Risky Business: Contraception and Pregnancy in Congenital Heart Disease

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Objectives

• Review congenital heart disease and their sequela
• Identify the contraceptive and obstetric challenges for the CHD patient
• Apply this knowledge to the counseling and management of the CHD patient
It will be a long day when......

- 17 year old sexually active patient
- She has had two heart surgeries but does not know details
- On lisinopril
- She “never” wants to have children and wants to discuss options for contraception
- Her mother wants to ask about risk of pregnancy

&%#@**!!
Your thoughts

- What is her cardiac defect?
- What was her cardiac surgery?
- What effective contraceptive can be used safely?
- What is her long term prognosis?
- What is risk of pregnancy in the future if she “changes her mind”?
Gaps in Knowledge

Adolescents with CHD / parents often do not know:

- Nature of **heart defect**
- Function and side effects of **medication**
- Impact on **exercise** capacity and limitations if any
- **Risk factors** for infective endocarditis
- **Hereditary** nature of CHD
- Risk of **pregnancy** and **birth control options**
- Need for **life long** cardiac care

Kovacs et al.  JACC 52:7:2008: 577-86
Fernandes S et al. Pediatrics 2011;128:1489-95
Cheuk DKL et al.  Heart 2004;90:435-9
Gaps in Parental Knowledge

- 156 parents of children with simple CHD
- Questionnaire of ten items including nature of the heart defect, impact on exercise and the hereditary nature of CHD
  
  64% Correct
  8% Incorrect
  26% Don’t know

Cheuk DKL et al.  Heart 2004;90:435-9
What Adolescents Understand

- 91 adolescents (median age 17) after transfer
- 78% knew they could be sexually active
- 20% knew of hereditary nature of CHD
- Women: (47%)
  - 14% and 35% did not know if IUD or OCP suitable
  - 63% of women did not know if risk with pregnancy
  - 25% did not know correct answer

Van Deyk et al. Am J Cardiol 2010;106c:1803-07)
Sexual behavior / Reproductive concerns

- Survey 144 adolescents (16-18) and 212 young adults (19-20) with moderate and complex CHD
  - Fewer adolescents (14%) were sexually active than healthy peers (33 to 42%)
  - Fewer young adults (48%) were sexually active than healthy peers (58 to 64%)
  - But 72% of sexually active adolescents and 36% young adults were engaging in potentially risky sexual behavior

Reid et al. Int J Cardiol. 2008;125:332-8
Reproductive concerns

- Patient concern about fertility

Approximately 10% of men were moderately or very concerned about their fertility.

Over 30% of women with complex CHD were moderately or very concerned about their fertility.

Approximately 10% of men were moderately or very concerned about their fertility.

What women are not told

- Questionnaire to determine knowledge gap:
  - 116 women (mean age 31)
  - 51% recalled information about birth control
  - 37% denied knowing increased pregnancy risk
  - 18 patients with high risk pregnancy: Only 9 had received information
  - 98 patients with low risk for pregnancy: 18 had been advised to avoid pregnancy
What Patients Know

- 536 women with CHD aged 29 (18-75)
- 76% had moderate or severe defects
- Level of information on 1-10 scale regarding:
  - Sexuality was 3.5 and contraception was 5
- Information from physician about:
  - Pregnancy 52% and contraception 57%
  - Discussion initiated by physician in 56%
- Two thirds had NOT discussed sexual health issues, contraception or pregnancy

Not only cardiac patients

- Patients with other chronic disease including CF and rheumatologic diseases

- Rheumatologists report time constraints, organizational issues and physician beliefs as barriers to having these conversations

Johannesson et al. Patient education and counseling 1998;34:15-123
Who should have this conversation?

- Congenital cardiologist?
- Cardiologist and family planning doctor?
  - Monthly regional clinic in Newcastle
- Pediatrician or primary care provider?
  - One third of all adolescents have annual visit without any mention of sexuality issues
- Adolescent medicine clinic/ gynecologist?
- Family with medical guidance?

Rogers et al. J Fam Plann Reprod Health care 2007;33: 17-21
Epidemiology

Incidence is 1/100 live births
Risk CHD with subsequent pregnancy (3%)
Risk of infant with CHD if parent has CHD (10-15%)
Survival patterns changing
> 90% of newborns with CHD expected to survive to adulthood
Now more adults than children with CHD
Survival and surgical milestones in CHD

% Survival to Adulthood

- BT-Shunt
- Fallot
- Atrial Switch
- Fontan
- Arterial switch
- Norwood


Pediatric vs Adult Prevalence

US estimate: 1.4 million adults with CHD

Marelli et al, Circulation, 2014;
Gilboa et al, Circulation, 2016
CARDIAC DEFECTS
Classification

- **Acyanotic / Cyanotic**

- **Obstructive/ Shunting/ Mixing**

- **Simple/ Moderate/ Complex**
<table>
<thead>
<tr>
<th>Simple</th>
<th>Moderate</th>
<th>Complex</th>
</tr>
</thead>
<tbody>
<tr>
<td>ASD (secundum)</td>
<td>Aortic stenosis</td>
<td>Cyanotic heart disease</td>
</tr>
<tr>
<td>VSD (small)</td>
<td>Coarctation of the aorta</td>
<td>Transposition great arteries</td>
</tr>
<tr>
<td>Small PDA</td>
<td>Tetralogy of Fallot</td>
<td>Pulmonary atresia</td>
</tr>
<tr>
<td>Mild pulmonary stenosis</td>
<td>Moderate pulmonary stenosis</td>
<td>Tricuspid atresia</td>
</tr>
<tr>
<td>Repaired ASD with no residual</td>
<td>VSD with associated defect</td>
<td>Single ventricle</td>
</tr>
<tr>
<td>Repaired VSD with no residual</td>
<td>AVSD</td>
<td>Eisenmenger syndrome</td>
</tr>
</tbody>
</table>
Atrial Septal Defect

- May be diagnosed during adolescence
- Risk of atrial arrhythmias
- Closed surgically or by device
- If residual ASD, risk of a paradoxical embolus
- **Avoid estrogen** if residual ASD
- May be at risk for pulmonary hypertension
- Pregnancy usually well tolerated

Ventricular Septal Defect

- May be small with no sequela
- At risk of small residual shunt and paradoxical embolus
- **Avoid estrogen** if residual VSD
- Evaluate for pulmonary hypertension
- If closed surgically with good result, pregnancy well tolerated

Pulmonary valve stenosis

- May be mild with no intervention
- If moderate to severe: balloon valvuloplasty or surgical valvotomy
- No contraindication to Estrogen in isolated PS
- Pregnancy well tolerated if mild-moderate PS
- If severe, valvuloplasty before pregnancy

Tetralogy of Fallot

- Usually well after surgery in infancy but may need pulmonary valve replacement
- Estrogen tolerated unless residual VSD or poor function
- Pregnancy usually well tolerated with 8% risk of CV event — mostly arrhythmia
- Pregnancy associated with increased RV size

Coarctation of the aorta

- May present in adolescence and adulthood
- Associated with BAV and aortic dilation
- Risk of aortic dissection or aneurysm rupture
- Common defect in Turner patients
- Pregnancy is usually well tolerated with BP fluctuation at higher baseline
- **Consider in difficult to treat hypertension**

Aortic stenosis

- Present with heart murmur
- Symptoms of heart failure/ poor cardiac output
- Estrogen tolerated unless depressed function
- Surgical or catheter valvuloplasty
- Avoid mechanical valves ....
- Severe: pregnancy is contraindicated. Increased cardiac output on pressure loaded LV
- Consider intervention before pregnancy
D-TGA

- After arterial switch (Jatene)
  - Described in 1975 but more since 80s
  - Risk of coronary disease
  - Estrogen and pregnancy well tolerated

- After atrial switch (Mustard or Senning)
  - Increased risk of systemic (RV) failure and atrial arrhythmia

L-TGA

- May be diagnosed during in adolescence or adulthood
- At risk of heart block
- Estrogen tolerated unless poor function
- Need pre pregnancy evaluation and metabolic stress test

Single ventricle

- Mixture of cardiac defects
- Multiple surgeries culminating in Fontan
- Increased risk of thrombosis
- Estrogen contraindicated
- Risk of atrial arrhythmias and heart failure
- High risk pregnancy and risk of prematurity
- Collaborative peripartum care

Eisenmenger/pulmonary hypertension

- Estrogen contraindicated
- Fluid retention with progesterone
- Vagal reaction with IUD
- Pregnancy “contraindicated” with high mortality
- Risk of hyperviscosity and paradoxical embolus
- Recent improved outcome with pulmonary vasodilator therapy

“Contraindications”

- **ESTROGEN**
  - Pulmonary hypertension
  - Eisenmenger syndrome
  - Decreased LVEF (< 40%)
  - Single ventricle
  - Cyanosis

- **PREGNANCY**
  - Severe aortic stenosis
  - Mitral stenosis
  - Pulmonary hypertension
  - Eisenmenger syndrome
  - Marfans with dilated aortic root
  - Decreased LVEF (< 40%)
  - ?? Single ventricle
  - Cyanosis
Risk to fetus?

- Determined by maternal functional status
- 30 to 50% in NYHA Class IV (based on acquired heart disease)
- Cyanosis threatens growth, development and fetal viability
- Rate of spontaneous abortion is high and increases with worsening hypoxemia
- Increased risk of CHD in infant
CONTRACEPTION
# WHO Risk Classification

<table>
<thead>
<tr>
<th>WHO CLASS</th>
<th>Risk for Contraceptive method</th>
<th>Risk for Pregnancy</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>No restriction</td>
<td>No increased risk of mortality or morbidity</td>
</tr>
<tr>
<td>2</td>
<td>Advantages outweigh risks</td>
<td>Small increase M &amp; M</td>
</tr>
<tr>
<td>3</td>
<td>Risk outweigh advantages: use alternative unless patient accepts</td>
<td>Significant increased M &amp; M</td>
</tr>
<tr>
<td></td>
<td>risks and risk of pregnancy is high</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>UNACCEPTABLE HEALTH RISK</td>
<td>Extremely high risk of M &amp; M</td>
</tr>
</tbody>
</table>

Thorne et al. J Fam Plann Reprod Health Care 2006;32: 75-81
Combined hormonal contraceptives

- Adverse cardiac effects:
  - Increased risk of thromboembolic complications
  - Reduce HDL and increase LDL
  - Increased BP

- Contraindicated if:
  - Cyanosis related to any intracardiac shunt
  - Fontan
  - Cirrhosis (consider in Fontan)
  - Poor cardiac function
Combined hormonal contraceptives

- Consider other **medication** and metabolism
- Consider other **risk factors**: obesity, smoking, DM and hypertension

Questions:
- What about patients on warfarin?
- Prior thromboembolic event and now on Warfarin
- Should we check for coagulopathy?
Progestin only formulation

- **Less thrombotic** so more suitable for CHD
- But **high failure rate** (5 to 10% in first year)
- **Side effects:**
  - Fluid retention and effect INR
  - Hematomas if on anticoagulants
  - Bosentan may reduce effectiveness
Intrauterine devices

• Decreasing blood loss can control symptoms of heart failure
• Risk of vagal reaction with device placement
• “Contraindicated”: (??)
• Pulmonary hypertension
• Fontan
Barrier methods

- Safe for women with CHD but potentially high failure rate (15 to 32% first year failure)
Sterilization/ permanent forms

- Increased risk of GA, increased intra-abdominal pressure and vagal response
- Vasectomy? May be an issue for partner and future relationships
- Must be discussed in detail with patient, partner and cardiologists / gynecologist
Emergency forms

- Levonorgestrel or Ullapristal
- **Risk of fluid retention**
- If on **Bosentan** (endothelial receptor agonist used in pulmonary artery hypertension) may need higher dose
# Cardiac risks of contraceptives

<table>
<thead>
<tr>
<th>Method</th>
<th>Intra-cardiac shunt</th>
<th>Cyanosis</th>
<th>Heart failure</th>
<th>Specific Precautions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Combined OCP</td>
<td>C/I</td>
<td>C/I</td>
<td>C/I</td>
<td>Thrombosis</td>
</tr>
<tr>
<td>Progestin only</td>
<td>OK</td>
<td>OK</td>
<td>C/I</td>
<td>Fluid retention</td>
</tr>
<tr>
<td>IUD</td>
<td>OK</td>
<td>Caution</td>
<td></td>
<td>Vagal</td>
</tr>
<tr>
<td>Barrier methods</td>
<td>OK</td>
<td>OK</td>
<td>OK</td>
<td>Decrease STD</td>
</tr>
<tr>
<td>Sterilization</td>
<td>OK</td>
<td>OK</td>
<td>OK</td>
<td>GA and vagal</td>
</tr>
</tbody>
</table>
For the cardiologists

- When recommending birth control, must consider:
  - Risk of birth control and pregnancy given patients cardiac anatomy and status
  - Available contraceptive options
  - Risks and benefits of various contraceptive options
  - Failure rate of contraceptives and consequences of pregnancy
  - Patient preference
For the gynecologists

• Understand the cardiac anatomy
• Cardiac defects are classified as:
  • Simple: ASD, VSD, pulmonary stenosis
  • Moderate: TOF, AVSD
  • Complex: Single ventricle
• Is the patient at an increased risk of a paradoxical embolus?
  • If residual shunts or intra-cardiac lesion
What do we do for our pregnant patients?
Counseling

Pregnancy Counseling:

What is the effect on the mother?
Will the effect change with time or treatment?
What is the long-term prognosis?
Optimize cardiac status
May need intervention or surgery
Is there a risk to the fetus? Recurrence, medication, poor growth?
Before pregnancy

- Adjust any teratogenic medication
- Address anticoagulation if appropriate
- Metabolic stress test to evaluate cardiac reserve and heart rate response
- Any necessary intervention (surgery or cath)
- Correct factors that affect cardiac reserve (thyroid, anemia, arrhythmias…)
- Genetic counseling
During pregnancy

- Early cardiac evaluation and follow up determined by cardiac defect/symptoms
  - Frequent discussion regarding L and D
  - High risk OB care
  - Fetal echocardiogram

- If intracardiac R to L shunt:
  - Risk of paradoxical air embolus

- DVT prophylaxis, early ambulation & compression
How do we change our patient situation to:

17 year old sexually active patient. She had a VSD closure and coarctation repair. She has discussed the cardiac risks of contraception with her cardiologist and wants to discuss this further with you. She does not want to have children but is aware that it is safe to do so in the future if she changes her mind.
Patient Education

- Provide **accurate medical information**
- Ongoing discussion **privately** and with family
- Culturally sensitive
- Gradually and regularly during teenage years
- Encourage patients to **discuss concerns**
- Preparation for **self care is an ongoing process**
- Defining roles and expectations
- Clear **communication** with patient/ medical team
Your Initial Patient Encounter

Assess patient knowledge during initial clinical evaluation and intermittently

- What can you tell me about your medical history?
- What do you know about your condition and the need for future care?
- Do you exercise? Do you know if you have any restrictions?
- Have you talked with your cardiologist about contraception and pregnancy risk?
Clear Communication is KEY

• If you say: You can’t get pregnant.

• What patients hear?
  You shouldn’t have children.
  OR
  You are infertile.
In Summary

- Increasing number of adolescents with CHD who have significant gaps in knowledge regarding sexuality, contraception & pregnancy
- Appropriate contraception and pregnancy is tolerated in most women with CHD
- Discussion should start early and each unique patient should be counseled
Thank you