Fetal cardiac arrhythmias: Diagnosis and Management

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Objectives

- Discussion and image demonstration of fetal tachycardia
- Review of treatment of fetal tachycardia
- Discussion and image demonstration of fetal bradycardia
- Review of treatment of fetal bradycardia
- Discussion of fetal heart block
Fetal arrhythmia mechanisms 1

• Disturbances of rhythm production:

  – Sinus bradycardia, tachycardia
  – Atrial extrasystoles, ectopic atrial tachycardia
  – Junctional extrasystoles, junctional tachycardia
  – Ventricular extrasystoles, ventricular tachycardia

Fetal arrhythmia mechanisms 2

• Disturbance of impulse conduction-propagation

  • Reentry
    – In atrial myocardium (atrial flutter, atrial fibrillation)
    – In the AV node (AV nodal reentrant tachycardia)
    – Through an accessory AV pathway (AV reentrant tachycardia)

  • Block
    – Sinoatrial
    – Atrioventricular
Echocardiographic analysis of fetal cardiac rhythm: M-Mode echo

Echocardiographic analysis of fetal cardiac rhythm: Doppler
Clinical manifestation 1: Irregular cardiac rhythm

- Atrial extrasystoles
- Junctional extrasystoles
- Ventricular extrasystoles
- 2\textsuperscript{nd} degree AV block (Wenckebach type)

PACs: Conducted and non-conducted
Ventricular bigeminy

Clinical manifestation 2:
Fetal bradycardia

- Heart rate <100 bpm
- Sinus bradycardia
- Atrial or junctional bigeminy with non-conducted extrasystoles
- AV block (high grade or complete)
Persistent bradycardia: Differential diagnosis

Sinus bradycardia

Atrial bigeminy

3rd Degree AV block
Sinus bradycardia

- Transient (vagal, e.g. from transducer pressure)
- Persistent
  - Fetal distress
  - Maternal hypothermia
  - Sinus node disease
    - Primary (genetic etiology)
    - In the context of heterotaxy syndrome (left atrial isomerism/polysplenia syndrome)
  - Long QT syndrome

AV block

- Isolated
- In the context of congenital heart disease
Isolated congenital AV block

- Usually autoimmune mediated
- Maternal collagen vascular disease (SLE, Sjögren’s)
- 1:20,000 births
- Anti-Ro (SSA), anti-La (SSB) antibodies
- Possibility of AV block in the presence of maternal antibodies: 2-5%
- Possibility of appearance in subsequent pregnancies: 15-20%

Autoimmune-mediated AV block

- Age of diagnosis: 18-24 weeks
- Occasionally progressive
- Survival: 75% in isolated CCAVB
- Indices of poor prognosis: Ventricular rate < 55 bpm, endocardial fibroelastosis, myocardial dysfunction, hydrops fetalis
Congenital AV block in the context of CHD

- AV discordance (L-TGA, isolated ventricular discordance)
- Heterotaxy syndrome (Left atrial isomerism)
- LV non-compaction

Congenital AV block in the context of CHD: Prognosis

- Much worse than isolated CCAVB
- 19/123 fetuses with CCAVB and CHD survived neonatal period
- 10/19 had L-TGA
- No hydropic fetus with CCAVB and CHD survived
- Heart rate <55 bpm: very poor prognosis
**CCAVB: Treatment**

- Conflicting results in terms of steroid use, plasmapheresis
- Possible benefit of steroids in 1\textsuperscript{st} and 2\textsuperscript{nd} degree AV block, hydrops, myocardial dysfunction
- Increase of HR by 10-15\% with sympathomimetics (terbutaline), but without significant impact on survival
- Ventricular pacing: Transient success without long term benefit (mostly experimental data)
- In hydropic fetuses >34 wks: Deliver and pace

**Prospective evaluation of fetuses with autoimmune-associated congenital heart block followed in the PR Interval and Dexamethasone Evaluation (PRIDE) Study**

- Multicenter, open-label, nonrandomized study involving 30 pregnancies treated with DEX (22 with third-degree block, 6 with second-degree block, 2 with first-degree block) and 10 untreated (9 with third-degree block, 1 with first-degree block).
- There was no reversal of third-degree block with therapy or spontaneously. In fetuses treated with DEX, 1/6 with second-degree block progressed to third-degree block and 3 remained in second-degree block (postnatally 1 paced, 2 progressed to third degree); 2 reverted to normal sinus rhythm (NSR; postnatally 1 progressed to second degree).
- Prematurity and small size for gestational age were limited to the DEX group.

Friedman DM, Am J Cardiol 2009;103(8):1102-6
Medical treatment of CCAVB

Fetal tachycardia

- HR > 160 bpm
- Sinus (160-200 bpm)
- Pathologic mechanisms (180-280 bpm)
Sinus tachycardia

- Multiple etiologies
- Fetal distress
- Anemia
- Infection
- Maternal use of sympathomimetics
- Fetal thyrotoxicosis

Pathologic fetal tachycardia

- Atrial tachycardia
  - EAT
  - Atrial flutter
- Junctional tachycardia
  - JET
  - AVNRT
- AV reentrant tachycardia
  - Usual AV accessory pathway
  - PJRT
- Ventricular tachycardia
  - Ectopic focus
  - Abnormal substrate (e.g. ventricular aneurysm, tumors)
  - Related to LQTS
Atrial flutter

Short VA tachycardia: Accessory pathway
Long-VA tachycardia differential diagnosis: Sinus tachycardia, EAT, PJRT

Fetal tachycardia: Etiology

- 70-90% AV reentry
- Check for congenital anomaly (Ebstein’s, L-TGA, rhabdomyoma)
- Majority of AV reentry: Classic AP with rapid conduction
- Minority: Incremental retrograde only AP with long VA conduction (PJRT). “Incessant” tachycardia with frequent brief terminations
Fetal tachycardia: Atrial

• Ectopic atrial tachycardia (<10%)
  – 1:1 conduction, long VA
  – Warm-up and cool-down
• Atrial flutter (20%)
  – More As than Vs
  – Atrial rate 300-500 bpm
  – r/o accessory pathway, CHD
• Refractory to therapy, may need combination of drugs. Rate control may be the only achievable goal sometimes

Fetal tachycardia: Junctional

• Junctional: very rare in fetus
• AV nodal reentrant (AVNRT)
  – Very short VA interval (simultaneous atrial and ventricular depolarization, cannon A waves)
  – Very rare in fetus (Dual AV nodal physiology develops later in life)
  – Theoretically easier to control
• Junctional Ectopic Tachycardia (JET)
  – Persistent
  – Variations in heart rate
  – Extremely rare
  – Resistant to medical therapy. Rate control the realistic goal
Fetal tachycardia: Ventricular

- Ventricular tachycardia: relatively rare
- HR 180-300 bpm
- AV dissociation, V>A
- May be difficult to differentiate from JET
- Usually automatic, non-sustained
- Rare underlying etiology: Long QT, LV non-compaction, rhabdomyoma, fibroma, LV aneurysm

Fetal tachycardia: Ventricular

- Long QT syndrome
  - Possible cause of intrauterine death
  - High suspicion if:
    - Constant bradycardia(110-120 bpm), low HR variability
    - 2\textsuperscript{nd} degree AV block (functional)
    - torsade des pointes
    - Diagnosis: Fetal magnetocardiography
- Treatment: B-blockers, other meds (mexiletine, Mg for TdP).
Fetal tachycardia: Treatment

• In the final stages of pregnancy: Brief therapeutic attempt, if unsuccessful, delivery (preferably with CS), direct neonatal therapy
• Fetuses <34 weeks with sustained tachycardia intensive medical therapy to avoid complications of prematurity

Fetal tachycardia: Maternal F/U

• Usually start therapy in hospital
• Maternal ECG, echo: Check for WPW, LQT, cardiomyopathy
• Check electrolytes, renal, thyroid function
• Drug levels (esp. digoxin)
Treatment of fetal tachycardia

- **Digoxin**
- Initially high doses (0.25-0.5 mg tid) until drug levels 2-2.5 ng/ml, then adjust according to needs
- Fetal levels 70-100% of maternal
- Watch for drug-drug interaction (Flecainide, propafenone, amiodarone, verapamil may increase Dig levels)
- 50-70% success rate in non-hydropic fetuses

- **Flecainide**
  - Fetal levels: 80% of maternal
  - Therapeutic levels: 200 - 1000 ng/ml
  - Dosage: 100 mg q 6-8 hrs
  - Possible toxicity: pro-arrhythmia, worsen myocardial dysfunction
  - Time to effect: 1-14 d.
Drug Combinations

• Digoxin + Flecainide, Digoxin + Amiodarone
• In hydropic fetus: direct fetal therapy
  – Intraumbilical digoxin, adenosine, amiodarone
  – Intramuscular digoxin
Other arrhythmias

- Sustained VT: Amiodarone, Sotalol
- Atrial flutter: Dig +/- amio or sotalol
- PJRT: usually resistant to dig, better effect with: Flecainide, Amiodarone, Sotalol

Conclusions

- Fetal arrhythmia can be diagnosed with high degree of accuracy with fetal echocardiographic methods
- Treatment, depending on etiology, severity, can be delivered either transplacentally or intraumbilically, or directly after emergent delivery