Congenital Heart Disease and Pregnancy

Marla A. Mendelson, M.D.
Director
Northwestern Heart Disease and Pregnancy Program
Northwestern Adult Congenital Heart Center

Management Issues

- Expected hemodynamic changes of pregnancy
- Preconception evaluation
- Risk stratification
- Cardiac risks
- Risks to fetus
- Cardiac diagnosis during pregnancy
- Cardiac medications during pegnancy
- Cardiac intervention during pregnancy
- Labor and Delivery

Expected Hemodynamic Changes

Hemodynamic Alteration	Time of Peak Effect	Potential Risks
Cardiac output ↑30-50%	20-24 weeks	Women with limited cardiac function or reserve may develop congestive heart failure
Stroke volume ↑20%	20-24 weeks	Increase preload is a problem for obstructive lesions (mitral or aortic stenosis) or ventricular dysfunction
Heart rate ↑10-20%	Third Trimester	Tachycardia causes palpitations and impairs ventricular filling
Blood volume ↑40%	20-24 weeks	"Physiologic" anemia of pregnancy caused by less increased in erythrocyte mass
Peripheral vasodilatation	Throughout	↓blood pressure; ↓valvular regurgitation
↑Minute ventilation	Second Trimester	Sensation of tachypnea or dyspnea

Preconception Evaluation

Subjective functional capacity
Objective functional capacity
Natural/Un-natural history of cardiac problem
Right ventricular function
Pulmonic insufficiency
Arrhythmias/Risk of sudden death
Pulmonary artery pressure
Risk to offspring of CHD
Current drug therapy

Risk Stratification of Cardiac Disease:

Maternal Morbidity and Mortality

Immediate Risk (5-15%)

- **Mitral stenosis (moderate)**
- **Aortic stenosis (moderate)**
- **Repaired cyanotic congenital heart disease**
- **Coarctation of the aorta**

Risk Stratification of Cardiac Disease

Maternal Morbidity and Mortality

High Risk (25-50%)

Pregnancy contraindicated

Eisenmenger's
Primary pulmonary hypertension
Cyanotic heart disease
Marfan's Syndrome (aortic dilatation)
Aortic stenosis, severe
Dilated cardiomyopathy
NYHA function classes III, IV

Predictors of Pregnancy Complications

1802 women with CHD (ZAHARA)
1302 pregnancies

Cardiac Complications:	arrhythmias	4.7%
	heart failure	1.6%
O.B. Complications:	hypertension	12.2%

Risk Factors:
- Cyanotic CHD (repaired/uncorrected)
- Use of medication prior to pregnancy
- Left heart obstruction
- Mechanical valve
- Pulmonary A V valve regurgitation

Neonatal Complications:	premature birth	12%
	SGA	14%
	Mortality	4%

Associations: Cyanotic CHD, Mechanical valve; smoking; multiple gestations; cardiac medications

Drenthen, et al: Eur Heart J:2010:2124

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

Cardiac Diagnosis During Pregnancy

- Signs and Symptoms
- ECG-based studies
- Echocardiographic evaluation
- Stress testing
- Cardiac catheterization and angiography
- Nuclear isotopes

Congenital Heart Disease & Pregnancy
Shunt Lesions

Atrial Septal Defect

Unrepaired: theoretic risk of paradoxical embolus
arrhythmia
ventricular dysfunction
increase in pulmonary artery pressure
Eisenmenger: contraindicated (ACC/AHA:IA)

Repaired: residual shunt
arrhythmia

Risk to offspring: 8-10%

Congenital Heart Disease & Pregnancy
Shunt Lesions

Ventricular Septal Defect

Unrepaired: risk of endocarditis
increase pulmonary artery pressure
increase in shunt

Eisenmenger's: contraindicated (ACC/AHA:IA)
50% maternal mortality
risks persist after delivery and with termination
higher rates of fetal loss
worsening cyanosis

Repaired: residual shunt
associated valve disease
abnormal ECG (BBB)
persistent elevation of pulmonary artery pressure

Congenital Heart Disease & Pregnancy
Shunt Lesions

Patent Ductus Arteriosus

Unrepaired: Risk of pulmonary hypertension

Repaired (ligated): Pregnancy well-tolerated

Risk to offspring of CHD

Congenital Heart Disease & Pregnancy
Valvular Lesions

Mitral Valve

Stenosis – usually rheumatic in origin
risks: atrial fibrillation
CHF
thromboembolism

Insufficiency – usually well tolerated
may be seen after ASD pressure repair
may decrease during pregnancy
if severe may benefit from valve repair prior to pregnancy

Tricuspid Valve (insufficiency)

May be associated with other lesions
Normally well-tolerated

Valvular Heart Disease in Pregnancy
Mechanical valve

- Preconception evaluation
- Treatment options
- Controversy
- Your side of the pond

Congenital Heart Disease & Pregnancy
Valvular Lesions

Pulmonic Valve

- Stenosis
  - Mild/Moderate – pregnancy usually tolerated
  - Severe – consider valvotomy prior to pregnancy
  - May be associated with other CHD (post-repair)

- Insufficiency
  - Often/after complex CHD surgery such as Tetralogy of Fallot
  - Effect on pregnancy determined by RV size and function

Congenital Heart Disease & Pregnancy
Valvular Lesions

Bicuspid Aortic Valve

Aortic stenosis:

Pregnancy discouraged if AVA < 1.0cm^2

Supravalvular Aortic Stenosis

William's syndrome: family history of HTN, CAD or CVA

Discourage pregnancy if significant obstructive coronary involvement or aortic disease (ACC/AHA:2C)

Aortic Insufficiency

May decrease during pregnancy

Usually well-tolerated if LV function and size are normal

Aorta – may dilate with pregnancy (needs to be followed)

Congenital Heart Disease & Pregnancy
Aortic Disease

Coarctation of the Aorta

Associated with bicuspid aortic valve/aortic valve pathology

Risk of aortic dissection

Unrepaired: lower body supplied by network of collaterals

Repaired: persistent hypertension
risk of re-stenosis as adult
should have preconception evaluation

Congenital Heart Disease & Pregnancy

Aortic Disease

The Marfan Syndrome and Related Syndromes

Aortopathy of Bicuspid Aortic Valve

Pregnancy discouraged if aortic root > 40mm

Maternal Risks: aortic dissection/rupture
worsening of mitral regurgitation

Risk to Fetus: autosomal dominant inheritance

Tetralogy of Fallot: An Illustrative Case History: Part One

- The patient was born with TOF

  Infancy: Waterston-Cooley shunt

  Age 5: Intracardiac repair with a transannular patch

  Age 8: Pulmonic Carpentier-Edwards valve placed

  LPA stenosis repair

  Age 12: Balloon valvuloplasty of porcine pulmonic valve

- Subsequently she did well

Tetralogy of Fallot: An Illustrative Case History: Part Two

The patient presented during her first pregnancy

- Pregnancy #1: Serial echoes revealed increasing pulmonic pressures associated with worsening fatigue
- Postpartum: Found to have branch pulmonary stenosis which was stented
- Pregnancy #2: She becomes pregnant months after stent placed while on aspirin. Develops endocarditis of porcine prosthetic valve at 19 weeks. Treated with antibiotics
- Postpartum: Pulmonic valve replaced for increased gradient across valve
- Pregnancy #3: Uncomplicated
- Pregnancy #4: Chooses to deliver at home – lost to follow-up

Congenital Heart Disease & Pregnancy
Complex Congenital Heart Disease

Tetralogy of Fallot

Unrepaired: In setting of cyanosis, pregnancy not advised

Repaired: Preconception evaluation for residual abnormal hemodynamics and/or ventricular dysfunction

Assess for risk of arrhythmia/heart block

Severe pulmonary insufficiency-

tolerated if right ventricle normal at rest and after exercise

Increased risk of fetal loss and CHD

Associated with 22q11 – deletion (DiGeorge's)

Pregnancy After Tetralogy of Fallot Repair

204 women (CONCOR) (ZAHARA)
74 had 157 pregnancies
123 completed pregnancies

CV events		8.1%
	SVT, CHF, thromboembolic events	
Obstetric (HTN/DM/etc)		58.9%
Fetal/Neonatal events		33.9%
	mortality	6.4%
Predictors:	Cardiac medications prior to pregnancy	
	Arrhythmia prior to pregnancy	
	CHF associated with severe PI and RV dysfunction	

Balci, et al: Am Heart J: 2011:307

Congenital Heart Disease & Pregnancy
Complex Congenital Heart Disease

Ebstein's Anomaly

Preconception evaluation (ACC/AHA:IC)

Maternal risks:	increased tricuspid insufficiency RV failure arrhythmia/heart block cyanosis
Fetal risks:	low birth weight and fetal loss associated with maternal cyanosis CHD risk 6%

Congenital Heart Disease & Pregnancy
Complex Congenital Heart Disease

D-Transposition of the Great Vessels

Full evaluation prior to pregnancy (ACC/AHA:IC)

Clinical, functional & echocardiography:
AV valve regurgitation
Atrial arrhythmia
Systemic ventricular function

After atrial switch (Mustard, Senning) procedure

- risk of further RV (systemic) dysfunction

After Rastelli procedure (LV to Aortic baffle/closure of VSD)

assess for LV or RV obstruction
assess systemic ventricular function

After arterial switch (Jatene) procedure

case reports
assessment should include coronary arteries

Pregnancy after Arterial Switch

74 Women (ASO-1979-1989)
17 Pregnancies in 9 women (2 Centers)

13 term pregnancies
2/13 complications: mechanical MV thrombosis
VT in woman with LV dysfunction

Anticipated complications: coronary, sudden death, aortic root dilation

Problems with study: small population
mixed pathology
retrospective

Tobler, et al: Am J Cardiol: 2010: 417

Tricuspid Atresia/Single Ventricle: Pregnancy

- Preconception evaluation
- Risks during pregnancy
  - Arrhythmia
  - LV dysfunction
  - Edema
  - Ascites
  - Risk of anticoagulation
  - Spontaneous abortion
  - Premature birth
  - CHD in offspring

Congenital Heart Disease & Pregnancy

Complex Congenital Heart Disease

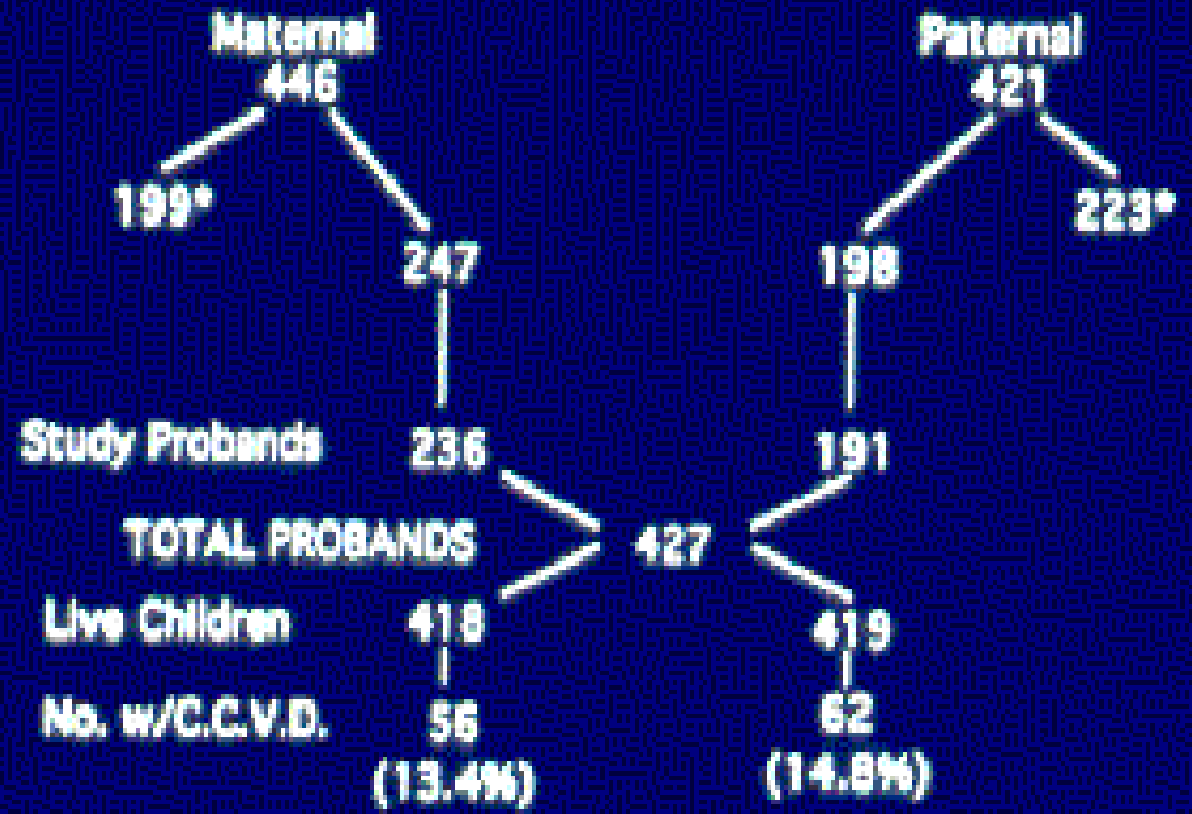
Single Ventricle Variants: Tricuspid Artesia
Double Outlet Right Ventricle

After Fontan:

Preconception Evaluation (ACC/AHA:IC)

Case Reports

Maternal Risks:	atrial arrhythmias; heart block
	ventricular dysfunction
	edema
	ascites
Fetal Risk:	spontaneous abortion
	premature birth
	warfarin embryopathy



Study group. *Nonparticipants. No. w/C.C.V.D. = number with congenital cardiovascular defect.

JACC, 1994
23:1459–67

Risk of Congenital Heart Disease to Fetus:

	Whittemore et al, AJC 50; 1982	Rose et al, JACC 6, 1985
Female patients studied	233	119
Pregnancies	482	385
Infants born	372	205
Overall incidence of CHD	16.1%	10.4%
Specific material lesions		
Pulmonary stenosis	7%	N/A
Aortic coarctation stenosis	12%	16.5%
Patent ductus arteriosus	9%	N/A
Ventricular septal defect	17%	N/A
Atrial septal defect	6%	12.3%
Cyanotic CHD (all types)	6%	N/A

*includes postoperative patients

Abbreviations: CHD, congenital heart disease, N/A data not available

Intervention During Pregnancy

- Medical therapy-relative safety of drugs
- Catheter-based interventions
  coronary and valvular
- Cardiac surgery-risks to fetus

Labor and Delivery

- General management recommendations
- SBE prophylaxis controversy
- Hemodynamic monitoring
- Mode of delivery
- Postpartum care and concerns

Management of Labor and Delivery
General Management

- Left lateral decubitis position
- Shortened second stage of labor
- Antibiotic prophylaxis
- Mode of delivery determined by obstetric indications
- Anesthetic management varies with cardiac disease

Management of Labor and Delivery Functional Class I & II Patients

General management
Clinical observation
Adequate analgesia
Maintain volume and blood pressure

Management of Labor and Delivery

Functional Class III & IV
Pulmonary Hypertension
Cardiomyopathy

Oxygen administration

Hemodynamic monitoring

Monitor at least 24 hours post-partum

Prophylactic heparin until ambulatory

Monitoring During Labor & Delivery

Indications: Invasive HemodynamicMonitoring

- NYHA III & IV
- Poor ventricular function
- Mitral stenosis
- Aortic stenosis
- Pulmonary hypertension
- Pulmonary edema/PPCM
- Severe hypertension