Consensus Statement

The North American Fetal Therapy Network Consensus Statement

Management of Complicated Monochorionic Gestations

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The North American Fetal Therapy Network is a consortium of 30 medical institutions in the United States and Canada with established expertise in fetal therapy and other forms of multidisciplinary care for complex fetal disorders. This publication is the third in a series of articles written by NAFTNet about monochorionic pregnancies. In this article, we provide the general obstetric practitioner with information regarding management options available for complications of monochorionic gestations. This information may be useful for a better understanding of the pathophysiology of the various conditions, for better patient counseling, for timely referral to a regional treatment center, and for ongoing comanagement after treatment.

(Obstet Gynecol 2015;0:1–10)

DOI: 10.1097/AOG.0000000000000994

The North American Fetal Therapy Network recently has published multidisciplinary, evidence-based, and consensus-driven recommendations for the surveillance and management of uncomplicated monochorionic gestations.[1,2] The goal of this publication is to provide the general obstetric provider with information about the management of various complications of monochorionic gestations. This information may be useful for the health care provider's own understanding, but also for better counseling their patients who have been diagnosed with a complication of their monochorionic pregnancy. Understanding of management options may aid in the timely referral to a regional treatment center. Finally, understanding of management may aid in the ongoing prenatal care of patients after treatment.

BACKGROUND

Because of their placental angioarchitecture, monochorionic twins are subject to unique pregnancy complications that can threaten the life and health of both fetuses and therefore impose a disproportionate disease burden on overall perinatal morbidity and mortality. This burden is a consequence of "chorioangiopagus," or sharing of a single placenta through intraplacental vascular anastomoses, which appropriately implies that the fetuses are hemodynamically connected; what happens to one fetus can directly affect the other. Survival after loss of a co-twin is linked to a high incidence (24–45%) of neurologic injury, likely from acute pressure and volume shifts from the living twin to the deceased twin through patent vascular channels.[3] An understanding of chorioangiopagus is therefore fundamental to understanding management strategies: all effective treatments must account for it.[1,2]

COMPLICATIONS OF MONOCHORIONIC GESTATIONS AND THEIR MANAGEMENT

For a detailed description of the pathophysiology of complications of monochorionic gestations, please refer to the recent North American Fetal Therapy Network publication by Bahtiyar and colleagues.[1] In this article we present contemporary management strategies, anticipated outcomes, and potential complications. With this information, primary obstetric providers may have a better understanding of the risk–benefit discussion that their patients experience while at a treatment

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Financial Disclosure

The authors did not report any potential conflicts of interest.

ISSN: 0029-7844/15

center. The objective of the risk–benefit discussion is to provide directive counseling when evidence of benefit exists and nondirective counseling when evidence of benefit is lacking.[4,5] Ultimately, the decision of whether and how to proceed lies with the patient.

I. Twin–Twin Transfusion Syndrome

Twin–twin transfusion syndrome affects approximately 10% of monochorionic twins and accounts for the majority of morbidity and mortality in monochorionic gestations.[6] Management options depend on gestational age, stage of disease, and availability of resources and include:

1. Fetoscopic laser photocoagulation has been shown to be superior to amnioreduction in a randomized controlled trial and is considered the treatment of choice for severe twin–twin transfusion syndrome.[7] The procedure is usually offered for Quintero stage II–IV disease between 16 weeks 0 day and 26 weeks 0 day of gestation (the upper threshold of gestational age has been set by U.S. Food and Drug Administration approval of fetoscopes imported from Europe). Some centers will offer laser therapy at stage I for symptomatic polyhydramnios, recipient cardiomyopathy based on specific echocardiographic scoring systems, or a shortened cervix.[8] The procedure is typically performed with intravenous sedation and local anesthesia or under regional anesthesia. Patients are usually admitted for 24-hour postoperative observation. Maternal complications such as placental abruption (1%) and intraabdominal leakage of amniotic fluid (3%) are relatively low, and recovery is generally swift. Preterm premature membrane rupture complicates approximately 30% of cases.[9]
   In severe disease, overall perinatal survival without treatment has been reported to be approximately 30%.[10] As with most surgical procedures, a clear learning curve has been documented for laser photocoagulation.[11,12] Emerging centers report dual survival rates of approximately 50%, whereas more experienced centers achieve a dual survival rate approaching 70%.[12,13] Importantly, the incidence of major neurologic abnormalities improves with surgical experience, achieving a rate as low as 6% in survivors, near the baseline for uncomplicated monochorionic gestations.[14] This is likely the result of the fact that intertwin connections on the chorionic surface of the placenta are visually identified and cauterized, thereby disrupting intertwin flow (ie, treating chorioangiopagus). Complete placental separation ("dichorionization") is unlikely, however, because anastomoses often occur below the chorionic plate and are therefore not visible at fetoscopy.[15] Incomplete separation is evidenced by recurrent twin–twin transfusion syndrome (14%) and the development of twin anemia polycythemia sequence (13%).[16] Recent technical improvements such as laser coagulation of the entire vascular equator, or "Solomonization," have led to incrementally improved outcomes with an overall survival rate of 75% and an incidence of recurrent twin–twin transfusion syndrome and twin anemia polycythemia sequence of 1% and 3%, respectively.[17] Nonetheless, ongoing ultrasound surveillance in treated pregnancies is warranted, and Solomonization is not possible in every case.[18] Laser treatment may exacerbate discordant donor–recipient placental share (the recipient possesses roughly 70% of the placenta, whereas the donor has roughly 30%) and results in uteroplacental insufficiency of the already compromised donor. Similarly, in stage III disease when the donor has critically abnormal Doppler waveforms, loss of the donor within 30 days after treatment is approximately 50%.[13] Perioperative maternal oral nifedipine may increase recipient survival in high-stage disease as a result of potential amelioration of recipient hypertension.[19]
   After treatment, weekly ultrasound surveillance with middle cerebral artery peak systolic velocity measurements to assess for recurrent twin–twin transfusion syndrome or the development of twin anemia–polycythemia sequence for 6 weeks is recommended and then ultrasound surveillance every 2 weeks as with uncomplicated monochorionic gestations.[20] Currently, there are no recommendations regarding initiation, timing, or method of antenatal testing for laser-treated twin–twin transfusion syndrome twins. Most treatment centers recommend delivery of treated twins at 36 weeks of gestation, whereas singleton survivors may progress to 39 weeks of gestation. Patients should be made aware that the risk of an adverse outcome such as single or dual demise, treatment failure, or long-term survivor morbidity is significant.[21] Nonetheless, laser photocoagulation offers the highest likelihood of intact twin survival for severe disease.
2. Serial amnioreduction is associated with an approximate 60% survival rate and a 25% risk of neurologic morbidity among survivors.[22] The intent of amnioreduction is to decrease intraamniotic pressure, thereby decreasing the risk of preterm labor or preterm premature membrane rupture, improving uteroplacental perfusion, and

decreasing maternal symptoms. The advantages of amnioreduction are that it is nearly universally available and relatively easy to perform. Disadvantages include a cumulative risk of procedure-related complications such as membrane rupture and labor (15% per procedure)[22] and the possibility of making laser photocoagulation more difficult if not impossible. Intraamniotic hemorrhage can obscure visualization of the placental anastomoses at the time of fetoscopy, incidental septostomy precludes easy access to the vascular equator of the placenta, and preterm premature rupture of membranes precludes the ability to perform fetoscopy. The major disadvantage of amnioreduction, however, is that it does not address the underlying pathophysiology of chorioangiopagus. Nonetheless, it may be the only available option in lower-resource areas or for pregnancy prolongation beyond 24–26 weeks of gestation when pregnancy termination and selective feticide are no longer available as a result of state laws, and laser photocoagulation is not available resulting from U.S. Food and Drug Administration device restrictions in the United States. Although high-quality evidence on which to base a recommendation is lacking, intensive surveillance after amnioreduction, typically twice-weekly ultrasonograms with multivessel Doppler interrogation (umbilical artery, umbilical vein, ductus venosus), is important to assess for evidence of disease progression. Should this occur, referral to a regional treatment center is warranted.

3. Selective feticide is most commonly performed when the twins are discordant for a major structural abnormality or one of the twins is moribund. The death of one of a pair of monochorionic twins can result in neurologic damage to the surviving co-twin. In high-stage disease, selective feticide may provide the highest likelihood of intact survival by allowing pregnancy prolongation.[23,24] Other indications for selective feticide include severe selective fetal growth restriction or to prophylactically avoid the potential complications of monochorionic gestation. Selective feticide in choriopagus twins is more complex than in dichorionic twins as cardioplegic drugs can pass from one twin to the other. Therefore, the procedure requires complete interruption of the intertwin vascular connections. Methods include bipolar cord coagulation, laser cord photocoagulation, intrafetal laser photocoagulation, or intrafetal radiofrequency ablation.[25] Most authorities wait until at least 16 weeks of gestation when there has been fusion of the amnion and chorion before undertaking the procedure. Intrafetal radiofrequency ablation is technically easier to perform than bipolar cord coagulation and is associated with a lower rate of procedure-related complications.[23] For example, intrafetal radiofrequency ablation is performed under ultrasound guidance with a 17-gauge device, whereas laser photocoagulation requires a 3-mm port to be introduced into the uterus to allow for placement of the fetoscope (preterm premature rupture of membrane risk of 17% compared with 30%, respectively).[9] In certain circumstances such as advanced-stage disease combined with selective fetal growth restriction or severe recipient cardiomyopathy, fetoscopic laser photocoagulation may have the same outcome as radiofrequency ablation (ie, single survivor), but with a higher procedure-related complication rate. Robust outcome data are not yet available, but preliminary data suggest an 83% likelihood of co-twin survival and a median gestational age of delivery of 35 weeks.[26] Because neonatal morbidity in survivors of twin–twin transfusion syndrome is strongly linked to prematurity, pregnancy prolongation may result in a better long-term outcome of the remaining twin.[27] There is insufficient evidence to recommend a management strategy for ongoing surveillance after selective feticide, but intensive surveillance such as that with amnioreduction is probably not necessary because the intertwin vascular circuit has been completely interrupted. There is also insufficient evidence to inform decision-making regarding timing of delivery, but most centers recommend allowing the pregnancy to proceed to term. Routine monitoring for maternal coagulopathy is not indicated.[28]
4. Expectant management: This may be appropriate for early disease (stage I), or late in gestation, and when frequent ultrasound surveillance is available. The optimum management of stage I disease has not been determined. A small retrospective study demonstrated improved neurologic function, but not overall survival, with fetoscopic laser photocoagulation for stage I disease.[29] Expectant management (ie, nonintervention) may be the only available option when operative intervention before viability is unavailable.
5. Delivery: The peak incidence of twin–twin transfusion syndrome is 19 weeks 6 days of gestation, but can occur any time thereafter.[20] Delivery is a good option in the mid- to late third trimester.

Iatrogenic delivery may, however, be the only available option when other alternatives for safe pregnancy prolongation are unavailable and there is concern for imminent demise of one twin or maternal health declines.

6. Pregnancy termination: Patients should be made aware of the option of pregnancy termination before viability and consistent with applicable law and organizational policy. All remaining management options will involve significant risk of an adverse outcome for one or both newborns as well as risks, iatrogenic and otherwise, to the mother.[30]

II. Twin Anemia Polycythemia Sequence

Chronic slow and unbalanced blood transfusion between monochorionic fetuses results in twin anemia polycythemia sequence. It is characterized by anemia of the "donor" twin and polycythemia of the "recipient" twin without amniotic fluid discordance.[31] Prenatal diagnosis of fetal anemia and polycythemia is possible by middle cerebral artery peak systolic velocity measurements by Doppler ultrasonography. Diagnosis of twin anemia polycythemia sequence is made when middle cerebral artery peak systolic velocity of one twin is greater than 1.5 MoM and the peak systolic velocity of the second twin is less than 1.0 MoM without polyhydramnios and oligohydramnios. It complicates approximately 3–5% of spontaneous monochorionic twin gestations,[31] but, up to 13%, after fetoscopic laser for twin–twin transfusion syndrome.[32] It is believed to develop as a result of very small-caliber, less than 1 mm in diameter, unidirectional arteriovenous connections, which are located close to the placental edges along with a paucity of arterio-arterial vascular connections.[33,34] Spontaneous twin anemia polycythemia sequence can occur any time during pregnancy but is frequently diagnosed after 26 weeks of gestation.[35] An ultrasound-based staging system has been proposed based on severity of anemia and polycythemia.[36]

Optimum prenatal treatment has not been established. Depending on gestational age, certain management alternatives can be considered including expectant management, selective feticide, delivery, intrauterine transfusion with or without partial exchange transfusion for the polycythemic fetus, and fetoscopic laser photocoagulation.

The utility of intrauterine transfusion was investigated in the past for symptomatic relief of the anemic fetus. Opponents of this therapy point out that the underlying vascular connections continue to exist and the polycythemia may worsen in the recipient twin. Furthermore, there are reports of skin necrosis in cases treated with multiple intrauterine transfusions.[16] To overcome these concerns about rapid transfusion of blood from the donor to the recipient, intraperitoneal blood transfusion has also been used.[37]

In select cases, partial exchange transfusion of the fetus with polycythemia combined with intrauterine transfusion of the fetus with anemia might eliminate previously reported complications of polycythemia.[28] Furthermore, mathematical modeling of intrauterine transfusion combined with prenatal exchange transfusion showed a reduction in polycythemia in the recipient.[29]

Slaghekke et al[38] reported their experience in 52 cases treated with fetoscopic laser, intrauterine transfusion, or expectant management. In this series, there was no difference in perinatal survival by treatment group: laser (94%), intrauterine transfusion (85%), expectant management (83%) (P=.3). Although there was no statistical difference, cases treated with laser had lower neonatal morbidity: 7%, 38%, and 24% in laser, intrauterine transfusion, and expectantly managed groups, respectively. Furthermore, no severe hematologic complications were detected in the laser group compared with intrauterine transfusion and expectantly managed groups (72% and 52%, respectively). Finally, the median time from diagnosis to delivery was significantly prolonged in the laser group: 11 weeks compared with 5 weeks in the intrauterine transfusion group and 8 weeks for the expectantly managed group.

In summary, in appropriately selected cases, either fetoscopic laser or intrauterine transfusion with recipient partial exchange transfusion may be the best treatment options at present, but management must be individualized based on a myriad of considerations.

III. Selective Fetal Growth Restriction

Selective fetal growth restriction is increasingly recognized as a major complication of monochorionic twin gestations, because it is significantly associated with fetal death and poor neurologic outcome.[39–41] Selective fetal growth restriction affects approximately 10–15% of monochorionic twin gestations.[42,43] At present there is no consensus on diagnostic criteria. Although some investigators use an estimated fetal weight below the 10th percentile[39,44] with or without significant growth discordance (greater than 25%), others solely use growth discordance greater than 25%[45] to diagnose the disease. Optimum delivery timing for pregnancies complicated by selective fetal growth restriction is

unknown. There are three types based on umbilical artery Doppler ultrasound findings.[46]

Type I

In this group, the umbilical artery has forward diastolic flow at all times. Type I tends to develop later in gestation (mean 23 weeks of gestation), deliver later in gestation (mean 35.4 weeks of gestation) and, compared with other types of selective intrauterine growth restriction, have lower intertwin growth discordance (29%). In one series, the risk of fetal death was approximately 3.0–4.3% with 0% of the surviving twins developing neurologic damage.[46]

Overall, type I selective fetal growth restriction has a favorable outcome. Expectant management with frequent monitoring is an appropriate choice. Although Doppler ultrasound findings might change over time, most of the cases will continue to have normal Doppler findings throughout the pregnancy.

Type II

In this group, umbilical artery Doppler shows persistent absent or reversed end diastolic flow. Studies have shown that these Doppler changes may be present for longer intervals before clinical deterioration of the fetus when compared with similar Doppler findings in the singleton gestation.[47,48] The growth-restricted fetus has a smaller share of the placenta and fewer intertwin anastomoses. Sequential deterioration of the growth-restricted fetus (abnormal umbilical artery Doppler followed by abnormal ductus venosus Doppler and umbilical venous Doppler and eventually declining biophysical profile score) is seen in 90% of cases.[46] Mean gestational age at delivery is approximately 28–30 weeks.[41,46] In one series, intact survival of the growth-restricted twin was 37.0% with a nearly 50% perinatal mortality rate.[41] In this same series, intact survival of the normally growing twin was 55%.[41] In another series, brain damage in the growth-restricted fetus was as high as 14.4%. Brain damage of the normally grown fetus was reported to be as low as 3%.[46]

Optimum treatment of type II selective fetal growth restriction is not yet established. In patients with early diagnosis, selective feticide can be considered. Laser photocoagulation in these cases should be considered investigational and has been reported to be associated with 70% intrauterine fetal death in the growth-restricted twin. Patients diagnosed later in pregnancy should be frequently monitored for early signs of deterioration in fetal well-being. Antenatal testing should guide decisions on timing of delivery.

Type III

In this group, intermittently absent or reversed end diastolic flow, which is the result of a large artery–artery anastomosis, creates an unstable hemodynamic environment for both twins.[49,50] Evolution of the disease is unpredictable. There are no known prognostic factors. Demise of the co-twin is reported to be as high as one in three cases.[46] Mean gestational age at delivery is 31–32 weeks.

Type III selective growth restriction poses an important treatment dilemma. There are no established treatment options. If diagnosed early in pregnancy, selective feticide or fetoscopic photocoagulation could be discussed. However, it should be noted that, in the absence of fluid discordance, fetoscopic laser photocoagulation might be technically difficult or impossible.[51] If expectant management is chosen, the frequency of antenatal testing with serial Doppler assessment and biophysical profiles should be increased. There are no data on how often these pregnancies should be monitored. Patients should be included in determining when to start antenatal testing. Antenatal testing could be started when the smaller twin reaches a point when neonatal survival is possible.

IV. Monoamniotic Twin Gestation

Monoamniotic twin gestations are at high risk of perinatal mortality, ranging from 10% to 40% of cases.[52] In addition, there is up to a 26% prevalence of congenital anomalies.[53] Recent literature shows that most of the mortality is the result of prematurity and congenital anomalies, debunking previous theories that cord entanglement was to blame.[54]

There are no randomized trials on which to base a recommendation on the optimal method of prenatal surveillance. A multicentered retrospective review of 96 monoamniotic twins found a rate of intrauterine fetal demise of 15% in the group managed as an outpatient; no fetal deaths occurred in the group admitted for inpatient monitoring.[55] More recent data from a single center revealed similar rates of in utero loss with outpatient compared with inpatient monitoring (2.1% compared with 4.7%). Of note, the outpatient group underwent fetal cardiotocograms four times a week in conjunction with once- to twice-weekly ultrasonograms. Fifty percent of the pregnancies required admission and 20% were delivered for fetal indications.[52] A survey of maternal-fetal medicine specialists in the United States found that 84% recommended elective admission at 26–28 weeks of

gestation for inpatient monitoring.[56] There are also little data to suggest the interval for fetal monitoring. Eighty-one percent of maternal-fetal medicine specialists recommended surveillance two to three times daily.[56] Continuous fetal monitoring has been recommended by some, although this is not practical with one study indicating that interpretable tracings on both fetuses could only be obtained 52% of the time.[57] Fetal heart rate tracings should be reviewed for the presence of variable decelerations, which indicate cord compromise. The frequency, nadir, and duration of variable decelerations that warrant delivery are subject to interpretation.

A recent Cochrane review concluded that there is insufficient evidence to draw a strong conclusion about the best strategy of expectant management compared with delivery at a preterm gestational age.[58] There are considerable institutional variations in prenatal management, inpatient compared with outpatient status, and timing of delivery. The recent survey of maternal-fetal medicine specialists indicated a median recommended gestational age for delivery of 33 weeks.[56]

The risk of fetal death appears to exceed the risk of a postnatal nonrespiratory complication at 32 weeks 4 days of gestation.[52] Recent literature support that there is no difference in the incidence of death or a nonrespiratory neonatal complication between fetuses managed as outpatients (13.2%) compared with inpatients (10.5%, P=.55).[52] The authors concluded that if close fetal surveillance is instituted after 26–28 weeks of gestation and delivery takes place at 33 weeks of gestation, the risk of fetal or neonatal death is low, no matter the surveillance method.[52] Many centers deliver by cesarean delivery at 32–34 weeks of gestation after a course of corticosteroids for fetal lung maturity.

Monoamniotic placentas have significantly greater numbers of both superficial and deep anastomoses than do uncomplicated monochorionic diamniotic pregnancies.[59] Monoamniotic twins are at risk for twin–twin transfusion syndrome and twin anemia polycythemia sequence[60] as well as all other complications of monochorionicity.[61] If selective feticide is planned and cord entanglement is detected, many experts recommend cord transection after selective feticide, typically with combined bipolar and laser as a means of reducing the risk of sudden death of the co-twin from a cord accident.[62]

V. Twin Reversed Arterial Perfusion

Twin reversed arterial perfusion results when a living fetus (the "pump twin") perfuses a nonviable twin (the "acardiac twin") through patent placental vascular channels. This abnormal vascular circuit can result in shunt physiology and high-output heart failure in the pump twin. Mortality in the pump twin approaches 50%.[63] As with other complications of monochorionic twin gestations, the goal of treatment for twin reversed arterial perfusion is to completely interrupt the intertwin vascular circuit. This can be achieved with several techniques such as interstitial laser in the late first or early second trimester or bipolar cord coagulation or fetoscopic laser cord photocoagulation. Intrafetal radiofrequency ablation of the circuit within the acardiac twin holds the advantage of lower procedure-related complications as a result of the small diameter of the device (17-gauge needle) compared with the 3-mm diameter of bipolar forceps and fetoscope.[64] Previously, intervention was recommended only if the size of the acardiac twin was 70% or more of that of the pump twin on ultrasonography.[63] With increasing diagnostic and interventional experience, many treatment centers are moving toward a proactive approach of treatment once continued growth of the acardiac twin is documented as opposed to waiting for overt evidence of deterioration (hydrops fetalis) of the pump twin.[65,66] A recent series documented an 82% pump twin survival rate with a median gestational age at delivery of 37 weeks.[65] As with selective feticide, a surveillance strategy of serial ultrasonograms for assessment of fetal growth should be considered. There is insufficient evidence to inform decisions regarding timing of delivery, but many centers allow progression to term.

VI. Discordant Anomalies

Structural anomalies are more common in monochorionic twins (6–8%) than in dichorionic gestations (1–2%).[6] Some anomalies may predispose the entire gestation to preterm birth, whereas others predispose to intrauterine demise of the affected twin. Loss of a monochorionic twin exposes the co-twin to demise (10–25%) or neurologic injury (24–45%).[3] Selective feticide by complete interruption of the intertwin communication may therefore increase the likelihood of intact survival of the unaffected twin by avoiding dual demise and allowing for pregnancy prolongation.[27] Considerations regarding ongoing pregnancy management and delivery timing are similar to those discussed under the Selective Feticide section of twin–twin transfusion.

VII. High-Order Monochorionic Multiples

High-order multifetal gestations containing a monochorionic twin pair are at increased risk of adverse

pregnancy outcomes. For instance, dichorionic triplets have lower birth weight ($P<.001$), lower gestational age at delivery ($P<.001$), and spend more days in the neonatal intensive care unit ($P=.05$) than trichorionic triplets.[67] Geipel and colleagues[68] documented an increased risk of intrauterine death, selective growth restriction, delivery less than 32 weeks of gestation, and a lower survival rate in dichorionic triplets than in trichorionic triplets. For these reasons, the pregnant woman should be counseled about the option of reducing the monochorionic pair.[69] For reasons described previously, reducing one of the monochorionic twins is significantly more complicated than reducing the pair because it would require interruption of the placental vascular connections. Reducing the monochorionic pair, on the other hand, can be done with injection of a cardioplegic medication and can be performed in the late first trimester. The procedure-related loss rate for the entire pregnancy is approximately 5%, similar to that seen in multifetal pregnancy reduction for other indications.[70]

Twin–twin transfusion syndrome and other complications of monochorionic gestations can occur within dichorionic triplet or higher-order gestations. Management strategies will need to account for chorioangiopagus among fetuses sharing a single placenta. Laser photocoagulation has been described in dichorionic triamniotic triplets with similar success rates but earlier delivery than in monochorionic diamniotic twins.[71] As with twins, ongoing ultrasound surveillance after treatment is warranted.

VIII. Conjoined Twins

Because of the rarity of the phenomenon, there are no evidence-based recommendations for pregnancy management. Long-term outcomes are largely predicated on the degree of sharing of vital organs, which can be determined prenatally by ultrasonography, echocardiography, and ultrafast magnetic resonance imaging.[72] There is an increased incidence of structural anomalies not associated with conjoined organs.[73] Mortality is high with only 18% of prenatally diagnosed conjoined individual twins surviving.[72] This information can be used to inform prenatal decision-making (pregnancy termination or location, timing and route of delivery).

IX. Loss of One Twin

As discussed in our May 2015 publication,[2] "a loss of a monochorionic twin exposes the surviving co-twin to a high risk of death and severe cerebral injury."[3] The suspected mechanism is acute exsanguination of the surviving twin into the low-pressure vascular circuit of the deceased twin through patent vascular anastomoses, resulting in sudden and profound hypotension, hypovolemia, and anemia with consequent tissue hypoxia and acidosis.[74] Urgent delivery after an unwitnessed twin death is unlikely to improve the co-twin's outcome and may unnecessarily expose the survivor to complications of prematurity. An earlier theory of intertwin embolization as an ongoing mechanism of injury to the co-twin has largely been abandoned. Expectant management to term is favored. Routine monitoring for coagulopathy is not indicated,[28] although consideration should be given to antenatal and postnatal neuroimaging.[75]

DISCUSSION

Because of their placental angioarchitecture, monochorionic twins are subject to unique pregnancy complications that can threaten the life and well-being of both fetuses. Early diagnosis and intervention can improve on the natural history of these conditions. Consultation with a maternal-fetal medicine specialist and prompt referral to a regional treatment center are critical once a complication is suspected. Effective management options address the underlying pathophysiology of chorioangiopagus. Nonetheless, the risk of adverse pregnancy outcomes remains relatively high. An often complex risk–benefit assessment must be undertaken, focusing on the wishes of the patient, especially when the outcomes are uncertain. Pregnancy outcomes will likely continue to improve substantially for complicated monochorionic gestations such as twin–twin transfusion syndrome, twin reversed arterial perfusion, discordant anomalies, and higher-order monochorionic multiple gestations once the concept of chorioangiopagus is widely appreciated. Significant challenges remain, however, in the management of twin anemia polycythemia sequence and selective fetal growth restriction. Ongoing multicenter, multidisciplinary collaborative research through the North American Fetal Therapy Network and other organizations will hopefully improve patient outcomes.

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