Complications of Monochorionic Twins

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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
Monoamniotic Twins

- Forked cord
- Abnormal vessel number
Monoamniotic Twins

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%
Conjoined Twins

- Rare event
- Results from cleavage at 13-15 days
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?
What is twin reversed arterial perfusion sequence?

• Complication of monochorionic twins
  - Prevalence: 1:100 monochorionic twins
  - 75% diamniotic, 25% monoamniotic

• Vascular disruption during early embryogenesis
  - Results in aberrant arterioarterial anastomoses between twins
  - Acardiac twin lacks direct placental perfusion
TRAP Sequence

Acardiac twin is dependent on retrograde arterial supply of relatively deoxygenated blood from its co-pump twin

- Asymmetric circulation leads to abnormal development of acardiac twin, favors growth of caudal aspects

- Differential diagnosis
  - Demise of one twin
  - Discordant twin growth
  - Cystic hygroma
  - Acrania / anencephaly
  - Placental teratoma
TRAP Sequence

How it is diagnosed?

• Abnormal early development of one twin of monochorionic pair

• Acardius acephalus most common
  - Well-formed fetal pelvis and lower limbs
  - Absent head/skull and usually without thoracic organs and arms
TRAP Sequence

- Acardiac twin lacks a normal heart but may have a rudimentary structure with pulsations
- Paradoxical arterial flow towards the acardiac twin from pump twin
What do you recommend?

Pump twin is at risk for:
- anomalies (5-10%)
- aneuploidy (10%)
- two vessel cord (65%)
- hemodynamic compromise (30%)
  - cardiac dysfunction, cardiomegaly, hydrops

Simpson, Decision Support in Medicine, 2014
**TRAP Sequence**

**Size matters!**

- Ratio of acardiac/pump twin >0.5-0.7 increases risk for:
  - cardiac failure (30%)
  - polyhydramnios (50%)
  - preterm delivery (90%)

Estimate size of acardiac twin:

- $\text{EFW (g)} = \text{length} \times \text{width} \times \text{height} \times 0.52$
- $\text{EFW (g)} = (-1.66 \times \text{length}) + (1.21 \times \text{length}^2)$

*Lee et al (NAFTNet), 2013; Jellin et al, 2010; Oliver et al, 2013*
TRAP Sequence

Consider invasive cord occlusion therapy when . . .

- High acardiac-to-pump ratio (≥50%)
- Rapid growth of acardiac twin
- Hemodynamic compromise of pump twin

Lee et al (NAFTNet), 2013
Aitken et al, 2014

- 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

Simpson et al. ACOG 2011:204:145
Twin-Twin Transfusion Syndrome

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
# Twin-Twin Transfusion Syndrome

<table>
<thead>
<tr>
<th>Stage</th>
<th>Ultrasound Assessment</th>
<th>Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Amniotic fluid</td>
<td>MVP &lt;2 cm in donor sac; MVP &gt;8 cm in recipient sac</td>
</tr>
<tr>
<td>II</td>
<td>Fetal bladder</td>
<td>Nonvisualization of fetal bladder in donor twin over 60 minutes of observation</td>
</tr>
<tr>
<td>III</td>
<td>Doppler studies</td>
<td>Absent or reversed umbilical artery diastolic flow, reversed ductus venous a-wave flow, pulsatile umbilical vein flow</td>
</tr>
<tr>
<td>IV</td>
<td>Fetal hydrops</td>
<td>Hydrops in one or both twins</td>
</tr>
<tr>
<td>V</td>
<td>Fetal cardiac activity</td>
<td>Fetal demise in one or both twins</td>
</tr>
</tbody>
</table>

*Quintero et al, 1999*
*Simpson et al. ACOG 2011:204:145*
Twin-Twin Transfusion Syndrome

What do you recommend?

- Pregnancy termination
  - Early, advanced stage TTTS

- Amnioreduction
  - Beyond 26 weeks
  - Declines fetoscopic laser therapy
  - Fetoscopic laser therapy unavailable

- Laser photocoagulation of communicating vessels
  - 18-26 weeks
  - Advanced stage TTTS

- Delivery
  - Late presentation
<table>
<thead>
<tr>
<th>TTTS Outcomes</th>
<th>Stage I</th>
<th>Stage II</th>
<th>Stage III</th>
<th>Stage IV</th>
<th>Normal Neurologic Outcome at 6 months and 6 years</th>
</tr>
</thead>
<tbody>
<tr>
<td>Resolution</td>
<td>27%</td>
<td>10%</td>
<td>1%</td>
<td>–</td>
<td></td>
</tr>
<tr>
<td>Amnioreduction</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>31%     70%</td>
</tr>
<tr>
<td>At least one survivor</td>
<td>90%</td>
<td>80%</td>
<td>48%</td>
<td>26%</td>
<td></td>
</tr>
<tr>
<td>Two survivors</td>
<td>50%</td>
<td>48%</td>
<td>20%</td>
<td>6%</td>
<td></td>
</tr>
<tr>
<td>Laser</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>52%     82%</td>
</tr>
<tr>
<td>At least one survivor</td>
<td>86%</td>
<td>84%</td>
<td>78%</td>
<td>62%</td>
<td></td>
</tr>
<tr>
<td>Two survivors</td>
<td>92%</td>
<td>82%</td>
<td>66%</td>
<td>52%</td>
<td></td>
</tr>
</tbody>
</table>


- 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 2011:204:145
Fetoscopic Laser Therapy
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 ≤10\textsuperscript{th} percentile of one twin
Unequal Placental Sharing

When should you be suspicious?

Disparate CRL’s and AC’s early sonographic signs of subsequent discordant growth and/or sIUGR in MC twins
Unequal Placental Sharing

- Development of sIUGR in monochorionic twins may be function of
  - Unequal placental sharing / placental insufficiency
  - Intertwin anastomoses and intertwin flow patterns

- 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
Unequal Placental Sharing

How it is diagnosed?

<table>
<thead>
<tr>
<th>Feature</th>
<th>UPS</th>
<th>TTTS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Growth discordance ≥20%</td>
<td>++++</td>
<td>++</td>
</tr>
<tr>
<td>EFW ≤10&lt;sup&gt;th&lt;/sup&gt; %ile</td>
<td>++++</td>
<td>++</td>
</tr>
<tr>
<td>Cardiac dysfunction</td>
<td>Smaller sIUGR twin</td>
<td>Larger recipient twin</td>
</tr>
</tbody>
</table>
| Amniotic fluid discrepancy           | Oligohydramnios in IUGR twin  
Normal fluid in AGA twin               | Oligohydramnios in one sac (MVP ≤2 cm)  
Polyhydramnios in other sac (MVP ≥8 cm) |

- Must have oligohydramnios-polyhydramnios sequence for diagnosis of TTTS
- Must have growth discordance +/- sIUGR for diagnosis of UPS
UPS versus TTTS

UPS with sIUGR

TTTS laser case
# UPS with sIUGR Staging

<table>
<thead>
<tr>
<th>Type</th>
<th>UA Dopplers</th>
<th>Placenta</th>
<th>Intertwin Flow Via Anastomoses</th>
<th>IUFD Risk</th>
</tr>
</thead>
<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) Decrease risk with serial surveillance/ early delivery</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) Unstable hemodynamics due to large AAAs</td>
</tr>
</tbody>
</table>

Gratacos et al, 2007
Valsky et al, 2010
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, gestational age, parental decisions, technical issues
UPS with sIUGR

Role for cord occlusion?

- Option in select cases to prevent death/injury of co-twin
  - Early detection of type II or III
  - High risk of spontaneous demise
    - *Cardiac dysfunction*
    - *Abnormal venous Dopplers*
    - *Associated anomalies*
- Prognosis related to GA at reduction – lower survival rate if performed <18 weeks
- Survival >80%, normal neurologic outcome in >90%

Role for laser?

- Higher obstetric risks
- Technical challenges
  - Lack of polyhydramnios
  - PCIs in close proximity (type III)
  - Large, diameter AAAs (type III)
- Substantial risk for demise of sIUGR twin anyway (65-75%)
- May be reasonable when selective feticide not an option
  - Gestational age, availability, parental decision

*Parra-Cordero et al, 2015; Chalouhi et al, 2013; Valsky et al, 2010; Gratacos et al, 2008; Quintero et al, 2001*
**UPS with Type III sIUGR**

<table>
<thead>
<tr>
<th></th>
<th>No Laser N=31</th>
<th>Laser N=18</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>GA diagnosis</td>
<td>22.0 wk</td>
<td>22.0 wk</td>
<td>0.21</td>
</tr>
<tr>
<td>GA delivery</td>
<td>31.0 wk</td>
<td>32.6 wk</td>
<td>0.32</td>
</tr>
<tr>
<td>Discordance</td>
<td>33%</td>
<td>38%</td>
<td>0.22</td>
</tr>
<tr>
<td>IUFD overall</td>
<td>14.5%</td>
<td>36.1%</td>
<td>0.02</td>
</tr>
<tr>
<td>IUFD smaller twin</td>
<td>19.4%</td>
<td>66.7%</td>
<td>0.001</td>
</tr>
<tr>
<td>Both alive</td>
<td><strong>80.6%</strong></td>
<td>27.8%</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

*Gratacos et al, 2008*

*Laser therapy for sIUGR needs further investigation*
TAPS

What it is?

• Twin anemia polycythemia sequence
• Chronic form of fetofetal transfusion
**TAPS**

**Spontaneous** (3-5% of MC twins)

**Iatrogenic** (10-15% post-laser)
- Residual anastomoses in 5-30% of laser cases
- Solomon technique reduces risk

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*Slaghekke et al, 2010, 2014*
*Lopriore et al, 2009, 2010*
TAPS

How is it diagnosed?

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

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

Lopriore et al, 2009, 2010
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
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

Lee et al, 2008
Barigye et al, 2005
Single MC Twin Demise: Acquired CNS Injury

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

<table>
<thead>
<tr>
<th>Chorionicity</th>
<th>Co-twin death after single demise</th>
<th>Abnormal CNS imaging in surviving co-twin</th>
<th>Neurodevelopmental impairment in surviving co-twin</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dichorionic</td>
<td>3%</td>
<td>16%</td>
<td>2%</td>
</tr>
<tr>
<td>Monochorionic</td>
<td>15%</td>
<td>34%</td>
<td>26%</td>
</tr>
</tbody>
</table>

Griffiths et al, 2015; Hillman et al, 2011
Take Home Message

Never take your eyes off MC twins