Preconception Evaluation of Women with Complex Congenital Heart Disease

“Getting to Yes”*

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*Fisher and Ury
Objectives

• Assess risk for pregnancy
• Identify need for intervention prior to pregnancy
• Identify the women at risk for pregnancy and discuss alternatives
• Plan a safe pregnancy for mother and baby
• Anticipate cardiovascular problems during pregnancy
• Maintain functional status after pregnancy
• Plan subsequent pregnancies
• Formulate discussion with the patient and her family
Complex Congenital Heart Disease by WHO Classification

- Pulmonary arterial hypertension
- Aortic Stenosis with Left Ventricular out flow tract obstruction
- Bicuspid aortic valve with aortic dilation >45 mm
- D-Transposition of the Great Arteries
  - After Mustard-atrial switch with systemic right ventricle (EF <30%)
  - After Arterial switch
- L-Transposition of the Great Arteries
- Single Ventricle Variants
  - After Fontan repair
- Unrepaired Cyanotic Congenital Heart Disease
- Severe Native Aortic Coarctation
Preconception Evaluation

Assess maternal risk clinically
  CARPREG
  ZAHARA
  WHO score
Anatomic evaluation
Functional testing
Medication changes
Need for Intervention
Cardiac Risk Prediction: CARPREG Investigators

- Prior cardiac event (CHF, TIA, CVA) or arrhythmia
- NYHA functional class >II or cyanosis
- Left heart obstruction: MVA<2cm; AVA<1.5 cm; LVOT gradient >30 mmHg
- Systemic ventricular dysfunction: EF <40%

Maternal Cardiovascular Risk

0 pts  5%
1 pt   27%
>1 pt  75%

EHJ, 2011
# CARPREG Scores in Complex Congenital Heart Disease Patients


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Contraindication-When to say no

Pulmonary arterial hypertension
Uncorrected, complex cyanotic congenital heart disease
Systemic ventricular dysfunction
Failing Fontan
$O_2 < 85\%$

Contraception in Women with Complex CHD

Compliance and access issues
Early initiation or at least discussion begins at menarche
Estrogen-contraindicated with a history or risk of thromboembolic disease
Progestosterone-only-may be optimal if thrombosis risk or cyanotic
IUD
Sterilization: tubal occlusion or ligation
Pregnancy Risks

- Thrombosis with increasing hypercoaguability
- Worsening of ventricular function
- Impaired functional capacity during and after pregnancy
- CHD in offspring
- SGA baby due to maternal cyanosis
- Arrhythmia
- Infection
- Medications
Preconception Evaluation

History

Symptoms
Prior surgeries
Sequelae of surgery
Medications
Family history
Functional capacity (subjective)
Preconception Cardiac Evaluation

Diagnostic Testing

Echocardiography
Arrhythmia detection (AV block; QTc; RBBB; ischemic changes)
Stress testing for objection functional testing:
  - with echocardiography for functional reserve
  - cardiopulmonary for aerobic threshold
MR imaging
Cardiac catheterization
Transposition of the Great Vessels

D-TGA/Mustard Repair
D-Transposition of the Great Vessels

Atrial Switch

Case

Mustard repair at age 8 weeks

Pregnancy #1
  PCE: Stress echo: 12:30 RV 30-35% with normal response to exercise
  Mild AV regurgitation
  Uncomplicated; episodic junctional rhythm
  Vaginal delivery
  Fetal echo negative for CAD

Pregnancy #2
  PCE: Stress echo 13:46- normal RV response
  Uncomplicated pregnancy on Digoxin
  Vaginal delivery

Postpartum
  Echo: RV 45-50%

Third child- She adopted a child with CHD
D-Transposition of the Great Vessels

Atrial Switch

Mustard or Senning Repair

Clinical issues

- Systemic ventricular function
- Systemic AV valve regurgitation
- Risk of arrhythmia; sudden death
  - Atrial fibrillation
- Risk of heart block
- Pacemaker/AICD
- Baffle obstruction

Medications

- Anticoagulation
- Aspirin
- Afterload reduction

Effects of the hemodynamic changes of pregnancy

- Volume load
- Tachycardia
- Hypercoaguable state
Preconception Evaluation (ACC/AHA 1C)

Clinical assessment
- functional capacity
- medications
- symptoms

Objective functional assessment
- stress echo
- monitor for arrhythmias and/or heart block
D-Transposition of the Great Vessels: Arterial Switch
D-Transposition of the Great Vessels

Clinical Issues (to determine need for intervention)
- Coronary artery abnormalities
- Neoaortic valve regurgitation
- Pulmonary homograft
- Dilated aortic root (>55 mm)
- Pulmonary artery stenosis
- RVOT obstruction (gradient (>50 mmHg)

Effects of pregnancy
- Hypercoaguoble state
- Aortic wall changes
- Volume overload
- Arrhythmias
D-Transposition of the Great Vessels

Arterial Switch

Preconception Evaluation

Clinical assessment/functional capacity
Medications-assess for changes in anticipation of pregnancy
Pacemaker function
Echocardiography
Stress echocardiography
Risk of CHD in offspring and need for fetal echo.
L-Transposition of the Great Arteries (Congenitally Corrected Transposition)
L-Transposition of the Great Vessels

Preconception evaluation
- Access systemic ventricular function
- Access medications
- Access pacemaker
Risk of CHD in offspring
Potential complications
- Arrhythmia
- Pacemaker failure
- Congestive heart failure
- Cyanosis
- Stroke
Hypoplastic Left Heart Syndrome
Fontan Repair
Pregnancy Risks and the Fontan Patient

• Specific risks of pregnancy discussed
  – Thrombosis with increasing hypercoaguability
  – Worsening of ventricular function
  – Right-sided failure
  – Impaired functional capacity during and after pregnancy
  – CHD in offspring
  – SGA baby due to maternal cyanosis
  – Arrhythmia
  – Infection
  – Medications
Elements of the Discussion during Preconception Evaluation

• Maternal risk during pregnancy
  – Cardiac complications: arrhythmia, CHF
  – Medications
  – Interventions required
  – During Labor, Delivery and postpartum

• Fetal risk
  – CHD
  – Preterm delivery
  – Small-for-gestational age (maternal cyanosis)
  – Genetic (ex: 22q11)

• Maternal cardiac and functional status after pregnancy
  – Cannot predict

• Recommendations for treatment prior to conception

• Contraception

• Full evaluation prior to subsequent pregnancies
Thank You