Maternal Congenital Heart Disease in Pregnancy

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Outline
- How common?
- Our role in preconception period
- Risks
  - Fetal
  - Maternal
- Prediction of risk
- Cardiovascular changes in pregnancy
- Specific lesions
- Labor & Delivery management pearls
- What to watch for

Maternal congenital heart disease
- Up to 85% of patients with congenital heart disease now survive to childbearing age
  - Advances in medical and surgical treatment
- Congenital heart disease is most prevalent form of structural heart disease (66%) affecting pregnancy outcomes
- Despite previous counseling to avoid pregnancy, not all do... and many women have sufficient cardiac function to safely carry a pregnancy

Maternal congenital heart disease
- Unfortunately remains a cause of maternal morbidity and mortality
- US 1995-2006
  - 1.4% delivery hospitalizations complicated by chronic heart disease
  - Severe complications during hospitalizations for delivery among women with chronic heart disease were more common in 2004-2006 than 1995-1997

Cardiovascular disease in pregnancy
- Cardiovascular disease complicates 1-4% of all pregnancies
  - Hypertension is most common acquired heart disease
  - Congenital is most common preexisting condition
- Cardiovascular disease is the leading cause of maternal mortality in the United States and California
  - >33% of all pregnancy-related deaths in US (2002-2006)

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Cardiovascular disease in pregnancy

- Data from the California Pregnancy Associated Mortality Review (CA-PAMR) of deaths occurring from 2002-2006
  - Only a small fraction of these women had a known diagnosis of cardiovascular disease prior to death
  - Most women who died had presented with symptoms either during pregnancy or after childbirth
  - A significantly higher proportion of women sustain short- and long-term morbidity due to undiagnosed or delayed diagnosis of cardiovascular disease as evidenced by the fact that one of every three intensive care admissions in pregnancy and postpartum period are related to cardiac disease.
  - 25% of these deaths may have been prevented if heart disease was diagnosed earlier.

https://www.cdc.gov/reproductivehealth/maternalinfanthealth/pregnancy-relatedmortality.htm

Maternal congenital heart disease

- Death is rare
- Risk of maternal complications
- Risk of fetal complications

Preconception counseling

- Many women with CHD are unaware of pregnancy risks
- Ideally preconception counseling with risk stratification
  - Possible repair prior to pregnancy
  - Determine cardiac function
    - ECHO, stress test, oxygen saturation
  - Discuss medication use
  - Discuss maternal risks
    - Long-term outlook
    - Discuss fetal risks
  - Delivery recommendations
  - Pregnancy alternatives such as adoption or surrogacy

In the real world...

- For patients who present already pregnant
  - We have to ask history to allow risk stratification
    - Low, moderate and high risk
  - Example: termination in the first trimester appears to be safer than pregnancy continuation in setting of Eisenmenger’s
  - Some women with congenital heart disease should be treated with anticoagulant medication
American Heart Association Scientific Statement:
Management of Pregnancy in Patients with Complex CHD

- Important that all practitioners who will be managing pregnant women with congenital heart disease have current information
  - Preconception counseling
  - Diagnostic evaluation to determine maternal and fetal risk
  - How to manage
  - When to refer to a regional center with expertise

What are the risks?

- Cyanotic maternal CHD increases risk for miscarriage, growth restriction and preterm delivery
  - 96 pregnancies in women with cyanotic CHD
    - 43% resulted in live birth
    - 37% premature
    - Rate of SAB increased in parallel with maternal hypoxemia
    - Mean term birth weight 2575g compared to 3500g
  - Even when cyanosis is mild, R to L shunt increases in response to fall in systemic vascular resistance thus risk of fetal loss is still important
  - Maternal medications may impact fetus

Fetal congenital heart disease

New York Heart Association Functional Classification

- Class I: Asymptomatic: no fatigue, palpitations or dyspnea
- Class II: Slight limitation in physical activity: symptoms with more than normal activity (>2 blocks)
- Class III: Marked limitation in physical activity. Symptoms with normal activity (>2 blocks)
- Class IV: Unable to carry on any physical activity without discomfort. Symptoms at rest

What are the risks?

- Inheritance
  - Background risk for fetal CHD is 0.8%
    - If one parent has CHD, risk to fetus is 3-6%
      - Higher if parent is mother
      - Higher risk if sibling has CHD
  - Underlying genetic syndrome like Marfan or 22q11 deletion syndrome
    - Up to 50% chance of passing to fetus
    - Variation among lesions in rate of recurrence of same lesion
      - VSD: 55% concordance (17 of 31 recurrences)
      - Coarctation: 13% (2 of 15 recurrences)
      - Hypoplastic left heart: 33% (4 of 12 recurrences)

How to classify maternal risk: CARPREG

1. NYHA class II or cyanosis
2. Obstruction – A/A <1.5 cm2, MVA <2 cm2 or peak LVOT gradient 30 mm Hg
3. Previous history – CHF, arrhythmia, TIA, or stroke
4. Ejection fraction <60%


4%
27%
62%

WHO Classification of risk

• Class I: no detectable increased risk of maternal mortality and no or mild increase in morbidity
  – Small PDA
  – Mild pulmonic stenosis
  – Mitral valve prolapse
  – Successfully repaired VSD, ASD, PDA
  – Isolated atrial or ventricular ectopic beats

WHO Classification of Risk

Class II: Small increased risk mortality, moderate morbidity
• Unrepaired ASD/ VSD
• Most arrhythmias
• Repaired Tetralogy of Fallot without pulmonic stenosis or regurgitation
• Hypertrophic cardiomyopathy
• Repair of coarctation
• Marfan with aortic dimension <40 mm

Class III: Significantly increased risk of mortality, severe morbidity
• Mechanical valve
• Right ventricle serving as systemic ventricle
• Fontan circulation
• Unrepaired cyanotic heart disease
• Other complex CHD
• Bicuspid aortic valve with ascending aorta 40-45 mm
• Marfan with aortic dimension 40-45 mm

Class IV: Extremely high risk of mortality and severe morbidity
• Severe mitral stenosis
• Symptomatic severe aortic stenosis
• Bicuspid aortic valve with aorta >50 mm
• Marfan with aorta >45 mm
• Severe systemic ventricular systolic dysfunction (EF <30%, NYHA III & IV)
• Native severe coarctation
• Significant pulmonary hypertension of any cause
Pregnancy is contraindicated
Caveats

- Recommended that even women with structural heart disease who have undergone "repair" not be considered corrected
  - Residual disease possible
  - Response to physiology of pregnancy can be unpredictable

Reducing risk

- Women with moderate or high risk lesions
  - Followed by MFM and Cardiology
  - Monthly ultrasounds for fetal growth surveillance
  - Detailed fetal echocardiogram
  - Antepartum fetal monitoring
    - As early as 26 weeks gestation
  - Anesthesia consultation
    - Need adequate preloading, slow onset
    - Delivery planning
      - Vaginal delivery preferred due to smaller shift in blood volume, decreased risk for blood clots and infection
      - Cesarean reserved for certain high risk patients
      - Marfan syndrome with dilated aortic root
      - Hemodynamic monitoring
        - Arterial lines, Swan-Ganz pulmonary artery catheters

What happens to the heart during pregnancy?

- Cardiovascular changes begin as early as 5-8 weeks and peak in late second trimester
- Cardiac decompensation in women with preexisting CHD often coincides with this peak
- Bottom line: the heart works harder

Final Common Pathway

- Heart failure
- Arrhythmias
- Death

During labor and delivery: Higher risk

- Hemodynamic changes are profound!!
- A uterine contraction displaces 300-500 cc of blood into circulation
- Stroke volume increases
- Cardiac output increases 50% with each contraction
  - Up to 75% above baseline
  - Heart rate and blood pressure may rise due to pain and anxiety
- Mean arterial pressure rises
- Blood loss, extreme fluid shifts
Postpartum can be the highest risk period for many of these women

- Relief of IVC compression
  - Increased venous return
  - Increased cardiac output
  - Autodiuresis
- Thromboembolic events

Atrial septal defect

- Most common repaired or unrepaired congenital cardiac defect in pregnancy
- Diagnosis may be missed
  - Physiologic flow murmur normal during pregnancy
- Usually well tolerated during pregnancy
- Rarely large ASDs may develop
  - Heart failure
  - Arrhythmias
    - Especially with advancing maternal age
  - Paradoxical embolus

Ventricular septal defect

- Common but most close by 2-5 years old
- Pregnancy usually tolerated well
- Rarely can develop
  - Heart failure
  - Arrhythmias
- 4-11% recurrence in infants born ↑ with VSD

Eisenmenger’s syndrome

- Reversal of left to right shunt (with ASD, VSD, PDA) due to pulmonary hypertension
- Pulmonary hypertension limits adaptive responses to circulatory changes
- Only 15-25% pregnancies progress to term
  - Significant fetal risks with cyanosis
  - IUGR
  - Preterm delivery

Eisenmenger’s syndrome

- Reported mortality 30-50%
  - Unchanged in past 50 years
- Most deaths occur during delivery or first week postpartum
  - Fall in SVR augments R to L shunt: Fatal hypoxemia
  - Thromboembolism
  - Volume depletion
    - Augments R to L shunt and leads to intense cyanosis
    - Preeclampsia
- Fixed pulmonary arterial resistance cannot accommodate to the hemodynamic fluctuations of labor, delivery, and puerperium

Cyanotic congenital heart disease

- Secondary erythrocytosis
  - Phlebotomy may be indicated in women with hematocrit > 65%
- Hyperviscosity symptoms: headache, loss of concentration, fatigue, myalgias
- “Relative” anemia is not well tolerated
  - Can have third-spacing volume overload
Mitral valve prolapse
- Rarely causes any complications during pregnancy
- NOT an indication for endocarditis prophylaxis

Mitral regurgitation
- Usually well tolerated in pregnancy
  - Decreased SVR partially compensates for volume overload due to regurgitant valve
  - But new arrhythmia or severe HTN can disrupt this

Aortic stenosis
- Most common etiology in women of childbearing age: congenitally bicuspid valve
- Bicuspid aortic valve and aortic dilation are at risk for spontaneous aortic dissection
  - Prophylactic repair recommended if:
    - Dilated to 40-45 mm before pregnancy
    - Progressive aortic regurgitation
    - Rate of increase dilation >5 mm per year
  - Need aggressive blood pressure control

Aortic stenosis in pregnancy
- Fixed cardiac output
- Asymptomatic
  - Restrict physical activity
  - Close observation
- Symptomatic
  - Mechanical relief
    - Aortic valve balloon valvuloplasty
    - Aortic valve replacement
  - Cardinal rule: Avoid hypotension!!
    - Spinal/epidural may cause disastrous vasodilation
    - Postpartum hemorrhage can be deadly
    - The answer is not lasix

Aortic stenosis in pregnancy: Case
- 36 yo Hispanic G2P1 presented at 16 3/7 weeks
- NYHA functional class II
- EKG: Normal sinus rhythm
- Echocardiogram:
  - EF 66%
  - Bicuspid aortic valve: AVA 0.9 cm², Peak gradient 64 mm Hg
- Symptoms of CHF at 27 weeks
- Percutaneous aortic balloon valvuloplasty @ 28 weeks
  - AVA 1.2 cm², Peak gradient 40 mm Hg
- Delivery at 36 weeks

Coarctation of the aorta
- Narrowing in region of ligamentum arteriosum
  - May present with resistant hypertension in childhood
- Can be associated with
  - ASD and VSD
  - Intracranial aneurysms
  - Hypertension
  - Dissection
- Hypotension in distal vascular beds can compromise uteroplacental blood flow
- Cardinal rule: Avoid hypotension
Marfan syndrome

• Autosomal dominant
  – 50% risk to fetus but variable penetrance
• Estimated 1% risk serious cardiac complications
• Hypervolemic and hyperdynamic circulatory state and/or hormonal effects of pregnancy can contribute to risk of dissection
• Mortality risk increases if aortic root > 40 mm
  – > 45 mm: 10% risk mortality
  – 2011 European Society of Cardiology strongly recommends preconception repair of aortic root aneurysms > 45 mm

Marfan syndrome

• Serial echocardiogram
• Aggressive treatment of hypertension with beta blockers
• Cardinal rule: Avoid valsalva
  – One category of women where elective cesarean under general anesthesia may be recommended to maximize hemodynamic control
• Risk of dissection appears to peak 3-20 days postpartum

Management of Labor & Delivery

• If concerns regarding heart function
  – Consider induction under controlled circumstances
• Mechanical methods of cervical ripening preferred if patient cyanotic to avoid drop in SVR or BP
• Labor in left lateral decubitus position, minimizes compression of aorta/IVC

Risk of infection

• Retrospective review 1985-2006
  – 2491 pregnancies in women with CHD
  – Rate of endocarditis 0.5% (1372 completed preg)
• But there are high rates of maternal and fetal mortality with endocarditis in pregnancy
• However, routine antimicrobial prophylaxis for bacterial endocarditis is not recommended in most women with CHD during pregnancy and delivery

Endocarditis prophylaxis

- Highest risk with prosthetic valve, history of prior infective endocarditis
- Recommended for:
  - Repaired CHD with prosthetic material or device during first six months after the procedure
  - Unrepaired cyanotic CHD (including with shunts and conduits)
  - Repaired CHD with residual defect at the site or adjacent to site of prosthetic device
  - Prosthetic heart valves

Cardinal rules

- Avoid hypotension in:
  - Aortic stenosis
  - Coarctation of aorta
  - Hypertrophic obstructive cardiomyopathy
  - Cyanotic heart disease
- Avoid tachycardia in all:
  - Especially in mitral stenosis

Cardinal rules

- Vast majority of cardiac cases present with sign and symptoms on more than one occasion
- Sudden cardiac deaths without prior warning are rare
- Thus there are certain symptoms that all providers of obstetrical care should be aware of

Physiologic changes

- Signs and symptoms of Pregnancy that mimic heart disease
  - Affect diagnostic tests

Common cardiac signs and symptoms

- Fatigue
- Shortness of breath
- Orthopnea
- Palpitations
- Light headedness/dizziness
- Edema
- JVD
- Murmurs
  - 96% pregnant women have “functional murmur”
  - Third heart sound is common

What’s not normal

- Exertional chest pain
- Paroxysmal nocturnal dyspnea
  - Waking up due to shortness of breath
- New orthopnea
  - Shortness of breath with lying down
- Diastolic murmurs
- Sustained arrhythmias
- Pulmonary edema
  - Bilateral infiltrates on CXR: consider cardiac unless proven otherwise
- New onset asthma in pregnancy: consider cardiac disease
Resources

- www.myheartsisters.com
- www.womenheart.com
- American Heart Association scientific statement on Management of Pregnancy in Patients with Complex Congenital Heart Disease
- European Society of Cardiology guidelines on management of cardiovascular diseases in pregnancy

What is the bottom line?

With proper management, most women with CHD can have a successful pregnancy. Talk with your ACHD cardiologist and high-risk OB before you get pregnant. Taking care of yourself is the best way to ensure the well being of your baby. Live a healthy life, avoid harmful substances, and have regular appointments with your ACHD cardiologist and OB where you can ask questions. Remember, constant communication with your healthcare team before, during and after pregnancy is key.