a more thorough review of the twin–twin transfusion syndrome, the reader is referred to the chapter by Drs. Harkness and Crombleholme.

Management of Monoamniotic Twins

Based on older literature, fetal mortality rates associated with monoamniotic twins have been reported to range from 30% to 70%.76-79 Although all of these reports are retrospective, there is a trend noted toward improved outcome over time. Carr and colleagues reported their experience with 24 sets of monoamniotic twins from 1967 to 1988. Although double survival occurred in only 46% of cases, they noted no intrauterine death after 30 weeks' gestation.80 Similarly, Tessen and colleagues reported on 20 sets of monoamniotic twins from 1961 to 1989, noting a 65% double survival rate and no stillbirths after 32 weeks' gestation.81 One theory suggested that, at a relatively late gestational age, movement of the fetuses was restricted thus conferring a protective effect. However, Tessen and colleagues noted after submission of their article a double death at 35 weeks, again casting doubt on optimal management. Rodis and colleagues reported on a group of 13 sets of monoamniotic twins from 1986 to 1996. With accurate prenatal diagnosis, serial sonography, and antenatal testing (9 of the 13 underwent daily nonstress tests), double perinatal survival occurred in 92%.82 Similar reports summarizing management of small case series (<34 sets of monoamniotic twins per article) from the late 1980s to early 2000 cited perinatal survival rates from 60% to 98% (excluding abnormal fetuses).61,83-86 More recently, Roqué and colleagues summarized the outcomes of 133 monoamniotic sets of twins from 1990 to 2002. Of note, perinatal mortality in infants without anomalies was 18%, with 7% versus 21.6% mortality rates for those with and without an antenatal diagnosis of cord entanglement. Also noted was a perinatal death rate of 10.2% for pregnancies that continued beyond 32 weeks. With respect to morbidity, 25% of the surviving twins after demise of the cotwin had evidence of neurological injury. Within this series, fetuses with anomalies were noted to have a 43% perinatal mortality rate. Heyborne and colleagues collected 96 monoamniotic sets from 1993 to 2003, summarizing outcome during a period reflecting contemporary neonatal intensive care unit (NICU) care and greater use of antenatal steroids for fetal benefit. The perinatal death rate excluding lethal anomalies was 12.6%. A subanalysis of 87 sets of twins that continued after 24 weeks' gestation compared twins that were either electively admitted for management with those that required an indicated admission. Elective admission patients underwent nonstress testing 1 to 3 times daily, whereas outpatient testing was performed 1 to 3 times weekly. No fetal deaths were noted in the 43 sets that were electively admitted, whereas a 14.8% (13/88) death rate was noted for the indicated admission group, with the latest death occurring at 33 weeks' gestation.73

Antepartum Management of Monoamniotic Twins

The occurrence of cord entanglement resulting in compression and occlusion is clearly the significant contributor to morbidity and mortality. Although the literature does not give clear recommendations, smaller case series describe reserving inpatient management only when indicated.83,85,86 On the other hand, the larger series have led investigators to recommend elective inpatient management in the hopes of providing rapid response to nonreassuring fetal testing.73,82 At our institutions, admission for daily testing is recommended at whatever gestational age the woman would desire intervention for nonreassuring fetal testing, usually at 26 weeks' gestation.

Several investigators have reported and/or incorporated the practice of medical amnioreduction by use of sulindac, a nonsteroidal antiinflammatory drug known to decrease fetal urine output with minimal adverse effect on the ductus arteriosus. Encouraged by the hypothesis that limiting movement within the uterine cavity may decrease cord compression and fetal death, Peek and colleagues reported 3 sets of monoamniotic twins where fetal lie did not change after administration of daily sulindac starting between 24 to 27 weeks' gestation, and all infants were delivered with good outcomes.88 Overton and colleagues also reported successful use of sulindac starting at 20 weeks' gestation in 2 cases, and Pasquini and colleagues successfully utilized it in 12 sets. However, Sebire and colleagues reported that, in 2 sets of twins treated with sulindac, only 1 infant survived. Although this does not prove a causal relationship, it is clear that sulindac does not prevent cord entanglement, and at this time there are not enough data to recommend its routine use.69,89,90

While nonstress testing has been used to monitor for cord compression, biophysical profiles and Doppler ultrasound have been utilized to assess the fetal well being of monoamniotic twins in addition to aiding in the diagnosis of cord entanglement.69,89,90 Abuhamad and colleagues noted that umbilical artery notching may suggest increased resistance to flow, suggestive of constriction of the vessel lumens; in severe narrowing, diastolic flow may be greatly reduced. The authors suggested that, although further study was needed, the presence of umbilical artery notching may be useful in dictating further management.91
Timing of Delivery and Intrapartum Management of Monoamniotic Twins

There is no clear consensus when to effect delivery. As NICUs have continued to improve their care and provide better outcomes, there is growing evidence to suggest that delivery effected at 32 weeks is reasonable. In their report, Roqué and colleagues looked at the percent perinatal loss per number of fetuses entering each 2-week gestational interval. There was a statistically significant increase in perinatal loss rates of 5.8%, 11%, and 21.9% for intervals 30 to 32 weeks, 33 to 35 weeks, and 36 to 38 weeks, respectively. Although serial antenatal steroid use for fetal benefit was practiced in many of the cases managed during the 1990s, more recent reports suggest that antenatal betamethasone treatment should be used judiciously, with limitations to the number of courses administered.

The optimal mode of delivery is also unclear. Cord entanglement is nearly ubiquitous, and cases of transection of a nuchal cord at delivery proved to be the cord of the second twin, necessitating its rapid delivery. Although many of the case series have a significant number of twins delivering vaginally, more recent reports recommend elective cesarean section to avoid emergent delivery, and inadvertent transaction of the second twin’s umbilical cord. When vaginal delivery is considered, the appropriate skilled personnel and equipment must be available to rapidly expedite delivery, be it emergency cesarean, or internal podalic version and/or breech extraction. At our institutions, elective cesarean delivery is recommended between 32 and 34 weeks’ gestation, after administration of betamethasone for fetal benefit.

Conjoined Twins

Incidence of Conjoined Twins

Conjoined twinning is an extremely rare complication of monozygotic twins that results from incomplete embryonic division occurring between 13 and 15 days after conception. Because conjoined twins develop after differentiation of the chorion and amnion, all conjoined twins are monochorionic–monoamniotic. Monochorionic–monoamniotic twins account for less than 1% of monozygotic twins, and conjoined twins are even less common, occurring in approximately 1 in 50,000 to 100,000 births. Conjoined twinning is 3 times more common in female fetuses than males. Conjoined twins are classified based on the most prominent site of union together with the suffix pagus, which means fixed. Ventral unions occur 87% of the time and are classified as: cephalopagus (11%), thoracopagus (19%), omphalopagus (18%), ischiopagus (11%), and parapagus (28%). Dorsal unions occur in 13% of conjoined twins and are defined as: craniopagus (5%), rachiopagus (2%), and pygopagus (6%).

Diagnosis of Conjoined Twins

The diagnosis of conjoined twins can be made by ultrasound in the first trimester. At this early gestational age, the diagnosis should be suspected if the embryonic pole appears bifid. However, the diagnosis should be made with caution in the first trimester and, when suspected, follow-up imaging should be performed to confirm the diagnosis. Additional sonographic features of conjoined twins that may be apparent in the first and second trimesters include an inability to separate the fetal bodies and skin contours, lack of a separating membrane between the twins, the presence of more than three vessels in the umbilical cord, heads remaining at the same level and body plane, extremities in unusual proximity, and failure of the fetuses to change their relative positions over time. The evaluation of conjoined twins should include a detailed ultrasound examination, including a fetal echocardiogram, at 18 to 20 weeks to determine the extent of shared organs and to exclude additional anomalies. Associated anomalies, even in organs unrelated to the conjoining, are not uncommon. Further information may be gained by MRI evaluation. A diagnosis of the anatomy of shared organs and the presence of additional malformations is essential for counseling families regarding outcome and planning postnatal surgical separation. Because chromosomal abnormalities are rare in conjoined twins, karyotyping is generally not indicated.

Management of Conjoined Twins

If the diagnosis of conjoined twins is made before viability, the option of pregnancy termination should be discussed. The prognosis of the fetuses can be based on the extent of fusion and the presence of additional malformations. Separation is rarely successful when there is cardiac or cerebral fusion. Additional malformations unrelated to the site of conjoining may also adversely affect prognosis. If a family elects to continue with the pregnancy, the objectives are to maximize the chance of survival of the twins and to minimize maternal morbidity. Serial ultrasounds may be useful to further define the shared anatomy and the effect on fetal development. Polyhydramnios, secondary to circulatory imbalance, is a common complication of conjoined twins that affects as many as 50% of cases. Amnioreduction may be necessary to decrease the risk of preterm labor or premature rupture of membranes, and to minimize maternal discomfort from an overdistended uterus. Elective cesarean delivery should be strongly considered once lung maturity is documented. In cases where one twin has a very low likelihood of survival and endangers the life of the cotwin, an EXIT procedure should be considered.

Outcome of Conjoined Twins

Survival of conjoined twins depends largely on the site of conjoining and the organs involved. In a series of 14 cases of prenatally diagnosed conjoined twins, 28% of cases died in utero, 54% died immediately after birth, and only 18% survived. Emergent surgical separation is performed in the event that one twin dies, or if a life-threatening condition exists in one of the twins that threatens the life of the cotwin. In these cases, the survival rate is reported as 30% to 50%. Elective separation, which usually occurs at 2 to 4 months of age, allows for stabilization of the twins, confirmation of anatomic relationships, diagnosis of previ-
ously unrecognized anomalies, and adequate planning of surgery. Most series report a survival rate of 80% to 90% for elective separation.103-105

Conclusion

The diagnosis and management of obstetrical complications in a multiple gestation pose a number of unique challenges. These complex clinical scenarios demand thorough counseling of the expectant parents, as well as care by physicians with expertise in the management of multiple gestations.

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