# Ultrasound in Twins

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#### Disclosures

Jodi S. Dashe, MD

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#### Learning Objectives

After completing this presentation, the learner will be able to discuss:

- 1. Sonographic characteristics that distinguish monochorionic from dichorionic twins
- 2. Complications unique to monochorionic twins
- 3. Evaluation for twin-twin transfusion syndrome
- 4. Ultrasound surveillance recommendations for monochorionic and dichorionic pregnancies

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#### Outline

- Twinning prevalence, associated morbidities
- Chorionicity and amnionicity
  - First trimester
  - Second and third trimesters
- · Discordance and growth
- Complications unique to monochorionic twins
   TTTS, TAPS, TRAPS, conjoined twins
- Surveillance recommendations

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# From 1980 to 2009, the age distribution of women giving birth to twins increased



#### Zygosity, Chorionicity, Amnionicity Dizygotic twins Monozygotic twins (2 oocytes/2 sperm) (1 oocyte/1 sperm) ~ 2/3 of twins ~ 1/3 of twins Always dichorionic If division < 3 days, morula Always diamniotic dichorionic diamniotic (~1/3) If division 3-8 days, blastocyst hatching monochorionic diamniotic (~2/3) If division 8-12 days, monochorionic monoamniotic If division has not occurred by 13 days, conjoined twins

#### Associated morbidities

- Twins are at increased risk for early loss, fetal death, preterm birth, low birthweight, neurological morbidity, and neonatal death
  - Monochorionic twins disproportionately affected
- · Structural abnormalities:

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- > 1:25 dichorionic twins
- 1:15 monochorionic diamniotic twins
- 1:6 monoamniotic twins

ISUOG Practice Guideline, Ultrasound Obstet Gynecol 2016;47:247-63.

## CDC: Births, Final Data for 2014

	Singletons	Twins
Preterm < 37 weeks	7.7%	58.7%
Preterm < 32 weeks	1.2%	10.6%
Low birthweight, < 2500 g	6.2%	55.2%
Very low birthweight, < 1500 g	1.1%	9.6%

National Vital Statistics Reports, Vol. 64, No. 12, Dec 2015.

# Fetal mortality (> 20 weeks) in singletons, twins, and triplets or higher order multiples, CDC 2005



#### Singleton and Twin Pregnancy Outcomes, Parkland Hospital, 2002-2012

	Singletons	Twins
Births	78,879	1700 (2%)
Stillbirths	406 5.1/1000	24 14.1/1000
Neonatal Deaths	253 3.2/1000	38 22.4/1000
Perinatal Deaths	659 8.4/1000	62 36.5/1000

# Loss of 1 twin

- "Vanishing twin" before 12 weeks
  - Reported in 36% of twin pregnancies and
     > 50% of triplets followed prospectively
- Fetus papyraceus observed after 12 weeks



Dickey RP et al, Am J Obstet Gynecol 2002;186:77-83.

#### Loss of 1 twin

- Second- or third-trimester loss of one or both twins occurs in up to 5%
  - Co-twin loss occurs in 15% of monochorionic and 3% of dichorionic pregnancies
- In a monochorionic gestation, 15% to 20% of co-twins suffer neurological morbidity following demise of 1 twin
  - Immediate delivery is *not* beneficial in absence of another indication

Hillman SC, Obstet Gynecol 2011;118:928-40. Ong SS, BJOG 2006;113:992-8. ACOG PB 144, Obstet Gynecol 2014;123:1118-32.

#### Neurological morbidity example

Development of intracranial hemorrhage with porencephaly after monochorionic co-twin demise



Immediate delivery is not beneficial unless other indication arises

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#### Chorionicity and amnionicity

- Diagnosis and evaluation of multiple gestation is considered an indication for first trimester sonography (NICHD, SMFM, AIUM, ACOG, ACR, SPR, SRU)
- Chorionicity assessment is at least 98% accurate in the 1st trimester
- Incorrect in up to 10% in 2<sup>nd</sup> trimester

Reddy U et al, Fetal Imaging Workshop, Obstet Gynecol 2014;123:1070-82. Lee YM, Am J Obstet Gynecol 2006;195:863-7. Blumenfeld Y et al (NICHD), J Ultrasound Med 2014;33:2187-92.

#### Antenatal sonographic prediction of twin chorionicity

Young Mi Lee, MD,<sup>3,\*</sup> Jane Cleary-Goldman, MD,<sup>a</sup> Harshwardhan M. Thaker, MD, PhD,<sup>b</sup> Lynn L. Simpson, MD<sup>a</sup> American Journal of Obstetrics and Gynecology (2006) 195, 863-7

Chorionicity assessed sonographically < 24 weeks in 410 twin pairs, 2001-2005, with postnatal pathology confirmation

Table I Stati monochorionicit	stical accuracy y	of antenatal	prediction of
	Sensitivity	Specificitiy	PPV NPV
	(%)	(%)	(%) (%)
Overall	88.9	97.7	92.6 96.5
1st trimester	89.8	99.5	97.8 97.5
2nd trimester	88.0	94.7	88.0 94.7

PPV, Positive predictive values; NPV, negative predictive values.

Chorionicity correctly assigned in 96%

# Accuracy of Sonographic Chorionicity Classification in Twin Gestations

- Secondary analysis of prospective trial by NICHD, 14 academic medical centers, 2004-2006
- 545 twin pairs, chorionicity assessed ≤ 20 weeks, postnatal pathology confirmation of placentation
- 17% monochorionic, 83% dichorionic
- Overall, sonographic assignment of chorionicity accurate in 94%
  - > 1<sup>st</sup> trimester 95%
  - > 2<sup>nd</sup> trimester 90%

Blumenfeld Y, et al (NICHD), J Ultrasound Med 2014;33:2187-92.

#### Early first trimester

- Gestational sac number equals the number of chorions
- Dichorionic twins have a thick band of chorion separating 2 gestational sacs
- · Monochorionic twins have a single sac
  - With monochorionic diamniotic twins, it may be difficult to visualize the thin intervening amnion before about 8 weeks

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Dichorionic diamniotic twins at 6 weeks Gestational sacs separated by thick band of chorion



Dichorionic diamniotic twins at 9 weeks Gestational sacs separated by thick band of chorion



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Monochorionic diamniotic twins at 8 weeks Single gestational sac, amnion visible as a fine membrane encircling each embryo (arrows)



Monochorionic diamniotic twins at 10 weeks Single gestational sac, amnion visible as a fine membrane encircling each fetus (arrows)



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Monochorionic diamniotic twins at 10 weeks Single gestational sac, amnion visible as a fine membrane encircling each fetus (arrows)



#### Monochorionic twins - yolk sacs

- If intervening membrane is difficult to see, the number of yolk sacs usually equals the number of amnions
  - But, there are reports of diamniotic gestations with 1 yolk sac and of monoamniotic gestations with 2 yolk sacs!
- If cord entanglement, monoamnionicity
- · If in doubt, perform follow-up sonography

Shen O, Ultrasound Obstet Gynecol 2006;27:53-5. Corbett, Ultrasound Obstet Gynecol 2012;39:607-8.



Monochorionic diamniotic twins at 8 weeks

#### From late first trimester onward

- Earlier is better
- 4 categories of findings
  - > Twin peak ( $\lambda$ ) sign or T-sign
  - Number of placental masses
  - > Thickness of dividing membrane
  - Fetal sex

# Twin peak (λ)- or T-sign

Twin peak or  $\lambda$ -sign: Wedgeshaped projection of placental tissue extending between layers of the inter-twin membrane and uterine wall in <u>a dichorionic gestation</u>

T-sign: Dividing membrane may form a "T" with the wall of the uterine cavity in a monochorionic diamniotic gestation



# Number of placental masses

- Dichorionic if 2 separate placental masses
  - Exception would be a bipartite monochorionic placenta (uncommon)
- Converse is not the case single placenta does not indicate monochorionicity
  - 2 adjacent masses may look like 1
  - Additional findings are necessary

# Number of placental masses



# Thickness of dividing membrane

- Membrane thickness is often > 2 mm with a dichorionic gestation
  - > 2 layers of amnion and 2 of chorion
- Dividing amnion layers in a monochorionic gestation tend to look "wispy," dichorionic membranes appear thicker
- Distinction more challenging with advancing gestation
- · Not diagnostic as single criterion

Membrane thickness examples





Monochorionic diamniotic twins
- Single placental mass
- Thin dividing membrane with T-sign





#### Fetal sex if visible

- Dichorionic twins same sex about  $\frac{1}{2}$  the time
- Monochorionic twins almost always same sex







#### Monoamniotic twins

- Incidence 1 per 10,000 pregnancies
   About 5% of monochorionic twins
- Usually just 1 yolk sac
- Invariably associated with cord entanglement
  - When systematically sought, identified in 98%
  - Look with color Doppler whenever a dividing membrane cannot be visualized

ACOG PB 144, Obstet Gynecol 2014;123:1118-32.

### Monoamniotic gestation, 20 weeks



The umbilical cord insertion sites are in close proximity to one another, with no visible intervening membrane. Color Doppler demonstrates cord entanglement.

# Monoamniotic twins

- Overall survival 89%, survival of  $\geq$  1 twin 93%
- Increased risk for prematurity, abnormalities, TRAP sequence
- Many offer inpatient hospitalization with daily fetal surveillance from 24 to 28 weeks
  - Optimal management uncertain
- ACOG and ISUOG recommend delivery via cesarean at 32-34 weeks in absence of other complications

Rossi AC, Ultrasound Obstet Gynecol 2013;41:131. ACOG PB 144, Obstet Gynecol 2014;123:1118-32.

# Monoamniotic gestation, 20 weeks





#### Fetal growth restriction

- Usually defined using 10<sup>th</sup> percentile
- If singleton nomogram is used, such as Hadlock, prevalence will be higher than if a twin-specific or chorionicity-specific nomogram is used
- Growth surveillance is recommended every 4 weeks, regardless of chorionicity
- As in singletons, management is guided by factors other than fetal weight alone – such as gestational age, amniotic fluid, Doppler findings, or other antepartum surveillance findings

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#### **Discordance**

- Difference in fetal weight (or birth weight) between larger and smaller twins, expressed as % of weight of the larger twin
  - Rationale: assuming normal size of the larger twin, may indicate abnormal growth of smaller twin
- Typically defined as  $\geq$  20% (ACOG) or  $\geq$  25% (ISUOG)

ACOG Practice Bulletin No. 144, Obstet Gynecol 2014;123:1118-32. ISUOG Practice Guideline, Ultrasound Obstet Gynecol 2016;47:247-63.

#### Fetal weight discordance

- "Whether growth discordant multifetal gestations – without a structural anomaly, aneuploidy, discordant infection, oligohydramnios, or fetal growth restriction – are at increased risk of adverse outcomes is debatable." - ACOG

ACOG Practice Bulletin No. 144, Obstet Gynecol 2014;123:1118-32.

# Selective fetal growth restriction

- Fetus with estimated weight below 10<sup>th</sup> percentile and discordance <u>> 25%</u>
- Dichorionic twins generally managed as singletons with FGR
- In monochorionic twins, limited evidence to guide management
  - sFGR believed to occur mainly due to unequal sharing of the placental mass and vasculature

ISUOG Practice Guideline, Ultrasound Obstet Gynecol 2016;47:247-63.

### Selective fetal growth restriction

- Gratacos (USOG 2007;30:28-34) has proposed staging sFGR according to smaller MC twin umbilical artery Doppler flow
   I: Positive end-diastolic flow
  - II: Absent or reversed end-diastolic flow III: Intermittent absent/reversed end-diastolic flow



# Complications unique to monochorionic twins

- · Twin-twin transfusion syndrome
- Twin-anemia polycythemia sequence
- Twin-reversed arterial perfusion sequence
- · Conjoined twins



#### Twin-twin transfusion syndrome

- 1-3/10,000 births, up to 10% of MC twins
- Diagnostic criteria
  - > Monochorionic diamniotic twin gestation
  - Hydramnios in one sac (> 8 cm pocket) and oligohydramnios in other sac (< 2 cm pocket)
- Blood is transferred from donor twin to recipient through placental arterio-venous anastemoses

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#### Twin-twin transfusion syndrome

- Surveillance: Sonography should be considered every 2 weeks from 16 weeks onward in all monochorionic twins to evaluate amniotic fluid and fetal bladders
- Quintero staging is recommended if TTTS is diagnosed

ACOG Practice Bulletin No. 144, Obstet Gynecol 2014;123:1118-32. Society for Maternal-Fetal Medicine. Am J Obstet Gynecol 2013;208:3-18.

# Quintero staging system

- I. Oligohydramnios in donor twin sac, hydramnios in recipient sac
- II. Urine no longer visible within bladder of donor twin
- Critically abnormal Doppler studies of umbilical artery, umbilical vein, or ductus venosus
- IV. Hydrops of either twin
- v. Demise of either twin

Quintero RA et al. J Perinatol 1999;19:550-5.









# Additional sonographic risk factors

asso	ciated with twin-twin transfusion syndrome
First-t	trimester findings
Cr	own-rump length discordance43
Nu	ichal translucency >95th percentile42,44 or discordance >20% between twins45,46
Re	eversal or absence of ductus venosus A-wave 47,48
Secon	id-trimester findings
Ab	odominal circumference discordance43
M	embrane folding <sup>28,42</sup>
Ve	lamentous placental cord insertion (donor twin)28
Pla	acental echogenicity (donor portion hyperechoic)49
SMFM	. Twin-twin transfusion syndrome. Am J Obstet Gynecol 2013.

Society for Maternal-Fetal Medicine. Am J Obstet Gynecol 2013;208:3-18.

First trimester CRL discordance



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# Enlarged nuchal translucency (progressed to severe TTTS)







#### TTTS natural history

Data limited

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- Stage I: > 75% of cases remain stable or regress without invasive interventions, reported perinatal survival about 86%
  - Progression confers poor prognosis
- Stage III or greater: Perinatal loss rate 70-100%, particularly when TTTS presents before 26 weeks

Society for Maternal-Fetal Medicine. Am J Obstet Gynecol 2013;208:3-18.

#### TTTS management

- For stages II, III, and IV, most consider fetoscopic laser photocoagulation to be the best available approach
  - However, meta-analysis data have not demonstrated survival benefit, neurologic outcome in Eurofetus trial not different than in controls
- Laser-treated TTTS is associated with 30-50% risk of perinatal death and 5-20% risk of neurologic handicap

Society for Maternal-Fetal Medicine. Am J Obstet Gynecol 2013;208:3-18.

## TAPS

- Characterized postnatally by hemoglobin difference between monochorionic twins
- Diagnosed prenatally based on MCA peak systolic velocity difference (elevated in anemia)
- May occur in up to 5% of monochorionic twins
   Role of surveillance has yet to be determined
- More common in TTTS 13% following laser photocoagulation of placental anastemoses
- Staging has been proposed

Lopriore E, et al. Prenat Diagn 2010;30:251-55. Slaghekke F, et al. Fetal Diagn Ther 2010;27:181-190.

# Twin-anemia polycythemia sequence (TAPS)









#### TRAPS

- Complicates approximately 1:35,000 pregnancies
   About 1% of MC twins
- Morphologically-normal "pump" twin and an "acardius" – a recipient with complete or partial absence of the heart and other abnormalities
  - Acardius anceps: complete absence of heart but at least partial presence of other structures
  - > Acardius acephalus: absence of head and heart
  - > Acardius amorphous: no recognizable structure

Napolitani FD, Schreiber I. Am J Obstet Gynecol 1960;80:582-9.

#### TRAPS

- Umbilical Doppler evaluation demonstrates pulsatile flow moving toward the acardiac twin, presumed due to large A-A and V-V anastomoses within placenta
- Deoxygenated blood from the pump twin flows in a reversed direction through the acardius
  - The lower body of the acardius is more likely to develop
- The pump twin has the burden of perfusing the acardiac twin and is at risk for hydrops from high-output cardiac failure





#### TRAPS

- Historically associated with > 50% mortality due to high-output cardiac failure or prematurity
- Risk is directly related to size of acardius • Ellipsoid volume estimate, L x W x H x  $\pi/6$
- NAFTNet Review, 98 RFA cases 1998-2008
  - » RFA performed at 20.2 ± 2.4 wks
  - Acardius to pump-twin ratio averaged 90%
  - > 80% survival to 30 days (16 fetal losses, 4 NND)

Lee H et al. Fetal Diagn Ther 2013;33:224-9.





#### Conjoined twins

- Estimated prevalence 1:70,000 births
- Monozygotic twins, do not fully separate by 12 days post-fertilization, continue to develop
- Term "Siamese" from Eng and Chang Bunker (1811-1874), joined at lower thorax
- In a review of 21 international surveillance programs (1968-2006), 50% were liveborn, 25% stillborn, and 25% were terminated

Mutchinick OM et al. Am J Med Genet 2011;157(C):274-287.

Types	Definitions	
Cephalopagus	There are two faces and are joined from the top of the head to the umbilicus	
Thoracopagus	Are joined face-to-face from the upper thorax to the upper part of the abdomen and always involve the heart	
Omphalopagus	The fusion includes the umbilicus region frequently at the lower thorax, but never the heart	
lschiopagus	The union usually includes the lower abdomen and duplicated fused pelvic bones, and external genitalia and anus are always involved	
Parapagus	Are laterally joined, regularly share the pelvis. Varieties of parapagas conjoined twins are parapagas dithoracic (separated thoraces), parapagas dicephalus (one trunk two separate heads), and parapagas diprosopus (one trunk, one head, and two faces)	
Craniopagus	Joined by the skull, share meninges but rarely the brain surface and do not include the face and trunk	
Pygopagus	Are dorsally fused sharing the perineal and sacrococcygeal areas, has only one anus but two rectums	
Rachipagus	Dorsally fused, the defect may involve the donsolumbar vertebral column and rarely the cervical vertebrae and the occipital bone	
Other symmetrical	Includes CT that some authors classify differently and also a variety of rare types of symmetrical CT	
Asymmetric	Parasitic CT and fetus in fetus	

## Conjoined twins

- Characterization of fused organs and associated anomalies is particularly important for prognosis
- Thoracopagus twins are the most common, and in 90% there is a shared pericardium, almost always associated with cardiac fusion
  - > Typically precludes surgical separation

# Conjoined twins



Thoracopagus twins at 13 weeks





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## First trimester sonography

Gestational age	Indication	Comment
7-10 weeks (ideally)	Gestational age assessment	
11-14 weeks	Nuchal translucency	Increased with aneuploidy, anomalies, TTTS
Earlier is better!	Evaluation of chorionicity	

Reddy U et al, Fetal Imaging Workshop, Obstet Gynecol 2014;123:1070-82. (Guidelines from NICHD, SMFM, AIUM, ACOG, ACR, SPR, SRU)

# Twin Sonography Recommendations

Gestational age	Indication	Comment
18-20 weeks	Detailed evaluation of anatomy and placenta	
24 weeks onward	Every 4 weeks to assess growth	If discordant or other complication, may need more frequent evaluation

#### Second trimester – Monochorionic

Gestational age	Indication	Comment
16 weeks onward	Every 2 weeks to assess amniotic fluid and fetal bladders	Largest vertical pocket 2 to 8 cm is normal
16 weeks onward	Every 4 weeks to assess growth	If discordant or other complication, may need more frequent evaluation
18-22 weeks	Detailed evaluation of anatomy and placenta, fetal echocardiography	Cardiac anomaly in 5%, up to 8% if TTTS is present
Reddy U et al, F Emery SP, et al ,	etal Imaging Workshop, Obs NAFTNet consensus, Obste	stet Gynecol 2014;123:1070 t Gynecol 2015;125(5):1236

Doppler		
Indication	Vessel	Comment
Monoamniotic twins, for diagnosis (cord entanglement)	Umbilical cord	Cord entanglement may not affect prognosis
Twin-twin transfusion (diagnosed)	<ul> <li>Umbilical artery/vein and ductus venosus</li> <li>MCA</li> </ul>	<ul> <li>Stage III criteria</li> <li>TAPS surveillance</li> </ul>
FGR (diagnosed)	Umbilical artery	As for singletons
Other surveillance [Controversial]	Umbilical artery or MCA	Not specifically recommended unless an indication arises

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#### Conclusions

- · Approximately 1/3 of twins are monozygotic
  - > Of monozygotic twins, 2/3 are monochorionic
  - > Overall, about 20% of twins are monochorionic
  - Early sonography is best for chorionicity assessment
  - Accuracy up to 98% in the 1<sup>st</sup> trimester
    - \* Look at number of sacs, intervening chorion
    - Usually, yolk sac number reflects amnionicity
  - Accuracy about 90% in the 2<sup>nd</sup> trimester
    - λ-sign or T-sign, placental masses, thickness of dividing membrane, fetal sex

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# Key References

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Emery SP, Bahtiyar MO, Dashe JS, Wilkins-Haug LE, Johnson A, Paek BW, et al. The North American Fetal Therapy Network consensus statement: prenatal management of uncomplicated monochorionic gestations. Obstet Gynecol 2015;125(5):1236-43.

Society for Maternal-Fetal Medicine, Simpson LL. Twin-twin transfusion syndrome. SMFM Clinical Guideline. Am J Obstet Gynecol 2013;208(1):3-18.

# Conclusions

- Monochorionic twins are at increased risk for abnormalities and specific complications
  - Monoamniotic twinning (cord entanglement), TTTS, TAPS, TRAPS, conjoined twinning
- Sonography is recommended in the 1st trimester
- Sonographic is recommended every 4 weeks to assess growth in 2<sup>nd</sup> and 3<sup>rd</sup> trimesters
  - TTTS surveillance is recommended every 2 weeks in MC twins, starting at 16 weeks