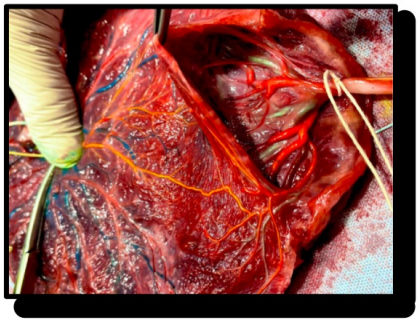
Society for Maternal-Fetal Medicine

Consult Series #72

# Twin-twin transfusion syndrome and twin anemia-polycythemia sequence



Monozygotic twins account for 30% of spontaneously occurring twins → 2/3 of monozygotic twins are monochorionic (MC), possessing a single placenta → >95% of monochorionic-diamniotic (MCDA) pairs share intertwin placental circulation



The placental surface is visualized, with forceps (image right) retracting the intertwin membrane. A thumb overlies 1 placental cord insertion, with yellow (arterial) and blue (venous) dyes injected into the superiorly located placental territory. Near the image bottom, a placental cord insertion is observed into the inferiorly located placental territory, with red (arterial) and green (venous) dyes injected into that circulation. Toward the left side of the image, near the location where the dividing membrane and placenta meet, intertwin anastomoses are demonstrated by a mixing of the colored dyes.
*[Image courtesy E. Bergh, MD].*

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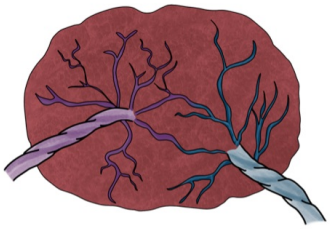
Common placental mass + Shared intertwin placental circulation → Risk of twin-twin transfusion syndrome (TTTS) and twin anemia-polycythemia sequence (TAPS)

Nine-week monochorionic-diamniotic twin pregnancy



A single gestational sac is visualized, with each twin occupying a distinct amniotic space.
*[Image courtesy R. Miller, MD].*

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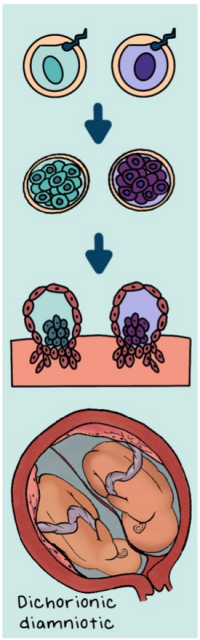
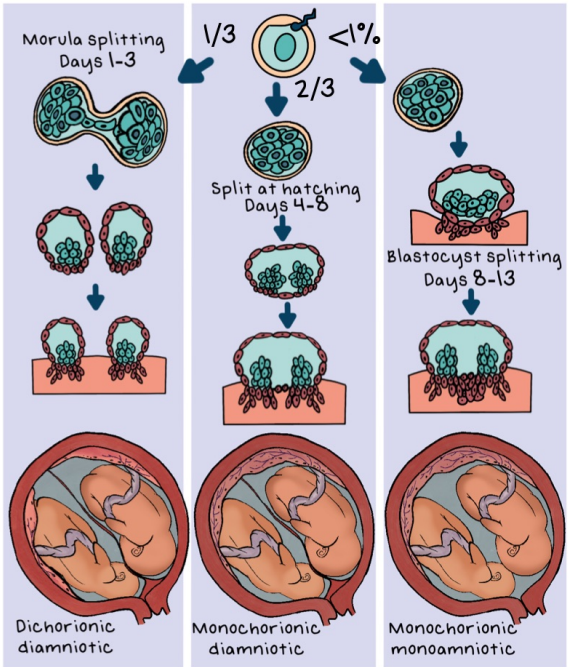
## CHORIONICITY

We recommend routine first-trimester sonography determination of chorionicity and amnionicity

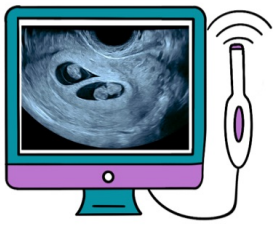
### Dizygotic twinning




### Monozygotic twinning




### Dizygotic



Evidence of 2 distinct gestational sacs on transvaginal ultrasound performed before 10 weeks of gestation indicates dichorionicity (DC)

Two distinct gestational sacs are visualized.
*[Image courtesy R. Miller, MD].*

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### Monozygotic




| | |
|---|---|
| <10 weeks | Determination of amnionicity is less accurate |
| 10-14 weeks of gestation | Reassess for intervening membrane |

"T" sign = perpendicular attachment of intervening twin membrane to the placenta; indicates MC placentation

**A,** 7-week monochorionic twin pregnancy; no dividing membrane is visible. **B,** Same monochorionic twin pregnancy at 11 weeks of gestation; a dividing membrane and "T" sign are now visible.
*[Image courtesy R. Miller, MD].*

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"Lambda" sign indicates DC placentation

Labels "AAA" and "BBB" refer to twins A and B, respectively, and "MEMBRANE" demonstrates the dividing membrane. The "twin peak" sign is the triangular projection of placental tissue in the base of the inter-twin membrane.
*[Image courtesy R. Miller, MD].*

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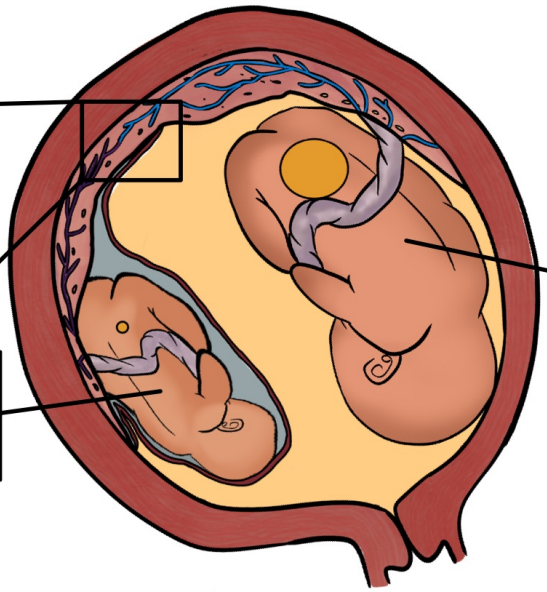
# TWIN-TWIN TRANSFUSION SYNDROME

Imbalanced sharing of blood and vasoactive substances across a common placental circulation through vascular anastomoses

Impacts 8-12% of MCDA twin pregnancies



Hypervolemic hypertensive recipient twin at risk for heart failure with polyuria and polyhydramnios

Hypovolemic donor twin with oliguria and oligohydramnios

## DIAGNOSIS/ STAGING

**TABLE 1**
**Quintero staging of twin-twin transfusion syndrome[22]**

| Stage | Ultrasound assessment | Criteria |
|---|---|---|
| I | Amniotic fluid | Maximal vertical pocket <2 cm in donor sac and maximal vertical pocket >8 cm in recipient sac |
| II | Fetal bladder | Nonvisualization of fetal bladder in donor twin over 60 minutes of observation |
| III | Doppler studies | Absent or reversed umbilical artery end-diastolic velocity, reversed ductus venosus a-wave flow, pulsatile umbilical vein flow |
| IV | Fetal ascites or hydrops | Ascites or hydrops in 1 or both twins |
| V | Fetal cardiac activity | Fetal demise in 1 or both twins |

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## SURVEILLANCE

We recommend that ultrasound surveillance for TTTS begin at 16 weeks of gestation for all MCDA twin pregnancies and continue at least every 2 weeks until delivery, with more frequent monitoring indicated for clinical concern

We recommend that ultrasound surveillance for TTTS minimally include assessment of amniotic fluid volumes on both sides of the intertwin membrane and evaluation for the presence or absence of urine-filled bladders, and ideally incorporate Doppler study of the umbilical arteries

+ Fetal weight assessments at least every 4 weeks
+ Fetal echocardiography

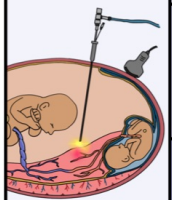
*Also consider middle cerebral arteries (MCA) Doppler

### Monitoring after laser

We suggest weekly surveillance for 6 weeks followed by resumption of every-other-week surveillance thereafter, unless concern exists for post-laser TTTS, post-laser TAPS, or FGR

## MANAGEMENT

We recommend that all patients qualifying for laser therapy be referred to a fetal intervention center for further evaluation, consultation, and care

| | | |
|---|---|---|
| 16 - 26 weeks of gestation  | Stage I TTTS asymptomatic | We recommend expectant management with at least weekly fetal surveillance |
| | Stage I TTTS complicated by additional factors such as maternal polyhydramnios-associated symptomatology | We recommend consideration of fetoscopic laser surgery |
| | Stage II - Stage IV TTTS | We recommend fetoscopic laser surgery as the standard treatment |
| Early- and late-presenting TTTS | | We recommend an individualized approach to laser surgery |

## DELIVERY

Following the resolution of TTTS after fetoscopic laser surgery and without other indications for earlier delivery, we recommend delivery of dual-surviving MCDA twins at 34 to 36 weeks of gestation

In TTTS pregnancies complicated by posttreatment single fetal demise, we recommend full-term delivery (39 weeks) of the surviving co-twin to avoid complications of prematurity unless indications for earlier delivery exist

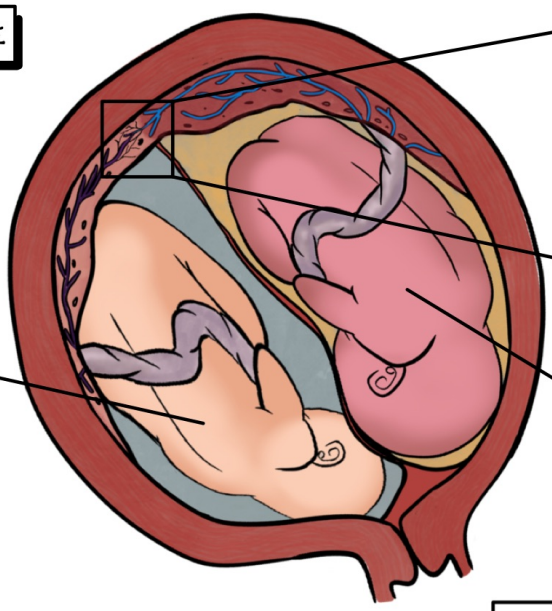
We recommend that fetoscopic laser surgery not influence the mode of delivery

# TWIN ANEMIA-POLYCYTHEMIA SEQUENCE

Chronic, insidious form of fetofetal transfusion with an imbalanced red blood cell transfusion

Naturally occurring TAPS impacts 2-5% of MCDA twin gestations

After laser surgery, TAPS occurs more frequently and can progress rapidly




## DIAGNOSIS/STAGING

**TABLE 2**
**Prenatal staging of twin anemia-polycythemia sequence**[85,95]

| Stage | Criteria | Intertwin criteria |
|---|---|---|
| 1 | MCA-PSV >1.5 MoM in donor and MCA-PSV <1.0 MoM in recipient | Δ MCA-PSV >0.5 MoM without cardiac compromise of donor[a] |
| 2 | MCA-PSV >1.7 MoM in donor and MCA-PSV <0.8 MoM in recipient | |
| 3 | Stage 1 or 2 with cardiac compromise of donor[a] | |
| 4 | Ascites or hydrops of donor | |
| 5 | Single or double fetal demise | |

*MCA-PSV*, middle cerebral artery Doppler peak systolic velocity; *MoM*, multiples of the median.

Adapted from Slaghekke et al,[85] 2010 and Tollenaar et al,[95] 2019.

[a] Cardiac compromise defined as absent or reversed end-diastolic flow in umbilical artery, pulsatile flow in umbilical vein, or reversed a-wave in the ductus venosus.

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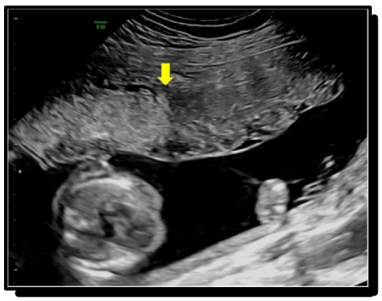
Accompanying ultrasound findings in up to 86% of TAPS cases, including:

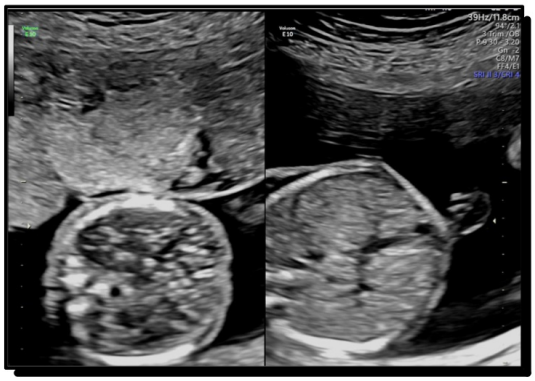
Recipient twin cardiomegaly

Discordant placental echogenicity



A distinction in placental echotexture exists, with a hyperechoic donor placental territory on the left and a hypoechoic recipient placental territory on the right.
*[Image courtesy R. Miller, MD]*

"Starry sky" appearance of recipient liver



On the left, axial view of recipient twin liver with "starry sky" appearance. On the right, axial view of donor liver with normal echotexture.
*[Image courtesy R. Miller, MD]*

## MANAGEMENT

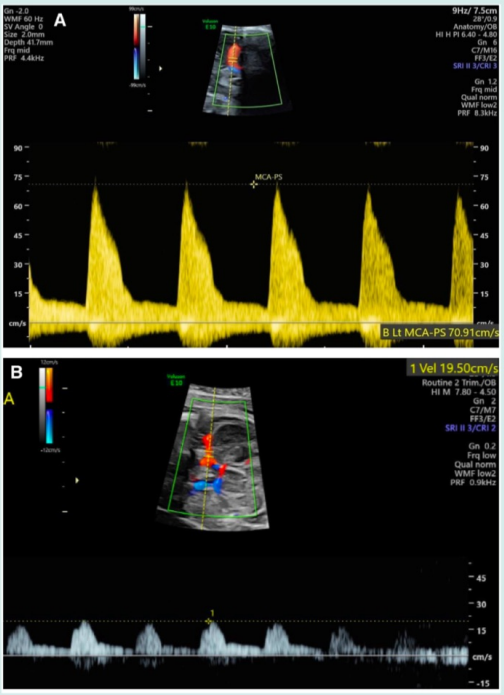
Consultation with a specialized fetal care center is recommended when TAPS progresses to a more advanced disease stage (stage≥II) before 32 weeks of gestation or when concern arises for coexisting complications such as TTTS

TAPS presentation beyond 32 weeks may be considered for delivery and do not necessarily require outside referral

## SURVEILLANCE

We recommend that providers consider incorporating Doppler MCA-PSV into all MC twin ultrasound surveillance beginning at 16 weeks of gestation

Detection of TAPS during pregnancy should prompt care escalation that may include: heightened surveillance, fetal care center referral, fetal therapy, or delivery




A. Donor twin elevated MCA-PSV (1.8 MoM) in a case of stage II TAPS at 29 weeks of gestation.
B. Recipient twin decreased MSA-PSV (<0.7 MoM) in a case of stage III TAPS at 23 weeks of gestation.
*[Image courtesy J. Miller, MD and R. Miller, MD, respectively]*

## Prognosis

Outcomes appear to correlate with disease severity

The natural history is incompletely understood