US Evaluation of Twin Pregnancies: Importance of Chorionicity and Amnionicity

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Abbreviations: DA = diamniotic, DC = dichorionic, MA = monoamniotic, MC = monochorionic, MCA = middle cerebral artery, MoM = multiples of the median, PART = polyhydramnios affecting recipient-like twin, PSV = peak systolic velocity, sFGR = selective fetal growth restriction, TAPS = twin anemia polycythemia sequence, TRAP = twin reversed arterial perfusion, TTTS = twin-twin transfusion syndrome

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SA-CME LEARNING OBJECTIVES

After completing this journal-based SA-CME activity, participants will be able to:

- Identify the chorionicity and amnionicity of multiple pregnancies in first, second, and third trimesters.
- Review imaging features of common complications effecting MC twin pregnancies, including TTTS, TAPS, PART, TRAP sequence, and sFGR.
- Recognize imaging findings that require referral to fetal treatment centers and possible fetal interventions.

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The twin birth rate is increasing in the United States. Twin pregnancies can be dichorionic or monochorionic (MC). MC twins account for 20% of twin pregnancies but 30% of all-cause pregnancy-related complications. This article describes the imaging findings that establish chorionicity and amnionicity. Ideally, these are established in the first trimester when accuracy is high, but they can also be determined later in pregnancy. Complications unique to MC twin pregnancy include twin-twin transfusion syndrome, twin anemia polycythemia sequence, twin reversed arterial perfusion sequence, and selective fetal growth restriction. The US features, staging systems, and management of these complications are reviewed, and the consequences of MC twin demise are illustrated. Ongoing surveillance for these conditions starts at 16 weeks gestation. Monoamniotic (MA) twins are a small subset of MC twins. In addition to all of the MC complications, specific MA complications include cord entanglement and conjoined twinning. Radiologists must be able to determine chorionicity and amnionicity and should be aware of potential complications so that patients may be referred to appropriate regional specialized centers. A proposed algorithm for referral to specialized fetal treatment centers is outlined.

Online supplemental material is available for this article.

Introduction

The twin birth rate in the United States in 2017 was 33.3 per 1 000 total births. Overall, the twinning rate (births in twin deliveries per 1 000 total births) increased 76% from 1980 to 2009 (1). Twin pregnancies can be monozygotic or dizygotic in nature. Dizygotic twinning results from two separate fertilizations, while monozygotic twinning occurs when a morula from a single fertilization divides and results in two embryos. Early division, up to 1–3 days postfertilization, results in two complete cell lines and dichorionic (DC) twinning. If the morula splits after 4 days or later, the postfertilization cells are already committed to forming the chorion, resulting in monochorionic (MC) twins. Based on the duration of postfertilization after which the split occurs, MC twins can be diamniotic (DA) (4–8 days after fertilization) or monoamniotic (MA) (8–13 days after fertilization) and can even result in conjoined twins with late splitting (after 13 days) (Fig 1). In the era of assisted fertility, the incidence of MC twins has steadily increased over the past few decades (2).

All twin pregnancies have a higher risk of preterm labor, maternal hypertensive disorders, diabetes, and premature rupture of membranes than those of singleton pregnancies, but chorionicity determines the prognosis of multiple gestations. Although MC twins account for only 20% of twin pregnancies, they account for 30% of all-cause pregnancy-related complications. Among multiple pregnancies, the stillbirth rate in MC twins compared with that of DC twins is 44.4 versus 12.2 per

TEACHING POINTS

- All twin pregnancies have a higher risk of preterm labor, maternal hypertensive disorders, diabetes, and premature rupture of membranes than those of singleton pregnancies, but chorionicity determines the prognosis of multiple gestations. Although MC twins account for only 20% of twin pregnancies, they account for 30% of all-cause pregnancy-related complications.
- Cord entanglement is a hallmark finding for MA pregnancy. Conjoined twins can only happen in MA pregnancy
- The diagnosis of TTTS is usually made in the second trimester, with the hallmark observation being oligohydramnios in one sac and polyhydramnios in the other.
- Amniotic fluid discrepancy is not a feature of TAPS and should never manifest in TAPS.
- In any MC pregnancy with an anomalous twin, check the direction of flow in the umbilical artery. If it is toward the abnormal fetus, that is evidence of a TRAP diagnosis.

1 000 births, respectively (relative risk, 3.6). Similarly, the rate of neonatal mortality in MC twins compared with that of DC twins is 32.4 versus 21.4 per 1 000 births, respectively (relative risk, 1.5). A major fetal anomaly affecting only one twin occurs in around one in 25 DC twins, one in 15 MC-DA twins, and one in six MC-MA twins (3,4).

Hemodynamic connections between MC fetuses sharing a single placenta produce the unique complications of MC twin pregnancy. These include unequal placental sharing, selective fetal growth restriction (sFGR), twin-twin transfusion syndrome (TTTS), twin anemia-polycythemia sequence (TAPS), and twin reversed arterial perfusion (TRAP) sequence (Table 1). Because of the connected vasculature, any adverse event affecting one twin can also compromise the co-twin. Death of one twin can result in death or permanent disability of the co-twin. MC-MA twin pregnancies have a unique subset of complications, including conjoined twinning, cord entanglement, higher rates of anomalies, and higher rates of intrauterine fetal demise.

Accurate and early determination of chorionicity is critical for appropriate triage and management. Once a twin pregnancy is identified, both the American College of Radiology (2) and the Society for Maternal-Fetal Medicine (5) recommend determining chorionicity as early as possible and ideally in the first trimester, as much as possible. Frequent US surveillance is required to manage MC twin pregnancies to allow the early detection of complications and appropriate referral and intervention to improve outcomes. The management of MC twin pregnancies is much different than that of singleton pregnancies or even that of DC twin pregnancies. (The original slide presentation for this article from the RSNA Annual Meeting is available online.)

Establishing Chorionicity

The best time to establish chorionicity (and amnionicity) is in the first trimester when US is up to 98% accurate (6–10). Early in the gestation period, even up to 7–9 weeks, determining amnionicity may be challenging because the thin amnion may be below the level of detection, even at transvaginal US (9).

Chorionicity and Amnionicity in the First Trimester

The gestational sac visualized in early pregnancy is created by the chorion, the precursor to the placenta; thus, the number of gestational sacs equals the chorionicity of a pregnancy (Figs 2–4) (2,10). In a DC twin pregnancy, two complete and separate gestational sacs are present (Fig 2a) (2). In an MC twin pregnancy, a single rim of chorionic tissue creates a single gestational sac (Figs 2b, 2c, 3, 4). Early on, up to 7–9 weeks postfertilization, the amnion may not be detectable (Figs 3,4) (9,11). In this scenario, amnionicity can be suggested on the basis of the number of yolk sacs (Figs 2b, 2c, 3, 4). Two yolk sacs are suggestive of DA twins; one yolk sac suggests MA twins (Figs 2b, 3, 4). Amnionicity must be verified at follow-up US examinations, as discrepancies have been reported in both directions, with a single yolk sac visualized in DA twins and two yolk sacs visualized in a pregnancy that was eventually found to be an MA twin pregnancy (12,13).

At 10–14 weeks gestation, when the placenta becomes a distinct structure, the number of placental masses represents the chorionicity of the pregnancy (Fig 5). A thick membrane composed of two chorions and two amnions is depicted between DC twins (Fig 5a), whereas a thin membrane composed of only two layers of amnion separates MC twins (Fig 5b). In DC twins, the chorionic tissue extending from the placenta into the base of the thick membrane creates the twin-peak sign (Fig 5a), which helps establish the true chorionicity even when there is a single placental mass (14,15). In MC-DA twins, the amnions abut the placenta without intervening chorion, creating a T sign (Fig 5b) (16). In MC-MA twins, both embryos or fetuses are in a common space surrounded by a single chorion and a single amnion (Figs 2c, 4).

Chorionicity and Amnionicity in the Second and Third Trimesters

In the second trimester, while the number of placental masses, thickness of the membrane, and the twin-peak sign remain viable options for determining chorionicity, fetal sex is also an important determinant of chorionicity. When the fetuses are different sexes, the pregnancy is almost always dizygotic and therefore must be DC. In extremely

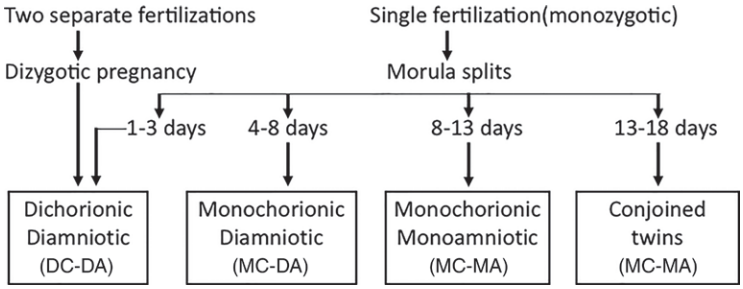

Figure 1. Diagram shows the embryological development of DC and MC pregnancies.

Table 1: Differentiating Features of MC Twin Pregnancy Complications

Entity	Δ EFW	Amniotic Fluid	Umbilical Artery Doppler US	MCA Doppler US
TTTS	±	Oligohydramnios in the donor Polyhydramnios in the recipient	Used for grading	Not used for grading
TAPS	NA	N in one twin	NA	Donor <0.8 MoM Recipient >1.5 MoM
PART	NA	N in one twin Polyhydramnios in the other	N	NA
I-oligo	NA	Oligohydramnios in one twin N in the other	N	NA
sFGR	≥25%	± oligohydramnios in the smaller twin	Used for grading	May help diagnose an abnormal CPR
TRAP	±	NA	Flow toward abnormal fetus	NA

Note.— CPR = cerebroplacental ratio, Δ = difference in twin EFWs, EFW = estimated fetal weight, I-oligo = isolated oligohydramnios in one twin, MCA = middle cerebral artery, MoM = multiples of the median, N = normal, NA = not applicable, PART = polyhydramnios affecting recipient-like twin.

rare circumstances, postzygotic nondisjunction can occur; hence, the sex can be different in MC-MA twins. As gestation progresses, membrane thickness lessens, and this thinning makes it a less reliable indicator, emphasizing the importance of establishing chorionicity at the time of the earliest US examination (17,18). Cord entanglement (Fig 5c) is a hallmark finding for MA pregnancy (19). Conjoined twins can only happen in MA pregnancy (Fig 6).

Once the chorionicity is established, each embryo or fetus must be identified to accurately and consistently assess growth, amniotic fluid, and bladder status, as alterations in these features are key for recognizing specific MC complications. Each fetus needs to be uniquely identified and referred to as *fetus A*, *fetus B*, and so on. This nomenclature should be consistently maintained throughout the pregnancy. The presence of unequal growth or unique anomalies confirms fetal identity, but in the absence of such discriminating features, placental cord insertions are best used to identify the fetuses (17,18). Hence, once the placenta has developed, the location of the cord insertions should be recorded. Twins should be labeled as specifically as possible. By convention the presenting twin is twin A, but additional details such as inferior location, maternal right location, sex, etc, should be recorded to ensure confident recognition at follow-up examinations.

In higher-order multiple pregnancy, the determination of chorionicity is of even greater importance, as up to 20% will contain at least one MC pair (20). Nontrichorionic triplets have high rates (up to 84%) of complications, including fetal demise,TTTS, growth restriction, major anomalies, and early preterm delivery at less than 29 weeks gestation (20). The outcome of these pregnancies can be much improved by the reduction of the MC pair (17).

If there is discordant size in the first trimester, most experts recommend using the larger twin to date the pregnancy and assess concordance with clinical or menstrual dating. This minimizes the chance of missing early growth restriction or anomalies (2,21).

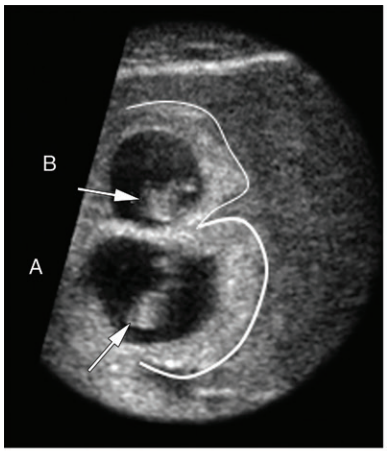

Figure 2. Determining chorionicity and amnionicity in the first trimester. **(a)** US image shows two thick chorionic sacs (outline) surrounding embryo A and B (arrows), consistent with a DC twin pregnancy. **(b)** US image shows one thick chorionic sac (black arrow) surrounding both embryos (white arrow). Each embryo is surrounded by its own thin amnion (blue arrows), a finding consistent with MC-DA twin pregnancy. Each amniotic cavity has its own yolk sac *(Y)*. In most cases, the number of yolk sacs corresponds to the amnionicity of the pregnancy. **(c)** US image shows one thick echogenic chorion (outline) and one thin amnion (blue arrow), with embryos A and B (white arrows) inside a single amniotic sac, a finding consistent with MC-MA twins. MC-MA is also abbreviated as *MoMo* in the literature.

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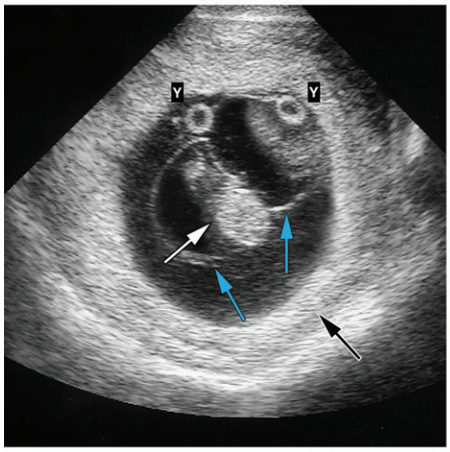

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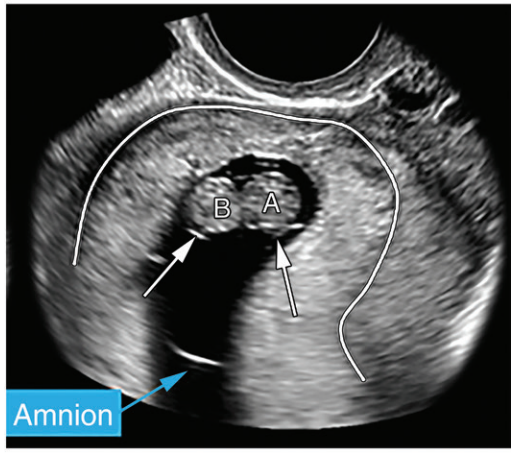

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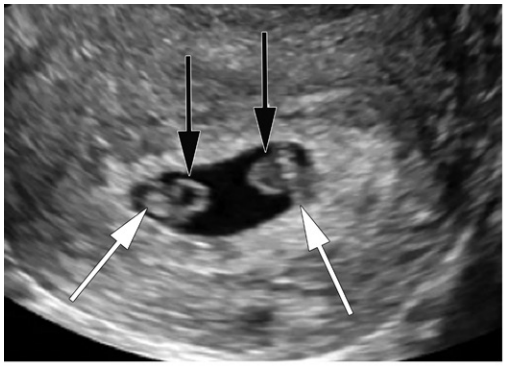

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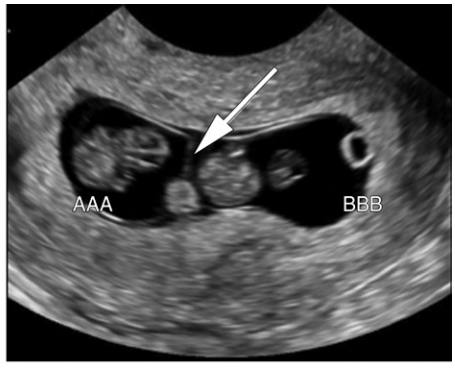

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Figure 3. Early MC-DA twin pregnancy diagnosis as a pitfall for MC-MA twin pregnancy. **(a)** US image at 5 weeks 5 days gestation shows a single chorionic sac, indicating an MC twin pregnancy. Two embryos (white arrows), each with their own yolk sac (black arrows) without a visible intervening membrane, are depicted. **(b)** Subsequent US image at 11 weeks 3 days gestation shows a thin membrane separating the two amniotic sacs (white arrow), confirming the DA nature of the pregnancy. Early in gestation, the thin membrane may be below the limits of resolution for US and cannot be visualized, creating a potential diagnostic pitfall. However, the presence of two yolks, each with its own embryo, is most suggestive of a DA twin pregnancy. *AAA* = twin A, *BBB* = twin B.

US Surveillance for MC Twin Pregnancies

All twin pregnancies are higher risk than singleton pregnancies, but as MC twins have a higher rate of complications, increased surveillance is merited. Some complications such as TRAP sequence and conjoined twins (Fig 6) may even be apparent in the first trimester, but for timely detection of TTTS, sFGR, and TAPS, US surveillance should be performed every 2 weeks from 16

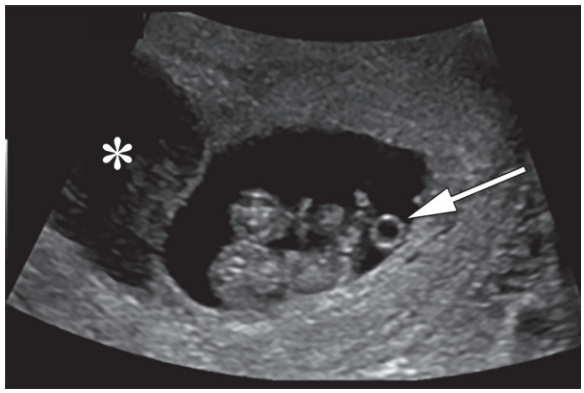
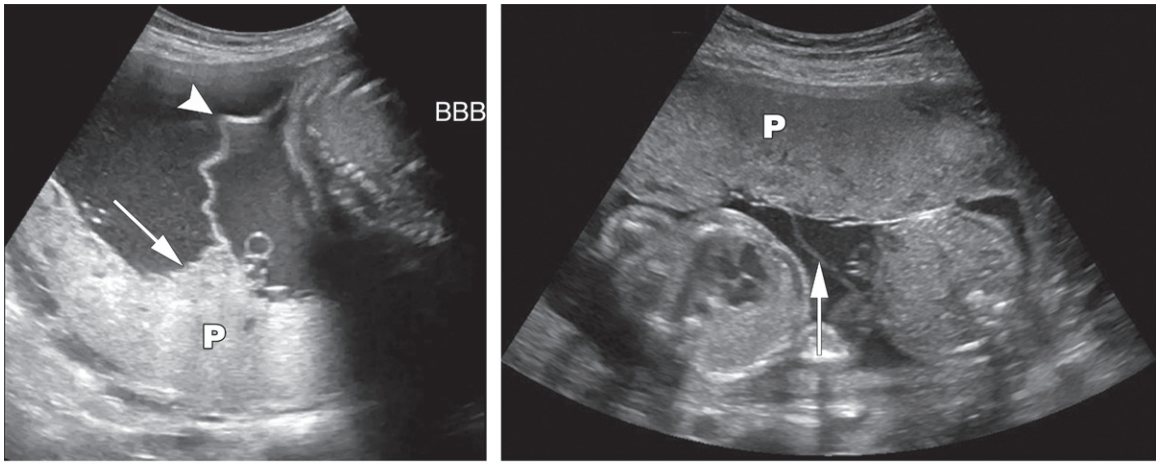


Figure 4. US image obtained at 8 weeks 0 days gestation after single-embryo-transfer in vitro fertilization shows an MC-MA twin pregnancy. A single chorionic sac is a finding compatible with an MC twin pregnancy. Two embryos with a single yolk sac (arrow) are depicted, a finding most compatible with an MA twin pregnancy. An important concern at this stage is the possibility of conjoined twins. Follow-up images obtained later in pregnancy are necessary to exclude the possibility of conjoined twins. Note the subchorionic hemorrhage (*) depicted superior to the gestational sac.



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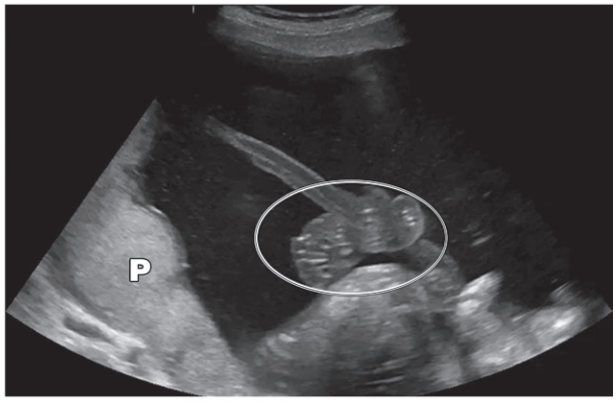

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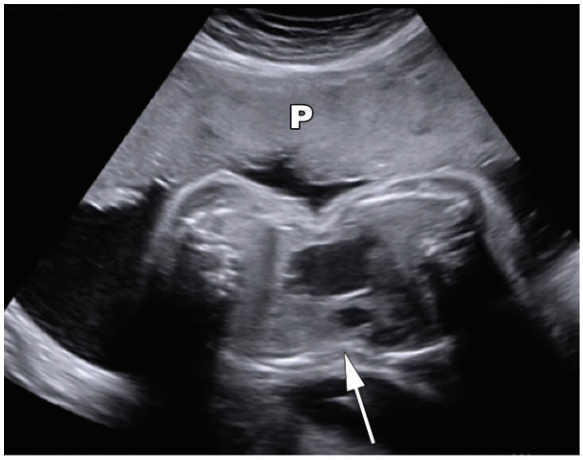
Figure 5. Determining chorionicity and amnionicity in the second trimester. *P* = placenta. **(a)** US image shows DC-DA twins with a fused DC placenta. Note that part of the placenta (arrow) is interposed in a thick membrane (arrowhead), often referred to as a *twin-peak sign*. Other US views demonstrated that the twins were different sexes. **(b)** US image shows a single shared anterior placenta with a thin membrane (arrow) separating the twins, consistent with an MC twin pregnancy. This is referred to as the *T sign*, which is visualized in MC twins. Both twins are the same sex. **(c)** US image shows entangled cords (oval), which is a hallmark finding of MC-MA twins with a single shared placenta.

weeks until the end of gestation (17,18,22). No randomized control trials have evaluated this interval, and this recommendation is based on expert consensus. Similarly, the appropriate interval for serial assessment of fetal growth is not well established. The North American Fetal Therapy Network recommends measuring growth every 4 weeks, with intervening US examinations every 2 weeks to assess fluid and bladder volumes. At the authors' institution (P.J., T.A.M), growth US examinations are performed at least every 2 weeks, along with the surveillance examinations for fluid and bladder. Growth is not assessed at intervals of less than 2 weeks. Like every other pregnancy, a complete anatomic examination on the basis of the American Institute of Ultrasound in Medicine guidelines should be performed between 18 and 22 weeks.

Doppler US examinations of various vessels, including the umbilical artery, ductus venosus, MCA, and umbilical vein, are performed as needed for the evaluation of TTTS, TAPS, and sFGR. Formal fetal echocardiography is also recommended at 18–22 weeks, as MC twins have a higher prevalence of cardiac anomalies, both congenital and as a result of the altered hemodynamics in TTTS (2). Specifically, in MC twins with TTTS, the recipient twin has been reported to demonstrate cardiac functional abnormalities.



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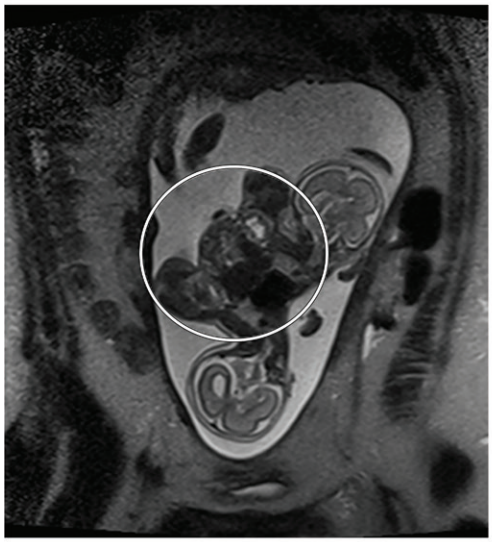

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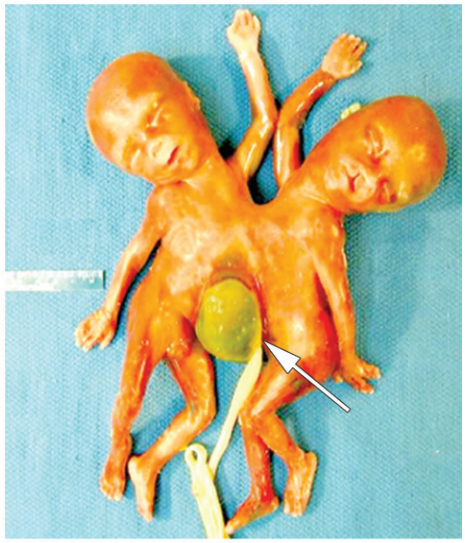


c.

Figure 6. Conjoined twins in three patients. **(a)** US image shows MA twins with conjoined cardiothoracic structures (arrow) (thoracopagus type). Note the single shared placenta *(P)*. **(b)** Oblique axial MR image shows conjoined hearts of MA thoracopagus conjoined twins (circle). **(c)** Gross specimen photograph after pregnancy termination shows thoraco-omphalopagus twins. Note the separate omphalocele (arrow). There is a high prevalence for other congenital abnormalities in conjoined twins, even when the anatomy is not conjoined.

In recent studies, researchers have shown that structural abnormalities leading to right ventricular outflow obstruction may develop in the recipients even in later gestation (3%–10% of cases), either before or after laser coagulation therapy for TTTS (23). Right ventricular outflow obstruction occurs usually in TTTS recipient twins but can rarely be present in TTTS donors and larger twins of pregnancies complicated by sFGR (23). Hence, attention should be paid to cardiac anatomy and contractility at each follow-up examination, and a low threshold for referral to fetal echocardiography should be maintained if any cardiac abnormalities become apparent.

MC twins are considered genetically identical but they may be phenotypically discordant, even for major congenital abnormalities. Structural abnormalities in MC twins are more common than in singletons or DC twins (22). Major anomalies occur in 6%–8% of MC twins but in only 1%–2% of DC twins (4). Given the single shared placenta, discordant anomalies may also negatively affect the healthy co-twin (4,24,25). Once the placenta has developed, intrauterine demise of one twin puts the co-twin at risk of demise (10%–25%) or cerebral damage (24%–45%).

Specific Complications of MC Twin Pregnancies

Multiple vascular connections in the single shared placenta result in unique issues complicating MC twin pregnancies; these include TTTS, TAPS, and TRAP sequence. The possible vascular connections are arterio-arterial, arterio-venous, and veno-venous (22,26) (Fig 7). Neither arterio-venous nor veno-venous connections are readily demonstrable at US, but with a diligent search, arterio-arterial anastomoses can be visualized in some cases (27). These arterio-arterial connections are thought to protect against the development of TTTS (27).

sFGR occurs in both DC and MC pregnancies. In MC twins, the cause is thought to be unequal placental sharing, the outcome of which is influenced by the type of vascular connections that

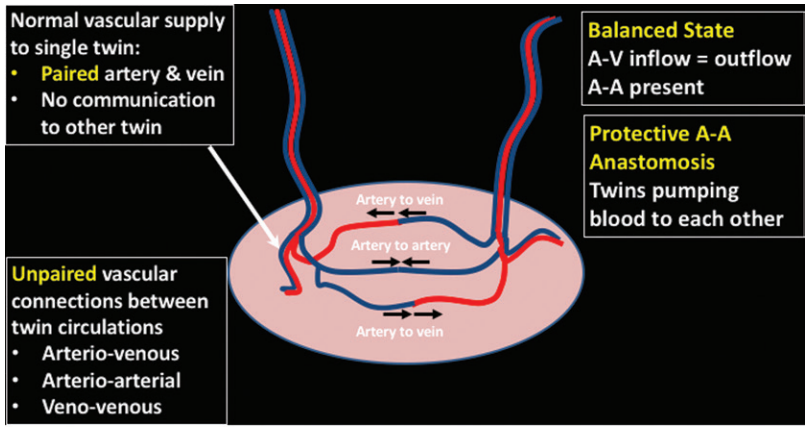


Figure 7. Possible intraplacental connections between twin circulations in an MC placenta. *A-V* = arterio-venous, black arrows = direction of flow in unpaired vascular connections between the two cords, blue lines = artery, red lines = vein, white arrow = normal paired connections. **(a)** Diagram shows a balanced state in an uncomplicated MC twin pregnancy, with equal inflow and outflow between twins created by balanced arterio-venous and arterio-arterial *(A-A)* connections. **(b)** Diagram shows an unbalanced state in an MC pregnancy complicated by TTTS. In this condition, unbalanced arterio-venous flow occurs from one twin to the other. *X* = lack of flow compensation by arterio-venous and arterio-arterial connections.

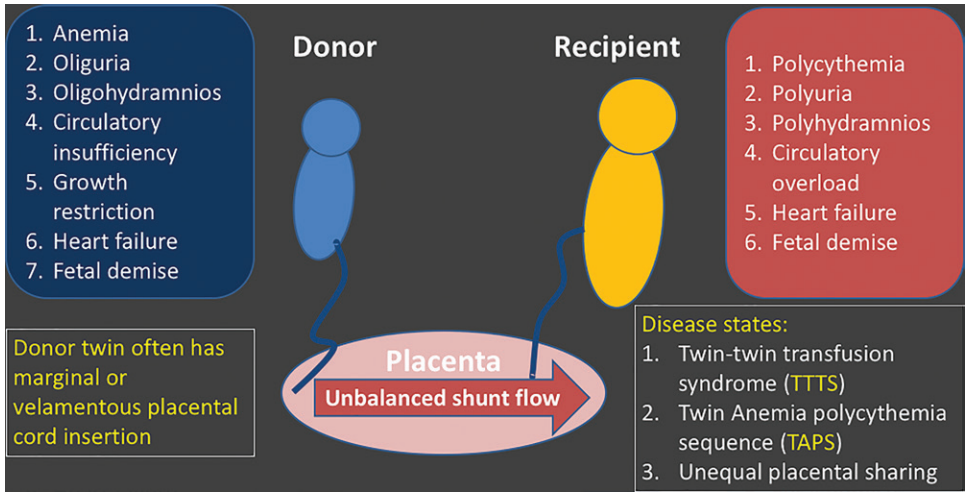
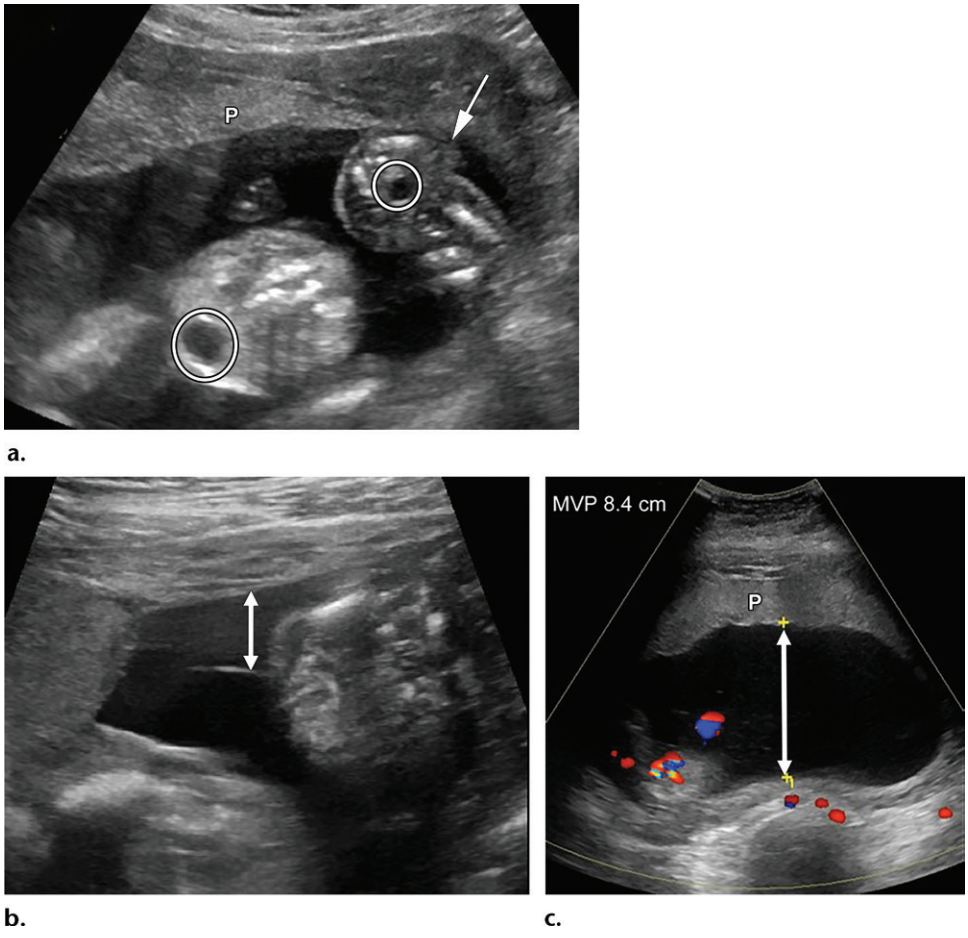


Figure 8. Schematic diagram shows the manifestations of abnormal unbalanced blood flow in a donor and recipient MC twin pair.

predominate. Conjoined twining is unique to MC-MA twin pregnancy; it is a complication of the late division of a monozygotic pregnancy rather than abnormal vascular connections (Fig 6). Umbilical cord entanglement can only occur when the fetuses are within the same amniotic sac (Fig 5c). This is a hallmark finding for an MC-MA pregnancy but can rarely occur following intervention in an MC-DA pregnancy, complicated by rupture of the intertwin membrane (28,29).

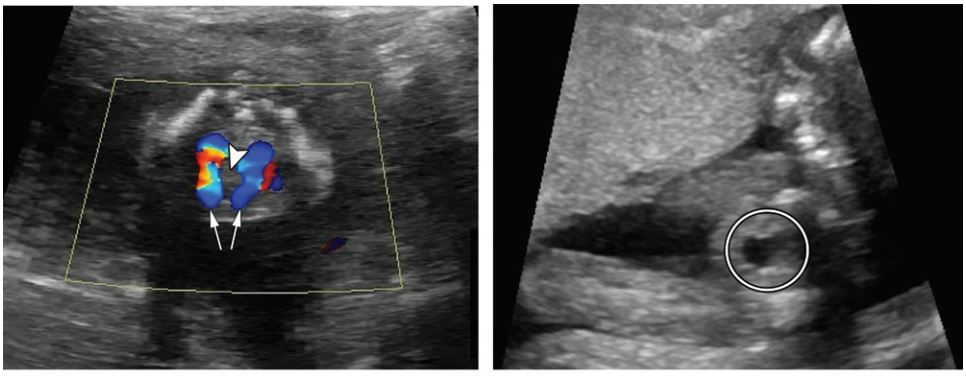
Twin-Twin Transfusion Syndrome

TTTS is one of the most serious consequences of the anomalous connections in the shared MC placenta (Figs 7b, 8–10). It affects 10%–15% of MC pregnancies (30) but causes more than half



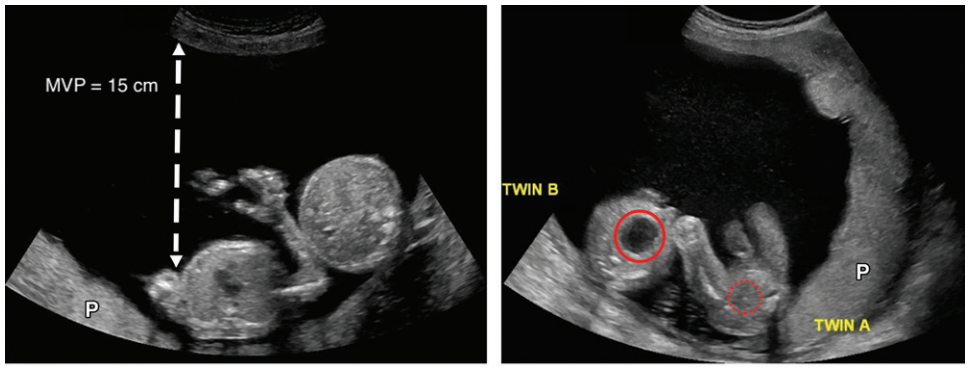
a. b. c.

Figure 9. Stage 1 TTTS in MC-DA twins with a single shared anterior placenta. **(a)** US image shows the stuck-twin appearance of the donor twin (arrow). Instead of being situated at the bottom of the gestational sac, the fetus is apposed to the superior uterine wall. With the continued lack of amniotic fluid, the fetus becomes wrapped, or shrink-wrapped, in the amnion. A bladder (circles) is depicted in each twin. *P* = placenta. **(b)** US image shows oligohydramnios in the donor twin, with a maximum vertical pocket (MVP) (double-sided arrow) of 1.7 cm. Note the difference in the echogenicity of the fluid between the two sacs. **(c)** US image shows polyhydramnios in the recipient twin, with an MVP (double-sided arrow) of 8.4 cm. *P* = placenta. Overall, the combination of oligohydramnios and polyhydramnios in the twin pair with a bladder depicted in each twin are findings consistent with stage 1 TTTS.



a. b.

Figure 10. Stage 2 TTTS complicating an MC-DA twin pregnancy with a single shared anterior placenta. **(a)** Color Doppler US image shows, in addition to oligohydramnios, the donor twin with an absent bladder (arrowhead). To adequately depict the absence of the bladder, a color Doppler US image of the bilateral umbilical arteries (arrows) without a fluid-filled bladder (arrowhead) in between should be obtained. **(b)** US image shows a normal bladder (circle) in the recipient twin. Polyhydramnios was also present (not shown). With the absent bladder in the donor, as well as oligohydramnios and polyhydramnios, these findings now constitute a stage 2 TTTS diagnosis.



a. **b.**

Figure 11. Stage 2 TTTS complicating an MC-MA twin pregnancy. *P* = placenta. **(a)** US image shows a single shared posterior placenta. The presence of a single amniotic space makes diagnosing TTTS challenging. However, polyhydramnios with a maximum vertical pocket *(MVP)* (double-headed dashed arrow) of 15 cm provides a clue to a potential TTTS diagnosis. **(b)** US image shows that the bladder is absent in twin A (dotted circle), which helps to identify the donor twin. A normal bladder is depicted in the recipient twin (solid circle). The presence of polyhydramnios and an absent bladder in one twin helps confirm stage 2 TTTS in an MC-MA twin pregnancy.

of deaths in MC-DA twins (4). If left untreated, pregnancy loss rates are as high as 70%–100% for severe disease diagnosed early in gestation (31). Even surviving infants have a 10%–30% chance of neurologic morbidity (32).

Pathogenesis of TTTS

The exact cause of TTTS is not well understood. However, vasoactive substances are thought to play a role (22). Unidirectional flow through a dominant arterio-venous anastomosis makes one twin the donor (with findings of anemia, oliguria, and oligohydramnios) and the other the recipient (with findings of polycythemia and polyhydramnios) (22) (Figs 7–10). Eventually, both fetuses develop progressive cardiovascular decompensation. In particular, increased afterload in the recipient causes myocardial hypertrophy with abnormal diastolic and systolic function (33).

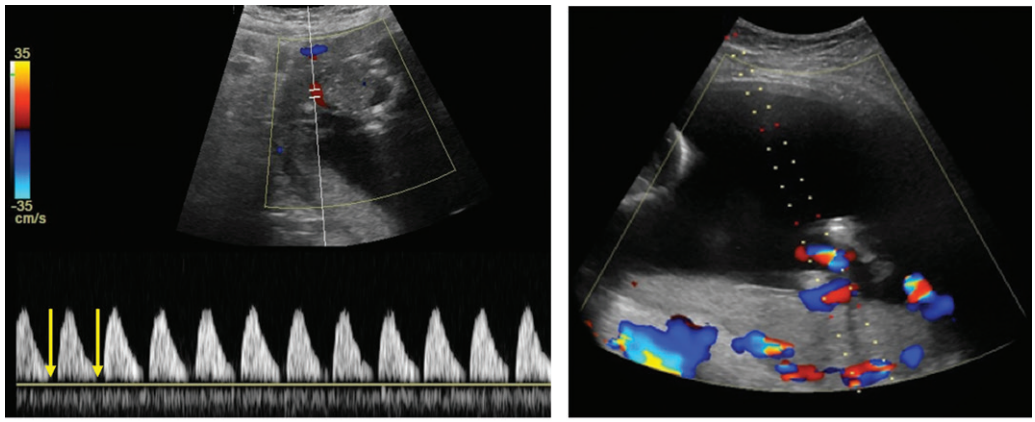
The diagnosis of TTTS is usually made in the second trimester, with the hallmark observation being oligohydramnios in one sac and polyhydramnios in the other (Figs 9b,9c). In the first trimester, reversal of the A wave in ductus venosus waveforms may indicate an increased risk for TTTS development (34). Discordant nuchal translucency and crown-rump lengths have also been suggested as potential first trimester markers for TTTS, but the specificity is low (35,36). Once diagnosed, TTTS is staged on the basis of imaging findings, with the Quintero staging system being the most commonly used (Table 2). The stages of the Quintero system are described in the following sections.

Stage 1: Oligohydramnios-Polyhydramnios Appearance.—In stage 1 TTTS, oligohydramnios effects the donor, with the maximum vertical pocket measuring less than 2 cm (Fig 9b). Polyhydramnios affects the recipient twin, with the maximum vertical pocket measuring greater than 8 cm (Fig 9c). In the absence of true oligohydramnios-polyhydramnios (or oli-poly) appearance, the presence of fluid discordance (with one twin having low normal and one twin having high normal maximum vertical pockets) increases the risk for subsequent development of TTTS, and such cases should be followed up sooner than a 2-week interval (37–39). A normal donor bladder is present (Fig 9a). The diagnosis is much harder to make in MC-MA twins, as there is only one amniotic space. TTTS usually manifests with polyhydramnios in these patients (Fig 11) (22).

Stage 2: Absent Bladder.—In addition to the combination of oligohydramnios and polyhydramnios, the donor bladder is absent in stage 2 TTTS (Fig 10). Usually, this is associated with anhydramnios, which creates the stuck twin appearance (Fig 9a). With no substantial amniotic fluid remaining in the sac, the donor twin is shrink-wrapped in the membranes and becomes apposed to the uterine wall.

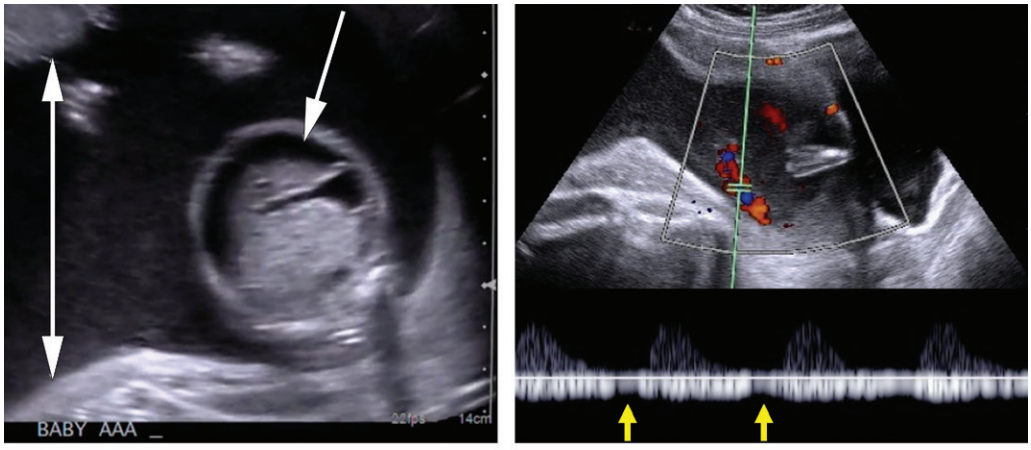
Stage 3: Doppler US Abnormalities.—In cases of more severe disease, Doppler US abnormalities become evident, including absent diastolic flow in the umbilical artery (Fig 12), most commonly in the donor twin. Reversed flow in the ductus venosus can also be depicted. Umbilical vein notching can be depicted, which is a premorbid waveform (Fig 13).

Stage 4: Hydrops Affecting One or Both Fetuses.—Hydrops (Fig 13a) is signified by the presence of fluid in two body compartments,



a. b.

Figure 12. Stage 3 TTTS complicating an MC-DA pregnancy with a single shared posterior placenta. **(a)** Spectral Doppler US image with waveform of the donor umbilical artery shows absent end-diastolic flow (yellow arrows). The donor twin had an absent bladder with oligohydramnios. Polyhydramnios was present in the recipient sac. **(b)** Color Doppler US planning image shows the US window for intraoperative guidance for a laser ablation procedure. A window free of placenta and at least 2 cm from the placental edge is required. The dotted lines indicate a potential track for the laser device.



a. b.

Figure 13. Stage 4 TTTS complicating an MC-DA twin pregnancy. **(a)** US image shows the recipient twin *(BABY AAA)* with polyhydramnios (double-headed arrow), hydrops, and ascites (arrow), findings consistent with stage 4 TTTS. **(b)** Color Doppler US image with recipient umbilical vein waveform shows notching (arrows). Notching in the umbilical vein is a premorbid waveform, with an extremely poor prognosis. The recipient umbilical artery waveform also showed absent end-diastolic flow (not shown).

Table 2: Quintero Staging System for TTTS

Stage I	Oligohydramnios in donor twin and polyhydramnios in recipient twin (“oli-poly”)
Stage II	Absent bladder in donor twin
Stage III	Doppler US abnormalities in umbilical artery and/or umbilical vein
Stage IV	Fetal hydrops
Stage V	Fetal demise

which includes body wall edema, pleural effusion, large pericardial effusion, and ascites.

Stage 5: Fetal Demise.—Single or dual twin demise may occur as an end result of TTTS. The Quintero staging system does not use cardiomyopathy for deciding severity of TTTS, despite the fact that cardiac dysfunction is an important component of the process (33). Nevertheless, ventricular wall hypertrophy and cardiac contractility should

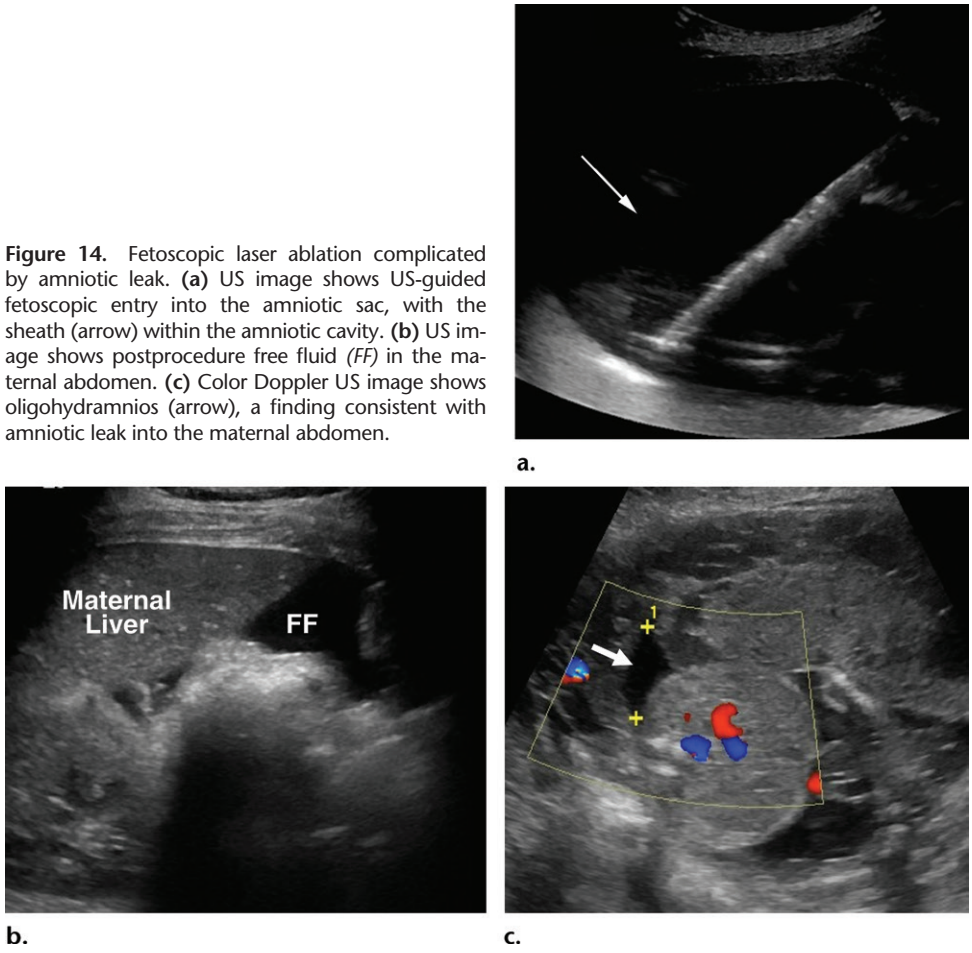


Figure 14. Fetoscopic laser ablation complicated by amniotic leak. **(a)** US image shows US-guided fetoscopic entry into the amniotic sac, with the sheath (arrow) within the amniotic cavity. **(b)** US image shows postprocedure free fluid *(FF)* in the maternal abdomen. **(c)** Color Doppler US image shows oligohydramnios (arrow), a finding consistent with amniotic leak into the maternal abdomen.

be assessed at each surveillance US examination. Some well-known limitations of this staging system are that not all cases follow the rules and skipping between stages is a well-known occurrence (2). Hence, a thorough evaluation for all of the possible abnormalities should be performed, and grading should be determined on the basis of the most abnormal findings.

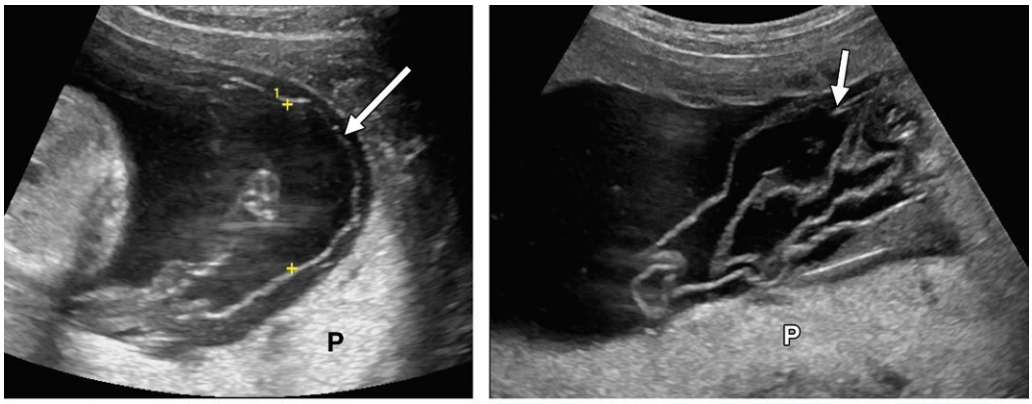
Management of TTTS

Expectant management with close surveillance is recommended for stage 1 TTTS. Early gestational age at detection and intermittent absence of the donor bladder are poor prognostic markers, indicating a greater risk of progression (40). Arterio-arterial anastomoses are thought to protect against the progression to a higher stage of TTTS (40). With expectant management, nearly three-fourths of pregnancies remain stable, and most have survival of at least one twin.

Fetoscopic laser ablation is the most commonly accepted treatment of TTTS (Figs 12b, 14). Outcomes are better than with amnioreduction alone, before 26 weeks gestation (41). The goal is dichorionization of the placenta by laser ablation of abnormal vascular anastomoses, but many connections are deep in the placenta and invisible with a fetoscope (17). When viewed in utero, the visible abnormal aterio-venous anastomoses appear as large-caliber unpaired vessels traveling on the surface of the placenta. The fetoscope, owing to its size, cannot transgress the placenta, so a window at least a few centimeters away from the placental edge is required (Fig 14), and the U.S. Food and Drug Administration restrictions preclude laser ablation after 26 weeks of gestation. It is a relatively safe procedure, but premature rupture of membranes occurs in up to 18%–30% of cases (42,43), especially with early gestational age at intervention (less than 17 weeks) (44).

The risk of maternal complications, such as placental abruption (1%) and intra-abdominal leakage of amniotic fluid (3%) (Fig 14b), are low (42). Chorioamniotic separation can occur as well (Fig 15). For 6 weeks after laser treatment, weekly US with MCA peak systolic velocity (PSV) is used to assess for recurrent TTTS or the development of TAPS. Thereafter, patients are monitored every 2 weeks, as with uncomplicated MC twin pregnancies (45).

Selective feticide (usually by means of radiofrequency ablation) is another option for managing TTTS, usually reserved for conditions in which one of the co-twins has discordant



a. b.

Figure 15. Chorioamniotic separation and shredded membranes after laser ablation. *P* = placenta. **(a)** Postprocedure US image shows a membrane (arrow) outlining the amniotic surface of the placenta and continuing along the uterine body, representing the amnion separated from the chorion, with fluid in the chorioamniotic space. The chorion and amnion are separate in early pregnancy and fluid is present between them. Eventually, the chorioamniotic space is obliterated, and fluid can reenter this space as a procedural complication. The correct method of measuring the maximum vertical pocket after chorioamniotic separation is shown by the calipers. Space outside the amnion should not be included in the measurement. **(b)** US image shows the shredded membranes (arrow).

(potentially lethal) congenital anomalies, severe intrauterine growth restriction affects either of the twins, or one of the twins is moribund (17). Selective feticide is thought to offer benefit over spontaneous in utero demise in terms of longer continued gestation and reduced prevalence of neurologic injury in the survivor (46). This is usually offered after 16 weeks to allow for complete opposition of the amnion and chorion and hence minimizes complications such as chorioamniotic separation.

Twin Anemia Polycythemia Sequence

TAPS is a consequence of chronic unbalanced flow between the twins (Fig 16). Similar to TTTS, it is characterized by anemia of the donor twin and polycythemia of the recipient twin. It occurs spontaneously in 3%–5% of MC twins after 26 weeks gestation (47) and in approximately 13% of cases treated with fetoscopic laser ablation for TTTS because of incomplete dichorionization (48).

TAPS Pathogenesis

TAPS is thought to result from submillimeter-caliber unidirectional arterio-venous connections, which are usually located close to the placental edges. Lack of arterio-arterial connections is believed to further accentuate this condition (49–51).

TAPS Diagnosis

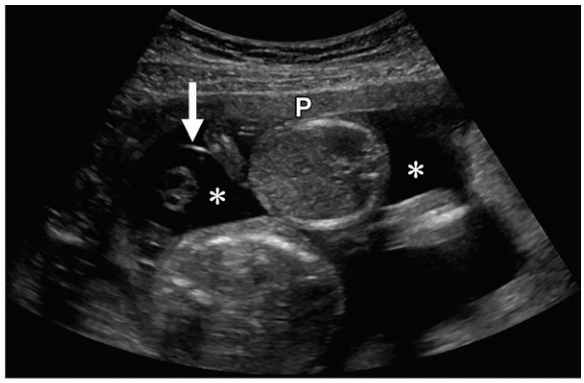
Doppler US allows for a noninvasive US diagnosis of fetal anemia and polycythemia by measuring MCA PSV(Fig 16) (49,50). TAPS is diagnosed when the MCA PSV of one twin is greater than 1.5 multiples of the median (MoM), indicating anemia, and the MCA PSV of the second twin is less than 1.0 MoM, indicating polycythemia. Online calculators such as those available at *perinatology.com* can be used to calculate the MoM corresponding to measured spectral Doppler US velocity (52). Amniotic fluid discrepancy is not a feature of TAPS (Fig 16) and should never manifest in TAPS (17). A US-based staging system has been proposed (Table 3) (53).

TAPS Management

Unlike TTTS, the management of TAPS is still evolving, and optimal prenatal treatment has not been established (17). Depending on gestational age and fetal comorbidities, options include expectant management, selective feticide, delivery, intrauterine transfusion with or without partial exchange transfusion for the polycythemic fetus, and fetoscopic laser photocoagulation (54). In initial studies, laser photocoagulation resulted in prolongation of pregnancy and decreased neonatal morbidity (54,55). However, more studies are needed to further guide management.

Polyhydramnios Affecting Recipient-like Twin

PART is a condition in which isolated polyhydramnios affects one twin of the MC-DA pair, with normal fluid in the co-twin. Polyhydramnios is likely due to polyuria. However, there is no true donor, as the co-twin has normal fluid (27). The bladder and Doppler US findings are normal. While previously considered a forme fruste for TTTS by some authors, more recent studies have shown that PART goes hand in hand with weight discordance, and the fluid discordance is likely related



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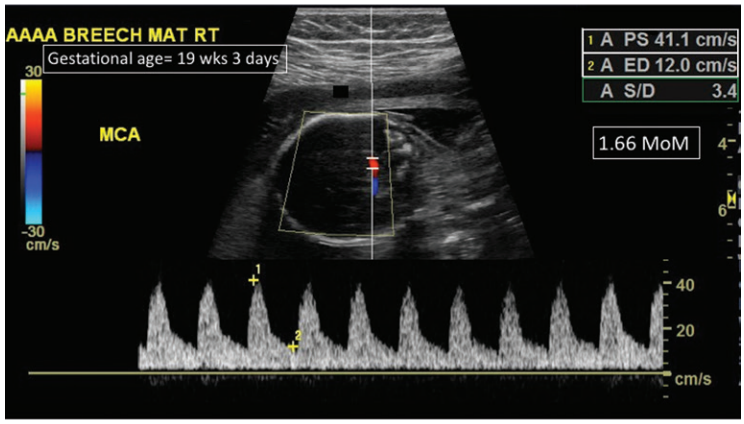

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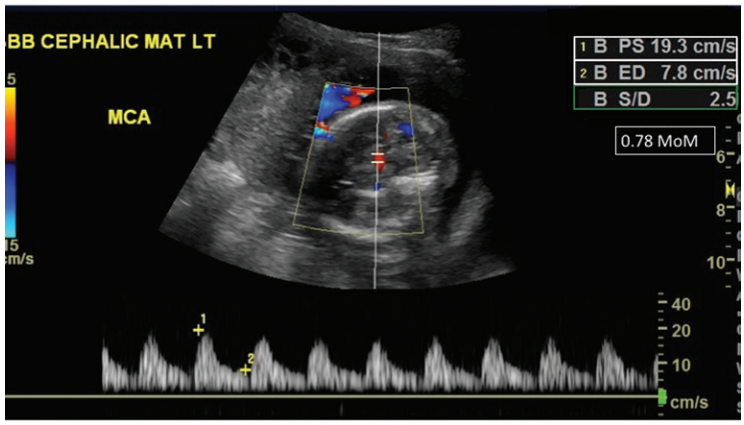


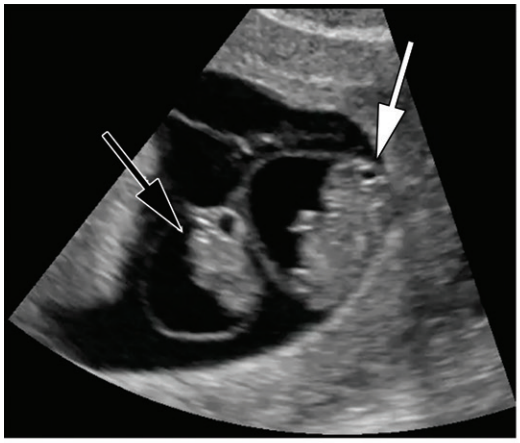
c.

Figure 16. MC-DA twin pregnancy complicated by TAPS. **(a)** US image shows an intervening thin membrane (arrow), a finding consistent with MC-DA twins. Normal fluid (*) is present in both sacs. *P* = placenta. **(b, c)** Color Doppler duplex US images show discordant MCA PSV *(PS)* measurements of 41.1 cm/sec *(cm/s)* (corresponding to 1.66 MoM, which indicates anemia) in twin A *(AAAA)* **(b)** and an MCA PSV of 19.3 cm/sec in the other twin *(BB)* **(c)**. The MCA PSV in twin B corresponds to 0.78 MoM, which constitutes polycythemia. A normal bladder was depicted in each twin (not shown). Fetal anemia and polycythemia in combination with a normal bladder and amniotic fluid constitutes a TAPS diagnosis. *ED* = end-diastolic velocity, *MAT* = maternal, *S/D* = systolic to diastolic ratio, *wks* = weeks.

Table 3: Antenatal US-based Staging of TAPS

Stage 1	MCA PSV of the donor is > 1.5 MoM, and MCA PSV of the recipient is < 1.0 MoM, without other signs of fetal compromise
Stage 2	MCA PSV of the donor is >1.7 MoM, and MCA PSV of the recipient is < 0.8 MoM, without other signs of fetal compromise
Stage 3	Includes the findings of stage 1 or 2, with cardiac compromise of the donor, defined as critically abnormal flow (which includes absent or reversed end-diastolic flow in the umbilical artery, pulsatile flow in the umbilical vein, increased pulsatility index or reversed flow in the ductus venosus)
Stage 4	Hydrops of the donor
Stage 5	Intrauterine demise of one or both fetuses preceded by TAPS

Source.—Reference 53.



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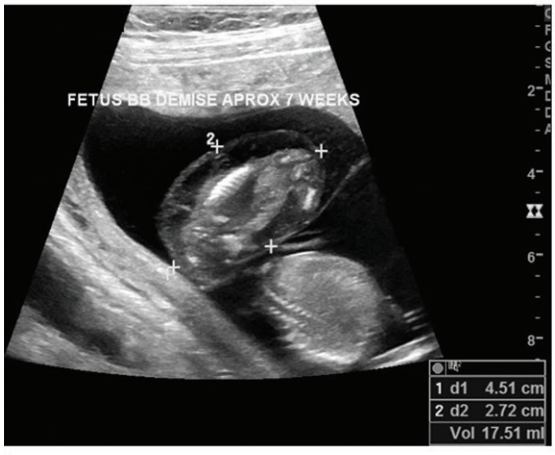

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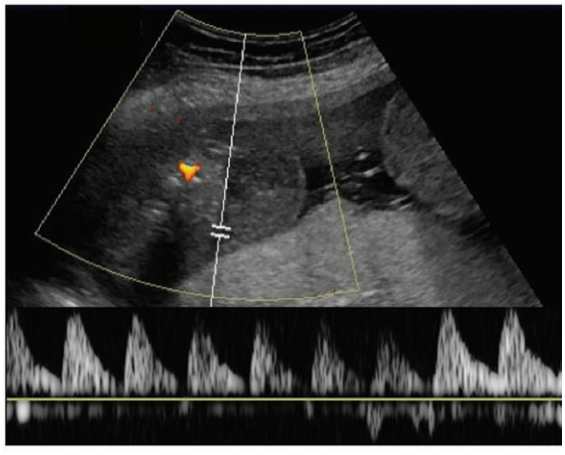


c.

Figure 17. TRAP sequence. **(a)** US image obtained early in pregnancy shows MC-DA twins. A well-formed living embryo (white arrow) and an amorphous demised embryo (black arrow) without cardiac motion are depicted. **(b)** US image obtained at 6-week follow-up shows unexpected interval growth of the anomalous embryo *(BB)*, without cardiac motion. The embryo is severely edematous (calipers) and has identifiable anatomy, such as a spine and other osseous structures, which are highly suspicious for TRAP sequence. *Aprox* = approximately, *d1* = distance 1, *d2* = distance 2. **(c)** US Doppler image and waveform interrogation obtained at the umbilical artery of the anomalous fetus shows reversed umbilical arterial flow, flowing toward the anomalous twin instead of toward the placenta. This is a pathognomonic finding of TRAP sequence. The reversal of flow is a consequence of perfusion of the anomalous twin by the normally developed pump twin.

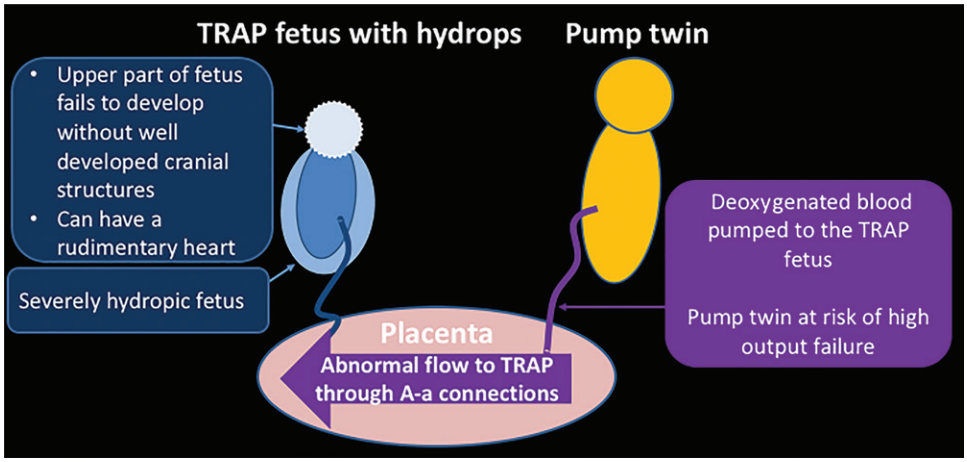


Figure 18. Schematic diagram shows the TRAP sequence in a twin pair. *A-a* = arterio-arterio.

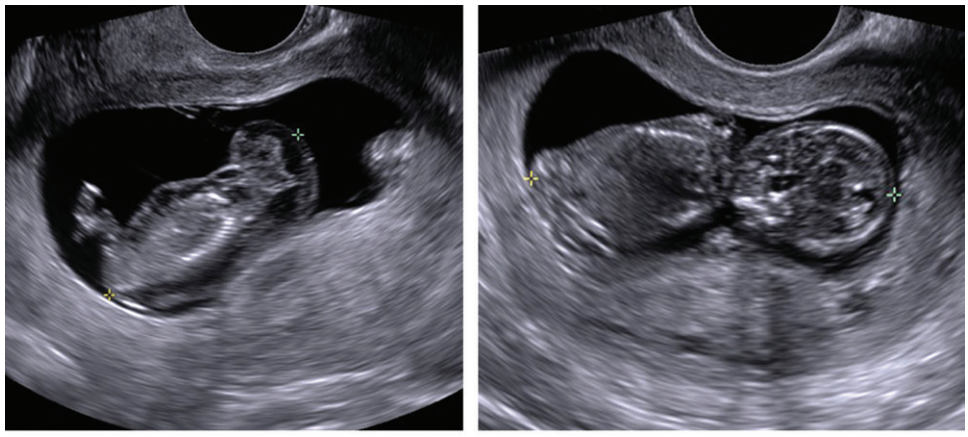
to discrepant fetal weights (27). Less than 25% of patients with PART actually progress to TTTS, and overall the prognosis is good. The presence of arterial-arterial anastomosis is protective against progression to more severe disease (27).

TRAP Sequence

TRAP sequence, previously referred to as acardiac twinning, is a rare complication specific to MC twin pregnancies (Figs 17–20). It occurs in at about 1% of MC twins (56), mostly in MC-DA twins, but it can also occur in MC-MA twins (57) or in any multigestational pregnancy that includes an MC pair.

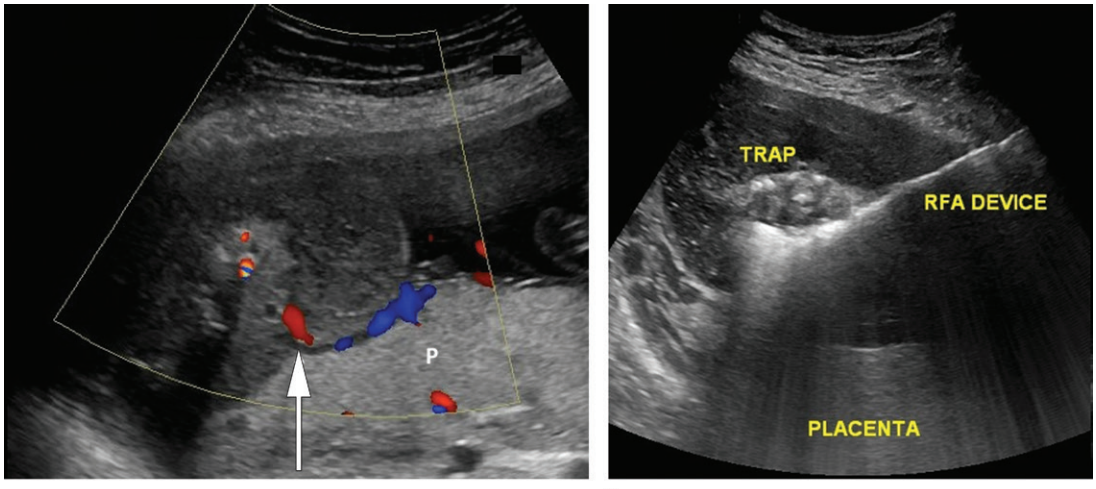
Pathogenesis of TRAP Sequence

In TRAP sequence, one twin develops with a normal phenotype, while the second twin



a. b.

Figure 19. MC-DA twin pregnancy with TRAP sequence and demise of the pump twin at 12 weeks gestation. **(a)** US image shows an anomalous hydropic fetus lacking normal cranial structures but with identifiable osseous structures. **(b)** US image shows hydropic changes present in the demised pump twin. The abnormal twin cannot survive ex utero, and management of this condition is focused on the well-being of the pump twin. Hence, radiofrequency ablation of the abnormal twin is considered.



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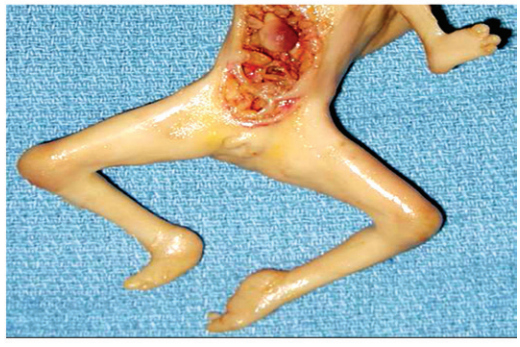


c.

Figure 20. Radiofrequency ablation of an abnormal TRAP sequence twin. **(a)** Color Doppler US image shows a hydropic anomalous co-twin, with perfusion and flow within the cord (arrow). Spectral Doppler US waveform (not shown) confirmed umbilical artery flow toward the abnormal twin, a finding consistent with TRAP sequence. *P* = placenta. **(b)** US image shows the correct placement of the radiofrequency ablation *(RFA)* device, with the tip at the abdominal cord insertion. Unlike the fetoscope, the device can be placed through the placenta owing to its smaller size. **(c)** Gross photograph after delivery shows the anomalous fetus with well-formed lower extremities and an anterior abdominal wall defect following radiofrequency ablation.

abnormally develops with a broad spectrum of appearances (Fig 17). At one extreme end of the spectrum, the entire fetus may be amorphous, while at the other end, there can be quite well-developed and recognizable structures, particularly the spine and lower extremities (Fig 17). A rudimentary heart that contracts irregularly may sometimes also be present; hence, the nomenclature of acardiac twinning is not favored.

The hallmark finding is that blood flow in the umbilical artery at its insertion into the abdomen of the abnormal twin is reversed in that it flows toward the twin, not toward the placenta (Fig 17c). The abnormal twin is entirely perfused by the normally developed co-twin, referred to as the

pump twin (Figs 17, 18). The TRAP sequence fetus blood supply comes from direct arterio-arterial and veno-venous connections, with the pump twin in a steal-like phenomenon. Normal umbilical arterial flow is away from the fetus, carrying deoxygenated blood toward the placenta for replenishment.

In TRAP sequence, the umbilical arterial flow is directed from the pump twin into the abnormal fetus (Fig 17c). The blood then leaves the TRAP sequence twin through the umbilical vein and bypasses the placenta, returning to the pump twin in a further deoxygenated state. There is 100% nonsurvival of the abnormal twin, but persistent perfusion allows it to grow, and the larger it gets the more negative impact it has on the pump twin, with mortality as high as 55% (58). Negative impact on the pump twin results from the high output state (Fig 18), which can lead to congestive heart failure, intrauterine fetal demise (Fig 19b), polyhydramnios, premature rupture of membranes, and preterm labor.

Imaging TRAP Sequence

A detailed anatomic survey should be performed for the pump twin, as identification of anomalies is critical to guide parental counseling and decision making. Evidence of strain to the pump twin includes enlargement of the heart, abnormal contractility, and hydrops, which can include body wall edema, pleural or pericardial effusion, and/or ascites. Doppler US of the pump twin should include evaluation of the umbilical artery, MCA, and ductus venous. In the setting of high-output cardiac failure, the ductus venous waveform can become abnormal and progress from deepening to absence to eventual reversal of the A wave.

US evaluation of the TRAP sequence twin includes an assessment of the morphology, with documentation of recognizable structures. The TRAP sequence twin volume is calculated on the basis of the measurements obtained in three orthogonal planes, which must be performed with care to elongate the fetus for accurate and reproducible serial measurement. In any MC pregnancy with an anomalous twin, check the direction of flow in the umbilical artery. If it is toward the abnormal fetus, that is evidence of a TRAP sequence diagnosis. In some cases, autoinfarction occurs, and there is no demonstrable flow into the anomalous twin. These cases have a favorable prognosis, as there is no longer any strain on the pump twin, and such cases are allowed to progress to term.

Be aware of twinkle artifact simulating blood flow at color Doppler US. This artifact occurs owing to osseous structures, and hence true perfusion should be confirmed with spectral Doppler US waveform analysis, which will demonstrate only noise with twinkle artifact. If surgical intervention is contemplated, it is important to map the location of the placenta and record any uterine abnormalities. In the absence of intervention, follow-up examinations are not standardized but are continued weekly until the TRAP sequence twin has no flow or is unchanged or smaller in size for two consecutive US examinations (59).

TRAP Sequence Management

When the TRAP sequence twin is small, there is almost uniform survival of the pump twin with expectant management only. In MC-DA pairs, survival is 100% (56). Expectant management can be used when the volume of the TRAP sequence twin is less than 50% of that of the pump twin. The 50% threshold was proposed by Moore et al (60) who demonstrated a linear relationship between the volume of the acardiac twin and heart failure and premature labor in the pump twin (59,60).

Treatment is aimed at stopping blood flow to the abnormal twin and has included open hysterotomy, fetoscopic ligation, selective delivery, laser or microwave coagulation techniques, alcohol or radiofrequency ablation (Fig 20b), and high-intensity focused US (HIFU) (56,57). Treatment is preferably performed in the second trimester owing to lower risk of pregnancy loss. Earlier interventions during the first trimester (both with fetoscopic laser and radiofrequency ablation) have shown poor outcomes owing to high pregnancy loss, similar to those losses that occur in other reductions performed for twinning (61).

Radiofrequency ablation is the most widely performed treatment procedure. A targeted device applies a current that causes tissue coagulation at the abdominal cord insertion of the abnormal twin (Fig 20b). Survival of the pump twin is greater than 90% in MC-DA pairs (57). Complications include hematoma, chorioamniotic separation, shredded membranes, premature labor, preterm premature rupture of the membranes, oligohydramnios, and thermal injury (56,57). HIFU is currently being investigated for the treatment of TRAP sequence. Unlike other methods, HIFU is completely noninvasive and requires no uterine puncture, greatly decreasing the risk of complications to the mother and the fetus. However, success in vessel occlusion can be partial, with efficacy of up to only 83% (62).

Selective Fetal Growth Restriction

Another complication of twin gestation is unequal growth. When the smaller twin meets the criteria for growth restriction, this is referred to as sFGR. It occurs in both DC and MC twin pregnancies but is more concerning in the

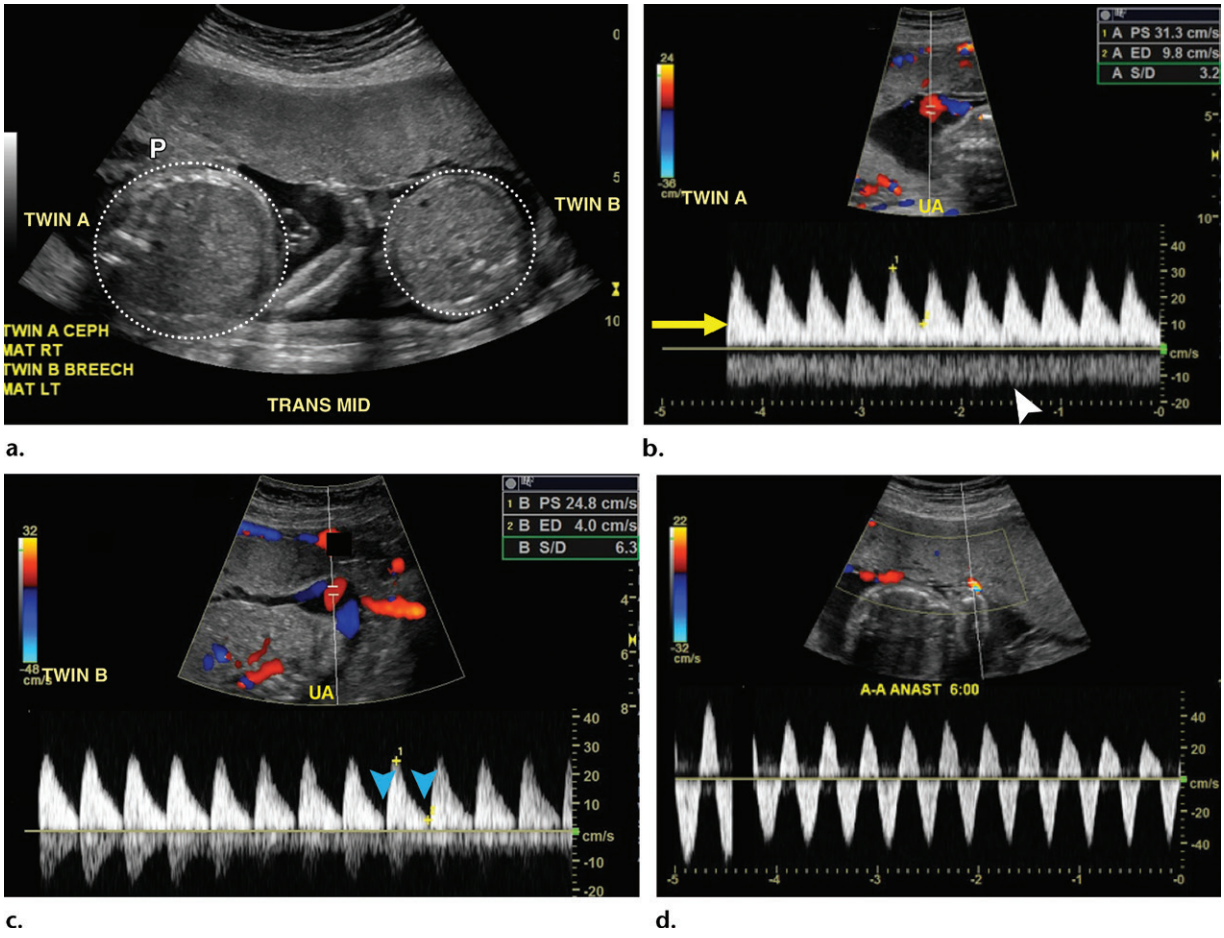


Figure 21. MC-DA twin pregnancy with unequal placental sharing. **(a)** US image shows MC-DA twins (dotted circles) with a single shared anterior placenta *(P)*. Visual discrepancy in the size of the twins is evident. A 30% weight discordance was determined. The smaller twin, twin B, has a velamentous cord insertion, which is a known risk factor for size discordance between twins. *CEPH* = cephalic, *MAT* = maternal, *LT* = left, *RT* = right, *TRANS MID* = transverse middle. **(b)** US Color Doppler image shows a twin with a normal umbilical artery *(UA)* waveform, with forward diastolic flow (arrow). Normal antegrade umbilical vein flow is present without pulsatility (arrowhead). *ED* = end-diastolic velocity, *PS* = PSV, *S/D* = systolic to diastolic ratio. Keys are the same for Figure 21c. **(c)** Color Doppler US image shows occasional loss of end-diastolic flow (arrowheads), sometimes diagnosed in the setting of an arterial-arterial anastomosis. **(d)** US Doppler image of the placental surface shows an arterial-arterial anastomosis *(A-A ANAST)* with to-and-fro flow. To visualize arterial-arterial anastomosis, search along the placental surface with color Doppler US for possible vessels with aliasing. Spectral interrogation of this focus should be pursued to evaluate for to-and-fro flow.

Table 4: Criteria for Diagnosis of sFGR*

Diagnosis	EFW Percentile	Δ EFW	AC Percentile	UA PI Percentile	Number of Criteria Needed for Diagnosis
MC pregnancy	<10th	≥25%	<10th	>95th	Three of four
DC pregnancy	<10th	≥25%	NA	>95th	Two of three

Source.—Reference 65.
*An EFW less than the 3rd percentile in one twin is sufficient to make an sFGR diagnosis; otherwise these criteria are used in combination for diagnosis. AC = abdominal circumference, Δ EFW = difference in twin EFWs, EFW = estimated fetal weight, NA = not applicable, UA PI = umbilical artery pulsatility index.

10%–15% of affected MC twins because demise of the smaller twin has potentially catastrophic consequences for the larger twin (63). Unequal placental sharing is thought to be the cause of sFGR in MC twins. Discordant cord insertions (eg, central vs velamentous, or velamentous vs marginal) correlate with weight discordance, and velamentous cord insertion in one or both twins has been shown to increase adverse outcomes, including TTTS in MC twin pregnancies (64).

Table 5: Classification and Outcome for Types of sFGR in MC Twins

Type	Umbilical Artery Doppler US Findings	Risks	Perinatal Survival
I	EDF present	Generally considered to have benign prognosis	97%
II	AEDF or REDF	Risk of hypoxia and demise of the smaller twin	50%
III	Cyclical flow*	Risk of demise of the smaller twin Neurologic injury to the normally grown twin	80% but unpredictable clinical course

*Variable positive, absent, and REDF.
Note.—AEDF = absent end-diastolic flow, EDF = end-diastolic flow, REDF = reversed end-diastolic flow.

Consensus criteria for the definition of sFGR have recently been published (65) (Fig 21) (Table 4). Once the diagnosis of sFGR is established in MC twins, umbilical artery Doppler US findings in the smaller twin are used for further classification (66). In type I sFGR, there is normal end-diastolic flow in the umbilical artery, and in type II the end-diastolic flow is absent or reversed. In type III, end-diastolic flow is intermittently absent or reversed (Fig 21). Type III flow is also called cyclical flow.

Additionally, as with any fetus affected by growth restriction, altered cerebroplacental ratio, or brain sparing, may be observed. In this case, the systolic-to-diastolic ratio as measured at MCA Doppler US becomes less than that of the umbilical artery. This reflects that the brain is being perfused preferentially over the rest of the body. The prognosis varies with subtype, as outlined in Table 5.

Type 1 sFGR can be managed expectantly, but there is a potential role for fetal intervention with either attempted laser dichorionization of the placenta (as used in the treatment of TTTS) or selective reduction of the smaller twin (67). Despite inconsistencies in reported studies, the data on perinatal outcome of MC twin pregnancies complicated by sFGR show the importance of recognition and appropriate referral (67).

It is important to note that there is overlap between sFGR and TTTS, but in the latter the observation of oligohydramnios in the donor and polyhydramnios in the recipient is the hallmark finding. There may or may not be sFGR of the donor or abnormal Doppler US findings in higher grades. In sFGR, the smaller twin may have oligohydramnios but the normally grown twin has normal fluid volume.

In Utero Twin Demise

When one MC twin dies in utero in the second or third trimester, the surviving twin has a high likelihood of death and severe cerebral injury (Fig 22) as a consequence of acute exsanguination into the low-pressure system of the dead twin. Expectant management is preferred, as the damage occurs at the time of co-twin demise. Emergent delivery only adds to the risk of prematurity to the surviving twin, and the mother may have to undergo cesarean delivery, with ultimate demise of both twins.

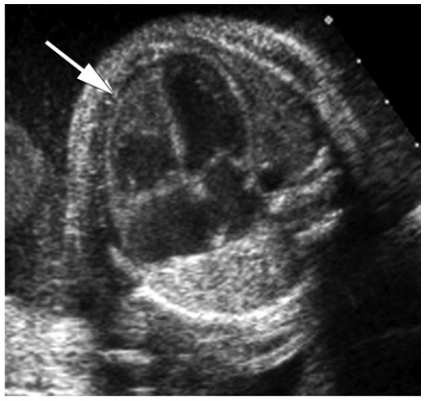
A previous theory that emboli from the twin who died cause ischemic injury in the surviving twin has been largely abandoned. First-trimester co-twin demise does not cause similar adverse consequences as does demise in the second trimester or later. This is likely related to placental vascularity, and hence the vascular anastomosis between the two twins is not well developed this early in gestation.

MA Twin Pregnancy

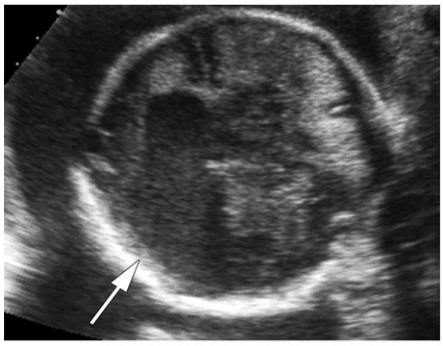
MA twins are all MC and share a single placenta. A recent study conducted in the United Kingdom showed that the estimated total incidence of MC-MA twin pregnancies was 8.2 per 1 000 twin pregnancies and the birth incidence was 0.08 per 1 000 pregnancies overall (singleton and multiple) (68). Cord entanglement and conjoined twinning are specific complications of MC-MA twins. Congenital anomalies, TTTS, and prematurity are also common.

MC-MA twins are at a high risk of perinatal loss in the third trimester, but most losses seem to occur as unexpected events; thus management is controversial (69). Traditionally, daily fetal monitoring and serial growth US examinations were initiated at viability, often with hospitalization at 24–28 weeks gestation to facilitate surveillance, but the MONOMONO study (70) showed no difference in mortality with inpatient versus outpatient surveillance. Additionally, there were no intrauterine fetal deaths or neonatal deaths between 32 weeks and 36 weeks 6 days, challenging the dogma that MC-MA twins should be delivered at 32–34 weeks by cesarean delivery (70).

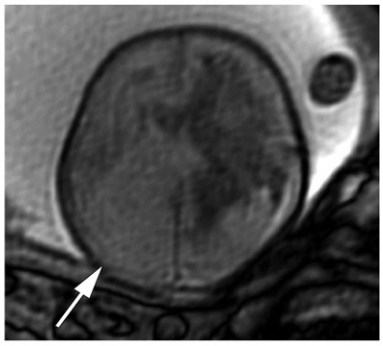
Conjoined twinning (Fig 6) is the rarest complication of MA twin pregnancy. A recent study predicted the total incidence of conjoined twinning to be around 1.47 per 100 000 births (71), with a significant female predominance of the



a.



b.



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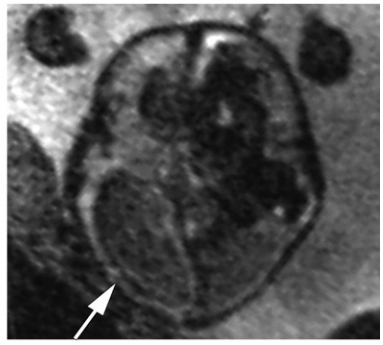

d.

Figure 22. Effects of co-twin demise on the surviving twin in MC twin pregnancy. **(a)** US image shows a dilated heart (arrow) in the surviving fetus, representing ischemic cardiomyopathy. This twin was also diagnosed with ischemic brain injury (not shown). **(b)** US image in another patient shows a surviving twin with a large right parietal infarct (arrow) and diffuse left cerebral hemisphere ischemia. The brain was previously structurally normal. **(c, d)** Corresponding fetal T2-weighted **(c)** and diffusion-weighted **(d)** MR images from the same patient as in **b** show large right parieto-temporal encephalomalacia (arrow), which corresponds to the US findings.

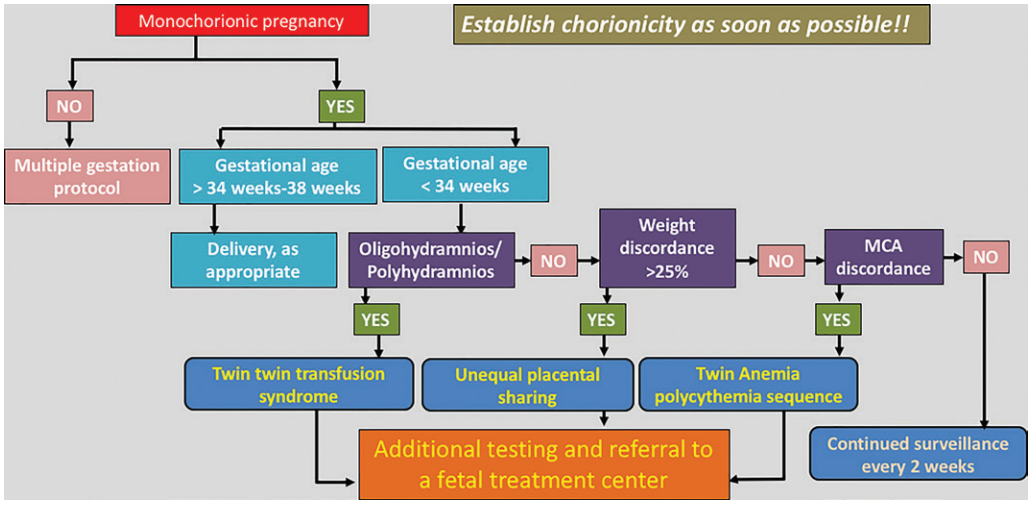

Figure 23. Flowchart shows an algorithm for diagnosis, management, and referral pathways to fetal treatment centers for cases of MC twin pregnancies.

thoracopagus type and significant male predominance in parapagus and parasitic types (71). In addition to conjoined organs, there is an increased prevalence of structural anomalies in the nonconjoined organs (71,72). Mortality is high, with only 18% of prenatally diagnosed conjoined twins surviving (72).

Conclusion

US surveillance is an integral part of the diagnosis and management of MC twin pregnancy. Early determination of chorionicity and amnionicity is imperative to institute surveillance for complications such as TTTS, PART, TRAP sequence, and sFGR. Early detection offers multiple advantages, including early referral to regional specialized centers and early intervention. This allows ample time for patient information and education about the complex disease process affecting the gestation and empowers patients to make informed decisions about pregnancy management. A proposed algorithm for referral

to specialized fetal treatment centers is outlined in Figure 23.

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