Monochorionic Twin Pregnancies

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This guideline has been updated to change the recommendation for diagnosis and management of selective Fetal Growth Restriction (sFGR), isolated fetal growth discordance, and TAPS.

Highlights

- Twin-twin transfusion syndrome (TTTS) occurs in 8-10% of MCDA twin pregnancies. Ultrasound screening should occur every 2 weeks starting at 16 weeks. Patients with findings concerning for TTTS should be referred to the TCH Fetal Center for evaluation.
- **BCM OB/Gyn Perinatal Guidelines Committee recommends using the following definition for selective FGR (sFGR): A condition in which one fetus has EFW < 10th centile or AC < 10th centile.**
- Fetal weight discordance ≥ 25th centile without growth restriction also is associated with adverse perinatal outcomes and warrants closer surveillance.
- Twin Anemia Polycythemia Sequence (TAPS) more often occurs following laser photocoagulation of the placenta for TTTS. TAPS is diagnosed when the Middle Cerebral Artery (MCA)-Peak Systolic Velocity (PSV) is >1.5 Multiples of the Median (MoM) in the anemic fetus and <1 MoM in the polycythemic fetus or delta MCA-PSV >0.5 MoM. Fetal Center referral is recommended if MCA-PSV> 1.7 MoM and <0.8 MoM in the anemic and polycythemic fetuses, respectively, or delta MCA-PSV is >1.

Background

The general prevalence of twin-twin transfusion syndrome (TTTS) is 1-3 per 10,000 births.[1] Approximately 8-10% of monochorionic, diamniotic (MCDA) twin pregnancies are complicated by TTTS.[2,3] The prenatal diagnosis, management, and treatment of TTTS have been previously described.[4-6] This guideline is based on recommendations from the Society for Maternal-Fetal Medicine[6] and on expertise within our own Baylor OBGYN Department.

General Principles

Optimal prenatal assessment of MC twins begins with a precise diagnosis of placental chorionicity. Identification of well-dated MC twins is best accomplished during the 11-14 week scan when direct binding between two thin amniotic membranes can usually be visualized (i.e. "t-sign").[7,8] The management goals for TTTS can differ:

15-28 weeks: early detection and/or treatment of serious complications mainly amenable to fetal therapy [TTTS, twin anemia polycythemia sequence (TAPS) and selective fetal growth restriction (sFGR).

29-36 weeks: early detection of complications mainly (not exclusively) amenable to early delivery and/or elective surgery that includes: 1) late onset TTTS; 2) late sFGR; and 3) TAPS.

Surveillance and Management of TTTS

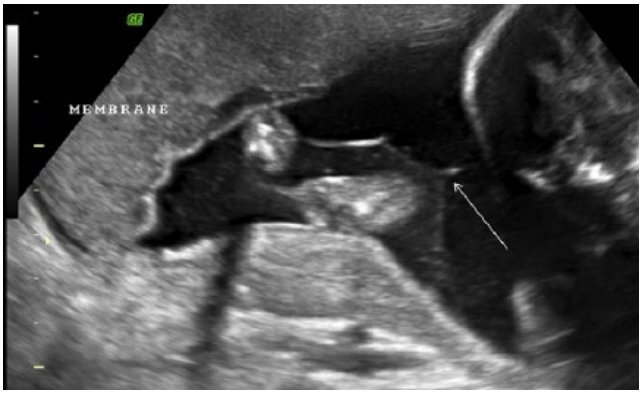
SMFM recommends ultrasound surveillance at 2-week intervals starting from 16-weeks until delivery to identify early occurring TTTS that may develop days following a normal scan. The basic pathophysiology is caused by abnormal placental vascular connections between fetuses. Placental blood flow imbalances lead to volume depletion in the donor twin (oliguria and decreased amniotic fluid) and volume overload in the recipient twin (polyuria and hydramnios).

Sonographic Findings Associated with TTTS[9]

First Trimester Findings:
- Crown-rump length discordance
- Nuchal translucency > 95th percentile or discordance > 20% between twins
- Reversal or absence of ductus venosus A-wave

Second Trimester Findings:
- Abdominal circumference discordance
- Membrane folding
- Velamentous placental cord insertion (donor twin)
- Placental echogenicity (donor portion hyperechoic)
- 20% EFW discordance

Some TTTS predictors have included ductus venosus Doppler, nuchal translucency, membrane folding, and abdominal circumference discordance although the sensitivity of these signs is not extremely high.[5,10,11]

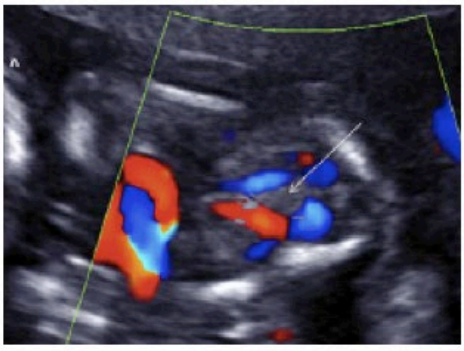


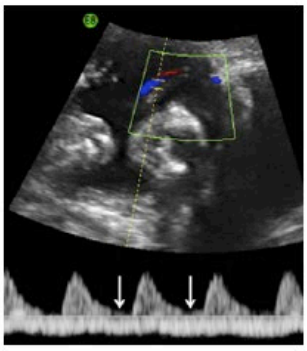
Membrane Folding Example

TTTS Staging

The Quintero classification system[8] is the most widely used for establishing fetal prognosis for TTTS on the basis of severity and provides a standardized method to compare outcomes using different therapeutic approaches.

Stage	Ultrasound Parameter	Categorical Criteria
I	MVP of amniotic fluid	MVP < 2 cm in donor sac; MVP > 8 cm in recipient
II	Fetal bladder	Non-visualized fetal bladder in donor twin over 60 minutes of observation
III	Doppler waveforms umbilical artery/vein, ductus venosus	Absent or reversed umbilical artery diastolic flow Reversed ductus venosus a-wave flow Pulsatile umbilical vein flow
IV	Fetal hydrops	Hydrops in one or both twins
V	Absent fetal cardiac activity	Fetal demise in one or both twins


Stage II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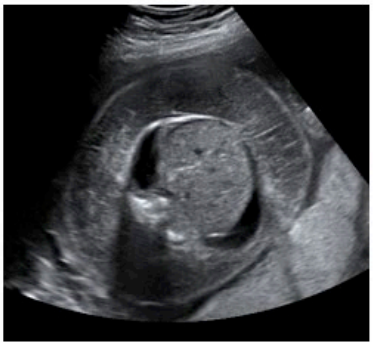
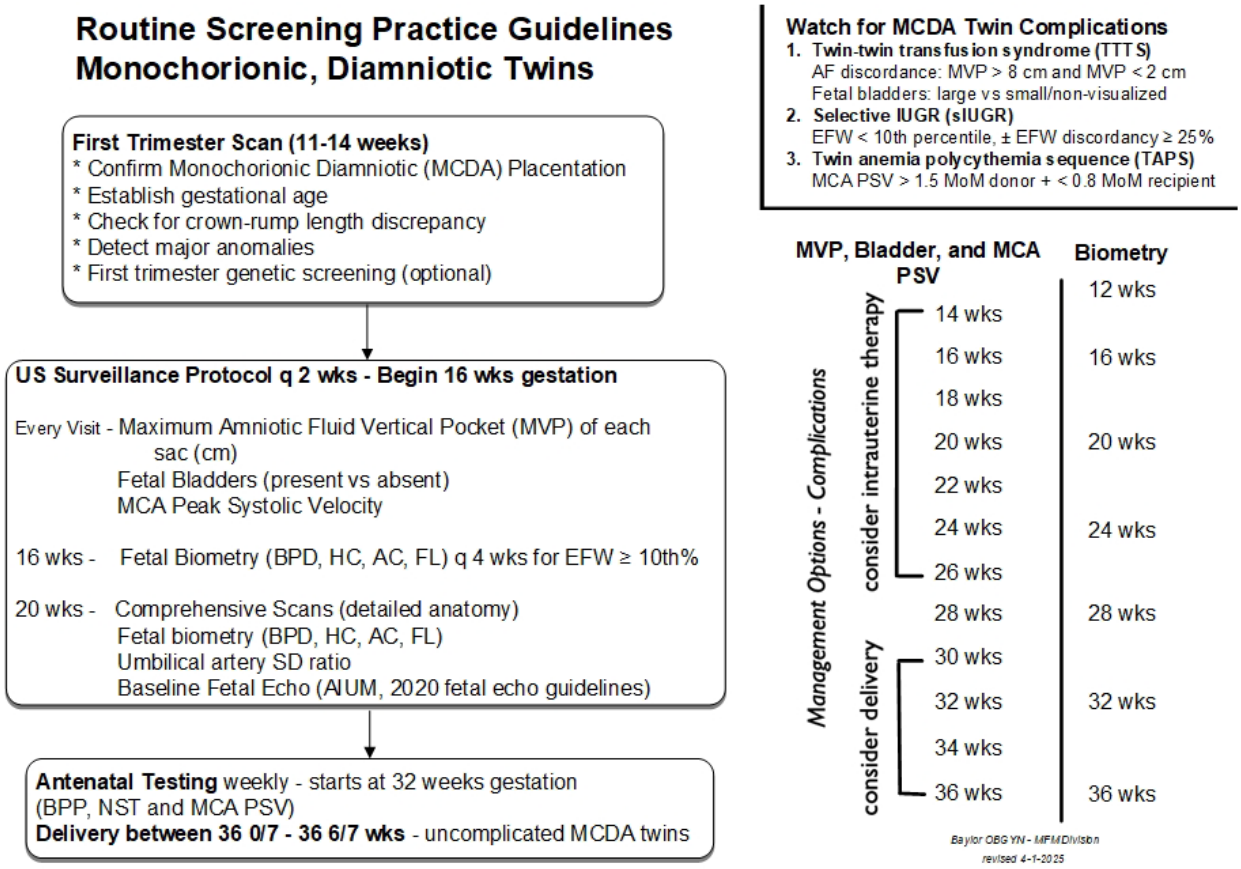

Stage IV

Ultrasound monitoring

Pregnancies not yet diagnosed with TTTS

Two different types of ultrasound scans are commonly used to monitor MCDA pregnancies for the potential development of TTTS. See Figure 1 for management algorithm for TTTS Screening

Figure 1. MCDA Screening Guidelines



Pregnancies affected by TTTS s/p laser ablation

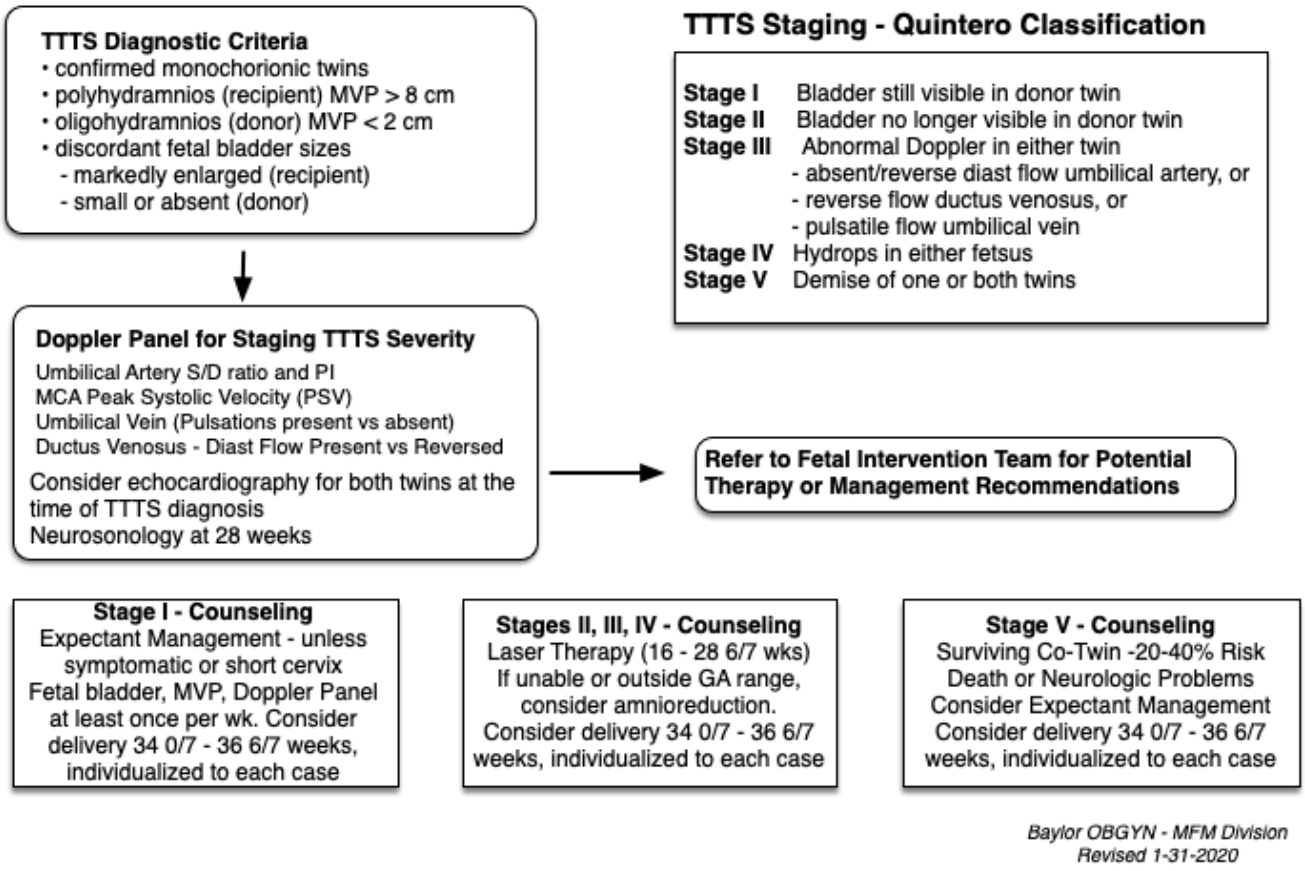
Postoperative follow-up scans should include **weekly** scans for 6 weeks postop before resuming at 2-week intervals provided there are no other concerns.

- ☐ Amniotic fluid assessment (maximum vertical pocket or MVP)
- ☐ Estimated fetal weights and abdominal circumference measurements
- ☐ Fetal bladder (present or absent)
- ☐ Middle cerebral artery peak systolic velocity (MCA-PSV).
- ☐ Umbilical artery Doppler should be evaluated after viability (at gestational age when delivery would be considered) in cases of sFGR
- ☐ Evaluation of possible complications related to surgery (chorioamniotic membrane separation, incidental septostomy).

Management of TTTS

Refer to Figure 2 for management of TTTS.

***Figure 2**. MCDA TTTS Management*



Delivery timing for pregnancies complicated by TTTS

In uncomplicated postoperative TTTS patients without evidence of sFGR, delivery at 36 weeks is reasonable as early elective delivery has not been shown to be protective against adverse neonatal outcomes in this group.[12] Furthermore, early delivery is associated with more complications related to prematurity. Delivery timing for cases complicated by sFGR and/or surgical complications should be individualized.

Management for other MCDA twin complications

In addition to TTTS, monochorionic twins are also at risk for developing selective fetal growth restriction (sFGR) and twin anemia polycythemia sequence (TAPS).[13]

Selective Fetal Growth Restriction (sFGR)

Diagnosis

MCDA twins are at risk for discordant twin growth that is more likely to be associated with either velamentous placental cord insertion or unequal placental sharing.[14] **BCM OB/Gyn perinatal guidelines committee recommends defining selective FGR (sFGR, formerly sIUGR) as a condition in which one fetus has EFW <10th centile or AC <10th centile.[15] Fetal weight discordance ≥ 25th centile without growth restriction is also associated with adverse perinatal outcomes. See management for growth discordance ≥ 25% centile (without growth restriction) for recommended management.**

In cases of early sFGR, the presence of inter-fetal anastomoses causes the smaller twin to receive well-oxygenated blood from its co-twin as a “rescue transfusion” phenomenon. The number and type of vascular anastomoses has led to three main well-defined clinical patterns as reported from Denmark.[16] About 5% of

sFGR cases occur later during the third trimester. Late sFGR has placental angioarchitecture that is distinct from early sFGR.[17] The following table summarizes this sFGR classification.[5]

Table 3. Type of sIUGR in MC twins according to the characteristics of umbilical artery Doppler in the small twin, and main clinical and placental features of each type [67]

Type	Clinical features	Placental features
Type I: normal Doppler (positive diastolic flow)	– Benign course with smaller degree of weight discordance – Very low risk of IUFD and co-twin brain damage – IUGR evolves well (normally >34 weeks' GA)	– Unequally shared placenta – High anastomotic diameter which largely compensates for placental territory imbalance – No or small AA anastomosis
Type II: absent/reverse end-diastolic flow (constantly)	– High risk of deterioration and IUFD of IUGR twin (predictable by Doppler evolution in most cases) – Very low risk of intrauterine brain injury of normal co-twin – Mean GA at delivery (due to deterioration of IUGR) 29 weeks	– Very unequally shared placenta – Smaller anastomotic diameter, which compensates for severe placental territory imbalance, but for a shorter time – No or small AA anastomosis
Type III Intermittent absent/reverse end-diastolic flow	– Low risk of hypoxic deterioration of IUGR twin which commonly survives until 32 weeks' GA and beyond – 10–15% risk of unexpected IUFD of IUGR twin (non-predictable) – 10–15% risk of brain injury in normal co-twin	– Very unequally shared placenta – A large AA anastomosis which largely compensates for unequal sharing and allows long in utero survival of IUGR twin, but carries high risk of acute feto-fetal transfusion accidents

Standardized protocol of expectant management of sFGR according to type:

The following protocol has been suggested for management of sFGR in monochorionic twins (based on expert opinion) in our institution:

Type I:

- ☐ Fetal Doppler (umbilical artery, and middle cerebral artery) and amniotic fluid levels checked once weekly
- ☐ Fetal growth assessment every 3 weeks (using Hadlock in-utero weight formula[3])
- ☐ Fetal echocardiography at initial presentation and again if there is evidence of worsening fetal Doppler changes or as otherwise suggested by the Pediatric Cardiology specialist.

Type II and III:

- ☐ Refer to Fetal Intervention
- ☐ Fetal Doppler (umbilical artery, ductus venosus and middle cerebral artery) and amniotic fluid levels checked twice weekly (to identify candidates who may benefit from fetal intervention)
- ☐ Fetal growth assessment every 3 weeks
- ☐ Fetal echocardiography at initial presentation and again if there is evidence of worsening fetal Doppler changes or as otherwise suggested by the Pediatric Cardiology specialist.
- ☐ Neonatology consultation at 23-24 weeks gestation to discuss the chances of survival of both twins in the view of estimated fetal weights
- ☐ Consultation with the Fetal Center is recommended

Presence of ominous signs of fetal deterioration (**persistent** absence/reversal of "a" waves in ductus venosus, fetal hydrops, or significant echocardiographic findings such as impaired cardiac function) are reasons to discuss the option of fetal intervention (laser photocoagulation if feasible or out of the state options). After fetal viability, delivery would be considered (fetal intervention after viability would be considered in the form of laser photocoagulation if feasible in selected cases with expected very low chances of survival of the FGR fetus).

Criteria for hospital admission

Patients with type II or III sFGR should be offered admission to the hospital between 26-28 weeks' gestation (26 weeks' gestation if there is evidence of reversed diastolic flow in umbilical artery or persistent abnormal blood flow pattern in the ductus venosus).

During admission:

- ☐ Administer course of antenatal corticosteroids for fetal lung maturation

- ☐ Neonatology consultation
- ☐ BPP twice weekly
- ☐ NST every 12 hours (frequency can be increased if initial tracings are non-reassuring)
- ☐ Magnesium sulfate for neuroprotection (before 32 weeks) and rescue course of antenatal corticosteroids (if 1-2 weeks have passed since the previous steroid dose) in case of imminent delivery

Management for growth discordance ≥ 25%centile (without growth restriction)

If there is evidence of is ≥ 25% discordance, the BCM OB/Gyn Perinatal Guidelines Committee recommends performing an umbilical artery Doppler study at the time of the growth scan. If the Doppler study is normal, recommend repeating the Doppler study at the next scheduled ultrasound (next q2 week TTTS check or next antenatal testing appointment). If the Doppler study is abnormal (PI > 95th centile), recommend initiation of weekly antenatal testing with Doppler studies.

Delivery timing:

Timing of delivery depends on fetal status

Table 1. Delivery timing by sFGR type

sFGR Type	**Delivery timing**
Elective delivery, reassuring fetal wellbeing	
Type I	34w0d-36w0d
Type II	32w0d
Type III	32w0d-34w0d *34 weeks if fetal status stable (NST/BPP remains reassuring)*
Emergency Delivery indications	
BPP ≤ 4/10	At diagnosis
Category 3 fetal heart tracing	At diagnosis
Sinusoidal pattern	At diagnosis
Moderate variability with accelerations and any of the following: • Recurrent late decelerations • Recurrent variable decelerations (FHR <80 bpm, more than 3/10 minutes) • Fetal bradycardia or recurrent prolonged decelerations	If recurrent and not responding to a trial of in utero resuscitation (IV fluid bolus and left lateral decubitus positioning)

Twin Anemia Polycythemia Sequence (TAPS)

The incidence of spontaneous TAPS in MCDA twins is approximately 5%. Following laser surgery for TTTS, this incidence can be as high as 13%. TAPS results from chronic transfusion across smaller arteriovenous anastomoses.

Diagnosis

SMFM recommends routine evaluation of MCA-PSV during ultrasound surveillance for TTTS and recommends that diagnosis of TAPS should be based on either the classical TAPS criteria (MCA-PSV>1.5 MoM in anemic fetus and and <1 MoM in the polycythemic fetus) or delta MCA-PSV >0.5 MoM.[9,18,19]

However, utilizing these criteria may be associated with increased number of referrals to fetal centers due to either false positive, transient MCA-PSV discordance or early cases of TAPS. Accordingly, the SMFM recommends fetal center referral for cases with stage II TAPS and above (MCA-PSV >1.7 MoM in the anemia fetus and <0.8 in the polycythemic fetus) as they are potential surgical candidates.

Management

The BCM Fetal Intervention team recommends referral to the Fetal Center for cases with stage II TAPS using the classical criteria or significant discordance of MCA-PSV (defined as delta MCA-PSV>1) and above. For cases of suspected or early-stage TAPS (defined as MCA-PSV >1.5 MoM but < 1.7 MoM in the anemic twin and <1 MoM but > 0.8 MoM in the polycythemic twin or delta MCA-PSV 0.5 - 1.0 MoM), we recommend evaluation of MCA-PSV, umbilical artery and Ductus venosus Doppler and at least weekly surveillance). Referral to a fetal center is recommended in early-stage TAPS if findings persist on repeat evaluation.

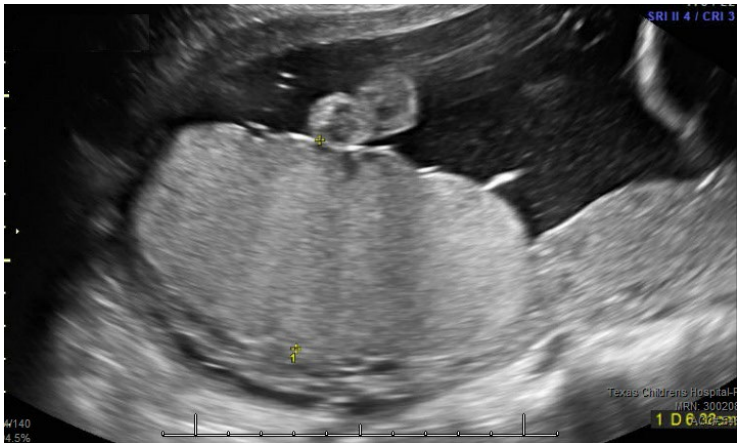
Stages of TAPS

	MCA-PSV in donor (MoM)	MCA-PSV in recipient (MoM)
Stage I	>1.5	< 1
Stage II	>1.7	<0.8
Stage III	Evidence of fetal cardiac compromise	
Stage IV	Hydrops	
Stage V	Single or dual fetal demise	

Associated ultrasound criteria in cases of TAPS

- There are fetal liver and placental changes suggestive of TAPS

The placenta of the anemic fetus appears thick and hyperechogenic, while the placenta of the polycythemic fetus appears thin and translucent . The liver of the polycythemic fetus has a "starry sky" appearance.



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