Fetal Counseling Provider Information
Last Updated: 10/08/2021

DIAGNOSIS: Complete Heart Block

- With a primary focus on autoimmune-mediated heart block

Incidence

- 1:15,000 live births
- 30% with structural heart disease
- 70% isolated
    - Of those 80-95% is autoimmune-mediated (maternal anti-Ro/SSA autoantibodies, can be seen in SLE, Sjogren's or with no symptoms
    - Maternal clinical disease does not correlate with risk of AV block
- Estimated prevalence of anti-SSA antibodies in the general population - 0.5%
- For those with antibodies, ~2-3% risk of conduction disease in their fetus
- For those with a previously affected child, risk goes to 15-20%
- AV block can develop from normal rhythm in < 24 hours

Differential Diagnosis – Fetal Bradycardia

1. Sinus Bradycardia (maternal hypothyroid, LQTS, fetal distress)
2. Blocked atrial bigeminy
3. Complete Heart Block
    - Maternal anti-SSA/SSB antibodies
    - Congenital heart disease
        - L-looped ventricles
        - Heterotaxy (particularly left atrial isomerism)
    - Myocarditis

Available Fetal Interventions

Prevention

1. Hydroxychloroquine
    - HCQ significantly reduces the recurrence of CHB below the historical rate by >50% in women with prior pregnancy affected by CHB (Izmurly)
2. Fetal echocardiographic surveillance for PR prolongation is reasonable beginning at 16-18 weeks, and continuing at 1-2 week

intervals (more frequent with previously affected child) until 28 weeks.

3. In patients with normal conduction, complete heart block MAY be predicted by first- and second-degree AV block (as described by abnormal mechanical PR intervals) (Glickstein, Andlefinger). Normal mechanical PR intervals vs gestational age are published (Nii)

4. Early ID of 1°AV block by Doppler AV interval
    - Controversial definition but probably AV interval > 160 ms
    - Maternal anti-Ro/SSA levels may risk stratify need for echo
5. Ambulatory fetal heart rate monitoring by mother
    - Is feasible and mothers can detect abnormal fetal cardiac rate and rhythm
    - Once irregular rhythm/bradycardia detected by monitoring , rapid echo confirmation of 2°AVB is needed for treatment to be effective

Therapies without proven benefit and possible harm

1. Eliminate maternal Ab (plasmapharesis)
2. Reduce fetal antibody exposure (transplacental IVIG)
3. Prophylactic Dexamethasone
    - Not effective for general prophylaxis without evidence of conduction abnormality
    - Side effects as below

Treatment

1. Fluorinated Glucocorticoids
   (Dexamethasone 4 mg daily or titrating dose scheme from 8-2 mg, Betamethasone 3 mg daily)
    - Very controversial in treating first-degree heart block if AV interval <150 ms)
    - More accepted in treating second-degree heart block
    - Not found to reverse complete heart block, but may play a role in treatment of myocarditis, effusions, EFE
    - Significant maternal side effects (HTN, infection, insulin resistance, gestational diabetes) and fetal side effects (IUGR, oligo) Data on

neurocognitive issues showed no impairment in school aged children. (Kelly)

2. Beta-agonists (Terbutaline)
    - Limited data to show improved HR, stroke volume when FHR is < 50-55 bpm
    - May prolong pregnancy for more mature delivery.

Fetal Imaging Predictors of Postnatal Interventions/Outcomes

In patients with heart block, predictors of poor outcome are:

- Fetal heart rate (ventricular rate) less than 55 bpm
- Endocardial fibroelastosis
- Ventricular dysfunction

In patients with heart block and structural heart disease, prognosis is dismal

Prognosis

Mortality rate 17-20%

- 20-50% in CHD (depending on type)
- 33% HR<55 bpm
- 52% if delivered before 34 weeks' gestation
- 70-100% with fetal hydrops

Neonatal pacemaker placement – 63-93% get a pacemaker

- 6% of those will develop a cardiomyopathy

Associated Problems

Associated defects/syndromes

- Dilated cardiomyopathy and endocardial fibroelastosis, AV chordal rupture leading to severe AVV insufficiency
- Consideration of other manifestations of neonatal lupus
    - Skin lesions, hematologic disease, hepatic dysfunction

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