Using Ultrasound to Manage Twins

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Learning Objectives

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

• Diagnosis and dating in twin pregnancies
• Sonographic characteristics that distinguish dichorionic from monochorionic twins
• Prenatal ultrasound screening in twins
• Complications unique to monochorionic twins
• Ultrasound surveillance recommendations for twins

Diagnosis and Dating of Twins

• Diagnosis best in the first trimester and dating optimal using crown-rump length
  - 20% of first trimester twin pregnancies result in singleton live births
  - “Vanishing twin” is associated with favorable prognosis of surviving twin if dichorionic
• If discrepancy in dates between the twins, date using the larger twin
  - Avoids missing diagnosis of IUGR

Importance of Pregnancy Dating

• Timing for screening and diagnostic testing
• Accurate interpretation of twin growth
• Scheduling of twin deliveries

Types of Twins

• All dizygotic twins are dichorionic
• All monochorionic twins are monozygotic
• Not all monozygotic twins are monochorionic

Determination of Chorionicity

• Optimal in first trimester
  - close to 100% accuracy
• Incorrect assignment in up to 10% of cases when chorionicity determined in second trimester

Lee et al, 2006
Blumerfeld et al, 2014
Determination of Chorionicity

- Gestational sacs
- Amniotic sacs
- Placenta number
- Intertwin membrane
- Gender

Intertwin Membrane

Importance of Chorionicity

- Aneuploidy
- Higher-Order Multiples
- Malformations
- Discordant Growth

Prenatal Diagnosis

Goals in twin gestations are the SAME as for singletons

- To identify fetal abnormalities that could change a couple’s decision to continue a pregnancy or alter obstetric care
- To identify fetuses that might benefit from fetal or early neonatal therapy
- To provide reassurance that twins are developing normally

First Trimester Risk Assessment

Trisomy 21 Screening in Twins:
Detection Rate for 5% False Positive Rate*
Importance of Nuchal Translucency in Twins

- Aneuploidy
- Structural malformations
- Twin-twin transfusion syndrome
  - PPV 30%

Twin Anatomy: Fetal Anomalies

Background risk for singletons
- 2% overall
Rate same per fetus for dizygotic twins
- 2% per fetus
- 4% overall
Rate 2-3 times higher for monozygotic twins
- 4-6% per fetus
- 8-12% overall

Congenital Heart Disease: Singletons vs Twins

- CHD is the leading malformation contributing to infant mortality and morbidity
- Background risk in singletons
  - Prevalence in mid-trimester: 10 per 1000 singletons
  - Prevalence at birth: 8 per 1000 live births
  - Major cardiac defect at birth: 3-4 per 1000 live births
- Rate higher for monozygotic twins
  - 2-3% per fetus
  - 6% overall

Indications for Fetal Echocardiography

Maternal
- Autoimmune antibodies
- Familial inherited disorders
- In vitro fertilization
- Metabolic diseases
- Teratogen exposure
  - Retinoids
  - Lithium

Fetal
- Abnormal cardiac screen
- First-degree relative with CHD
- Abnormal heart rate or rhythm
- Fetal chromosomal anomaly
- Extracardiac anomaly
- Hydrops
- Increased NT
- Monochorionic twins

Twins conceived by IVF at increased risk for CHD irrespective of chorionicity

Screening for Fetal Anomalies: Singletons vs Twins

- Imaging difficult with greater number of fetuses in variable positions
- Monochorionic twins may be complicated by other factors that impact imaging
  - Polyhydramnios-oligohydramnios sequence
  - Discordant twin growth / sIUGR
  - Monoamnionicity
- Overall, lower detection rate expected in twins compared to the 30-50% observed in singletons

TTTS: Acquired CHD

- Biventricular Hypertrophy: >50% of recipient twins
- Pulmonary Stenosis: 5% of recipient twins

Karantra et al, Heart 2008

AOGP practice guideline for the performance of fetal echocardiography. Ultrasound Med 2013
Bahtiyar et al, 2010; Reefhuis et al, 2009

Twins conceived by IVF at increased risk for CHD irrespective of chorionicity
**Discordant Anomalies**

- 1-2% of twin pregnancies face the dilemma of expectant management versus selective termination

**DC twins**
- 3% risk of procedure-related pregnancy loss with selective reduction via intracardiac KCl

**MC twins**
- 5% risk of procedure-related pregnancy loss with selective reduction via cord occlusive techniques
  - 3% neurologic morbidity in surviving co-twin
  - Must weigh against 20% risk of neurologic injury if spontaneous demise of abnormal MC twin

O'Donoghue et al., 2009

**Importance of Placental Evaluation**

- Placenta previa more common in twins
- Placental cord insertion more likely to be abnormal in twins
  - Marginal
  - Velamentous
  - Vasa previa

Ananth et al., 2003; ACOG 2011:204:145

**Velamentous Placental Cord Insertion**

- 10% of twins compared to 1% of singletons
- Marker for unequal placental sharing with discordant twin growth/sIUGR in MCDA twins and IUGR in dichorionic twins
- 2% of velamentous PCI associated with vasa previa
- Detection rate >90-95% with routine use of transvaginal ultrasound using color and pulsed Doppler in midtrimester
- Perinatal mortality of vasa previa
  - ~50% in undiagnosed cases
  - <5% in cases identified prenatally

Simpson et al., 2009; O'Sullivan et al., 2007

**Importance of Cervical Length**

- Identify patients at risk for preterm delivery
  - Mean gestational age for live born twins = 35.4 weeks
  - Potential clinical value for all twin gestations
  - Transvaginal approach proven to be optimal approach to assess cervix

Inamdar et al., 1997; Guzman et al., 2000

**PCI: Ultrasound and Pathology**

- Vasa Previa
- Velamentous PCI with intertwin anastomoses

Simpson et al., 2009; O'Sullivan et al., 2007

**Meta-Analysis of 21 Twins Studies**

- Transvaginal cervical length ≤20 mm at 20-24 wk
  - performed best as predictor of spontaneous preterm birth in asymtopic women with twins

<table>
<thead>
<tr>
<th>Spontaneous Preterm Birth</th>
<th>Sensitivity</th>
<th>Specificity</th>
<th>Positive LR</th>
<th>Negative LR</th>
</tr>
</thead>
<tbody>
<tr>
<td>28 weeks</td>
<td>35%</td>
<td>93%</td>
<td>5.2</td>
<td>0.69</td>
</tr>
<tr>
<td>32 weeks</td>
<td>39%</td>
<td>96%</td>
<td>10.1</td>
<td>0.64</td>
</tr>
<tr>
<td>34 weeks</td>
<td>29%</td>
<td>97%</td>
<td>9.0</td>
<td>0.74</td>
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</tbody>
</table>

- Cervical length >35 mm at 20-24 wk
  - high likelihood of delivery ≥34 wk, PPV >95%

Conde-Agudelo et al., 2010
Cervical Length

- Baseline assessment and serial assessments for patients at risk
  - All twin pregnancies
- Optimal cervical length threshold and frequency of follow-up assessments uncertain
  - ≤20 mm, two week intervals
- Management of patient with twins and a short cervix remains controversial
  - May be role for vaginal PG

Importance of Twin Growth

- Diagnosis of twin discordance
- Detection of intrauterine fetal growth restriction
- Identify cases for increased surveillance
- Twin growth impacts delivery planning

Twin Discordance

Discordance = \( \frac{\text{EFW of larger twin} - \text{EFW of smaller twin}}{\text{EFW of larger twin}} \)

- 20-25% discordance considered to be significant
- Disparate abdominal circumferences early sonographic sign
- Increased discordance associated with increased risk of fetal and perinatal death compared to concordant twins

Causes of Discordant Growth

- Structural anomalies
- Chromosomal abnormalities
- Genetic syndromes
- Discordant congenital infection
- Unfavorable placental implantation
- Unfavorable cord insertion site
- Placental abruption
- Complications of monochorionic placentaion

Potential Complications of Monochorionic Twins

- Monoamniotic twins
- Conjoined twins
- Twin reversed arterial perfusion (TRAP) sequence
- Twin-twin transfusion syndrome (TTTS)
- Unequal placental sharing (UPS)
  - Discordant twin growth
  - Selective intrauterine growth restriction (sIUGR)
- Twin anemia-polycythemia sequence (TAPS)
- Single twin demise in the second or third trimester

Monoamniotic Twins

- 1% of all monozygotic twins
- Results from cleavage at 8-13 days
**Monoamniotic Twins**

**How do you make the diagnosis?**
- Lack of separating membrane on serial exams
- Cord entanglement
  - Utilize color Doppler
  - Present in >80% of cases
- Single placenta with two cord insertions
  - Often in very close proximity
- Associated congenital anomalies
  - Present in 10% of cases

**What are your management recommendations?**
- Monthly growth scans
- Hospital admission at 24-28 weeks
- Serial surveillance
  - BPP
  - NST / continuous EFM
  - Doppler
- CD at 32-34 weeks

*Contemporary management has increased survival from 50-60% to over 90%*

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**Conjoined Twins**

**How do you make the diagnosis?**
- Monoamniotic placentation
- Same relative positions of twins to each other in all views
- Direct opposition of the twins
- Extreme extension of the fetal spines
- Shared organs, vascular connections, associated anomalies

**Management recommendations?**

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**TRAP Sequence**

**What is twin reversed arterial perfusion sequence?**
- Complication of monochorionic twins
  - Prevalence: 1:100 monochorionic twins
  - 75% diamniotic, 25% monoamniotic
- Aberrant arterioarterial anastomosis between twins
  - Acardiac twin lacks direct placental perfusion, dependent on retrograde flow from pump twin
  - Leads to abnormal development of acardiac twin

**How it is diagnosed?**
- Abnormal early development of one twin of MC pair
- Acardius acephalus most common
- Paradoxical arterial flow towards the acardiac twin on pulsed Doppler
**TRAP Sequence**

**What do you recommend?**

- Pump twin at risk for anomalies (5-10%), aneuploidy (10%), 2VC (65%), and hemodynamic compromise (30%)
- Ratio of acardiac/pump twin >0.7 increases risk for cardiac failure (30%), polyhydramnios (50%), and PTD (90%)

Estimate size of acardiac twin:
- EFW (g) = length x width x height x 0.52
- EFW (g) = (1.66 x length) + (1.21 x length^2)

Lee et al (NAFTNet), 2013; Jellin et al, 2010; Oliver et al, 2013; Simpson 2014

What do you recommend?

- Pump twin at risk for anomalies (5-10%), aneuploidy (10%), 2VC (65%), and hemodynamic compromise of pump twin
- Ratio of acardiac/pump twin >0.7 increases risk for cardiac failure (30%), polyhydramnios (50%), and PTD (90%)

Consider invasive cord occlusion therapy when . . .

- high acardiac-to-pump ratio (≥50%)
- rapid growth of acardiac twin
- hemodynamic compromise of pump twin

Variety of techniques available

- bipolar coagulation, radiofrequency ablation (RFA) most popular
- RFA targets intrafetal cord insertion within acardiac twin
- survival 80-90%, mean GA at delivery 34-36 weeks

**Twin-Twin Transfusion Syndrome**

- Complicates 8-10% of monochorionic diamniotic twin gestations
- Untreated TTTS developing before the third trimester has a perinatal mortality rate of >70%
  - 15-50% risk of handicap in survivors

**What it is?**

- Intertwin transfusion
  - Unequal sharing of blood
  - Changes in regional blood flow
  - Alterations in cardiac function
- Due to presence of vascular anastomoses in single placenta
  - 80-100% have intertwin anastomoses
  - Superficial bidirectional AA and VV
  - Deep unidirectional AV

**Stage** | **Ultrasound Assessment** | **Criteria**
---|---|---
I | Amniotic fluid | MVP <2 cm in donor sac; MVP >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 arterial diastolic flow, reversed ductus venosus a-wave flow, pulsatile umbilical vein flow
IV | Fetal hydrops | Hydrops in one or both twins
V | Fetal cardiac activity | Fetal demise in one or both twins

Quintero et al, 1999; Simpson et al. ACOG 2011:204:145

What do you recommend?

- Pregnancy termination
  - Early, advanced stage TTTS
- Amnioreduction
  - Beyond 26 weeks
  - Declines fetoscopic laser therapy
- Laser photocoagulation of communicating vessels
  - 18-26 weeks
  - Advanced stage TTTS
- Delivery
  - Late presentation

Simpson et al, 2013; Aitken et al, 2014; Complicates 8-10% of monochorionic diamniotic twin gestations; Untreated TTTS developing before the third trimester has a perinatal mortality rate of >70% - 15-50% risk of handicap in survivors
Unequal Placental Sharing

What it is?
Pathologic discordance in territorial share of the common placenta in monochorionic twins

- Discordant twin growth
  - 20-25% discordance significant
- Selective IUGR
  - EFW ≤10th percentile of one twin

When should you be suspicious?

- Disparate CRL's and AC's are early sonographic signs of subsequent discordant growth and/or sIUGR in MC twins
- Velamentous PCI is an independent risk factor for UPS

Unequal Placental Sharing

• Abnormal umbilical artery waveforms of sIUGR twin may represent effects of
  - Placental resistance
  - Type and size of intertwin anastomoses
• Leads to substantial clinical differences in apparently similar cases

<table>
<thead>
<tr>
<th>Type</th>
<th>UA Dopplers</th>
<th>Placenta</th>
<th>Intertwin Flow Via Anastomoses</th>
<th>IUFD Risk</th>
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<tbody>
<tr>
<td>I</td>
<td>Positive DF</td>
<td>Small placental territory, many anastomoses</td>
<td>Compensates for small placental share</td>
<td>2-4% (unpredictable)</td>
</tr>
<tr>
<td>II</td>
<td>Persistent AREDF</td>
<td>Smaller placental territory, many anastomoses</td>
<td>Attenuates severity of sIUGR</td>
<td>0-30% (predictable)</td>
</tr>
<tr>
<td>III</td>
<td>Intermittent AREDF</td>
<td>Tiny placental territory, close PCIs, large AAAs</td>
<td>Enables survival of sIUGR twin but potential for acute, massive transfusion</td>
<td>10-20% (unpredictable)</td>
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Fetoscopic Laser Therapy

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<thead>
<tr>
<th>Resolution</th>
<th>Amnioreduction</th>
<th>Laser</th>
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<tr>
<td>Resolution</td>
<td>Resolution</td>
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<td>Stage I</td>
<td>Stage II</td>
<td>Stage III</td>
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<th>Normal Neurologic Outcome at 6 months and 6 years</th>
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<td>Resolution</td>
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• In over 1000 published cases of laser performed for TTTS from six different centers, there were:
  - Two survivors: 50%
  - Single survivor: 30%
  - No survivors: 20%
• Normal neurologic development at 2 years of age: 80-90%

Simpson et al. ACOG 2011:S44:145

Gratacos et al, 2007
Gratacos et al, 2007
Gratacos et al, 2007
**UPS with sIUGR**

**What are your management considerations?**

- Spontaneous demise of MCDA twin carries 10% risk of death of co-twin, 20% risk of neurologic injury
  - Acute anemia due to massive blood transfer from surviving twin into dead twin/placenta
- Presence of placental anastomoses may be protective for sIUGR twin – compensatory flow from its co-twin
- Management strategy remains a challenge!
  - Influenced by severity of sIUGR, coexisting TTTS, gestational age, parental decisions, technical issues

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**TAPS**

**What it is?**

- Twin anemia polycythemia sequence
- Chronic form of fetofetal transfusion

**How is it diagnosed?**

- Elevated PSV-MCA in one twin = anemia
- Decreased PSV-MCA in co-twin = polycythemia

**Stage**

<table>
<thead>
<tr>
<th>Stage</th>
<th>Criteria</th>
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</thead>
</table>
| 1     | PSV-MCA >1.5 MoM in donor  
PSV-MCA <1.0 MoM in recipient |
| 2     | PSV-MCA >1.7 MoM in donor  
PSV-MCA <0.8 MoM in recipient |
| 3     | Stage 1 or 2 with cardiac compromise of donor |
| 4     | Hydrops of donor |
| 5     | Single or double IUFD |

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**TAPS**

**What are your management recommendations?**

- Depends on gestational age, technical considerations, disease severity

**Options**

- Repeat laser: technical difficulties
- Fetal transfusion: not curative
- Cord coagulation
- Expectant management
- Early delivery

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**UPS versus TTTS**

**UPS with sIUGR**

**TTTS laser case**
Single MC Twin Demise

Retrospective cohort analysis of 1000 consecutive twins ≥24 weeks

- 804 DCDA − 1.1% stillbirth
- 198 MCDA − 3.6% stillbirth

Analysis of 151 normal MCDA twins ≥24 weeks

All demises within 2 weeks of a normal scan

Overall risk of late fetal death in uncomplicated MCDA twins:

- 4.6% per pregnancy
- 3.3% per fetus

Single MC Twin Demise: Acquired CNS Injury

Analysis of 151 normal MCDA twins ≥24 weeks

All demises within 2 weeks of a normal scan

Overall risk of late fetal death in uncomplicated MCDA twins:

- 4.6% per pregnancy
- 3.3% per fetus

- For single death in MC pair, 5 times more likely to have neurologic morbidity

Use of Ultrasound in Twins: Summary

Timing | Examination
--- | ---
1st trimester | Determination of chorionicity
11-13 weeks | Measurement of crown rump lengths for dating
16-26 weeks | Measurement of nuchal translucency for screening
18-20 weeks | Screen for structural, placenta and PCI abnormalities
20-22 weeks | Baseline cervical length
24 weeks to delivery | Fetal echocardiography for IVF and MC twins
24 weeks to delivery | Serial determination of twin growth and discordance
24 weeks to delivery | Assessment of cervical length as indicated
24 weeks to delivery | Antenatal fetal testing as indicated
24 weeks to delivery | Fetal presentation prior to delivery
24 weeks to delivery | Delivery of second twin

Take Home Message

In order to provide high-quality obstetric care of twin pregnancies, need to

- Diagnose early
- Counsel extensively
- Follow closely
- Manage using best available up-to-date evidence

which requires extensive use of ultrasound