FETAL ECHO IN TWIN PREGNANCY: MONOCHORIONIC TWINS

DELHI CHILD HEART CENTER & INDRAPRASTHA APOLLO HOSPITAL
NEW DELHI
Scope of this talk

- Twin to Twin Transfusion
- TRAP Sequence
- Congenital Heart Defects in Twins
- Heart Failure and Function Assessment

NOT INCLUDED

- Arrhythmia in Twins
- Conjoined Twins
Scope of Cardiac Assessment in Twins

• Assessment of
  – Normal function
  – Normal Heart physiology
  – Detection of Structural Abnormality
  – Follow up of Functional abnormalities
  – Monitoring and FU of invasive procedures
  – Issues specific to Twins
SPECIFIC TO MONOCH TWINS

• Evaluate Twin to Twin Transfusion
• Evaluate Cardiac Defects
• Evaluate TRAP Sequence
• Cardiac Pathology in conjoined Twins
TIMING OF FETAL ECHO IN TWINS

• Done at 18 weeks
• Visualization optimal
• Calcification does not help at 30 weeks
• Abd fat/Oligohydramnios:
  – make exam less reliable
  – Inaccurate
  – time consuming
  – difficult
Dizygotic (Dichorionic, Diamniotic)
Monozygotic
(Monochorionic, Monoamniotic)
Discordant Heart Anomalies in Twins

IRENE A. UCHIDA AND RICHARD D. ROWE
The Departments of Zoology and Pediatrics, University of Toronto, and The Research Institute of The Hospital for Sick Children, Toronto, Canada

<table>
<thead>
<tr>
<th>Cardiac Malformation</th>
<th>Monozygotic</th>
<th>Dizygotic</th>
<th>Totals</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Cyanotic</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>2</td>
<td>3</td>
<td>5</td>
</tr>
<tr>
<td>Transposition of great vessels</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Tricuspid atresia</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Aortic atresia</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Dextrocardia with pulmonic stenosis</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td><strong>Subtotal</strong></td>
<td>3</td>
<td>6</td>
<td>9</td>
</tr>
<tr>
<td><strong>Non-cyanotic</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>5</td>
<td>2</td>
<td>7</td>
</tr>
<tr>
<td>Auricular septal defect</td>
<td>2</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Pulmonic stenosis with normal aortic root</td>
<td>2</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Post-ductal coarctation of the aorta</td>
<td>0</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Primary endocardial fibroelastosis</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td><strong>Subtotal</strong></td>
<td>10</td>
<td>7</td>
<td>17</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>13</td>
<td>13</td>
<td>26</td>
</tr>
</tbody>
</table>
A study to determine the incidence of structural congenital heart disease in monochorionic twins

PRENATAL DIAGNOSIS 2006

- Risk of at least one of a MC twin pair having a structural congenital cardiac anomaly was 9.1% (15/165)
- MC/DA twins, this figure was 7.0% (11/158)
- MC/MA twins the risk for at least one affected twin was 57.1% (4/7).
- If one of a pair of MC twins was affected, the risk to the other twin for a structural cardiac anomaly was 26.7% (4/15).
TWIN TO TWIN TRANSFUSION SYNDROME
TTTS

- Only in Monochorionic Diamniotic Twins
- In 1/3 of all Monozygotic twins
- 15-20% of all MCh DiA twins have unbalanced blood flow
Quintero’s Staging

- **Stage I:** This is the initial way that TTTS is seen on ultrasound; oligohydramnios in the donor’s sac; an MVP of 2 cm and polyhydramnios in the recipient’s sac with a MVP of fluid of 8 cm. The bladder of the donor baby is still seen.
- **Stage II:** Polyhydramnios and oligohydramnios; bladder is no longer seen in the donor twin.
- **Stage III:** Blood flow in the umbilical cord and fetal ductus venosus reveals abnormal patterns in Stage III. These patterns can occur in either or both fetuses.
  - In the umbilical cord, the diastolic flow can be either absent or reversed in the umbilical artery. This pattern is usually seen in the donor twin.
- **Stage IV:** One or both babies shows signs of hydrops.
- **Stage V:** One or both babies have died.
Twin to Twin Transfusion

- MonoCh Twins: routine echo at 18 weeks if concordant growth
- If growth dischordant: echo to be performed at that stage
- TTTS results from unbalanced circulation from the same set of twins
- Hypervolemia in recipient and hypovolemia in donor
Hydrops: Etiologies

• Part of pathophysiology is volume shift
• Rest is related to increased resistance related to placental malfunction
• Discordant oncotic pressures due to different protein levels
## Echo in TTTS

<table>
<thead>
<tr>
<th>Index</th>
<th>Donor</th>
<th>Recipient</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heart Size</td>
<td>NI</td>
<td>Increased</td>
</tr>
<tr>
<td>Myocard</td>
<td>NI</td>
<td>Hyperkinetic</td>
</tr>
<tr>
<td>AtrioVentr V</td>
<td>NI</td>
<td>Increased</td>
</tr>
<tr>
<td>Doppler</td>
<td>MCA, UA</td>
<td>IVC, DV, UV</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Fnctn decreased, TR, MR, PE</td>
</tr>
</tbody>
</table>
TTTS- Recipient heart

- Recipient heart develops hypertrophy
- This is due to incr peripheral resistance and afterload
- Endothelin 1 levels increased in recipient
- BNP levels increased in recipient twin when CHF starts
- BNP counteracts Angiotensin II which remodels the heart
TTTS- DONOR HEART

- High Renin Angiotensin protein levels indicating chronic ischemia
- Increase peripheral resistance, placental insufficiency and restrict the growth
- Hypertension in recipient is due to transfer of renin from donor
- Major impact of renin is stimulation of catecholamines, aldosterone and vasoconstr and myoc remodeling
• These result in persistent myocardial load and finally CHF
• Donor heart usually is normal size and lacks any functional abnormality
• Donors have lower cardiac output as compared to recipients
First TM: Recipient Echo

- Increased Nuchal lucency
- Discordant amniotic fluid volumes
- Intertwin membrane folding

SECOND TRIMESTER

- Cardiac first amongst abnormalities noted
- Earliest abnormality of fetal hypervolemia is pulmonary insufficiency
Second Trimester

• The pulmonary insufficiency noted is due to increased pulmonary resistance
• Followed by myocardial hypertrophy and AV Valve regurgitation
• Fetal myoc hypertrophy affects right heart more than left
Second Trimester

- TR is present in 40% patients
- E and A wave fusion for TCV valve Doppler
- Recipients have higher Pulm and Aortic flow velocities
- Cardiomyopathy causes systolic and diastolic dysfunction
- This may result in right heart failure
- This may behave like Pulm Stenosis
- Systemic Hypertension
- Completely reversible after birth
Donor Heart

- Donor heart is normal
- Blood flow velocity across the AV valves is increased
- Hyperdynamic state
- Increased LV SF
- Significantly decreased values of AV valve flow values
Donor Heart

- Hydrops in donor heart is rare
- Cause is not clear
- Maybe:
  - Myocardial Ischemia
  - Dysfunction
  - Redirected blood flow
- Hydrops in donor after laser therapy is not rare: 25% : due to difficulty in accommodating to the higher volume
Third Trimester TTTS

- May occur without significance size discordance
- Manifests with milder symptoms of volume overload in recipient and hypovolemicemia in donor
- Hypovolemia is acute
Post Laser Therapy

• More hemodynamic disturbances maybe seen after laser therapy
Fetal echocardiographic findings are not predictive of death in twin-twin transfusion syndrome

- A cohort of 30 pregnancies with TTTS between 1990 and 2001 was included.
- Fetal echo findings:
  - Cardiomegaly
  - Right ventricular hypertrophy
  - Tricuspid regurgitation were evaluated
  - Approximately 80% power to detect a 2-fold increased risk of fetal death
TTTS
TWIN REVERSED ARTERIAL PERFUSION SEQUENCE
<table>
<thead>
<tr>
<th>Clinical setting</th>
<th>Connections</th>
</tr>
</thead>
<tbody>
<tr>
<td>No complications</td>
<td>no connections; multiple connections, including bidirectional a→vc,</td>
</tr>
<tr>
<td></td>
<td>or a→a plus a few a→vc, with equilibrated bidirectional transfusion</td>
</tr>
<tr>
<td>TTT</td>
<td>severe: single causative a→vc</td>
</tr>
<tr>
<td></td>
<td>mild: few, bidirectional a→vc,</td>
</tr>
<tr>
<td></td>
<td>perhaps with a→a</td>
</tr>
<tr>
<td>TRAP</td>
<td>a→a plus v→v</td>
</tr>
<tr>
<td>Brain-damaged survivor after</td>
<td>a→a, v→v, a→v from survivor to dead fetus</td>
</tr>
<tr>
<td>fetal demise of co-twin</td>
<td></td>
</tr>
</tbody>
</table>

**TTT**, twin-twin transfusion; **TRAP**, twin reversed arterial perfusion; a→vc, arteriovenous connection; a→a, arterioarterial; v→v, venovenous
TRAP Sequence
(Twin-Reversed Arterial Perfusion Sequence)
TRAP PATHOPHYSIOLOGY

MULTIPLE PREGNANCY
TRAP Sequence

- Twin reversed arterial perfusion
- 1% of monochorionic twins
- 1:35,000 pregnancies
- Reversed perfusion is used as blood enters acardic twin via UA and leaves via UV both connected to normal twin
- Twin with heart is referred to as PUMP twin
- Untreated, death in 50-70% pump twins
TRAP SEQUENCE

- 9% pump twin have chromosomal abnormality
- 51% have polyhydramnios
- 75% have premature labour
- Ratio of weight between >50% between acardic and pump twin predicts nearly 100% mortality
Twin-reversed arterial perfusion sequence: pre- and postoperative cardiovascular findings in the 'pump' twin


- 27 pregnancies, 4 yrs, retrospective
- Elevated indexed cardiac output noted with cardiac involvement
- Normalization post therapy noted
- RV dysfunction noted in these
FETAL CHF, MYOCARDIAL FUNCTION
CHF: Fetal Echo Assessment

• Cardiac Output Assessment
• Shortening Fraction
• Ejection Fraction
• Tei Index
• Ventricular Ejection Force
• Atrioventricular Flow
• Tissue Doppler
Cardiac Output Assessment

• Cardiac Output = Stroke Volume x HR
• Stroke Volume Assessment
  – Aortic/Pulm Valve area X Doppler VTI
  – M Mode End Diastolic Volume and Systolic Volume
  – 2-D Method using Disc diameters
  – 3-D Volume based STIC
M-MODE

A

B

LV

IVS

RV

EDD

ESD

LV

IVS

RV
DISC METHOD
Ejection & Shortening fraction

- **Ejection Fraction**
  - This has been abandoned in Fetal Medicine
  - Dependent on presumptions & formulae

- **Shortening Fraction**
  - Extensively used
  - Single point measurement
  - As helpful as “eyeballing”
Tei Index: Myocardial Performance Index

• Indirect reflection of systolic cardiac function + Diastolic Function

• ISOVOLUMIC CONTR T + ISOVOLUM RELAXATION T/EJECTION TIME
Fetal Echo Markers of CHF

Cardiomegaly: heart area/chest area ratio > 0.45
Heart circumference/chest circumference ratio > 0.55
Atrial enlargement
Holosystolic tricuspid valve regurgitation
Slow upstroke for tricuspid valve regurgitation (dP/dt)
Trivial and holosystolic mitral valve regurgitation
Pulmonary and aortic regurgitation
Decreased shortening fraction of right ventricle, left ventricle or both (normal values 28–40%)
Myocardial hypertrophy (wall thickness > 4 mm, sign of fetal hypertension)
Abnormal A/E ratio for mitral and/or tricuspid valve (in compromised fetus E = A, or E > A)
Dilatation of the inferior vena cava > 5 mm
Dilatation of the hepatic vein
Hepatomegaly
Reversal flow in ductus venosus
Abnormal pulsation in the inferior vena cava (A/S ratio > 0.15)
Pulsation in the umbilical vein
Ascites, pericardial effusion or hydrothorax
Polyhydramnios
Placentomegaly

P, pressure; t, time
Acknowledgements