ADULT CONGENITAL HEART DISEASE

HOW DO YOU MEND A BROKEN HEART?

Carey Winkler, MD

June 7, 2017
Practice gap: lack of knowledge on the special problems associated with pregnancy in patients with congenital heart disease.

Educational Goal: improve knowledge and provide some educational “pearls” on the evaluation and management of these patients.
"OK, the old one's in my right hand, the donor's in my left. Right?"
SCOPE OF THE PROBLEM

• Almost 1 in 100 babies born with CHD
• Now > 1 million adults living with corrected CHD
  • Pssst: many of these are women of reproductive age!!
• Growth of this population about 5% a year!!!
• Currently more adults have corrected CHD than children
• Unique problems in pregnancy
Fig. 1. Median age of patients with severe CHD over time in 1985, 2000, and 2010. (Data from Marelli AJ,
Fig. 3. Prevalence of CHD across the life span in infants, children, adults, and geriatric patients in Quebec, Canada. (Data from Marelli AJ, Mackie AS,
Fig. 2. (A) The number and proportions of adults and children in Quebec, Canada with all CHD over time in 2000, 2005, and 2010. (B) The number and proportions of adults and children in Quebec, Canada with severe CHD over time in 2000, 2005, and 2010. (From Marelli AJ, Ionescu-Ittu R, Mackie AS, et al. Lifetime prevalence of congenital heart disease in the general population from 2000 to 2010. Circulation 2014;130:753; with permission.)
"All right, so he dropped the heart. The floor is clean."
ISSUES TO BE ADDRESSED

• Effects of pregnancy on maternal cardiac situation: risks to mother
• Risk to the fetus
• Predictors for adverse outcome
• Basic management suggestions
WHAT IS THE RISK TO MY BABY, DOCTOR?

• Most CHD is a multi-factorial inheritance
• Some are autosomal dominant like Marfan’s
• Also based on specific abnormality
• Higher risk if mother has it
• General population: 8/1000
<table>
<thead>
<tr>
<th>Defect</th>
<th>Father affected</th>
<th>Mother affected</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrial septal defect</td>
<td>1.5</td>
<td>4–4.5</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>2</td>
<td>6–10</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>2.5</td>
<td>3.5–4</td>
</tr>
<tr>
<td>Atrioventricular canal</td>
<td>1</td>
<td>14</td>
</tr>
<tr>
<td>Pulmonary stenosis</td>
<td>2</td>
<td>4–6.5</td>
</tr>
<tr>
<td>Aortic stenosis</td>
<td>3</td>
<td>13–18</td>
</tr>
<tr>
<td>Coarctation of the aorta</td>
<td>2</td>
<td>4</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>1.5</td>
<td>2.5</td>
</tr>
</tbody>
</table>

CHD: congenital heart disease.
BUT I REALLY REALLY WANT A BABY!
IS PREGNANCY BAD FOR ME?

- Pregnancy is a huge stress on the cardiovascular system
- Cardiac output increases 30-40% (even more in labor)
- Blood volume increases by a similar amount
- SVR decreases
- \( CO = HR \times SV \)
  - Initial increase in CO is from increase SV, then mostly from HR
- These changes impact how various lesions may do
CARDIOVASCULAR CHANGES

- Increase in HR shortens diastolic filling time
- Decrease in SVR may close gap between pulmonary and systemic pressures
- Generally regurgitative lesions do better
  - Tolerate increase volume, shortened diastolic time, decrease in SVR well
- Generally stenotic lesions do worse
  - Don’t like shorter filling time, increased volume, increased heart rate
- Pulmonary hypertension with systemic-pulmonary communication can do very very bad
"Catch it...!"
PRE-CONCEPTION COUNSELING (ideally)

• Long term outlook/outcome for mother

• Is her condition getting worse
  • Need further surgery soon?
  • If so, consider sooner rather than later?
  • Different type of valve for childbearing....

• Effects of pregnancy on maternal cardiac condition
  • Risks to mother

• Effects on the fetus
  • Anticoagulants?
  • Prematurity?
PRE-CONCEPTUAL COUNSELING

- History and physical and EKG
- Possible CXR
- Echocardiogram to assess
  - Ventricular function
  - Valves
  - Vessels
  - PAp
  - Presence or absence of shunts
PRE-CONCEPTION COUNSELING

• Exercise tolerance testing to see clinical function
• One of the best predictors of bad outcome
  • Pulmonary hypertension
  • Pulmonary hypertension with systemic-pulmonary shunt

• If any doubt on PAp, consider cardiac cath
Fig. 2. Unique risk factors for pregnancy complications among women with CHD. Anatomic and physiologic complications combined with pregnancy physiology create a unique risk profile for each pregnant individual with ACHD.
ACHD: PREGNANCY (N=2491)
ACHD: PREGNANCY (N=2491)
POSSIBLE PREDICTORS OF GOOD/BAD OUTCOMES

• Simpler lesions
  • ASD
  • VSD
  • Coarctation
  • PS
  • AS (mild)
  • Epstein’s
• Well selected and managed
• Generally well tolerated
NYHA CLASSIFICATION

• Class 1: asymptomatic
• Class 2: symptoms with > than normal activity
• Class 3: symptoms with normal activity
• Class 4: symptoms at rest

***Class 1 or 2 have < 1% maternal mortality
***Class 3 or 4 have a 5-10% maternal mortality
NYHA

• Pretty simple
• Easy to remember and apply
• Doesn’t discern well between different lesions
• What is considered “more than normal activity”
• Probably is a pretty good predictor for mild, low risk lesions
• Less helpful for specific, more complex lesions
CARPREG RISK SCORE

• 562 patients with ACHD
• 599 pregnancies
• 13% of these pregnancies complicated by
  • Pulmonary edema
  • Arrhythmias
  • Stroke
  • Cardiac death
PREDICTORS OF CARDIAC EVENTS

• Prior arrhythmia, CHF, TIA, or stroke
• Baseline NYHA class > 2 or cyanosis
• Left heart obstruction
  • MVA < 2.0 cm or AVA < 1.5 cm
  • Peak LVOT gradient > 30 mm Hg by echo
• Reduced LV function with ejection fraction < 40%

• Risk index predicts CV event rate
Table 2. The CARPREG risk-predictor score.\textsuperscript{11}

<table>
<thead>
<tr>
<th>Predictors</th>
<th>Points</th>
</tr>
</thead>
<tbody>
<tr>
<td>Previous cardiac event: heart failure, transient ischaemic attack, cerebrovascular accident or arrhythmia</td>
<td>1</td>
</tr>
<tr>
<td>New York Heart Association class $\geq$ II or cyanosis</td>
<td>1</td>
</tr>
<tr>
<td>Left heart obstruction: Mitral valve area $&lt; 2$ cm$^2$ or aortic valve area $&lt; 1.5$ cm$^2$ or peak left ventricular outflow tract obstruction $&gt; 30$ mmHg on echocardiogram</td>
<td>1</td>
</tr>
<tr>
<td>Reduced left ventricular function (ejection fraction $&lt; 40%$)</td>
<td>1</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Risk score</th>
<th>Frequency of maternal cardiac complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>5%</td>
</tr>
<tr>
<td>1</td>
<td>27%</td>
</tr>
<tr>
<td>$\geq$2</td>
<td>75%</td>
</tr>
</tbody>
</table>
RISK ESTIMATE OF CV MATERNAL COMPLICATIONS

0 Points: 5%

1 Point: 27%

≥ 2 Points: 75%

74% of patients with congenital heart disease
WHO SANG HUNGRY HEART?
CARPREG

• CARPREG may overestimate risk
• Risk scores have limitations
  • Representation of risk factors in the population determines which risk factors and complications emerge
  • Pulmonary hypertension was under represented in CARPREG

• Calculation of risk scores should only be a part of pre-pregnancy evaluation/counseling
<table>
<thead>
<tr>
<th>Congenital heart disease</th>
<th>N</th>
<th>Cardiac complications</th>
<th>Obstetric complications</th>
<th>Neonatal complications</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>AR</td>
<td>HF</td>
<td>CE</td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>188</td>
<td>7</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Aortic coarctation</td>
<td>160</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>148</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Pulmonary valve stenosis</td>
<td>148</td>
<td>3</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>124</td>
<td>7</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Marfan syndrome</td>
<td>118</td>
<td>1</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Atroventricular septal defects</td>
<td>89</td>
<td>15</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>Aortic valvar stenosis</td>
<td>81</td>
<td>3</td>
<td>4</td>
<td>0</td>
</tr>
<tr>
<td>Complete transposition of the great arteries</td>
<td>52</td>
<td>11</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td>Ebstein malformation</td>
<td>22</td>
<td>2</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Congenital corr. transposition of great arteries</td>
<td>19</td>
<td>1</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Pulmonary atresia</td>
<td>12</td>
<td>3</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Pulmonary hypertension or Eisenmenger§</td>
<td>4</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Complex cyanotic heart disease</td>
<td>9</td>
<td>3</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Other</td>
<td>128</td>
<td>3</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>Overall</td>
<td>1302</td>
<td>62</td>
<td>21</td>
<td>17</td>
</tr>
</tbody>
</table>

N, number of pregnancies. AR, arrhythmias; HF, heart failure; CE, cardiovascular complications; EN, endocarditis; PI, pregnancy-induced hypertension; PE, preeclampsia; EC, eclampsia; HE, HELLP syndrome; PL, premature labour; PH, post-partum haemorrhage; PD, premature delivery; SG, small for gestational age; MO, foetal or neonatal mortality.

§Respectively, ASD related n = 2 and VSD related n = 2.
ZAHARA PREDICTORS

• History of arrhythmia event
• NYHA functional class > 2
• Left heart obstruction (peak gradient > 50 mm Hg)
• Mechanical valve prosthesis
• Moderate-severe AV regurgitation
• Use of cardiac medication pre-pregnancy
• Repaired or unrepaired cyanotic heart disease
<table>
<thead>
<tr>
<th>Predictors</th>
<th>Points</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. History of arrhythmias</td>
<td>1.50</td>
</tr>
<tr>
<td>2. Cardiac medication before pregnancy</td>
<td>1.50</td>
</tr>
<tr>
<td>3. New York Heart Association class prior to pregnancy ≥ II</td>
<td>0.75</td>
</tr>
<tr>
<td>4. Left heart obstruction (peak gradient &gt; 50 mmHg or aortic valve area &lt; 1.0 cm²)</td>
<td>2.50</td>
</tr>
<tr>
<td>5. Systemic atrioventricular valve regurgitation (moderate/severe)</td>
<td>0.75</td>
</tr>
<tr>
<td>6. Pulmonary atrioventricular valve regurgitation (moderate/severe)</td>
<td>0.75</td>
</tr>
<tr>
<td>7. Mechanical valve prosthesis</td>
<td>4.25</td>
</tr>
<tr>
<td>8. Cyanotic heart disease (corrected/uncorrected)</td>
<td>1.00</td>
</tr>
</tbody>
</table>

Total number of points: 0–13 points
<table>
<thead>
<tr>
<th>Risk score</th>
<th>Frequency of maternal cardiac complication (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0–0.50</td>
<td>2.9</td>
</tr>
<tr>
<td>0.51–1.50</td>
<td>7.5</td>
</tr>
<tr>
<td>1.51–2.50</td>
<td>17.5</td>
</tr>
<tr>
<td>2.51–3.50</td>
<td>43.1</td>
</tr>
<tr>
<td>&gt;3.51</td>
<td>70.0</td>
</tr>
</tbody>
</table>
### ZAHARA PREDICTORS

<table>
<thead>
<tr>
<th>Cardiac complications in % total no. pregnancies</th>
<th>Points</th>
</tr>
</thead>
<tbody>
<tr>
<td>History of arrhythmias</td>
<td>1.50</td>
</tr>
<tr>
<td>Cardiac medication before pregnancy</td>
<td>1.50</td>
</tr>
<tr>
<td>NYHA class prior to pregnancy ≥II</td>
<td>0.75</td>
</tr>
<tr>
<td>LHO (PG &gt;50 mm Hg or AVA &lt;1.0 cm²)</td>
<td>2.50</td>
</tr>
<tr>
<td>Syst AV valve regurgitation (mod/severe)</td>
<td>0.75</td>
</tr>
<tr>
<td>Pulm AV valve regurgitation (mod/severe)</td>
<td>0.75</td>
</tr>
<tr>
<td>Mechanical valve prosthesis</td>
<td>4.25</td>
</tr>
<tr>
<td>Cyanotic heart disease (corrected/uncorrected)</td>
<td>1.00</td>
</tr>
<tr>
<td><strong>Total number of points</strong></td>
<td><strong>0-13</strong></td>
</tr>
</tbody>
</table>

**Risk score**
- 0-0.50: 2.9
- 0.51-1.50: 7.5
- 1.51-2.50: 17.5
- 2.51-3.50: 43.1
- >3.51: 70.0

*Drenthen: Eur Ht J, 2010*
Overall cardiac complications
Occurred in 7.6% of pregnancies
ZAHARA

• Most common cardiac complications
  • Arrhythmias 4.7%
  • Heart failure 1.7%

• Factors independently associated with heart complications
  • Cyanotic heart disease (p < 0.0001)
  • Pre-pregnancy cardiac meds (p < 0.0001)
  • Left heart obstruction (p < 0.0001)
**ZAHARA**

- Neonatal complications
  - Prematurity 12%
  - SGA 14%
  - Mortality 4%
WHO SANG HEARTBREAKER?
PAT BENATAR
LIMITATIONS OF CARPREG/ZAHARA

• Highly population dependent
  • CARPREG: congenital and acquired heart disease
  • ZAHARA: congenital only

• Important risk factors (PAH and dilated aorta) no identified or were under represented

• Did not allow prediction of more severe events (i.e. heart failure) to less severe
WHAT ELSE IS THERE?
WHO
CLASSIFICATION
PREGNANCY RISK WHO I

- Uncomplicated, small or mild
  - Pulmonary stenosis
  - Patent ductus arteriosus
  - Mitral valve prolapse

- Successfully repaired simple lesions
  - Atrial & ventricular septal defect
  - Patent ductus arteriosus
  - Anomalous pulmonary venous drainage

- Atrial or ventricular ectopic beats, isolated
<table>
<thead>
<tr>
<th>WHO II</th>
<th>WHO II - III</th>
<th>WHO III</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unoperated ASD VSD</td>
<td>Mild LV dysfunction</td>
<td>Mechanical valve</td>
</tr>
<tr>
<td>Repaired TOF</td>
<td>HCM</td>
<td>Systemic RV</td>
</tr>
<tr>
<td>Most arrhythmias</td>
<td>Native or tissue valve ht disease (not WHO I or IV)</td>
<td>Fontan</td>
</tr>
<tr>
<td></td>
<td>Marfan without ao diln Ao &lt; 45 mm + BAV</td>
<td>Cyanotic ht disease</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Marfan ao 40 - 45 mm</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Ao 45 - 50 mm + BAV</td>
</tr>
</tbody>
</table>
- PAH of any cause
- Severe systemic ventricular dysfunction
  LVEF < 30%, NYHA III – IV
- Previous peripartum CM with residual impairment LV function
- Severe MS, severe symptomatic AS
- Marfan with aorta > 45 mm
- Aorta > 50 mm + bicuspid aortic valve
- Native severe coarctation
<table>
<thead>
<tr>
<th>Risk class</th>
<th>Risk by medical condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>No detectable ↑ risk maternal mortality &amp; no / mild ↑ in morbidity</td>
</tr>
<tr>
<td>II</td>
<td>Small ↑ risk maternal mortality or moderate ↑ in morbidity</td>
</tr>
<tr>
<td>III</td>
<td>Sig ↑ risk maternal mortality or severe morbidity Expert counseling required. If pgym – expert care</td>
</tr>
<tr>
<td>IV</td>
<td>Extremely high risk maternal mortality or severe morbidity: pgym c/i. If pgym, discuss termination</td>
</tr>
</tbody>
</table>
SUMMARY OF PREDICTORS

• Appears that the WHO system is an overall better predictor of maternal performance in pregnancy
• Seems to have a better sensitivity/specificity
• Better stratifies patients into risk categories and what the actual risk is
• ARE THERE ANY OTHER THINGS THAT MIGHT HELP?
NT-pro BNP at 20 WK GESTATION (n=213 pg/gy)

- NT-pro BNP available in 169 /213 pts
- 1° CV events in 22 pregnancies (10.3%)
- Pts < 100 pg/ml : no CV complications

Sensitivity > 128 pg/ml = 81.3%
Specificity > 128 pg/ml = 61.8%

Positive predictive value = 18.3%
82.4% of patients with complications had BNP > 128
Negative predictive value < 128 = 96.9%
QUESTIONS??
ABOUT
PREDICTION
CONCERNS AND MANAGEMENT OF SPECIFIC LESIONS

JUST THE BASICS!!!!
ASD (WHO I)

- Maternal risks
  - Contraindicated if PAH/Eisenmenger syndrome
  - If hemodynamically significant, closure before pregnancy
  - Arrhythmias more common if unrepaired, closed at a later age, or in patients over 30
  - Up to 5% risk of thrombo-embolic complications

- Vaginal delivery appropriate

- Concern for paradoxical emboli
  - Avoid stasis (sequential compression devices, early ambulation)
  - Consider prophylactic heparin for prolonged bedrest, other high risk conditions
VSD (WHO I)

• Maternal risks
  • If repaired, no increased risk of complications
  • If unrepaired and small with no LV dilatation, then low risk
  • If large, long standing, and has PAH, not WHO 1 but more likely III or IV

• Vaginal delivery is appropriate
• No need for alteration in pregnancy management
• No change in maternal/fetal surveillance
AV SEPTAL DEFECT (WHO I)

- Maternal risks
  - If repaired and minimal valve regurgitation, then well tolerated
  - If symptomatic valvular regurgitation or impaired LV function, pre-pregnancy surgery recommended
  - If AVSD with PAH, then class III or IV and would be high risk
- Arrhythmias, worsening NYHA class, and worsening regurgitation all have been reported in pregnancy
- Heart failure is more common especially postpartum
- If uncorrected, paradoxical emboli possible
- Vaginal delivery is appropriate
COARCTATION (WHO I)

• Well tolerated if repaired

• If unrepaired or still have hypertension after repair/residual coarctation or have evidence of aortic aneurysm, there is increased risk of aortic rupture and cerebral aneurysm rupture

• Other risks for this aortic root dilatation with bicuspid aortic valve

• Risk of cerebral aneurysm is 10%

• Important to control BP but not overtreat if still have residual coarctation as this may decrease downstream perfusion

• Vaginal delivery with epidural recommended
WHO SANG HARDEN MY HEART?
PS and PR  
(WHO I, II, III, OR IV)

• Mild/moderate PS are low risk lesions (WHO I or II) and well tolerated
• Severe PS (peak gradient > 64 mm Hg) much higher risk for right heart failure and arrhythmias
• Severe PR with abnormal RV function correlates with poor outcome and valve replacement prior to pregnancy is recommended
• Consider percutaneous valvuloplasty for severe PS
• Vaginal delivery for PS in NYHA 1 or 2 whether mild-severe
• If severe PS in NYHA 3 or 4, consider c-section
TOF (WHO II)

• If unrepaired, should be repaired prior to pregnancy
• Well corrected TOF is usually well tolerated with good outcomes
• Cardiac complications common (up to 12%)
  • Arrhythmias
  • Heart failure
• Also increase risk of thromboembolic events, progressive aortic root dilatation, endocarditis, persistent increase in RV size
• Should repair significant pulmonary regurgitation pre-pregnancy if associated with RV dysfunction
• Vaginal delivery recommended
TGA (WHO III)

• Most of data is on atrial switch (Mustard or Senning)
• Limited data on arterial switch but this appears very promising
• Atrial switch at increased risk for severe arrhythmias (many life threatening), heart failure, and 10% have permanent decline in RV function
• If have baseline mod-severe RV function or severe TR, then pregnancy is discouraged
• If asymptomatic and good ventricular function, vaginal delivery
• If ventricular function worsens, recommend early c-section
FONTAN CIRCULATION (WHO IV)

- Systemic right ventricle
- Even higher risk of Fontan circuit not optimal
- Arrhythmias and deterioration in NYHA are common
- High risk of prematurity, SGA, and fetal death (up to 50%)
- Controversial about anti-coagulation but most recommend therapeutic anticoagulation (many on ACE inhibitors and coumadin prior to pregnancy)
- Vaginal delivery is recommended
MARFAN’S (WHO II)

- Inheritance is 50% (AD)
- Risk of aortic dissection is < 1% if aortic root is normal
- Risk increases as root size increases but no safe level
- If < 4 cm, risk is acceptable
- If > 4.0 cm or root size is increasing in pregnancy, dissection risk increases
- If > 4.5 cm, pregnancy should be discouraged
- Low dose beta-blocker may be helpful in reducing risk
BICUSPID AORTIC VALVE

• Approximately 50% with bicuspid aortic valve and AS will have dilated aortic root
• Often times, the most dilated part of the aorta is in the distal part of the ascending aorta which is difficult to see on echo
• Lower risk of dissection than Marfan’s
• Unclear what is the concerning value
• If > 5.0 cm pre-pregnancy, encourage surgery or discourage pregnancy
MS

• Mod to severe MS (valve area < 1.5 cm) is poorly tolerated
• Heart failure occurs frequently particularly in 2\textsuperscript{nd} and 3\textsuperscript{rd} trimester
• Atrial fib may occur in 10-15\% of patients which also carries a risk of thrombo-embolic event
• Maternal mortality is up to 3\%
• If symptoms of failure or elevated PAP (> 50 mm Hg), consider beta1 selective blockers and diuretics
• Vaginal delivery is recommended with mild MS, no evidence of PAH, NYHA 1 or 2. Otherwise, consider c-section
AS

• Usually from bicuspid valve
• Often times first presents in pregnancy due to increased demands
• If known prior, consider exercise testing
• CV changes in pregnancy result in heart failure about 10% of the cases and arrhythmias in up to 25%
• Evidence of longstanding disease (hypertrophic LV, aortic root dilatation > 5.0 cm, high gradient) should not get pregnant
• Beta-blocker for rate control and diuretics of
• C-section under general anesthesia may be best for delivery
PROSTHETIC VALVES

• Best valve for pregnancy is a bioprosthetic valve
  • Excellent hemodynamic performance
  • No need for anti-coagulation
  • 50% of patients under 30 who have had their valve for 10 years will have deterioration, higher in mitral valve than aortic
  • Mortality on a redo is 0-5% depending on valve position and how emergent
  • Pregnancy may accelerate the deterioration of the valves

• Mechanical valves last longer
  • Need anti-coagulation therapy which increases complications significantly
  • Valve thrombosis is kind of a big deal
• Risk of thrombosis
  • OAC throughout pregnancy 3.9%  Mortality 2%
  • Heparin in 1\textsuperscript{st}, OAC in 2\textsuperscript{nd} and 3\textsuperscript{rd} 9.2%  Mortality 4%
  • Heparin throughout 33.0%  Mortality 15%

• Using LMWH is controversial
  • Best to check pre or post dosing anti-Xa levels?
  • Pre-dose levels often subtherapeutic when post dose is therapeutic
  • Increasing requirements during pregnancy so how often to check

• Risk of warfarin embryopathy low (2.6%) if dose was $< 5$ mg/day compared to 8% when $> 5$ mg/day
“YOU GOT TO ASK YOURSELF ONE QUESTION. DO I FEEL LUCKY. WELL DO YOU PUNK!!!”
DIRTY HARRY

You got to ask yourself one question. Do I feel lucky, punk?
• If OAC dose is < 5 mg/day, it is reasonable to continue throughout the pregnancy and alter dose based on PT

• If greater than 5 mg/day
  • D/C OAC from 6 – 12 weeks
  • Dose adjusted UFH to maintain PTT > 2.0 times control
  • For LMWH, check anti-Xa levels 4-6 hours after dose and have 0.8-1.2 U/ml
    • If cant check anti-Xa levels then don’t use LMWH
  • Levels should be checked weekly
  • Restart OAC after first trimester and continue until 36-37 weeks
  • Convert to UFH for the duration of pregnancy
  • Consider scheduled induction to optimize anti-coagulation therapy
PROSTHETIC VALVE

- If on OAC and presents in labor, recommend c-section due to the fact the fetus is anti-coagulated and reversing the mom may not adequately reverse the fetus
- In labor, have IV UFH drip and discontinue 4-6 hours before delivery
- Restart heparin 4-6 hours after delivery
- Transition to coumadin in the postpartum period
- Safe in breastfeeding
- Easier to monitor
QUESTIONS?
• Spontaneous labor is best but nice to have coordination
• Epidural is the ideal form of anesthesia
  • More narcotic than local anesthetic agent has less cardiovascular issues
• Phenylephrine is drug of choice for epidural induced hypotension because pure alpha stimulant, less effect on heart rate
• Often best to shorten 2\textsuperscript{nd} stage with operative vaginal delivery
• C-section generally for obstetrical indications except for severe and pretty rare heart defects
• Fluid status is critical but rare to use PA catheter
INTRAPARTUM STUFF

• Very few indications for SBE prophylaxis
  • Prior history of SBE
  • Prosthetic valve
  • New prosthesis material (within 6 months because not epithelialized) or incomplete repair with prosthesis material
  • Significant leaflet pathology or regurgitation in a transplanted heart

• Early epidural
• Really not much else
<table>
<thead>
<tr>
<th>Disease</th>
<th>Evaluate/Exclude</th>
<th>Potential Risks</th>
<th>Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>VSD</td>
<td>PAH</td>
<td>Arrhythmia</td>
<td>Antibiotics if unrepaired</td>
</tr>
<tr>
<td></td>
<td>Ventricular dysfunction</td>
<td>Endocarditis</td>
<td></td>
</tr>
<tr>
<td>ASD (unrepaired)</td>
<td>PAH</td>
<td>Arrhythmia</td>
<td>Low-dose aspirin</td>
</tr>
<tr>
<td></td>
<td>Ventricular dysfunction</td>
<td>Paradoxical embolism</td>
<td>thromboembolic prophylaxis (if bed rest)</td>
</tr>
<tr>
<td>Coarctation (repaired)</td>
<td>Re-coarctation</td>
<td>Pre-eclampsia/HTN</td>
<td>β-Blocker if HTN</td>
</tr>
<tr>
<td></td>
<td>Aortic/brain aneurysm</td>
<td>Aortic dissection</td>
<td>Avoid placental hypotension</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Left heart dissection</td>
<td>Consider C-section if aneurysm</td>
</tr>
<tr>
<td>TOF (repaired)</td>
<td>Severe RVOT obstruction</td>
<td>Arrhythmia/VT</td>
<td>Volume control</td>
</tr>
<tr>
<td></td>
<td>Severe PR</td>
<td>Right heart failure</td>
<td>Peridelivery</td>
</tr>
<tr>
<td></td>
<td>RV dysfunction</td>
<td></td>
<td>Consider preterm delivery if RHF</td>
</tr>
<tr>
<td></td>
<td>DiGeorge syndrome</td>
<td></td>
<td>Antibiotic prophylaxis</td>
</tr>
<tr>
<td>Mitral stenosis</td>
<td>Severe MS</td>
<td>Atrial fibrillation</td>
<td>β-Blockers</td>
</tr>
<tr>
<td></td>
<td>Pulmonary venous HTN</td>
<td>Thromboembolic event</td>
<td>Diuresis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Pulmonary edema</td>
<td>3rd-trimester bed rest</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Antibiotic prophylaxis</td>
</tr>
<tr>
<td>Condition</td>
<td>Risk Factors</td>
<td>Recommendations</td>
<td></td>
</tr>
<tr>
<td>--------------------------</td>
<td>---------------------------------------------------</td>
<td>-----------------------------------------------------</td>
<td></td>
</tr>
<tr>
<td><strong>AS</strong></td>
<td>Severe AS (peak &gt;80 mm Hg), Symptoms, ST depressions, LV dysfunction</td>
<td>Arrhythmia, Angina, Heart failure, Shock, Endocarditis, ± 3rd-trimester bed rest, Diuretics as needed, If HF: valvotomy or preterm, C-section, Antibiotic prophylaxis</td>
<td></td>
</tr>
<tr>
<td><strong>Systemic RV</strong></td>
<td>Ventricular dysfunction, Severe systemic TR, Arrhythmia, Symptoms, Venous pathway obstruction</td>
<td>Heart failure, Arrhythmia, Thromboembolic event, Restore NSR if needed, Aspirin 81 mg daily, ± Rhythm monitoring</td>
<td></td>
</tr>
<tr>
<td><strong>Cyanotic lesions without PAH</strong></td>
<td>Ventricular dysfunction, Saturation &lt;85%, Hemorrhage</td>
<td>Thromboembolic events, Increased cyanosis, Heart failure, Endocarditis, Bed rest and O₂, Thromboembolic prophylaxis</td>
<td></td>
</tr>
<tr>
<td><strong>Fontan circulation</strong></td>
<td>Ventricular dysfunction, Arrhythmia, Prior heart failure</td>
<td>Thromboembolic events, Increased cyanosis, Heart failure, Endocarditis, Anticoagulation or aspirin, Avoid dehydration during delivery, Restore NSR quickly, Avoid calcium channel blockers, Antibiotic prophylaxis</td>
<td></td>
</tr>
<tr>
<td><strong>Marfan syndrome</strong></td>
<td>Aortic root &gt;4.0</td>
<td>Type A dissection, β-Blocker, C-section if root &gt;4.0–4.5 cm</td>
<td></td>
</tr>
<tr>
<td><strong>Eisenmenger syndrome</strong></td>
<td>Ventricular dysfunction, Arrhythmia</td>
<td>30%–50% risk of death, Discuss termination, If late for termination: • O₂, • Bed rest, • Thromboembolic prophylaxis, • Prolonged postpartum stay, • Permanent contraception</td>
<td></td>
</tr>
<tr>
<td>Congenital Heart Disease (Ref. no.)</td>
<td>Arhythmias</td>
<td>Heart Failure</td>
<td>Cardiovascular Events</td>
</tr>
<tr>
<td>------------------------------------</td>
<td>------------</td>
<td>--------------</td>
<td>-----------------------</td>
</tr>
<tr>
<td></td>
<td>Events, n (%)</td>
<td>Pregnancies, n</td>
<td>Events, n (%)</td>
</tr>
<tr>
<td>ASD (1,20,27,33,45)</td>
<td>1 (0.8)</td>
<td>123</td>
<td>0 (0.0)</td>
</tr>
<tr>
<td>VSD (20,27,33,45)</td>
<td>0 (0.0)</td>
<td>66</td>
<td>0 (0.0)</td>
</tr>
<tr>
<td>AVSD (13,45)</td>
<td>9 (10.2)</td>
<td>88</td>
<td>1 (1.1)</td>
</tr>
<tr>
<td>PS (17,27,33,35,36,48)</td>
<td>0 (0.0)</td>
<td>100</td>
<td>0 (0.0)</td>
</tr>
<tr>
<td>Ebstein (9,10,20,27,35,36)</td>
<td>5 (3.9)</td>
<td>127</td>
<td>4 (3.1)</td>
</tr>
<tr>
<td>AOS (14,17,20,21,27,28,33-36)</td>
<td>4 (2.4)</td>
<td>168</td>
<td>14 (7.0)</td>
</tr>
<tr>
<td>CoA (3,20,27,29,32,33,35,36,41-43)</td>
<td>0 (0.0)</td>
<td>297</td>
<td>3 (1.0)</td>
</tr>
<tr>
<td>CC-TGA (8,20,36,39)</td>
<td>3 (3.6)</td>
<td>84</td>
<td>6 (7.1)</td>
</tr>
<tr>
<td>TGA (6,12,15,16,20,22-24,27,33,35,36,47)</td>
<td>27 (15.6)</td>
<td>173</td>
<td>19 (10.8)</td>
</tr>
<tr>
<td>TOF (20,25,27,33,35,36,40,45)</td>
<td>13 (6.4)</td>
<td>204</td>
<td>5 (2.4)</td>
</tr>
<tr>
<td>PAVSD (7,26,46)</td>
<td>1 (2.5)</td>
<td>40</td>
<td>8 (20.0)</td>
</tr>
<tr>
<td>Fontan (4,5,11,18)</td>
<td>4 (16.0)</td>
<td>25</td>
<td>1 (4.0)</td>
</tr>
<tr>
<td>Cyanotic CHD (30,31,33,35)</td>
<td>3 (4.8)</td>
<td>63</td>
<td>14 (18.9)</td>
</tr>
<tr>
<td>Eisenmenger (2,19,27,29,37,38,44)</td>
<td>0 (0.0)</td>
<td>4</td>
<td>4 (21.1)</td>
</tr>
<tr>
<td>Overall</td>
<td>70 (4.5)</td>
<td>1,562</td>
<td>79 (4.8)</td>
</tr>
<tr>
<td>Expected occurrence (%)</td>
<td>&lt;0.5</td>
<td>&lt;0.5</td>
<td>&lt;0.5</td>
</tr>
</tbody>
</table>

Expressed as number of complications/completed pregnancy (percentage per completed pregnancy).

AOS = aortic stenosis; ASD = atrial septal defect; AVSD = atrioventricular septal defects; CC-TGA = congenital corrected transposition of the great arteries; CHD = congenital heart disease; CoA = aortic coarctation; Ebstein = Ebstein's anomaly; Eisenmenger = Eisenmenger syndrome; Fontan = patients after Fontan repair; PAVSD = pulmonary atresia with ventricular septal defects; PS = pulmonary valve stenosis; TGA = complete transposition of the great arteries; TOF = tetralogy of Fallot; VSD = ventricular septal defect.
SUMMARY

- Preconceptual counseling is critical
- Optimizing maternal condition prior to pregnancy is key
- Risk predictors are really only to be used in non-pregnant state
- WHO is the best in predicting outcome
- Underlying heart disease can be unmasked (Russia??) for the first time during pregnancy
- Most of the complications are from arrhythmias or heart failure but maternal death is a real problem
SUMMARY

• Intrapartum and immediate postpartum can be the highest risk time due to the wide fluctuations in volume status, anesthesia, ...

• Ideal anticoagulation for mechanical heart valve is heparin derivative from 6-12 weeks, followed by OAC until 36 weeks, and then heparin until delivery

• Transition back to OAC immediately postpartum since this is much better in preventing valve thrombosis
WHO SANG
SHOT THROUGH THE
HEART AND YOU’RE
SO VAIN, YOU GIVE
LOVE A BAD NAME!!
“Quick! Somebody call a lawyer!”