

**American College of Radiology
ACR Appropriateness Criteria®
Multiple Gestations**

Variant 1: Known or suspected multiple gestations. Monochorionic or dichorionic. First trimester US.

Procedure	Appropriateness Category	Relative Radiation Level
US pregnant uterus transvaginal	Usually Appropriate	0
US pregnant uterus transabdominal	Usually Appropriate	0
US cervix transvaginal	Usually Not Appropriate	0
US duplex Doppler pelvis	Usually Not Appropriate	0
US assessment for TTTS	Usually Not Appropriate	0
US pregnant uterus biophysical profile	Usually Not Appropriate	0
US echocardiography fetal	Usually Not Appropriate	0

Variant 2: Multiple gestations. Dichorionic. Second trimester US. Anatomy scan.

Procedure	Appropriateness Category	Relative Radiation Level
US pregnant uterus transabdominal	Usually Appropriate	0
US cervix transvaginal	Usually Appropriate	0
US echocardiography fetal	May Be Appropriate	0
US duplex Doppler pelvis	Usually Not Appropriate	0
US pregnant uterus transvaginal	Usually Not Appropriate	0
US pregnant uterus biophysical profile	Usually Not Appropriate	0
US assessment for TTTS	Usually Not Appropriate	0

Variant 3: Multiple gestations. Monochorionic. Second trimester US. Anatomy scan.

Procedure	Appropriateness Category	Relative Radiation Level
US pregnant uterus transabdominal	Usually Appropriate	0
US assessment for TTTS	Usually Appropriate	0
US echocardiography fetal	Usually Appropriate	0
US cervix transvaginal	Usually Appropriate	0
US duplex Doppler pelvis	Usually Appropriate	0
US pregnant uterus biophysical profile	Usually Not Appropriate	0
US pregnant uterus transvaginal	Usually Not Appropriate	0

Variant 4: Multiple gestations. Dichorionic. Growth and antepartum surveillance.

Procedure	Appropriateness Category	Relative Radiation Level
US pregnant uterus transabdominal	Usually Appropriate	0
US pregnant uterus biophysical profile	Usually Appropriate	0
US cervix transvaginal	May Be Appropriate	0
US duplex Doppler pelvis	May Be Appropriate	0
US echocardiography fetal	Usually Not Appropriate	0
US pregnant uterus transvaginal	Usually Not Appropriate	0
US assessment for TTTS	Usually Not Appropriate	0

Variant 5: Multiple gestations. Monochorionic. Growth and antepartum surveillance.

Procedure	Appropriateness Category	Relative Radiation Level
US pregnant uterus transabdominal	Usually Appropriate	0
US pregnant uterus biophysical profile	Usually Appropriate	0
US duplex Doppler pelvis	Usually Appropriate	0
US assessment for TTTS	Usually Appropriate	0
US echocardiography fetal	May Be Appropriate	0
US cervix transvaginal	May Be Appropriate	0
US pregnant uterus transvaginal	Usually Not Appropriate	0

Variant 6: Multiple gestations. Known twin discordance. Monochorionic or dichorionic.

Procedure	Appropriateness Category	Relative Radiation Level
US pregnant uterus transabdominal	Usually Appropriate	0
US duplex Doppler pelvis	Usually Appropriate	0
US pregnant uterus biophysical profile	Usually Appropriate	0
US assessment for TTTS	Usually Appropriate	0
US cervix transvaginal	May Be Appropriate	0
US echocardiography fetal	May Be Appropriate	0
US pregnant uterus transvaginal	Usually Not Appropriate	0

MULTIPLE GESTATIONS

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Summary of Literature Review

Introduction/Background

Over the past 4 decades, the increased use of assisted reproductive techniques in the United States has been associated with a substantial rise in the rate of multiple births [1]. The rate of triplet and higher-order births has declined over the past decade in the context of a reduction in the transfer of three or more embryos during in vitro fertilization [1]. Multiple gestations are high risk compared with singleton pregnancies [2-6]. There is an approximate 5-fold increase in fetal death and 7-fold increase in neonatal death compared with singletons, which is primarily due to complications of prematurity [2]. The risk of preterm delivery and associated complications is proportional to the number of fetuses present. Growth restriction is also more common with multiple gestations. Multiple gestations are also at a higher risk for congenital anomalies, placenta previa, vasa previa, and velamentous insertion of the umbilical cord [4,7-13].

Twin pregnancies may be monozygotic or dizygotic [14]. Dizygotic twins (two-thirds of twin pregnancies) are always dichorionic, whereas monozygotic twins may be dichorionic-diamniotic, monochorionic-diamniotic, or monochorionic-monoamniotic depending on when the twins separated. Therefore, with rare exceptions, all monochorionic twins are also monozygotic. Monochorionic twins comprise 25% to 30% of twin pregnancies.

Most monochorionic twins are also diamniotic, with the twins sharing a single placenta. Unequal sharing of the placenta and vascular communications can result in various complications unique to monochorionic twins, including twin-to-twin transfusion syndrome (TTTS), twin embolization syndrome, and acardius, or twin-reversed arterial perfusion (TRAP) sequence [6,14]. Monochorionic diamniotic pregnancies have an overall mortality rate of about 10%, due largely to TTTS and fetal anomalies [15].

Monochorionic-monoamniotic placentation occurs in approximately 1% of all monozygotic twin pregnancies. The twins are in the same amniotic cavity, so entangled umbilical cords are typical and even a hallmark of monoamniotic twins [16]. These pregnancies are at further increased risk of fetal death. Earlier studies suggested mortality rates of 46% to 64%, but more recent studies have shown encouraging survival rates of greater than 90% with early diagnosis, serial ultrasound (US), and antenatal surveillance [17,18]. Most deaths in monoamniotic pregnancies are due to fetal malformations including conjoined twins, followed by TRAP sequence, TTTS, and preterm delivery or spontaneous miscarriage before 20 weeks gestation [16,17].

Overview of Imaging Modalities

Women with twin or higher-order pregnancies will typically have many more US examinations than women with a singleton pregnancy. The aim of each US varies with gestational age, and there is no accepted standard for the number of scans. However, the majority of women will have, as a minimum, a first trimester scan, a 12-week nuchal translucency (NT) scan, a fetal anatomy scan at 18 to 22 weeks, and one or more scans in the third trimester to evaluate growth.

First Trimester US

The role of US in the first trimester includes determination of chorionicity, pregnancy dating, and assessment of the NT. Ideally, dating is performed when the crown-rump length (CRL) measurement is between 45 to 84 mm at

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the time of the NT evaluation. Referral to a specialist is encouraged as early as the first trimester if there is a CRL discrepancy of $\geq 10\%$ or NT discordance is $\geq 20\%$. NT discordance $\geq 20\%$ is found in approximately 25% of monochorionic twins with an associated risk of severe TTTS or early intrauterine fetal demise (IUFD) up to 30% [19]. Intertwin discordance in CRL $\geq 10\%$ is significantly associated with pregnancy loss; however, the pooled predictive risk is only 52% [20,21].

Second Trimester Routine US

The role of US in the second trimester includes the anatomic survey, placental evaluation, and cervical length assessment. However, serial surveillance should be performed for pregnancies complicated by anomalies, cervical shortening, fetal growth disturbances, and amniotic fluid abnormalities [6].

- The **routine anatomic survey** occurs at the usual timing to evaluate for fetal anomalies, which are increased in twins. A major fetal anomaly affecting only 1 twin is present in around 1 in 25 dichorionic twins, 1 in 15 monochorionic-diamniotic, and 1 in 6 monoamniotic twin pregnancies [22,23]. It should be noted that fetuses can be assessed for the presence of any major anomalies at the first trimester scan. In monochorionic twins, there is an elevated risk of congenital cardiac disease; thus, cardiac screening assessment is recommended in this subgroup of twins.
- **Cervical Length US:** At the time of the routine anatomic survey, a cervical length assessment may be performed via transvaginal US in order to determine whether the patient should be triaged into a higher risk group for preterm delivery [24].
- **Placenta and Umbilical Cord Insertion:** Vasa previa and velamentous cord insertion are more commonly present in multiple gestations. Both of these conditions are associated with adverse pregnancy outcome and deserve a dedicated evaluation at this point in the pregnancy [25].

Serial Follow-up US and Third Trimester US

The role of third trimester scans is primarily to monitor fetal growth. However, ongoing surveillance may include fetal biometry, amniotic fluid assessment, and assessment for the development of TTTS, including twin anemia-polycythemia sequences (TAPS) and TRAPs, in addition to conventional evaluation of fetal well-being. Typically, surveillance begins at 16 weeks for monochorionic twins, with fetal biometry performed every 2 to 3 weeks and assessment for potential TTTS or other complications specific to monochorionic twins performed weekly. In contrast, a dichorionic pregnancy without complications is commonly followed every 3 to 4 weeks. When there is discordance in fetal size or amniotic fluid, regardless of chorionicity, closer surveillance may be warranted.

Selective intrauterine growth restriction (sIUGR), or selective fetal growth restriction, does not have a consistent definition amongst clinicians. A commonly used definition would be a condition in which one fetus has an estimated fetal weight (EFW) below the 10th percentile and the intertwin EFW discordance is $>25\%$. Some consider that a discordance of 20% is acceptable to triage the pregnancies at increased risk of adverse outcome. The formula for EFW discrepancy is $= (\text{EFW larger twin} - \text{EFW smaller twin}) / \text{EFW larger twin} \times 100$. It is important to consider other causes of fetal growth restriction such as viral infection or chromosomal abnormalities; nonetheless, the most common etiology would be related to unequal sharing of the placental mass and vasculature. Typically, charts used to monitor fetal growth restriction are the same in singletons and twins, but because there is a reduction in fetal growth in twins, particularly in the third trimester and even more so in the monochorionic group, close observation is warranted. When the umbilical artery Doppler waveform demonstrates intermittent or sustained absent or reversal of end-diastolic flow (EDF), there is a high risk of IUFD of the growth-restricted twin and potential for neurological morbidity in the surviving twin. If the pregnancy is dichorionic, sIUGR can be followed, similarly to its use for growth-restricted singleton fetuses. There is limited evidence to guide the management of monochorionic twins affected by sIUGR; however, a common follow-up strategy would be weekly assessment of fetal well-being to include Doppler of the umbilical artery and middle cerebral artery (MCA) with biweekly fetal biometry evaluations.

Monochorionic Twin Complications Assessment

Monochorionic twin pregnancies are, by definition, considered high-risk pregnancies with specific complications such as TTTS, TAPS, TRAP, monoamniotic pregnancy, and conjoined twinning. Approximately one-third of twin pregnancies are monochorionic. Virtually all of these contain a degree of vascular anastomoses connecting the two placental circulations [25]. The most commonly utilized classification system for TTTS is Quintero staging

[26], despite its acknowledged criticism that the staging may not always represent a chronological order of deterioration. Monitoring of monochorionic twins for TTTS begins at 16 weeks' gestation with subsequent biweekly scans. Features to evaluate at each US include biometry, the presence of discordant bladder size, and amniotic fluid volumes. From 20 weeks and onward, umbilical artery Doppler and MCA peak systolic velocity (PSV) should be obtained. TAPS occurs spontaneously in approximately 5% of monochorionic-diamniotic twins but may be as high as 13% post laser ablation. It is hypothesized to be related to microanastomoses resulting in a chronic form of TTTS. TRAP sequence is a rare complication of monochorionic twin pregnancies. The chance of survival of the pump twin is increased by techniques such as cord ligation or ablation techniques, preferably before 16 weeks' gestation. A common complication of monochorionic-monoamniotic twin pregnancies is cord entanglement. The presence of cord entanglement does not appear to contribute to morbidity and mortality; however, preterm delivery and premature rupture of membranes are more common than in monochorionic diamniotic pregnancies.

Fetal Echocardiography

Screening for congenital heart disease is warranted in all monochorionic twins as the risk of cardiac anomalies has been reported to be 2% in otherwise uncomplicated monochorionic twins and 5% in cases of TTTS, particularly among recipient twins [27-29]. Although controversial, there are some data to suggest that fetuses conceived by in vitro fertilization have a higher rate of congenital heart disease, in particular monochorionic twins [30]. The presence of TTTS increases the risk for congenital cardiac disease in monochorionic twins, thus development of TTTS may be an indication for fetal echocardiography in later gestation [29] if not performed previously or for functional cardiac assessment after development of TTTS [31]. TTTS occurs in 10% to 20% of monochorionic-monoamniotic twins. In these cases, the recipient twin has been reported to demonstrate cardiac functional abnormalities, and in recent studies structural abnormalities leading to right ventricular outflow obstruction may develop in later gestation in 3% to 10%, either before or after laser coagulation therapy of TTTS [32]. Recent data suggest that the right ventricular outflow obstruction may also develop in the donor twin and in monochorionic twins affected by selective intrauterine growth retardation. Selective IUGR or twin discordance complicates approximately 10% of all monochorionic twin pregnancies. These potentially high-risk groups may require surgery or catheter intervention in the newborn period.

Discussion of Procedures by Variant

The variants discussed are presented in approximate order of gestational age. This discussion is almost entirely focused on twin pregnancies because twins represent 98% of multiple gestations and the vast majority of data relate to twin pregnancies. It is recognized that triplets and higher-order pregnancies are at an even higher risk [33].

Variant 1: Known or suspected multiple gestations. Monochorionic or dichorionic. First trimester US.

Multiple gestations are usually first detected in the first trimester because of the widespread use of US, both for symptomatic and asymptomatic patients. First trimester NT screening at approximately 11 to 14 weeks has now been incorporated into most practice guidelines, so that twin pregnancies are usually diagnosed by this time, if not before. Caution is warranted in establishing viability of a twin during early pregnancy because the demise of one of the twins is relatively common, resulting in the so-called "vanishing twin."

Chorionicity and amnionicity should be determined as early as possible when a twin pregnancy is identified [7-11,13,34-36]. Determination of chorionicity is most accurate in the first trimester because the number of gestational sacs equals the number of chorions, with a reported accuracy of nearly 100% [37]. When there is a single gestational sac, evaluation of the amniotic sacs is also helpful because separate and distinct amnions should be visible for diamniotic pregnancies. However, because the amniotic membranes are thin and delicate, it is important to search diligently via transvaginal US for the presence of a membrane given the significant outcome differences between a monochorionic-diamniotic twin and monochorionic-monoamniotic twin pregnancy. The intertwin membrane is typically identified by 10 weeks on transvaginal US. The absence of identification of the intertwin membrane can be technical; thus, it is important to confirm the absence either by demonstrating umbilical cord entanglement (using color or pulsed wave Doppler to identify two different heart rates), or by short-term serial US. A single amniotic cavity containing two living embryos indicates a monochorionic-monoamniotic gestation. Although it has been suggested that a monoamniotic twin pregnancy has a single yolk sac [38], this may not always be the case [39,40], and determining the number of yolk sacs is not an absolutely

accurate indicator of amnionicity. It is encouraged to refer to a tertiary center for a monochorionic-monoamniotic twin pregnancy.

After 10 weeks, other features that may be helpful for determining chorionicity include number of placentas, the lambda or twin peak sign as seen in dichorionic gestations as opposed to the “T” sign as seen in monochorionic gestations, and, to a lesser degree, the dividing membrane thickness [37,41-44]. At the time of the 11 to 14 week scan, chorionicity was correctly assigned by US in 612 of 613 pregnancies, for an accuracy of 99.8% [41]. It is important to use a combination of features to accurately determine chorionicity rather than a single feature to ensure accuracy. If it is not possible to determine chorionicity on a transabdominal scan, a transvaginal scan should be performed. If it is still not possible, then either re-examination within a short time period or referral to a tertiary center may be appropriate.

By the second trimester, there may be thinning of membranes, loss of the lambda peak sign, and fusion of the placentas, thus, absolute confirmation of a dichorionic twin pregnancy may require confirmation of discordant gender (one male and one female) to confirm a dizygotic gestation. As up to 55% of twins are same gender, the assignment of chorionicity in first trimester when other signs are reliably present is crucial to make this important distinction.

Twin embryos in the first trimester are usually similar in size. When there is disparity in size early in the pregnancy, most authorities suggest using the larger twin for dating purposes to minimize the chance of missing a fetus that might present with growth restriction [5]. However, others have found that the smaller twin more closely reflects the true gestational age when using the charts of Robinson [45]. A significant discrepancy in embryo size increases the risk of underlying growth restriction, aneuploidy or congenital anomalies, and subsequent demise. Bora et al [34] found that regardless of chorionicity, there was a correlation between subsequent embryonic demise and size discrepancy between 7 weeks and 9 weeks 6 days. The likelihood of subsequent demise was 3% if the discrepancy was <20%, whereas it was 100% if the discrepancy was >60% [34]. Others have found that CRL discordance in the first trimester poorly predicted demise before 24 weeks [46], but this reflected less severe degrees of discrepancy.

At 11 to 14 weeks, significant discrepancy in fetal size has also been associated with aneuploidy or other anomalies and growth restriction [47,48]. Weissman-Brenner et al [48] reported that discordant CRL at the time of a NT scan at about 12 weeks could identify 5 of 21 pregnancies with birth weight discordance of more than 25%. Memmo et al [49] found that CRL discordancy could identify fetuses at risk for subsequent growth restriction but not TTTS.

Nuchal Translucency Scan and Aneuploidy Screening

NT screening at approximately 11 to 14 weeks is now widely accepted and can be performed for multiple gestations as well as singletons [50-55]. This subject is addressed in a separate ACR Appropriateness Criteria document, and a detailed discussion is beyond the scope of this article. The relative importance of the NT measurement in the first trimester increases in multiple pregnancies as the biochemistry is less useful since it is not possible to accurately assess the contribution of each fetus, and levels from the normal twin can mask abnormal levels in the affected twin.

Similar to singleton pregnancies, increased NT increases the risk for aneuploidy and other birth defects, and markedly increased NT also increases the risk of subsequent demise [50,52,53]. Among monochorionic twins, markedly discordant NT also can be a marker for early-onset TTTS [56-58]. Nonetheless, normal fetal anatomy and karyotype were the most common outcomes among monochorionic diamniotic twins with discordant NT [59]. Allaf et al [60] also found that NT and CRL discordances were not predictive of overall adverse outcomes in monochorionic diamniotic twin pregnancies, although this varies with the severity of discordancy.

Variant 2: Multiple gestations. Dichorionic. Second trimester US. Anatomy scan.

A fetal anatomy scan should be performed at 18 to 22 weeks for all pregnancies with the primary aim to screen for birth defects [2,61]. Congenital anomalies are more common in twin pregnancies, but this appears almost entirely due to the increased risk among monozygotic twins [62], which is estimated to be 2 to 3 times greater than singletons.

At the time of the fetal anatomy scan, it is important to evaluate the placenta, umbilical cords, and cervix. Placenta previa is more common in twin pregnancies, especially dichorionic twins, as one would expect due to greater placental surface area [63]. Marginal and velamentous cord insertion are more common in twin

pregnancies with the frequency of velamentous cord insertion also resulting in a higher frequency of vasa previa. The antenatal knowledge of adverse outcome predictors such as velamentous cord insertion of vasa previa may be useful in risk stratification and management of twin pregnancies. At the time of the routine anatomic survey, a cervical length assessment may be performed via transvaginal US to determine whether the patient should be triaged into a higher risk group for preterm delivery.

Variant 3: Multiple gestations. Monochorionic. Second trimester US. Anatomy scan.

Similar to dichorionic twin pregnancies, monochorionic twins should be scanned at 18 to 22 weeks for fetal anatomy. The risk of congenital anomalies appears to be higher for monozygotic twins that separate later, with conjoined twins representing the most extreme example. Also, the risk for at least one of a monochorionic-monoamniotic twin pair having a structural congenital cardiac anomaly is eight times that of a monochorionic-diamniotic twin pair. In addition, if a monochorionic twin is affected, the risk of the co-twin having a cardiac anomaly is higher [64]. For these reasons, fetal echocardiography should be considered in monochorionic gestations, especially in monochorionic-monoamniotic twins, as well as in dichorionic twin pregnancies conceived using assisted reproductive technologies based on the increased risk of congenital heart disease in these groups [65].

Although monochorionic twins are also monozygotic, monochorionic twins can be discordant for fetal anomalies and even karyotypic abnormalities, with the latter usually explained by mosaicism. The presence of a fetal anomaly increases the risk of the other normal twin for preterm delivery, low birth weight, and perinatal mortality [66].

The placenta, umbilical cords, and cervix should be evaluated at the time of the anatomy scan to assess for placenta previa and marginal or velamentous cord insertion. The latter two are more common among monochorionic pregnancies. Velamentous cord insertion may be seen in 22% of monochorionic twin pregnancies, but has not been shown to be associated with TTTS [67]. Nonetheless, velamentous cord insertion in monochorionic twins increases the risk of adverse outcome, including small for gestational age and sIUGR, lower gestational age at birth, and IUFD [68]. There is also a higher frequency of vasa previa when a velamentous cord insertion is found, which, if overlooked, will result in acute fetal hemorrhage, distress, and even death at the time of delivery. For this reason, sonographers should be aware of the possibility of vasa previa, especially in monochorionic pregnancies [69].

A baseline cervical length assessment can be performed using transvaginal US. This will help determine whether patients should be triaged into a higher risk group for preterm delivery.

Variant 4: Multiple gestations. Dichorionic. Growth and antepartum surveillance.

The most effective fetal surveillance system for multiple gestations is still not established. In current practice, the frequency of US evaluation in otherwise uncomplicated twin pregnancies is influenced primarily by chorionicity and growth pattern. Finberg et al [70] suggested follow-up scans every 4 to 6 weeks for dichorionic twins. Current trends in expert opinion appear to favor even closer surveillance with dichorionic twins followed every 3 to 4 weeks [5,71]. Certainly, closer follow-up is warranted when there is significant discordance in fetal size or amniotic fluid, regardless of chorionicity [72]. The risk of fetal demise is also low after 32 weeks among uncomplicated twins, even among monochorionic pregnancies [15]. At each US scan, the following should be assessed: fetal biometry, amniotic fluid volume, and umbilical artery Doppler after 20 weeks onward for both twins. The EFW discrepancy discordance should be calculated and documented at each scan from 20 weeks onward. To date, there is insufficient data in the literature to suggest that antenatal surveillance of twins with biophysical profile (BPP) is beneficial in the setting of a reactive nonstress test or in the absence of associated risk factors [73].

Variant 5: Multiple gestations. Monochorionic. Growth and antepartum surveillance.

Similar to dichorionic twins, the most effective follow-up evaluation of monochorionic twins is still not well-established. Finberg et al [70] suggested follow-up scans every 3 to 4 weeks for monochorionic twins with current trends in expert opinion appearing to favor even closer surveillance every 2 to 3 weeks beginning at 16 weeks [5,71]. Some clinicians monitor monochorionic twins every 2 weeks or even more frequently [5].

Monochorionic twins are at risk of complications related to vascular communications between the fetuses because of a common placenta. These include sIUGR, TTTS, TAPS, TRAP sequence, and IUFD. Selective IUGR due to discordant twin growth occurs in up to 25% of monochorionic pregnancies [74]. Although there is no real

consensus on what constitutes sIUGR, most agree that using an EFW of less than the 10th percentile, an EFW discordance of >25% between the twins, or a discordant fetal abdominal circumference of >10% would be acceptable to make the diagnosis [75]. The growth restriction can occur at any time during the pregnancy, and correlation with Doppler studies of the umbilical cord artery can help predict the outcome. Selective IUGR has been classified into three types based on Doppler findings in the growth restricted twin: type 1 shows constant EDF in the umbilical artery, type 2 shows constant absent or reversed EDF, and type 3 shows intermittent absent or reversed EDF. In a prospective study evaluating the perinatal outcome of monochorionic twins with sIUGR, Weisz et al [76] compared restricted twins with abnormal Doppler findings to those with normal Doppler findings. The authors found an overall higher incidence of neonatal complications (sepsis, central nervous system abnormalities, respiratory distress, and neonatal death) in sIUGR twins with absent or reversal EDF in the umbilical artery. Ishii et al [77] found that the additional finding of severe oligohydramnios or “stuck twin” phenomenon was a significant predictor of mortality in the growth-restricted twin with abnormal Doppler waveforms.

Probably all monochorionic twins have a mild degree of unequal sharing, but clinically significant TTTS affects 10% to 20% of monochorionic twins. The most severe cases are evident before 20 weeks, whereas milder degrees may not become apparent until 26 to 28 weeks. Untreated severe TTTS in the mid second trimester carries a very poor prognosis with mortality rate in excess of 70% [78].

Clinically significant cases are usually apparent by 20 weeks with polyhydramnios and a large urinary bladder in the recipient twin and oligohydramnios and a small urinary bladder in the donor twin [26]. Discordance in fetal size may be subtle on early scans. A pathognomonic sign for the diagnosis of TTTS is the appearance of the donor as the stuck twin, contained within the collapsed intertwin membrane because of anhydramnios. Severity is according to the Quintero classification, which consists of five stages with stage 1 of oligo-polyhydramnios sequence having the best outcome and stage 5 having the worst outcome of one or both twin demise [26]. Doppler studies may show absence or reversal of EDF in the umbilical cord artery of the donor, decreased ventricular function seen as tricuspid regurgitation or reversal of A wave in ductus venosus. Cardiac chamber enlargement in the recipient can be seen in more advanced stages of TTTS [26].

TAPS is an atypical form of TTTS characterized by significant intertwin hemoglobin differences but in the absence of oligohydramnios and polyhydramnios [79,80]. This condition may develop spontaneously in up to 5% of monochorionic twins or after incomplete laser treatment of TTTS in 10% of cases [6]. Because of the relatively low prevalence and lack of clinical awareness, the natural history is unclear and the antenatal treatment remains uncertain [79]. This condition can be monitored by assessing PSV of the MCA, with fetal anemia showing as accelerated velocity. The diagnosis can be suggested when the PSV of the MCA is >1.5 multiples of the median for the donor twin and <1 for the recipient, and the severity can also be graded by more discordant Doppler values [81].

TRAP sequence is a rare condition, occurring in approximately 1 in 30,000 pregnancies. It results from a parasitic arrangement in which a fetus with absent or nonfunctional cardiac function of its own (acardiac twin) receives systemic arterial supply through arterial-arterial anastomosis by the donor twin (pump twin). The acardiac twin grows, but is markedly anomalous, often lacking a head, upper extremities, and a trunk, and is usually edematous. The mortality rate of the donor twin is high (approximately 50%) due to cardiac overload. Fetal echocardiography should be performed in the pump twin to monitor its cardiac function as well as to look for congenital heart disease, which can be seen in up to 10% of cases. Treatment is based on interruption of the communicating vessels or the umbilical cord of the anomalous twin [82,83]. Fetoscopic laser coagulation of placental vascular anastomoses or the umbilical cord of the acardiac twin after 16 weeks is an effective treatment. Hecher et al [82] reported a survival rate of 80%, with 67% of surviving pump twins delivering at 36 weeks without other complications for patients treated by this method at a median of 18.3 weeks. However, because of the risk of spontaneous cessation of flow in the acardiac twin before planned intervention at 16 to 18 weeks with subsequent brain injury in the majority of survivors, Chaveeva et al [84] suggest that optimal outcome may be earlier elective intervention at 12 to 14 weeks.

Demise of one fetus occurs in up to 5% of twin pregnancies during the second and third trimesters [85]. A single fetal death is 3- to 4-fold more likely in monochorionic twins than in dichorionic twins [86]. It is also more common in higher-order multiples, complicating 14% to 17% of triplet pregnancies. In general, the prognosis of the surviving twin is excellent when co-twin demise occurs early in pregnancy. However, some studies have found a higher frequency of complications compared to singletons, including gestational diabetes, growth

restriction, low birth weight, and perinatal mortality [87,88] with an overall 50% to 80% of surviving twins being born preterm. Survivors of a monochorionic pregnancy have significant additional risks because of the vascular communications, as well as a 10% to 30% risk of developing neurologic injuries due to ischemic events [86]. In addition, death of a monochorionic twin may result in fetal demise of its co-twin in 10% of cases.

Variant 6: Multiple gestations. Known twin discordance. Monochorionic or dichorionic.

Studies have shown an association with increased mortality and morbidity when there are significant differences in birth weights between the twins [89,90]. Detection of growth restriction is important and relies on EFW percentile or measurement of the abdominal circumference and comparison to the expected for gestational age [91-98]. Significant discordancy in EFW is the most widely accepted method to determine differences in twin size, and the most commonly used threshold when estimated weights are discordant by 20% or more [5]. Some authors suggest that discordance should be defined as mild if weight estimates for the twins are 15% different, moderate if 20% different, and severe if 25% different or greater.

In a group of 300 monochorionic diamniotic pregnancies followed every 2 weeks from the first trimester, isolated twin weight discordance of 25% or greater was observed in 11.6% of cases [15]. Discordant growth may be predicted by earlier scans in the first or second trimesters. One multi-institutional study found that discordance in the abdominal circumference by more than 10% between 14 and 22 weeks was the single best predictor of subsequent adverse outcome for both monochorionic and dichorionic pregnancies [99]. In addition to evaluation of growth, amniotic fluid is important to assess. As mentioned previously, oligohydramnios in one sac may be a sign of TTTS, but more commonly indicates uteroplacental insufficiency or leakage of amniotic fluid [100].

Other tests for evaluating fetal well-being include nonstress test, Doppler velocimetry of the umbilical artery and ductus venosus, and BPP or modified BPP. A lengthy discussion on assessment of fetal well-being has already been the subject of a previously reviewed in the ACR Appropriateness Criteria “[Assessment of Fetal Well-Being](#)” [101]. At present, antepartum fetal testing in multiple gestations is recommended in all situations in which surveillance would ordinarily be performed in a singleton pregnancy (including suspected growth restriction). In the practice bulletin of the American College of Obstetricians and Gynecologists, the recommendation based on consensus and expert opinion was that the management of discordant growth restriction or death of one fetus in a high-order multiple gestation should be individualized, taking into consideration the welfare of the other fetuses [2].

Although there is no proven benefit of umbilical artery Doppler evaluation in uncomplicated twins, it has been shown to be helpful when growth delay is suspected and in monochorionic twins. Abnormal Doppler findings are usually seen in the third trimester, but can be detected earlier in the pregnancy at 16 to 20 weeks gestation and, not surprisingly, have been shown to be associated with an increased risk of adverse outcome and fetal demise [102]. Surveillance with a nonstress test or BPP for pregnancies complicated by abnormal fluid volumes, pregnancy-induced hypertension, fetal anomalies, growth abnormalities, monoamnicity, or other standard obstetric indications is as reliable in multiple gestations as in singleton gestations [103].

Summary of Recommendations

- Transabdominal and transvaginal US are recommended in the first trimester when a twin pregnancy is known or suspected. Chorionicity and amnionicity are most accurately evaluated in the first trimester.
- Transabdominal US is recommended for dichorionic twins when evaluating fetal anatomy. Transvaginal US of the cervix may help triage patients into higher risk group for preterm delivery. Fetal echocardiography may be useful in some instances, such as when twins are conceived through in vitro fertilization.
- Transabdominal US is performed in monochorionic twins for fetal anatomy and to screen for fetal anomalies and TTTS. Fetal echocardiography helps screen for structural congenital cardiac anomalies. Transvaginal US of the cervix may help triage patients into higher risk group for preterm delivery. Duplex Doppler velocimetry is recommended in cases of TTTS, velamentous cord insertion, and sIUGR.
- Transabdominal US is recommended for growth and antepartum surveillance for dichorionic twins with duplex Doppler velocimetry used in cases of growth discrepancy.
- Transabdominal US is recommended for growth and antepartum surveillance for monochorionic twins. Duplex Doppler velocimetry and BPP monitoring are helpful in cases of IUGR, TTTS, TAPS, TRAP sequence, and IUFD. Fetal echocardiography should be performed to look for congenital cardiac disease and monitor cardiac function.

- Transabdominal US, duplex Doppler velocimetry, and BPP monitoring are recommended for follow-up of known twin discrepancy. Fetal echocardiography is helpful in monochorionic-monoamniotic twins.

Summary of Evidence

Of the 108 references cited in the *ACR Appropriateness Criteria® Multiple Gestations* document, 2 are categorized as therapeutic references including 1 good-quality study. Additionally, 105 references are categorized as diagnostic references including 2 good-quality studies, and 43 quality studies that may have design limitations. There are 61 references that may not be useful as primary evidence. There is 1 reference that is a meta-analysis study.

The 108 references cited in the *ACR Appropriateness Criteria® Multiple Gestations* document were published from 1986-2016.

While there are references that report on studies with design limitations, 3 good-quality studies provide good evidence.

Safety Considerations in Pregnant Patients

Imaging of the pregnant patient can be challenging, particularly with respect to minimizing radiation exposure and risk. For further information and guidance, see the following ACR documents:

- [ACR-SPR Practice Parameter for Imaging Pregnant or Potentially Pregnant Adolescents and Women with Ionizing Radiation](#) [104]
- [ACR-ACOG-AIUM-SRU Practice Parameter for the Performance of Obstetrical Ultrasound](#) [105]
- [ACR Guidance Document on MR Safe Practices](#) [106]
- [ACR Manual on Contrast Media](#) [107]

Appropriateness Category Names and Definitions

Appropriateness Category Name	Appropriateness Rating	Appropriateness Category Definition
Usually Appropriate	7, 8, or 9	The imaging procedure or treatment is indicated in the specified clinical scenarios at a favorable risk-benefit ratio for patients.
May Be Appropriate	4, 5, or 6	The imaging procedure or treatment may be indicated in the specified clinical scenarios as an alternative to imaging procedures or treatments with a more favorable risk-benefit ratio, or the risk-benefit ratio for patients is equivocal.
May Be Appropriate (Disagreement)	5	The individual ratings are too dispersed from the panel median. The different label provides transparency regarding the panel’s recommendation. “May be appropriate” is the rating category and a rating of 5 is assigned.
Usually Not Appropriate	1, 2, or 3	The imaging procedure or treatment is unlikely to be indicated in the specified clinical scenarios, or the risk-benefit ratio for patients is likely to be unfavorable.

Relative Radiation Level Information

Potential adverse health effects associated with radiation exposure are an important factor to consider when selecting the appropriate imaging procedure. Because there is a wide range of radiation exposures associated with different diagnostic procedures, a relative radiation level (RRL) indication has been included for each imaging examination. The RRLs are based on effective dose, which is a radiation dose quantity that is used to estimate population total radiation risk associated with an imaging procedure. Patients in the pediatric age group are at inherently higher risk from exposure, both because of organ sensitivity and longer life expectancy (relevant to the long latency that appears to accompany radiation exposure). For these reasons, the RRL dose estimate ranges for pediatric examinations are lower as compared to those specified for adults (see Table below). Additional

information regarding radiation dose assessment for imaging examinations can be found in the ACR Appropriateness Criteria® [Radiation Dose Assessment Introduction](#) document [108].

Relative Radiation Level Designations		
Relative Radiation Level*	Adult Effective Dose Estimate Range	Pediatric Effective Dose Estimate Range
○	0 mSv	0 mSv
⊕	<0.1 mSv	<0.03 mSv
⊕⊕	0.1-1 mSv	0.03-0.3 mSv
⊕⊕⊕	1-10 mSv	0.3-3 mSv
⊕⊕⊕⊕	10-30 mSv	3-10 mSv
⊕⊕⊕⊕⊕	30-100 mSv	10-30 mSv

*RRL assignments for some of the examinations cannot be made, because the actual patient doses in these procedures vary as a function of a number of factors (eg, region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as “Varies”.

Supporting Documents

For additional information on the Appropriateness Criteria methodology and other supporting documents go to www.acr.org/ac.

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The ACR Committee on Appropriateness Criteria and its expert panels have developed criteria for determining appropriate imaging examinations for diagnosis and treatment of specified medical condition(s). These criteria are intended to guide radiologists, radiation oncologists and referring physicians in making decisions regarding radiologic imaging and treatment. Generally, the complexity and severity of a patient's clinical condition should dictate the selection of appropriate imaging procedures or treatments. Only those examinations generally used for evaluation of the patient's condition are ranked. Other imaging studies necessary to evaluate other co-existent diseases or other medical consequences of this condition are not considered in this document. The availability of equipment or personnel may influence the selection of appropriate imaging procedures or treatments. Imaging techniques classified as investigational by the FDA have not been considered in developing these criteria; however, study of new equipment and applications should be encouraged. The ultimate decision regarding the appropriateness of any specific radiologic examination or treatment must be made by the referring physician and radiologist in light of all the circumstances presented in an individual examination.