No Financial Disclosures

• I have no financial relationship with any manufacturer of any commercial product and/or provider of commercial services discussed in the conference.

• I do not intend to discuss an unapproved/investigative use of a commercial product or device in my presentation.
Arrhythmia Evaluation

• Rate
  – Fast or slow?

• Rhythm
  – Regular or irregular?
  – Atrial rhythm or ventricular rhythm?
Normal Conduction

- Sinoatrial (SA) node
- Atrioventricular (AV) node

RA = Right atrium
RV = Right ventricle
LA = Left atrium
LV = Left ventricle
Fetal Arrhythmia Background

- Complicates ~2% of all pregnancies
- Account for 10-20% of referrals to pediatric cardiologist
- Abnormal impulse generation or propagation
- Various techniques
  - Fetal ECG
  - Magnetocardiography (fMCG)

2:1 AV Block
Fetal Arrhythmia Background

• Complicates ~2% of all pregnancies
• Account for 10-20% of referrals to pediatric cardiologist
• Abnormal impulse generation or propagation
• Techniques
  – Fetal ECG
  – Magnetocardiography (fMCG)
  – Echocardiography

Assess with M-mode or spectral Doppler
Color and tissue Doppler may be helpful
M-Mode Assessment

- Place cursor across ventricular and atrial wall

1:1 AV Conduction
Color M-Mode Assessment

FR 6Hz
7.0cm

2D / MM
63% 63%
C 58
P Off
HRes

CF
77%
3.0MHz
WF High
Med

Atrial

Ventricular

M3 M4
+66.7

-51.9 cm/s

100mm/s
Spectral Doppler Assessment

• E = early passive ventricular inflow from atrium

• A = active atrial systolic contraction stimulated by SA node, mechanical event correlating with atrial electrical stimulation
Spectral Doppler Assessment

MV-LVOT
Spectral Doppler Assessment

SVC-Aorta
Tissue Doppler Imaging

- Measurement of myocardial velocity and timing
<table>
<thead>
<tr>
<th>Too Fast</th>
<th>Irregular</th>
<th>Too Slow</th>
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<tr>
<td>Sinus tachycardia</td>
<td>Premature atrial contractions (conducted, blocked)</td>
<td>Sinus bradycardia</td>
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<td>Supraventricular Tachycardia – Re-entrant</td>
<td>Atrial bigeminy (conducted)</td>
<td>Atrial bigeminy (blocked)</td>
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<tr>
<td>Atrial Flutter</td>
<td>Atrial trigeminy (conducted, blocked)</td>
<td>2:1 AV Block</td>
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<td>Junctional tachycardia</td>
<td>Premature ventricular contractions (conducted, blocked)</td>
<td>Third degree AV Block</td>
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<tr>
<td>Ventricular tachycardia</td>
<td>2\textsuperscript{nd} Degree AV block (Wenkebach)</td>
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# Irregular Rhythm

<table>
<thead>
<tr>
<th></th>
<th>Premature Atrial Contraction (blocked)</th>
<th>Premature Atrial Contraction (conducted)</th>
<th>Atrial Bigeminy (conducted)</th>
<th>Atrial Trigeminy (blocked)</th>
<th>2nd Degree AV Block Wenkebach</th>
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<td>Atrial Rate</td>
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<tr>
<td>A-A Interval</td>
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<td>regularly irregular</td>
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<td>regular</td>
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<td>A-V Relation</td>
<td>&gt;1:1</td>
<td>1:1</td>
<td>2:1</td>
<td>3:2</td>
<td>&gt;1:1</td>
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<td>Incidence</td>
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<td>minor, transient</td>
<td>minor, transient</td>
<td>minor, transient</td>
<td>minor, transient</td>
<td>may progress</td>
</tr>
</tbody>
</table>

E Jaeggi, Fetal Cardiology 2nd ed.
Premature Atrial Contraction - Conducted

- Isolated benign event
- Most common rhythm disturbance
- Causes
  - Maternal caffeine/medications
  - Floppy atrial septum
- 1-3% risk of SVT

Drawings courtesy of Dr. Edgar Jaeggi
Atrial Bigeminy - Conducted
Atrial Bigeminy - Conducted

19w3d

A PAC A PAC

V V V V

100mm/s
Atrial Trigeminy - Blocked
<table>
<thead>
<tr>
<th></th>
<th>Sinus</th>
<th>Blocked Atrial Bigeminy</th>
<th>2:1 AV Block</th>
<th>Third Degree AV Block</th>
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</thead>
<tbody>
<tr>
<td>Atrial Rate</td>
<td>75-90</td>
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<td>slow-normal</td>
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<tr>
<td>A-A Interval</td>
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<td>A-V Relationship</td>
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<td>Ventricular Rate</td>
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<td>V-V Interval</td>
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<tr>
<td>V-A Interval</td>
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<tr>
<td>Incidence</td>
<td>+</td>
<td>+</td>
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<tr>
<td>Relevance</td>
<td>depends on cause</td>
<td>minor, transient</td>
<td>major, may progress</td>
<td>major, irreversible</td>
</tr>
</tbody>
</table>

E Jaeggi, Fetal Cardiology 2nd ed.
Definition of Bradycardia

Classic definition of bradycardia => 110 bpm

Mitchell et al. Circulation 2012
Blocked Atrial Bigeminy

- A>V, slow
- BAB has “long-short-long” pattern
- Self-limited and benign
- May progress to SVT (0.5-3% of the time)
2:1 AV Block

• AV associated then non-conducted
• Regular A-A interval

Courtesy of Dr. Moon-Grady
Blocked Atrial Bigeminy vs 2:1 AV Block

- Assess for signs of immune mediated heart block
  - Take a thorough history
  - Effusions, AV valve regurgitation, endocardial fibroelastosis (more common with 2:1 AV block)
  - Draw SSA & SSB antibodies

- BAB is slower than sinus bradycardia but faster ventricular rate than 2:1 AV block
- BAB more irregular than 2:1 AV block
- BAB has multiple rhythms (sinus, isolated PAC, SVT...)
- Repeat evaluation with close follow up

Wiggins et al. Heart Rhythm 2013
Long QT Syndrome

- Prolonged repolarization creates prolonged QT interval
  - Abnormal potassium and sodium channels

- Family history
  - Autosomal dominant (50% recurrence risk)
  - Cardiac arrest or sudden death spontaneously or during loud noises, swimming, or while sleeping
  - Near drowning, seizures, syncope
Long QT Syndrome

- Prevalence: 1 in 2000 people
- Multiple genetic mutations identified in subtypes (cord blood testing available)
  - KCNQ1, KCNH2, SCN5A...
- Fetal rhythm: sinus bradycardia, 2:1 AV block, ventricular tachycardia
Complete Heart Block Background

• Prevalence – 1 in 20,000 pregnancies

• Etiology
  – Immune mediated
    • SSA and SSB antibodies
    • Maternal SLE, Sjogren’s, Rheumatoid Arthritis
  – Structural heart disease
    • Heterotaxy syndrome (left atrial isomerism)
    • Congenitally corrected TGA
  – Long QT “blocked” rhythm
Immune Mediated Heart Block

• Transplacental transfer of SSA and SSB antibodies
  – Mainly in second trimester

• Anti-SSA/SSB
  – 2% incidence of AV block
  – 16% incidence of AVB with affected prior sibling

• Bind to AV/SA node, Purkinje fibers, or myocardium
  – myocyte inflammation, apoptosis, and scar formation

• May cause cardiomyopathy
Immune Mediated - Third Degree AV Block

- $A>V$, slow, regular
- Patchy endocardial fibroelastosis
Third Degree AV Block

Elevated S/D ratio
Case of Fetal Bradycardia

- Presented @ 21 wks to outside hospital
- “go home and let nature take its course”
- 2\textsuperscript{nd} opinion in Phoenix
- FHx Lupus (sister), pt asymptomatic
- SSA 164 IU/mL; SSB neg
Immune Mediated - CHB

- Rx dexamethasone, terbutaline
- IVIG 70g x4 doses during pregnancy
- Delivered @ 33 wks; BW 3lbs3oz
- IVIG after delivery
- Temporary pacemaker > permanent
- Surgical ASD closure @ 4 1/2 years
  and doing great!

Hornberger et al. JACC 2011.
CHB – Prenatal Treatment

- Dexamethasone – ongoing inflammation
  - 8 mg/day
  - After two weeks, 4 mg/day
  - After 28 weeks, 2 mg/day and continue to birth
- β-sympathomimetics – HR <50 bpm
- IVIG – decrease inflammation
- Plasmapheresis – decrease antibody load
- Delivery at 24-30 weeks with early initiation of isuprel and external pacing
- Hydroxychloroquine reduces AV block/EFE by 64%

Immune Mediated Process – Key Points

- May be associated with sinus node dysfunction
- SSA titers may be helpful (15% incidence; ≥ 50 U/mL and 85% incidence; ≥ 85 U/mL Jaeggi 2011)
- Complete heart block is irreversible
  - Steroids used to prevent associated cardiomyopathy
- Weekly mechanical AV interval monitoring NOT effective (PRIDE study Friedman 2008)
- Prospective study of ability of home monitoring to predict AV block (Cuneo et al)
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<tr>
<th>Case</th>
<th>Year</th>
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<th>A rate (bpm)</th>
<th>V rate (bpm)</th>
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<td>16*</td>
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</table>
CAVB & CCTGA

- A-V and V-A discordance
- Rare: 0.5% of CHD
- Associated with VSD, LVOTO, Ebstein’s TV
- 40% CHB in 20 years
  - Anterior-superior displacement of the AV node

CHB & CCTGA

Normal 3 VV
CHB & Left Atrial Isomerism

- Absence/dysfunction of SA Node
  - When CHB present, only 1/18 survived at 1 year
CHB & Left Atrial Isomerism

- Azygous Vein
- Aorta
- Coronary sinus
LAI – Interrupted IVC/Azygous Vein

LSVC

Azygous Vein

Aorta
# Tachyarrhythmias

<table>
<thead>
<tr>
<th></th>
<th>Sinus</th>
<th>SVT (re-entry)</th>
<th>Atrial Flutter</th>
<th>JET/PJRT</th>
<th>Vent Tachy (+VA block)</th>
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</thead>
<tbody>
<tr>
<td><strong>Atrial Rate</strong></td>
<td>160-200</td>
<td>190-280</td>
<td>300-500</td>
<td>180-230</td>
<td>slow-normal</td>
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<td><strong>A-A Interval</strong></td>
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<td>regular</td>
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<tr>
<td><strong>A-V Relation</strong></td>
<td>1:1</td>
<td>1:1</td>
<td>primarily 2:1</td>
<td>1:1</td>
<td>&lt;1:1</td>
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<tr>
<td><strong>Ventricular Rate</strong></td>
<td>160-200</td>
<td>190-280</td>
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<td>180-230</td>
<td>160-260</td>
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<tr>
<td><strong>V-V Interval</strong></td>
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<td>regular</td>
<td>mainly regular</td>
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<td>regularly irregular</td>
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<td><strong>V-A Interval</strong></td>
<td>long VA</td>
<td>short VA</td>
<td>--</td>
<td>long VA</td>
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<tr>
<td><strong>Incidence</strong></td>
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<tr>
<td><strong>Relevance</strong></td>
<td>depends on cause</td>
<td>major, treatable</td>
<td>major, treatable</td>
<td>major, treatable</td>
<td>major, treatable</td>
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</tbody>
</table>

E Jaeggi, Fetal Cardiology 2nd ed.
Fetal Tachycardias

A. Sinus tachycardia
   - Atrial
   - AV conduction
   - Ventricular
   - Echo and ECG:
     - A-V ratio: 1:1, long VA
     - V-V: regular, same as atrial rate

B. AV re-entrant tachycardia
   - Atrial
   - AV conduction
   - Ventricular
   - Echo and ECG:
     - A-A: regular, 190 – 300 bpm
     - A-V: 1:1, short VA: stops with AV block
     - V-V: regular, same as atrial rate

C. Atrial flutter
   - Atrial
   - AV conduction
   - Ventricular
   - Echo and ECG:
     - A-A: regular, 300 – 500 bpm
     - A-V ratio: mainly 2:1 (1:1 – 4:1)
     - V-V: regular, 150–250 bpm

D. Permanent junctional reciprocating tachycardia
   - Atrial
   - AV conduction
   - Ventricular
   - Echo and ECG:
     - A-V ratio: 1:1, long VA (like AET)
     - V-V: regular, 180–220 bpm
SVT - Mechanism

- Immature myocardium
  - Persistence of accessory pathways
  - Macroreentry circuit

- Associations
  - Ebstein’s anomaly, rhabdomyomas, or myocarditis
Supraventricular Tachycardia

- FHR 190-280 bpm
- 1:1 A-V conduction, Short VA accessory pathway (orthodromic-down slow AVN and up fast APW)
- Sudden onset/termination
- Fast, brief/intermittent/sustained
- ~60% of fetal tachycardias
Supraventricular Tachycardia

Supraventricular = Short VA
Long VA: PJRT & EAT (possible Sinus Tach)

- FHR 180-230 bpm
- 1:1 A-V, fast, regular
  - PJRT 1:1, EAT 1:1 primarily
- ~10% of fetal tachycardias
Heart rate 195 bpm
Atrial Flutter

- A rate 300-500 bpm; V rate 150-250 bpm
- A>V, fast, mainly regular
- Irregular/Variable conduction (mainly 2:1)
- “Quivering” atria
- GA: 3\textsuperscript{rd} trimester, macroreentry circuit
- ~30% of fetal tachycardia
Atrial Flutter

Atrium

Ventricular
Comparison of SVT and AF
Ventricular Tachycardia

- HR 280-340 bpm
- V>A (with VA block), fast, regular
- Seen with rhabdomyomas, aneurysms/diverticuli, cardiomyopathy, long QT syndrome, immune mediated myocarditis, electrolyte imbalance
- Rx: sotalol, flecainide, propranolol, amiodarone, IV Mg (LQTS)
- ~1-2% of fetal tachycardias
Ventricular Tachycardia
Fetal Tachyarrhythmia - Treatment

• Multiple studies assessing different therapeutic options
  – Observation, antiarrhythmics, or delivery

• Comparison of Transplacental Therapy of Fetal Supraventricular Tachyarrhythmias with Digoxin, Flecainide, and Sotalol – Jaeggi et al 2011
  – 159 cases, 3 tertiary care centers

• Fetal Atrial Flutter & Supraventricular Tachycardia Therapy Trial (FAST Therapy Trial) – Jaeggi

Fetal Tachyarrhythmia - Treatment

- Assess fetal factors

Percent Freedom from Prenatal Conversion

Jaeggi et al. Circulation 2011
Fetal Tachyarrhythmia - Treatment

• Assess fetal factors

SVA Mechanism

Percent Freedom from Prenatal Conversion

Jaeggi et al. Circulation 2011
Fetal Tachyarrhythmia - Treatment

1st Line Drug Therapy

Freedom from prenatal conversion to sinus

Percent Freedom from Prenatal Conversion

Jaeggi et al. Circulation 2011
When to Treat?

• Transplacental therapy most common but direct fetal or umbilical cord therapy useful with hydrops
• Sustained tachycardia (>50% of the scan time)
• Typically treat if < 32 weeks
  – If > 35 weeks, consider early delivery
  – Concerns for late pre-term complications
• Signs of fetal compromise
• Treatment more effective without hydrops
Fetal Tachyarrhythmia – Treatment

• Maternal medical exam/cardiac history
• Maternal EKG, BMP, Mg, +/- TFTs
  – Correct K, Ca, and Mg
• Serum drug level
  – Evaluate for toxicity
  – Estimate fetal serum level
• Be cognizant of possible early delivery as a “treatment”
Digoxin

- Fetal:Maternal drug level
  - 0.6:1 (decreased to 0.2:1 with hydrops)

- Safety
  - Maternal: level >2 can be well tolerated
  - Safe in the fetus

- Efficacy
  - 32-71% conversion without hydrops
  - 10-41% with hydrops
  - Conversion rate SVT > A flutter
Flecainide

- Fetal:Maternal drug level
  - 1:1

- Safety
  - Fetal demise 18% (ventricular rapid response with a flutter)
  - Widen QRS and PR prolongation on maternal ECG
  - Avoid with maternal cardiac disease

- Efficacy
  - 90-100% without hydrops
  - 50-80% with hydrops
Sotalol

- Fetal:Maternal drug level
  - 1:1?

- Safety
  - 25% fetal deaths reported (but may be an overestimate)
    - Prolongs QT interval on maternal ECG

- Efficacy
  - 80-90% conversion

Oudijik, 2000
Amiodarone

- Fetal:Maternal drug level
  - 0.15:1 no hydrops, 0.028:1 with hydrops
- Safety
  - No reported fetal demise from drug
  - Hypothyroidism in mother
  - Prolongs QT interval on maternal ECG
  - Neonatal hypothyroidism
- Efficacy
  - 93% for SVT
  - Lower for atrial flutter
Summary

- Atrial bigeminy is typically benign and resolves.
- Fetal bradycardia requires early evaluation for associated structural heart disease.
- LQTS may present with sinus bradycardia and 2:1 AV block.
- Dexamethasone and hydroxychloroquine shown to be beneficial in SSA pregnancies.
- Medical therapy exists for immune mediated complete heart block.
Summary

- Fetal tachycardias are common and treatable
- Determine mechanism of tachycardia for treatment selection
- Structural heart disease may be the cause
- Ventricular tachycardia is rare and consider LQTS
The 8th
Phoenix Fetal Cardiology Symposium
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