Twins Are Not Just Twice the Work-The Importance of Diagnosing Twin Vascular Anomalies
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DIAGNOSING TWIN VASCULAR ANOMALIES

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Twin gestation pregnancies, especially monochorionic gestations, carry a significantly higher risk of complications and adverse outcome. Indeed, monochorionic twin pregnancies have a three- to five-fold higher potential perinatal morbidity and mortality compared with dichorionic twin pregnancies, with most of the complications due to shared vascular connections in the placenta. Sonographic assessment of chorionicity and amnionicity, as well as identifying these vascular pathways, is essential.

THE ZYGOSITY OF TWINNING

Twin gestations occur one of two ways:

Dizygotic twins: result from two ova which are fertilized by two sperm, producing two unique and different individuals. This type of twinning occurs about 70% of the time. All dizygotic twins have dichorionic placentation, and all dichorionic twins are diamniotic.

Monozygotic twins: result from one zygote that subsequently cleaves, producing two "identical" individuals. Monozygotic twins occur about 30% of the time, and have different variations of placentation and membranes.

Dichorionic / Diamniotic (DC/DA) – cleavage occurs in the morula stage, before differentiation of the blastocyst. Each blastocyst implants separately in the endometrium, although the sites of implantation may be close. If this happens, two discrete placentas may not be identified; indeed, they may abut and fuse, appearing as a single placental mass.

Monochorionic / Diamniotic (MC/DA) – 75% of monozygotic twins are MC/DA. Cleavage occurs sometime between the fourth and eighth days after fertilization, when the blastocyst has already formed but the amnion has not yet developed. There is one shared placenta, and almost all MC placentas have intertwin vascular connections.

Monochorionic / Monoamniotic (MC/MA) – rarely, cleavage occurs after the eighth day after fertilization. By this time the amnion has already formed, so the twins share not only a placenta but also an amniotic cavity. The embryonic disc is formed as well, and if the disc fails to divide, conjoined twins result.

Before beginning the discussion of sonographic features of these placental and membrane configurations, it should be mentioned that the only way to determine zygosity with certainty is if the fetuses are different gender. Dizygotic fetuses arguably have a 75% chance of being the same sex, so relying on gender is only valid when they differ. Monozygotic twins present as DC/DA close to 25% of the time, so chorionicity is not an indicator of zygosity.

Sonographic Features of Chorionicity and Amnionicity

First trimester sonography to determine chorionicity and amnionicity (prior to 14 weeks gestation) can render better than 95% accuracy! (Some studies report close to 100% accuracy.) In this timeframe, certain sonographic features are noted, as discussed below.

Dichorionic gestations:

- possibly two separate placental masses – if the implantation sites are not closely opposed, early identification of the decidua basalis and developing placental masses in different locations of the endometrium indicate two separate blastocysts have implanted. Both implantations may be close enough that individual placentae are not obvious (they are "fused"), and the other features need to be assessed for further determination.
- the "lambda" or "twin peak" sign – the membrane that separates the DC placenta contains fused chorionic leaves. Because chorion is contained in this membrane, it is possible for villi to grow into the junction of the membrane when the placental masses are fused. At the membrane base, a triangular region of tissue has a peaked appearance, hence the terms "lambda" or "twin peak." This feature, if present, is very reliable in early pregnancy, but it does not always persist later in gestation.
- the thickness of the intertwin membrane – a thick intertwin membrane is present in early DC gestations, because of the multiple layers of membranes (two layers of chorion and two layers of amnion). It should be noted that with increasing gestational age, membranes normally become thinner in appearance, so reliability on this feature alone is not advised.

Monochorionic gestations:

- a single placental mass – is unique to MC twins, but can be similar in appearance to two fused placentas. This is not a reliable feature on its own.
- absent twin peak sign – because the gestation is contained within one chorion, it is not possible for the twin peak sign to develop.
- a thin intertwin membrane – the membrane separating the twins consists of two layers of amnion only, and appears much thinner than in DC gestations. Early sonography will demonstrate two amnions inside of a single chorionic cavity, especially before the amnions fuse with the outer chorion.

Monoamniotic gestations:

- no visible membrane – because both embryos are within one amniontic cavity, no membrane will be identified. However, early amnions are normally not well demarcated, so relying on the absence of a membrane to determine monoamnionicity prior to 8 weeks is not appropriate.
- a single yolk sac – monoamniotic twins will share a single yolk sac in addition to sharing a placenta and amnion. The presence of only one yolk sac with two identifiable heart beats or embryos should prompt a follow-up sonogram after 9 weeks to assess amnionicity definitively.

In the **second and third trimesters**, definitive determination of chorionicity becomes increasingly difficult. Placental masses are more difficult to separate, the intertwin membrane normally thins, and certain pathologic conditions may make identification of the intertwin membrane even more challenging. While the twin peak sign may persist in the second trimester, non-visualization of this feature cannot exclude DC twinning. During these sonographic evaluations, the examiner must be diligent and patient, and possess knowledge of MC complications and the imaging features associated with them.

What to include on routine sonograms of twins

Use every tool and trick in your bag! Always attempt to document, in addition to routine fetal anatomy and growth assessment, the following:

- Placental location(s)
- Presence (or absence) of intertwin membrane and its thickness
- Amniotic fluid assessment – for both sacs, if there are two
- Fetal gender
- Fetal cord insertion sites into the placenta(s)

MONOCHORIONIC TWIN COMPLICATIONS

Placental vascular anastomoses

As stated earlier, almost all MC placentas demonstrate intertwin vascular connections on postpartum placental injection studies. These anastomoses are implicated in several syndromes of MC twins, and certainly can complicate the pregnancy. Three types of intertwin vascular connections can occur:

- Arterio-arterial (A-A) anastomoses: are frequent, direct, end-to-end connections on the surface of the placenta. They do not communicate with the parenchyma of the placenta, and are present in 75% of MC placentas, although there is seldom more than one connection per placenta. Doppler ultrasound is helpful in identifying the A-A connection; color Doppler locates the connection and spectral Doppler confirms the bidirectional pulsatile flow.

- Veno-venous anastomoses: are seen in approximately 5% of MC placentas, but are not detected sonographically. These direct end-to-end connections also course on the fetal surface of the placenta.

- Arteriovenous (AV) anastomoses: are common intertwin vascular connections and are seen in Twin-Twin Transfusion Syndrome (TTTS). It is a deep connection in the placenta, not a direct superficial connection on the fetal surface. The theory is that the absence of the superficial anastomoses (A-A and veno-venous), which maintain balanced blood flow, is the mechanism behind TTTS. Employing both color and spectral Doppler may help locate these AV anastomoses on the placental surface as well.

Unequal placental sharing

Fetal growth discordance is often caused by unequal placental parenchyma sharing, with one twin typically having a marginal or velamentous cord insertion site and a small parenchymal share while the larger twin has a more central cord insertion. The smaller twin, as it becomes growth restricted, will develop oligohydramnios, but this should not be confused with TTTS. Discordant twins should nonetheless be closely monitored.

Sonography is important for identifying cord insertion sites, for surveillance of fetal growth, and monitoring discordance if it develops. Therapeutic intervention is rarely indicated.

Twin-twin transfusion syndrome (TTTS)

Occurring in approximately 10% to 20% of MC twins, TTTS results from the vascular shunting between the circulations of twins who share a placenta. Blood is transfused from the donor, who becomes oligohydramniotic and growth restricted, to the recipient, who develops circulatory overload and resultant polyhydramnios. TTTS can present at any gestational age; earlier onset is often associated with poor outcome. If untreated, the prognosis is poor with a 60% to 100% mortality rate for both twins.

The sonographic determination of TTTS is difficult, and certain features must be identified in order to correctly make the diagnosis:

- Twins must be monochorionic / fetuses are of the same gender
- A single placenta is present
- Weight discordance exists
- Significant amniotic fluid discordance is present, often with one twin appearing "stuck"

Once these criteria are met, the evaluator can interrogate the surface of the placenta with color Doppler to locate connecting vessels. Spectral Doppler analysis will prove AV anastomoses. The absence of an A-A anastomosis is associated with a greater risk of developing TTTS. One study diagnosed TTTS in only 5% of cases in which an A-A anastomosis was found. Interestingly, MC/MA twins have predominantly A-A anastomoses, so TTTS is rare in this population of twins.

Twin embolization syndrome

A rare complication of MC pregnancies is twin embolization syndrome, which follows the in utero demise of one twin. It was theorized that thromboplastin-rich blood from the demised twin was transfused to the survivor via placental vascular anastomoses, or that clot or detritus from the dead twin embolized into the circulation of the surviving twin. More recently, researchers have postulated that injury to the surviving twin results in a sudden change of placental vascular territory perfused by the survivor. The impact of the co-twin demise and likelihood of injury to the survivor depends on the degree of placental sharing, the number and type of intertwin vascular connections, and the timing of the demise. It seems perinatal mortality is higher when superficial anastomoses (A-A or veno-venous) exist, and lower in cases of only AV anastomoses.

The prevalence of twin embolization syndrome, when one of a MC twin pair demises, is not firmly established. When it does occur, the prognosis is poor. Abnormalities of the surviving twin include brain lesions (i.e. hydranencephaly, porencephaly, cystic encephalomalacia, and hydrocephalus), renal necrosis, bowel and other organ infarcts.

It is likely that injury to the surviving twin is immediate, so by the time imaging happens, irreversible damage has occurred. Monitoring of these MC pregnancies may enable recognition of the characteristic structural defects of the survivor. MRI has been found to be useful in assessing brain injury, often not identifiable sonographically.

Acardiac parabiotic twin

This rare MC twin complication is characterized by the presence of one twin who, without its own functioning heart, continues to grow in utero. At the very least, an A-A and veno-venous

communication exists between the normal twin (the "pump" twin) and the acardiac twin, which circulates blood to and from the anomalous fetus. The large vascular connections are seen on the placental surface between the cord insertion sites. This circulatory system allows for blood to completely bypass the placental parenchyma and perfuse the abnormal twin with "used" blood from the normal twin. In the acardiac fetus, flow is reversed in the cord vessels, so that the umbilical vein carries blood away from the fetus and the umbilical artery flow is toward the fetus.

The most substantial risks for the normal pump twin are cardiac failure, polyhydramnios, hydrops, preterm labor and in utero demise. Sonographic factors cannot reliably predict which pregnancies are at highest risk for pump twin decompensation or demise, but it has been observed that the larger the anomalous acardiac twin, the more likely the pump twin will not survive.

Monoamniotic twins

Monochorionic monoamniotic twin pregnancies have the highest mortality rate of otherwise uncomplicated twin pregnancies, and for this reason sonographic diagnosis is exceedingly important. Because the amnion is difficult to visualize early in the first trimester, often the diagnosis is not usually possible until after 8 weeks. An adjunctive test for monoamniocity is performed using CT amniography: radiocontrast is injected into the amniotic cavity under ultrasound guidance, and if contrast is seen in the gastrointestinal tracts of both fetuses within hours of injection, monoamniocity is confirmed.

Lack of the separating membrane allows cord entanglement; demonstration of cord entanglement sonographically, rather than intertwined fetal extremities, is reliably diagnostic for MA twinning. True knots of the cord account for most of the increased mortality in MA twins, but it is unlikely that any form of monitoring will help predict an acute cord accident. However, knowledge of a MA condition will permit informed obstetric planning and cesarean delivery.

Conjoined twins

Conjoined twins develop from incomplete division of the embryonic disc. Prenatal diagnosis and characterization of the severity of the malformations is desirable for optimal obstetric management. Earlier diagnosis offers the option of termination via vaginal delivery, whereas later diagnosis will influence predictions of viability and decisions of delivery mode.

Sonographic diagnosis may be straightforward, but a careful approach is important to avoid misdiagnosis. A single placental is seen, and no separating intertwin membrane is identified. Findings include an inability to detect separate fetal skin contours, visualization of both heads persistently at the same level, no change in relative position of either fetus, bibreech or bicephalic presentation, backward flexion of the spine, a single umbilical cord with more than three vessels, and shared organs. Sonography is the most definitive method for diagnosis and characterization of conjoining, which predicts chances for postnatal survival.

Discordant anomalies

Chromosomal and anatomic abnormalities are more common in multiples than in singletons; lethal congenital defects are seen especially in monozygotic twins. Although rare, a major structural

anomaly may be detected in one of a MC pair, while the other co-twin is spared. Anomalies that have been seen include neural tube defects, diffuse lymphangiectasia, and diaphragmatic hernias. If the malformed fetus is likely to die in utero or cause difficulties during pregnancy or delivery, selective termination is considered. Thus correct determination of chorionicity is essential. DC twins are not typically at risk for injury, whereas a MC twin is at risk for injury at the time of co-twin demise (twin embolization syndrome).

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