MANAGEMENT OF MATERNAL CONGENITAL HEART DISEASE

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AIM CONFERENCE
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OBJECTIVES
- Physiologic cardiovascular changes of pregnancy
- Epidemiology of congenital heart disease (CHD)
- Complications & outcomes of maternal CHD
  - Specific examples of CHD
- Management of maternal CHD

DISCLOSURES
- No financial disclosures

CARDIOVASCULAR CHANGES ANTEPARTUM
- Plasma volume ↑50%
  - Peaks @30-34 weeks
  - Systemic vascular resistance
    - 2nd tri mean BP 105/60
  - Sodium and water retention
- RBC volume ↑30%
  - Physiologic anemia
  - Hb < 11 g/dL 1st & 3rd tri
  - Hb < 10.5 g/dL 2nd tri
- Term total blood volume
  - 100mL/kg

http://childrensheartinstitute.org
CARDIOVASCULAR CHANGES ANTENATAL

- Cardiac output ↑30-50%
  - ↓Afterload, ↑preload
  - ↓Stroke volume
  - ↑Heart rate
  - 15-20 beats/min

- Uterine artery blood flow
  - 1st tri 50-60 mL/min
  - 3-6% of CO
  - Term 450-750mL/min
  - 12% of CO

- 30° degree left lateral decubitus position
  - Preserves preload
  - ↑CO 25-30%

HEMODYNAMIC CHANGES DURING PREGNANCY

NORMAL ECHOCARDIOGRAM CHANGES IN LATE GESTATION

- Physiologic hypervolemia of pregnancy:
  - Valvular annular dilatation
  - Multivalvular regurgitation R > L
  - Chamber enlargement
  - Small asymptomatic pericardial effusion

- No change in:
  - Left ventricular ejection fraction
  - Pulmonary artery pressures

HEMODYNAMIC CHANGES PERIPARTUM

- Intrapartum
  - Pain: ↑HR + BP
  - Contractions: ↑Preload
    - +500mL intravascular
  - Stage II of labor
    - ↑CO +50%
    - Valsalva
      - ↑Preload + CO

- Postpartum
  - ↑CO +30%
  - +500mL intravascular
  - Autodiuresis
  - 24-48hrs postpartum
  - Return to pre-pregnancy physiology
  - 6-8 wks
EPIDEMIOLOGY OF CHD

- CHD: #1 type of severe congenital anomaly
  - Incidence = 8.8/1000 live births¹
- Only 10-15% of CHD due to identifiable cause
  - Aneuploidy, genetic syndromes and teratogens²,³
- Increasing success of pediatric cardiac surgery
- More women with CHD reaching reproductive age
- Maternal CHD
  - Significant cause of morbidity and mortality
- Risk of recurrence in offspring = 5 to 6%

¹Hoffman et al. Am J Cardiol 1978
²Schinzel, AA. Prog Med Genet 1983

OVERVIEW OF CHD & MATERNAL RISK IN PREGNANCY

- High risk of major complications or death (25-50%)
  - Pulmonary HTN
    - Eisenmenger syndrome
  - Uncorrected coarctation of the aorta w/ proximal aortic dilatation
  - Marfan syndrome
    - Aortic root diameter >4.5cm
  - Severe AS
    - 'Single ventricle' with poor systolic function

Harris, IS. Prog Cardiovasc Dis 2011

OVERVIEW OF CHD & MATERNAL RISK IN PREGNANCY

- Moderate risk of complications (5-15%)
  - Unrepaired cyanotic defects
  - Systemic RV (TGA)
  - Well-functioning Fontan circulation (single ventricle)
  - Palliated TOF w/ severe pulmonic valve + RV dysfunction

- Low risk of complications (<1%)
  - Isolated ASD, repaired or unrepaired
  - Isolated VSD, repaired or unrepaired
  - Repaired coarctation w/ normal proximal root size
  - Repaired TOF w/ normal pulmonic valve + RV function

Harris, IS. Prog Cardiovasc Dis 2011

MATERNAL CHD: SUMMARY OF PREGNANCY OUTCOMES

- Literature review: Maternal CHD pregnancy outcomes 1985-2006
- N = 2491 pregnancies, 48 retrospective publications

<table>
<thead>
<tr>
<th>Type of Complication</th>
<th>Overall risk (%)</th>
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<tbody>
<tr>
<td>Heart failure</td>
<td>5</td>
</tr>
<tr>
<td>Arrhythmia</td>
<td>5</td>
</tr>
<tr>
<td>Major cardiac event</td>
<td>2</td>
</tr>
<tr>
<td>Preeclampsia</td>
<td>3</td>
</tr>
<tr>
<td>VTE</td>
<td>2</td>
</tr>
<tr>
<td>PTD</td>
<td>16</td>
</tr>
<tr>
<td>SGA</td>
<td>8</td>
</tr>
<tr>
<td>Perinatal death</td>
<td>2</td>
</tr>
</tbody>
</table>

Drenthen et al. J Am Coll Cardiol 2007
PREGNANCY COMPLICATIONS & OUTCOMES OF SPECIFIC CHD LESIONS

- Aortic stenosis (AS)
- Eisenmenger syndrome
- Tetralogy of Fallot (TOF)
- Transposition of the great arteries (TGA)

CONGENITAL AORTIC STENOSIS

- **Epidemiology**
  - 5% of CHD
  - Males > females

- **Pathophysiology**
  - Left ventricular hypertrophy
  - Preload dependence
  - Fixed obstruction

<table>
<thead>
<tr>
<th>Degree of AS</th>
<th>Mean gradient (mmHg)</th>
<th>AV area (cm²)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild</td>
<td>&lt;25</td>
<td>&gt;1.5</td>
</tr>
<tr>
<td>Moderate</td>
<td>25 - 40</td>
<td>1.0 - 1.5</td>
</tr>
<tr>
<td>Severe</td>
<td>&gt;40</td>
<td>&lt;1.0</td>
</tr>
<tr>
<td>Critical</td>
<td>&gt;70</td>
<td>&lt;0.6</td>
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</tbody>
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Yap et al. Int J Cardiol/2008

CONGENITAL AORTIC STENOSIS

- **Pregnancy complications**
  - Cardiac events 9.4%
  - CHF & atrial arrhythmias
  - Rarely TIA, syncope
  - Severe AS
    - 2nd in balloon valvuloplasty

- **Pregnancy outcomes**
  - HTN 11%
  - PTD 13%
  - SGA 13%
  - IOL 36%
  - Elective C/S 13%
  - Assisted 2nd stage 25%
  - Neonatal CHD 4%

EISENMENGER SYNDROME

- **Contraindication to pregnancy**
  - High maternal mortality
    - 50% in older series

- **Epidemiology**
  - Increasingly rare

- **Pathophysiology**
  - L → R shunt
  - AVSD, VSD, PDA, ASD
  - Pulmonary HTN
  - Right volume overload
  - R → L shunt

Yap et al. Int J Cardiol/2008

Kahn, ML. NEJM 1993
EISENMENGER SYNDROME

- Pregnancy complications
  - Worsening hypoxia
  - Arrhythmia
  - Sudden cardiac death
- Major cardiac event
  - 33%
  - MI/CVA/Death
- CHF 21%
- VTE 19%

Pregnancy outcomes

- TAB <5%
- SAB 12%
- PTI 16%
- PTD 65%
- SGA 38%
- Neonatal death 18%

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TETRALOGY OF FALLOT

- Epidemiology
  - 10% of CHD
  - Most common type of cyanotic CHD
- Pathophysiology
  - 1. VSD
  - 2. Overriding aorta
  - 3. Pulmonic stenosis
  - 4. RVH
  - Cyanotic heart disease
  - R → L shunt

TOF COMPLICATIONS IN PREGNANCY

- Residual post-surgical abnormalities
  - Persistent RV outlet obstruction and RVH
- Post-operative sequel
  - Pulmonic insufficiency → RV overload
  - Right-sided heart failure + atrial arrhythmias
  - RV outflow aneurysms
  - VSD patch leak
  - Ventricular arrhythmias → sudden cardiac death
  - Aortic root dilation with aortic insufficiency
  - RV & secondary LV dysfunction
- Risk of a cardiac event = 7%
  - SVT, heart failure, RV dilation, PE w/ CV collapse

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TOF PREGNANCY OUTCOMES

- SAB 27%
- IUFD 1%
- Preeclampsia 2%
- Cesarean 23%
- PTD 1%
- SGA 9%
- Fetal anomaly 6%
- Fetal CHD 4%

 установлен на странице 2004 из J Am Coll Cardiol

Risk factors for poor pregnancy outcome

- Severe pulmonary valve insufficiency
- RV systolic dysfunction

 установлен на странице 2004 из J Am Coll Cardiol
TRANSPOSITION OF THE GREAT ARTERIES

■ Epidemiology
  ■ 5-7% of all CHD
  ■ Most common cyanotic CHD diagnosed in newborns

■ Pathophysiology
  ■ Parallel circulations
  ■ Shunts decrease hypoxemia
    ■ ASD, VSD or PDA

TGA CARDIAC COMPLICATIONS

■ Pregnancy
  ■ Cardiac event 36%
    ■ 3rd tri CHF 15%
    ■ 3rd tri arrhythmia 13%
  ■ Digitalis 35%
  ■ Antiarrhythmic 20%

■ Postpartum
  ■ CCU/ICU 16%
  ■ Cardiac event 18%
  ■ CHF 71%
    ■ 1 required transplant
    ■ 1 new atrial baffle leak
    ■ Decreased O2 saturation
    ■ 1 atrial fibrillation
    ■ 1 sudden death
      ■ 6 wks PP
      ■ Had 3rd tri CHF w/ PTD

TGA PREGNANCY OUTCOMES

■ SAB 14%
■ TAB 9%
■ Live births 77%
■ HTN 8%
■ PTD 39%
  ■ <34 wks 18%
  ■ LBW 31%
  ■ IOL 42%
  ■ Cardiac reason 70%
  ■ Cesarean 28%
  ■ Cardiac reason 53%
■ CCU/ICU 17%

MANAGEMENT OF CHD

■ Multidisciplinary team approach
  ■ Anesthesia
  ■ Cardiology
  ■ Maternal-Fetal Medicine
  ■ Nursing
■ Monthly meetings
PRECONCEPTIONAL COUNSELING

- Identify maternal risks
  - Previous cardiac event
  - NYHA Class III or IV
  - Left heart obstruction
  - LVEF <40%
- Quantify maternal risks
  - CARPREG index¹
    - 1 point per predictor
    - 0 points 5% risk
    - 1 point 27% risk
    - 2+ points 75% risk

- Untreated CHD
  - Repair prior to pregnancy
- Genetic counseling
  - Risk of fetal CHD 5-6%²

¹Siu et al. Circulation 2001
²Hoffman, JI. Pediatr Cardiol 1995

NYHA FUNCTIONAL CLASSIFICATION

<table>
<thead>
<tr>
<th>CLASS</th>
<th>DESCRIPTION</th>
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<tbody>
<tr>
<td>I</td>
<td>Unlimited physical activity</td>
</tr>
<tr>
<td>II</td>
<td>Slight limitation of physical activity: comfortable at rest but symptomatic with ordinary activity</td>
</tr>
<tr>
<td>III</td>
<td>Marked limitation of physical activity: symptomatic with less than ordinary activity</td>
</tr>
<tr>
<td>IV</td>
<td>Symptomatic with any degree of physical activity and may have symptoms at rest</td>
</tr>
</tbody>
</table>

ANTEPARTUM CARE

- Terminate pregnancy?
  - Surgical correction
  - Improve outcomes in subsequent pregnancies
- Cardiac bypass during pregnancy
  - Pregnancy loss
  - PTD

- Serial CV assessments
  - Symptoms
    - SOB
    - Easy fatigability
    - ↓exercise tolerance
    - LE edema
  - Maternal ECHO

ANTEPARTUM CARE: CONSULTATION

- Euvolemia
  - Diuretics
- Beta blockers
- Antihypertensives
- Anticoagulation
  - Prosthetic heart valves
- Avoid nifedipine
  - Significant AS
  - Cyanotic CHD
- Fetal ECHO
- Antenatal testing
  - Start at 28 wks if cyanotic
- Serial fetal growth US
  - Start at 28 weeks
**INTRAPARTUM PLAN**
- Timing of delivery – schedule IOL?
- Early neuraxial blockade
- Mode of delivery
  - Expectant management
  - Assisted second stage
  - Scheduled cesarean
- A-line
- Telemetry
- SBE prophylaxis
- ICU bed

**PAST GUIDELINES SBE PROPHYLAXIS**
- 1997 AHA Consensus Statement
  - “It is reasonable to consider SBE prophylaxis before vaginal delivery in select patients at highest risk”
    - Prosthetic valves
    - Unrepaired or palliated (shunts/conduits) cyanotic lesions

**CURRENT GUIDELINES SBE PROPHYLAXIS**
- 2007 AHA Consensus Statement
  - “Antibiotic prophylaxis solely to prevent infective endocarditis (IE) is not recommended for GU or GI tract procedures”

**WHY THE CHANGE?**
- Only small number of IE prevented by prophylaxis.
- IE most commonly due to bacteremia of daily activities
  - IE rarely due to bacteremia from GI or GU tract procedures.
- Risk of adverse events from antibiotics exceeds the benefit from prophylactic antibiotic therapy.

2008 ACOG COMMITTEE OPINION #421

- "IE prophylaxis is no longer recommended for vaginal or cesarean delivery in the absence of infection, regardless of the type of maternal cardiac lesion."
- "Mitral valve prolapse is no longer considered a lesion that ever needs IE prophylaxis."
- "Only cardiac conditions associated with the highest risk of adverse outcome from endocarditis are appropriate for any IE prophylaxis."
  - Prosthetic cardiac valve
  - Previous IE
  - Unrepaired or palliated (shunts/conduits) cyanotic CHD
  - CHD repair with prosthesis material < 6 months ago
  - Residual defects preventing endothelialization of a prosthetic device

ANTIBIOTIC PROPHYLAXIS FOR INFECTIVE ENDOCARDITIS

- IV antibiotic 30-60 min prior to procedure
  - First-line is Ampicillin 2g
  - Alternatives
    - Cefazolin 1g
    - Ceftriaxone 1g
    - Clindamycin 600mg
    - Enterococcus
      - Vancomycin 1g

POSTPARTUM CARE

- Slow infusion of IV oxytocin
  - <2 U/min avoids hypotension
- Euvolemia
  - Strict I/Os
- Prevention of VTE
  - SCD’s
  - Compression stockings
  - Early/frequent ambulation

POSTPARTUM COUNSELING

- Contraception
  - PPTL safe
  - Mirena preferred IUD
  - Decreased blood loss
  - OCPs may be contraindicated
    - Pulmonary HTN
    - Valvular lesion
    - Surgical baffle
  - DMPA concerning for fluid retention → CHF
- Surgical repair of CHD prior to next pregnancy
Why is supine position especially dangerous for pregnant women with Aortic Stenosis?

A. Interferes with respiration
B. Decreases preload and cardiac output
C. Increases preload and cardiac output

What are risk factors for an adverse cardiac outcome in pregnancy?

A. Previous cardiac event
B. NYHA Class III or IV
C. Left heart obstruction & LVEF <40%
D. All of the above

Should a pregnant woman with corrected Tetralogy of Fallot receive SBE prophylaxis for delivery?

A. True
B. False

THANK YOU

- UCSF Cardiology: Adult CHD Service
  - Dr. Ian Harris
  - Dr. Elyse Foster, Director of the Adult CHD Service
  - Valerie Bosco, FNP-C, USfD
- UCSF MFM Division
  - Dr. Mari-Paule Thiet, Division Director
- UCSF OB Anesthesia
  - Dr. Mark Rosen
  - Dr. Mark Rollins
- UCSF L&D Nursing
  - Molly Kilison, RN, MS, CNS