



Preconception Evaluation of Women with Complex Congenital Heart Disease

“Getting to Yes”*

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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

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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%
>1pt	75%

Siu SC, Sermer, M, et al Circulation 2001: 104:515-521
EHJ, 2011

CARPREG Scores in Complex Congenital Heart Disease Patients

Cauldwell, M, Ghonim, S, et al: Preconception counseling, predicting risk and outcomes in women with mWHO 3 and 4 heart disease. *In J Cardiol* 234 (2017):76-80

Classification	N	CAREPREG 0	CARPREG 1	CARPREG 2	CARPREG 3
Systemic right ventricle	32	16	15	1	0
Pulmonary hypertension	14	9	4	1	0
Fontan circulation	15	2	11	2	0
Mechanical valve	14	8	6	0	0
Aortic stenosis	11	0	10	1	0
Severe systolic dysfunction	7	0	3	3	1
Peripartum cardiomyopathy	4	3	1	0	0
Mitral stenosis	4	0	4	0	0
Aortopathy >40 mm	1	1	0	0	0

Contraindication-When to say no

Pulmonary arterial hypertension

Uncorrected, complex cyanotic congenital heart disease

Systemic ventricular dysfunction

Failing Fontan

O₂ <85%

Ref: Elkayam, U. et al. J Am Coll Cardiol. 2016;68(5):502-16

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

Progesterone-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

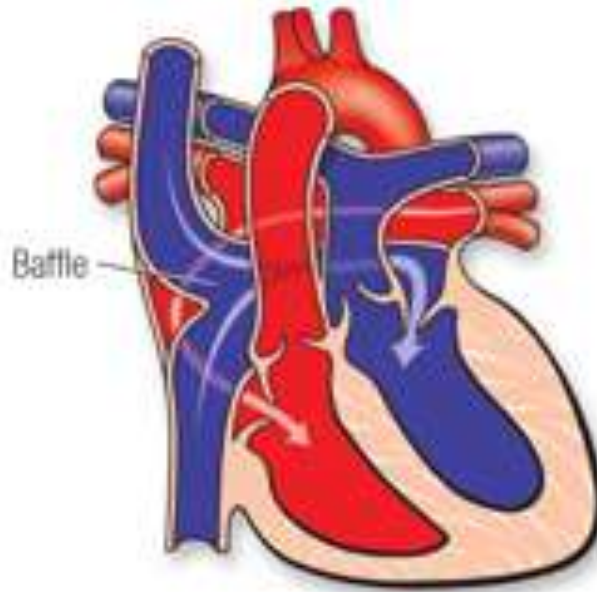
MR imaging

Cardiac catheterization

Transposition of the Great Vessels

D-TGA/Mustard Repair

Intra-Atrial Baffle (Mustard or Senning Procedure)



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

D-Transposition of the Great Vessels

Atrial Switch

Preconception Evaluation (ACC/AHA 1C)

Clinical assessment

- functional capacity

- medications

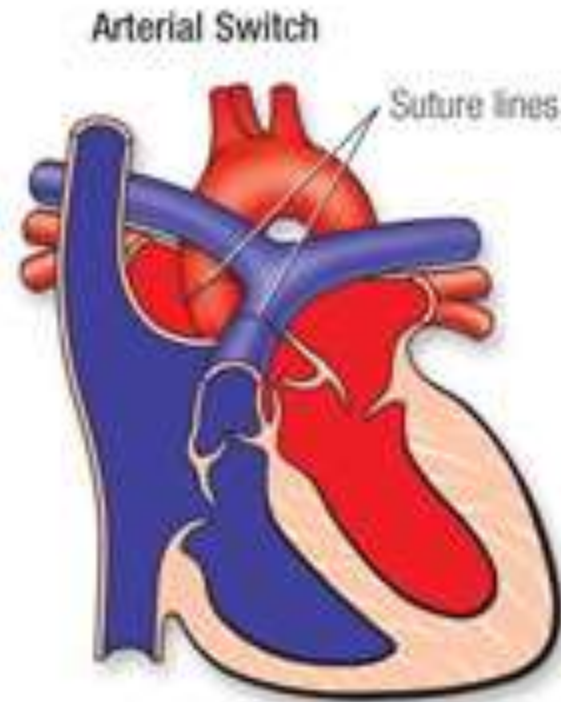
- symptoms

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

Arterial Switch

Clinical Issues (to determine need for intervention)

- Coronary artery abnormalities
- Neo-aortic valve regurgitation
- Pulmonary homograft
- Dilated aortic root (>55 mm)
- Pulmonary artery stenosis
- RVOT obstruction (gradient >50 mmHg)

Effects of pregnancy

- Hypercoagulable 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

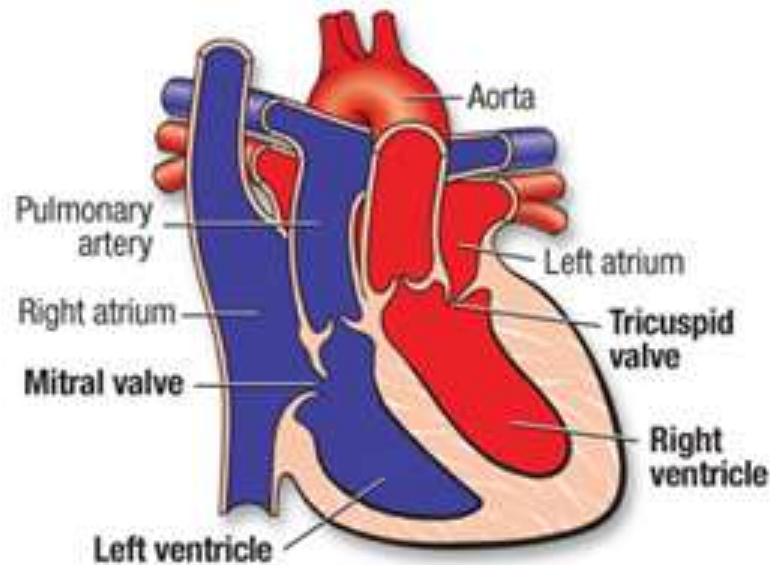
- Echocardiography

- Stress echocardiography

- Risk of CHD in offspring and need for fetal echo.

L-Transposition of the Great Arteries

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

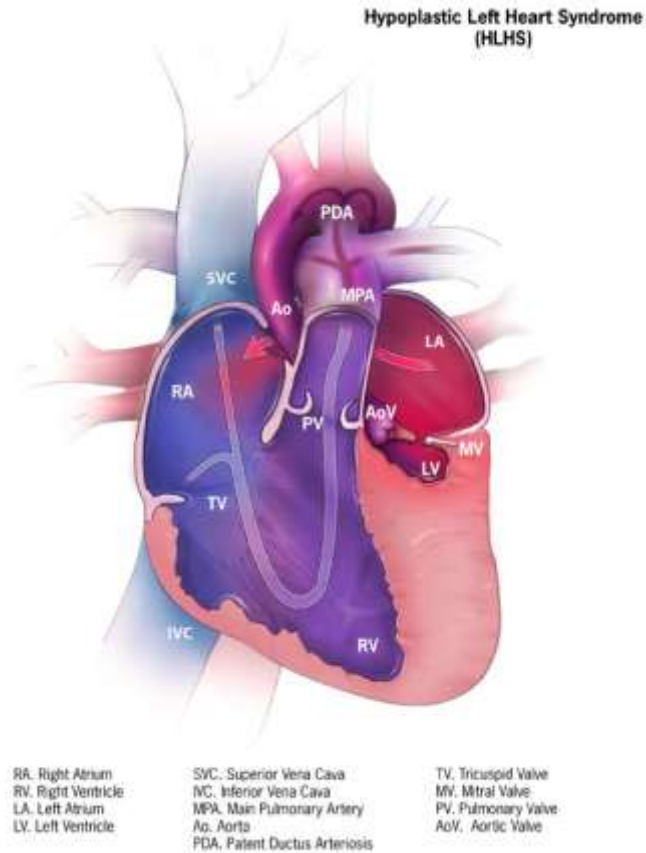
Pacemaker failure

Congestive heart failure

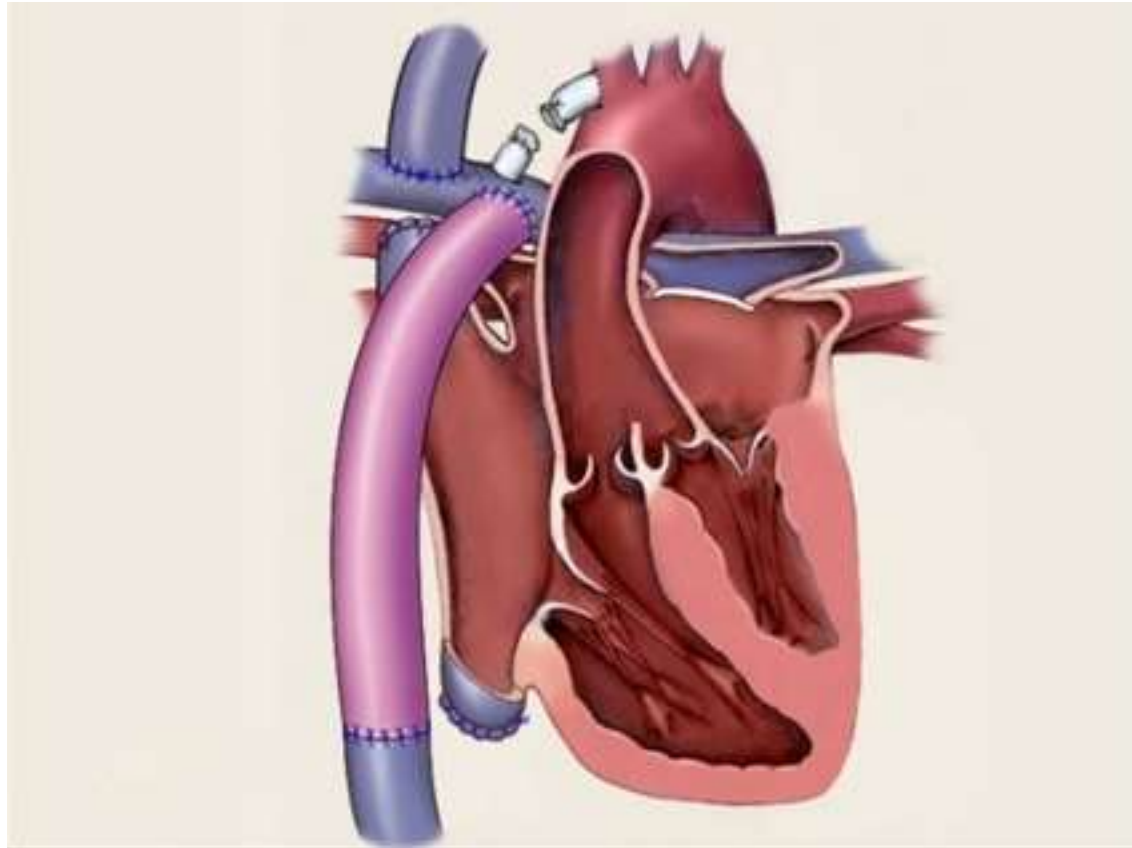
Cyanosis

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