Congenital Heart Disease and Pregnancy: A Growing Population

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Vanderbilt University

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Who are they?

HV: 28 yo G3P0020 at 12 weeks with history of double-inlet LV with left AV valve atresia and transposition of the great arteries, s/p fenestrated lateral tunnel Fontan with residual cyanotic heart disease

PH: 30 yo G1 with history of d-TGA and ASD who is s/p atrial septostomy at birth followed by a Senning atrial baffle

CP: 21 yo G1 with history of subaortic stenosis
Who are they, and where did they all come from?

*Past Medical History is Always Important*
History of Congenital Heart Surgery (Abbreviated)

1938: Ligation of the Ductus arteriosus  
     (Dr. Gross, Boston)
1944: Aortic coarctation repair  
     (Dr. Crafoord, Stockholm)
1944: Blalock-Taussig shunt for Tetralogy of Fallot  
     (Baltimore)
1953: “Heart-Lung Machine” intracardiac repairs  
     (Dr. Gibbon, Philadelphia)
1954: “Complete” repair of Tetralogy of Fallot  
     (Dr. Lillehei, Minneapolis)
Controlled Cross-Circulation

• A lesson from Maternal Fetal Medicine
• Lillehei March 26, 1954
• VSD closure
Fast Forward

• Oldest congenital heart survivors are in their 6th and 7th decades
• Oldest “complex” survivors are 65 (TOF)
• The surgeries for heart defects changed over time
  – TOF born in 1946 versus 1992
  – D-TGA born in 1965 versus 1980
Childhood Disease

• Once fatal diseases have grown into adulthood.
  – Cystic fibrosis – 40% of current pts are adult.
  – Childhood cancer – 80% survival to adulthood.
• 85% of CHD babies born today reach adult age.
• Greatest advances in complex disease.
  – Patient profile is changing: older and more complex.

Aggregate Survival
Changing Severity Distribution

% Survival to adulthood

Birth Year

1940-1959: Simple 90, Moderate 55, Complex 10
1960-1979: Simple 95, Moderate 65, Complex 50
1980-1989: Simple 95, Moderate 90, Complex 80

JACC 2001;37:1170
Impact of increasing survival

- More children with severe disease surviving into adulthood
- More women with severe disease are becoming pregnant
- Pregnancy in women with CHD remains a significant cause of morbidity and mortality
Impact of increasing survival

• From 2004-2006, 1.4% of delivery hospitalizations were complicated by chronic heart disease

• There has not been a substantial change in prevalence of chronic heart disease although increases have been seen in women with CHD at the time of delivery

• Increase in severe complications during delivery hospitalizations compared to early time periods

Kukina & Callaghan BJOG 2010
Tennessee’s Epidemiologic Demand Estimates

Tennessee’s population: 6,400,000 people

CHILDREN <18 years of age
- TN 2012 Children Population
  - 1.5 million
- 1% with CHD
  - 15,000
- 50% with moderate-complex disease
  - 7,500

ADULTS 18+ years of age
- TN 2012 Adult Population
  - 4.9 million
- 1% with CHD
  - 49,000
- 50% with moderate-complex disease
  - 24,500
  - Including mortality
  - 20,825

- Number of pediatric congenital heart patients reaching adulthood will continue to increase.
- In Tennessee, estimated 480 moderate-complex CHD patients reach the age of 18 annually.
- Approximately **50% of these are women**, many of whom will decide they want to have children and enter into our care.
And Not Many ACHD Programs Available in the Southeast

• 2010 ACHA Clinic Survey - patients served annually:
  - UT Memphis  91
  - Lexington  110
  - Arkansas  300
  - Louisville  420
  - UAB  430
  - Vanderbilt  1,392
  - Emory  1,533

• Makes sense that OB care is a reentry point
Vanderbilt Has Experienced a Decade of Growth

Average Annual Growth
2001-2009: 10% per year
2009-2011: 16% per year

Vanderbilt has experienced a current trend of 200 new patients per year over the last 5 years.
Maternal Cardiology Clinic

Disease Mix

- Congenital (71%)
- Arrhythmia (6.8%)
- Cardiomyopathy (6.8%)
- Marfan/CTD (6.8%)
- Other (8.5%)
General Maternal Cardiology Considerations

- Interplay of two complex systems
- What does the patient know about her disease?
- What is “heart failure” in pregnancy?
- Who can/should get pregnant?
- What can be done to make it successful?
Understanding Why & How Pregnancy can be “Severe”
Evaluation Process

• Identify residual lesions
• Determine the baseline functional level
• Recognize the hemodynamic changes of pregnancy and labor and delivery
• Extrapolate from what we know about these lesions in pregnancy
  – About individual lesions
  – About complex lesions (i.e. TOF and Truncus)
  – Risk models
### Traditional Heart Failure Scoring

#### New York Heart Association Classifications

<table>
<thead>
<tr>
<th>Class</th>
<th>Description</th>
</tr>
</thead>
</table>
| 1     | - No limitation of physical activity  
        - Physical activity does not cause fatigue, palpitation or shortness of breath |
| 2     | - Slight limitation of physical activity  
        - Comfortable at rest, but physical activity results in fatigue, palpitations or shortness of breath |
| 3-A   | - Limitation of physical activity  
        - Comfortable at rest, but ordinary activity causes fatigue, palpitations or shortness of breath |
| 3-B   | - Significant limitation of physical activity  
        - Comfortable at rest, but minimal activity causes fatigue, palpitation or shortness of breath |
| 4     | - Unable to carry on any physical activity without discomfort  
        - Symptoms of heart failure at rest |

**Pregnancy Week**

- 1-10
- 11-16
- 17-28
- 29-40
Heart Failure

• NYHA Functional Classification > 1
  – NOT normal for a 35 year old woman
  – NORMAL for 35 year old PREGNANT woman?

• Current projects
  – Pregnancy specific symptom score
    • Not likely to be dyspnea-based (NYHA)
  – Normal BNP values in pregnancy
Anticipated Cardiac Changes in Pregnancy
Hemodynamic changes in pregnancy

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Change</th>
</tr>
</thead>
<tbody>
<tr>
<td>Plasma Volume</td>
<td>Increase up to 40%</td>
</tr>
<tr>
<td>Systemic vascular resistance</td>
<td>Decrease 25-30%</td>
</tr>
<tr>
<td>Cardiac output</td>
<td>Increases 40%</td>
</tr>
<tr>
<td>Heart Rate</td>
<td>Increases 10-20bpm</td>
</tr>
<tr>
<td>Blood pressure</td>
<td>Decreases 1\textsuperscript{st} and 2\textsuperscript{nd} trimester Back to normal in 3\textsuperscript{rd} trimester</td>
</tr>
<tr>
<td>Central Venous Pressure</td>
<td>Unchanged</td>
</tr>
</tbody>
</table>

Nelson-Piercey, Handbook of Obstetric Medicine 2002
Antepartum Physiology

CO = HR \times SV

CO ↑ 30-50%

HR ↑ 10-15 bpm

SV ↑ 20-30%

Cardiac Output

\[ \text{CO} = \text{HR} \times \text{SV} \]

- CO: ↑ 30-50% (peaks 20-24 weeks)
- SV ↑ early (↑ plasma volume and LV dilation)
- HR ↑ 10-15 bpm by 32 weeks
Cardiac Output & Maternal Position

• Rolling from LEFT side to BACK will ↓ CO by 25%
  – Gravid uterus compresses IVC
  – ↓ Venous return to the heart
  – ↓ Ventricular filling (↓ SV)
  – ↓ CO
Physical Findings in Pregnancy

Normal
• Dyspnea
• Syncope
• Palpitations
• Fatigability
• Systolic murmur

Abnormal
• Severe dyspnea
• Syncope with exertion
• Arrhythmias
• Chest pain with exertion
• Diastolic murmur
• Loud, harsh systolic murmur
Normal Imaging Findings

• CXR
  – Enlarged heart silhouette

• EKG
  – Mild ST changes in inferior leads
  – PACs and PVCs

• Echocardiogram
  – Tricuspid regurgitation
  – LA size increased
  – LV outflow cross sectional area increased
Intrapartum Hemodynamics

• Labor – further increase in cardiac output
  – Uterine contractions → auto-transfusion of 300-500ml of blood back into circulation

• Pain → sympathetic response
  → increase in HR and BP
Postpartum Hemodynamic Changes

• Immediate rise in cardiac output
  (Relief of IVC obstruction and uterine contraction)

• Cardiac output increases 60-80% followed by rapid decline to pre-labor values within 1 hour of delivery

• Transfer of fluid from extravascular space increases venous return and stroke volume

Nelson-Piercey, Handbook of Obstetric Medicine 2002
What does this mean for us?

• Minimal data on more complex lesions
  – Must extrapolate
    • Art, not science
    • Understand “how the blood goes round”
  – Understanding of individual shunts and valve lesions often must be considered in combination

• Of assistance . . .
  – General hemodynamic principles
  – General risk stratification models
Review of Specific Cardiac Lesions and Severity
Benign in Pregnancy

ASD, repaired
VSD, repaired
TOF with PS

Serious in Pregnancy

Benign Cardiac

Serious Cardiac
Atrial Septal Defect

• Opening in the atrial septum allowing blood to shunt from left to right

• Involving septum primum around foramen ovale – secundum ASD
Atrial Septal Defect

- Defects sinus venosus – embryological origin of the junction of SVC and IVC into the right atrium – sinoseptal defects

- Involvement of endocardial cushion tissue – primum ASD
Atrial Septal Defect

- Most common congenital lesion
- Generally asymptomatic
- Tolerate pregnancy, labor and delivery well
  - Avoid fluid overload
  - Vaginal delivered preferred
- 3-10% risk fetal cardiac lesion
Ventricular Septal Defect

• Occur anywhere along the ventricular septum

• Clinical manifestations are determined by size and location

• Defects can be
  • Inlet
  • Muscular
  • Perimembranous
  • Outlet
Ventricular Septal Defect

- Small defects well tolerated
- Tolerate pregnancy, labor and delivery well UNLESS pulmonary hypertension
  - Avoid fluid overload
  - Vaginal delivery preferred
- 6-10% risk of fetal cardiac lesion
<table>
<thead>
<tr>
<th>Benign in Pregnancy</th>
<th>Serious in Pregnancy</th>
</tr>
</thead>
<tbody>
<tr>
<td>ASD, repaired</td>
<td>Mitral stenosis</td>
</tr>
<tr>
<td>VSD, repaired</td>
<td>Aortic stenosis</td>
</tr>
<tr>
<td>TOF with PS</td>
<td>Bicuspid aortic valve</td>
</tr>
<tr>
<td>Marfan with normal aorta</td>
<td></td>
</tr>
<tr>
<td>Uncorrected TOF</td>
<td>Previous MI</td>
</tr>
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</tr>
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</table>
Mitral Stenosis

• Most common rheumatic valve lesion
• Risk varies with severity
• FIXED CARDIAC OUTPUT
  – LA outflow obstruction ➔ ➔LV filling
  – LA and pulmonary vascular pressure ↑
    • Worsens in pregnancy due to ↑ volume
• 25% of patient experience heart failure for the first time in pregnancy
Mitral Stenosis

Mitral stenosis obstruction to left atrial emptying

- Difficulty in LV filling
- ↑ LA pressure → Change in LA function

↑ Pulmonary venous pressure

- Perivascular edema, luminal narrowing

Reversal of pulmonary blood flow

- Pulmonary compliance
  - ↑ Work of breathing

- ↑ Pulmonary artery pressure
  - ↓ Cardiac output

Severe pulmonary hypertension

- ↑ Pulmonary vascular resistance

Right ventricular overload

- Tricuspid regurgitation

Stable with mild symptoms
Mitral Stenosis

- Avoid tachycardia
- Maintain high normal PCWP
  - Avoid hypotension
  - Avoid fluid overload
- Treat atrial fibrillation
- Epidural strongly recommended
- Shortened second stage
Aortic Stenosis

- Due to bicuspid valve or rheumatic disease
- Worsens due to ↑ volume
- **FIXED CARDIAC OUTPUT**
- Significant if value is 1/3 normal size

Congenital 4-12% risk of fetal lesion
Aortic Stenosis

Aortic stenosis

Obstruction to LV ejection

Pressure overload

↑ LV mass

Early

LV compliance ↓
Contractility maintained

↓ Preload mechanism
↑ Atrial "booster pump"

Maintenance of normal stroke volume

Late

Fibrosis, contractility ↓

LV dilation

↓ Stroke volume
Aortic Stenosis

• Antenatal
  – Limit activity
  – Growth ultrasound – increased risk of IUGR
  – If severe – bedrest consider valve replacement

• Labor
  – Maintain high normal PCWP
    • Avoid hypotension
    • Avoid fluid overload
  – Epidural slow titration
  – Assisted second stage
Uncorrected Tetralogy of Fallot

- Findings
  - Pulmonary stenosis
  - Right ventricular hypertrophy
  - Overriding aorta
  - VSD

- Corrected – low risk lesion

- Uncorrected – moderate risk lesion
Uncorrected Tetralogy of Fallot

- 40% incidence of cardiac failure in pregnancy
- Maternal mortality 4-15%
- Fetal mortality 30%
- Risk of IUGR
- Prognosis worse if have pulmonary HTN

5% risk of fetal cardiac lesion
Uncorrected Tetralogy of Fallot

• Avoid hypotension and hypovolemia
  – Could cause shunt reversal
• Epidural anesthesia with slow titration
• Assisted second stage
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<td></td>
<td>Previous MI</td>
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- **Benign Cardiac**
  - ASD, repaired
  - VSD, repaired
  - TOF with PS

- **Serious Cardiac**
  - Moderate to severe regurgitation
Mitral Regurgitation (MR)

• Facts
  – Well-tolerated
  – Commonly secondary to mitral valve prolapse
  – Longstanding MR $\Rightarrow$ ventricular dysfunction or LA enlargement
MR (Cont)

• Workup:
  – Echo: severity of regurgitation, LV function, LA enlargement
  – EKG: A-fib

• Complications:
  – Pulmonary edema, arrhythmia (both rare)

• Medical treatment:
  – Treatment of arrythmias or decreased EF
  – Anticoagulation for dilated LA or A-fib
Aortic Regurgitation (AR)

• Facts:
  – Well-tolerated in pregnancy
  – Long-standing AR can → LV dysfunction

• Workup with echo to evaluate severity of AR and LA enlargement and EF

• Complications unlikely

• Medical therapy not indicated unless arrhythmia present
ASD, repaired
VSD, repaired
TOF with PS

Mitral stenosis
Aortic stenosis
Bicuspid aortic valve
Marfan with normal aorta
Uncorrected TOF
Previous MI

Marfan with aortic dilation
Dilated cardiomyopathy
Coarctation of aorta with valve involvement
Fontan/Mustard
Pulmonary HTN
Eisenmenger
Marfan Syndrome

• Defective fibrillin → connective tissue weakness
• Increased mortality related to aortic root involvement
• Autosomal dominant
  – 50% risk of fetal Marfan syndrome
  – Prenatal testing available if mutation is known
Marfan Syndrome

Aortic Root $>40$mm
- Strongly urge pregnancy termination
- $\beta$-blockers decrease pulsatile pressure on aorta
- Minimize activity
- Avoid HTN
- Cesarean delivery
- Risk of aortic dissection persists 6-8 wks postpartum
Marfan Syndrome

- Surgical intervention indicated when aortic root diameter of 5cm
- If replacement is due to valvular dysfunction, ascending aortic replacement should be done if diameter is 4.5cm
Cardiomyopathy

- Outcomes depend on degree of LV dysfunction
- Greatest risk in women with EF <45%, prior cardiac event or NYHA class III or IV
- Prior postpartum cardiomyopathy that has improved still at risk of heart failure
- Pregnancy termination should be considered in women with EF <45%
Cardiomyopathy

• Management
  – Bedrest
  – Na restriction
  – Digitalis
  – Diuretics
  – Consider anticoagulation depending on EF
  – Vaginal delivery preferred
Eisenmenger Syndrome
HIGHEST RISK LESION

Left-to-right shunt

Increased pulmonary blood flow
(shear stress/circumferential stretch)

Endothelial dysfunction and vascular remodeling
Smooth muscle cell proliferation, increase in extracellular matrix, intravascular thrombosis

Increase in PVR

Inverted shunt: right-to-left

Cyanosis (Eisenmenger syndrome)
Eisenmenger Syndrome

• Right ➔ left shunting increases in pregnancy due to decreased SVR
  – Less venous return
  – Less pulmonary perfusion
• Maternal mortality 50-75%
  – Risk highest immediately postpartum to 1 week postpartum
• Fetal mortality 50%
  – Increase risk of IUGR
Eisenmenger Syndrome

Antenatal
- Pregnancy termination and sterilization strongly recommended
- Hospitalization
- Serial ultrasounds for growth
- Epoprostoneol or Sildenafil
  - Improves right sided pressures by vasodilation

Labor
- PA catheter
- Vaginal delivery with assisted second stage
Single ventricle anomalies

- Tricuspid atresia
- Hypoplastic left heart syndrome
- Double inlet left ventricle
- Heterotaxy defects
- Some double outlet right ventricles

Common surgical correction leads to “Fontan physiology”
Fontan physiology

A SINGLE ventricle is doing the work of pumping oxygenated blood FROM the lungs TO the body while DEOXYGENATED blood returns from the body to the lungs via blood vessel connections only with no pumping chamber in the heart (bypasses the heart).
Fontan complications

Maternal
- Protein Losing Enteropathy; 10%
- Liver or kidney disease
- Arrhythmia
- Leaky valves
- Heart failure
- Thrombosis
- Cyanosis

Fetal
- SAB
- FGR
- Preterm birth
General Hemodynamic Principles
Right heart valve disease
• Obstruction well tolerated
• Regurgitation not as well tolerated
• Example: Tetralogy of Fallot

Left heart valve disease
• Obstruction (even mild) not as tolerable
• Regurgitation well tolerated
  – drop in SVR and placenta are better afterload reducers than ACE-Inhibitors
• Example: Bicuspid aortic valve disease
Anemia
• Obstructive Diseases, echo gradients worsen
• May or may not matter functionally
• WILL matter for anesthesia

Vascular abnormalities
• Marfan
• Bicuspid Aortic Valves

Cyanosis
• Outcomes worsen in relation to lower saturation
• Often due to intracardiac shunts
• Increased risk of thromboembolism
Can cardiac risk be stratified during pregnancy?
Existing Risk Stratification Models

- WHO
- CARPREG
- CARPREG + modifications
World Health Organization

• Laundry-list categorizations
• General guide, but can be difficult to extrapolate
  – extremely heterogeneous population
• Contraceptive guidelines reasonably helpful
  – more adaptable to broad categorization
  – Guidelines – not set in stone!
  • Weigh quality of life (i.e DUB)
  • Weigh risk of cardiac complication to risk of pregnancy
Clinical Investigation and Reports

Prospective Multicenter Study of Pregnancy Outcomes in Women With Heart Disease

Samuel C. Siu, MD, SM; Mathew Sermer, MD; Jack M. Colman, MD; A. Nanette Alvarez, MD; Lise-Andree Mercier, MD; Brian C. Morton, MD; Catherine M. Kells, MD; M. Lynn Bergin, MD; Marla C. Kiess, MD; Francois Marcotte, MD; Dylan A. Taylor, MD; Elaine P. Gordon, MD; John C. Spears, MD; James W. Tam, MD; Kofi S. Amankwah, MD; Jeffrey F. Smallhorn, MD; Dan Farine, MD; Sheryll Sorensen, RN;
on behalf of the Cardiac Disease in Pregnancy (CARPREG) Investigators

Circulation 2001
CARPREG

- 562 prospectively collected pregnant women with heart disease (10/1994-11/1999)
- 13% event rate
  - Pulmonary edema
  - Arrhythmia
  - Stroke
  - Cardiac Death
- Baseline status was good
  - 96% were NYHA I or II
  - 89% were on no cardiac medications
Maternal Morbidity (cont)

- Four *predictors* for maternal complications:
  - 1) Prior cardiac event (heart failure, TIA, stroke, arrhythmia)
  - 2) Prepregnancy NYHA class >II
  - 3) Left heart obstruction (mitral/aortic valve area <2 cm², peak left outflow gradient >30 mm Hg)
  - 4) EF<40%

Siu et al. 2001
Predictors Defined

• Prior events
  – Episode of heart failure
  – TIA or CVA
  – Arrhythmia

• Poor NYHA class = II or greater

• Cyanosis = resting saturation <88%
Predictors Defined

- **Left heart obstruction**
  - MV area <2cm²
  - AV area <1.5cm²
  - PEAK echo LVOT gradient >30mmHg

- **Reduced LV function**
  - EF less than 40%
## CARPREG Predictor

<table>
<thead>
<tr>
<th>Predictors</th>
<th>Risk of Cardiac Event</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>5%</td>
</tr>
<tr>
<td>1</td>
<td>27%</td>
</tr>
<tr>
<td>&gt;1</td>
<td>75%</td>
</tr>
</tbody>
</table>
Limitations of CARPREG

• Some limited representation of groups
• Risk index aimed at MOST sensitive assay—captured all events at the expense of specificity!
Pregnancy Outcomes in Women With Congenital Heart Disease

Paul Khairy, MD, PhD; David W. Ouyang, MD; Susan M. Fernandes, MPH, PA-C; Aviva Lee-Parritz, MD; Katherine E. Economy, MD; Michael J. Landzberg, MD

Background—Pregnant women with congenital heart disease are at increased risk for cardiac and neonatal complications, yet risk factors for adverse outcomes are not fully defined.

Methods and Results—Between January 1998 and September 2004, 90 pregnancies at age 27.7±6.1 years were followed in 53 women with congenital heart disease. Spontaneous abortions occurred in 11 pregnancies at 10.8±3.7 weeks, and 7 underwent elective pregnancy termination. There were no maternal deaths. Primary maternal cardiac events complicated 19.4% of ongoing pregnancies, with pulmonary edema in 16.7% and sustained arrhythmias in 2.8%. Univariate risk factors included prior history of heart failure (odds ratio [OR], 15.5), NYHA functional class ≥2 (OR, 5.4), and decreased subpulmonary ventricular ejection fraction (OR, 7.7). Independent predictors were decreased subpulmonary ventricular ejection fraction and/or severe pulmonary regurgitation (OR, 9.0) and smoking history (OR, 27.2). Adverse neonatal outcomes occurred in 27.8% of ongoing pregnancies and included preterm delivery (20.8%), small for gestational age (8.3%), respiratory distress syndrome (8.3%), intraventricular hemorrhage (1.4%), intrauterine fetal demise (2.8%), and neonatal death (1.4%). A subaortic ventricular outflow tract gradient >30 mm Hg independently predicted an adverse neonatal outcome (OR, 7.5). Cardiac risk assessment was improved by including decreased subpulmonary ventricular systolic function and/or severe pulmonary regurgitation (OR, 10.3) in a previously proposed risk index developed in pregnant women with acquired and congenital heart disease.

Conclusions—Maternal cardiac and neonatal complication rates are considerable in pregnant women with congenital heart disease. Patients with impaired subpulmonary ventricular systolic function and/or severe pulmonary regurgitation are at increased risk for adverse cardiac outcomes. (Circulation. 2006;113:517-524.)

Key Words: arrhythmia ■ heart defects, congenital ■ pregnancy ■ tetralogy of Fallot ■ transposition of great vessels
• Modification of CARPREG risk model
• Improved by including
  – Decreased subpulmonary ventricular systolic function
  – Severe pulmonary regurgitation
• Speaks to the Tetralogy of Fallot population
  – Numbers enough to impact outcome numerics (28% of study population)
  – Functional status typically good (high representation in reproductive cohorts)
# Pulmonary Insufficiency

Khairy Circ 2006

## TABLE 4. Maternal Predictors of Primary Cardiac Events During Pregnancy

<table>
<thead>
<tr>
<th>Predictor</th>
<th>OR</th>
<th>95% CI</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Univariate predictor</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Baseline NYHA class ≥2</td>
<td>5.4</td>
<td>1.2, 25.2</td>
<td>0.0320</td>
</tr>
<tr>
<td>Weight, kg</td>
<td>1.1</td>
<td>1.02, 1.14</td>
<td>0.0098</td>
</tr>
<tr>
<td>Prior history of heart failure</td>
<td>15.5</td>
<td>1.5, 163.6</td>
<td>0.0223</td>
</tr>
<tr>
<td>Smoking history</td>
<td>15.6</td>
<td>2.6, 92.7</td>
<td>0.0026</td>
</tr>
<tr>
<td>Pulmonary regurgitation</td>
<td>1.8</td>
<td>1.1, 3.1</td>
<td>0.0440</td>
</tr>
<tr>
<td>Severe pulmonary regurgitation</td>
<td>4.6</td>
<td>1.1, 19.5</td>
<td>0.0372</td>
</tr>
<tr>
<td>Depressed subpulmonary ventricular EF</td>
<td>7.7</td>
<td>1.5, 40.2</td>
<td>0.0159</td>
</tr>
<tr>
<td>Depressed morphological right ventricular EF</td>
<td>4.6</td>
<td>1.4, 15.2</td>
<td>0.0130</td>
</tr>
<tr>
<td><strong>Multivariate predictor</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Severe pulmonary regurgitation or depressed subpulmonary ventricular EF</td>
<td>9.0</td>
<td>1.5, 53.1</td>
<td>0.0158</td>
</tr>
<tr>
<td>Smoking history</td>
<td>27.2</td>
<td>1.9, 384.6</td>
<td>0.0145</td>
</tr>
</tbody>
</table>

EF indicates ejection fraction.
Delivery Modifications
Historic Recommendations

- No published systematic reviews on methodology to date
- “Prolonged second stage of labor”
- Increased use of cesarean section in older literature
- Liberal use of invasive monitoring
Toronto Experience

• 559 women with heart disease
• 1118 women without heart disease
• Quartenary care setting
• January 2000-April 2009
• 1677 pregnancies
  – 594 in 489 women with heart disease
  – 35 excluded for TAB, SAB, or multiple gestations.
• 88% were NYHA I, none were NYHA 4
• 5% on anticoagulation
Objectives

1) Compare the peripartum outcomes in women with heart disease to a similar obstetric population, but without heart disease

2) Determine if a less aggressive obstetric management approach, with its benefits, adversely impacts the mother or the neonate.
Types of Cardiac Cases

- Congenital (63%)
- Valvular (15%)
- Arrhythmias (11%)
- Cardiomyopathy (7%)
- Other (4%)
Congenital by Severity

Classifications per Connelly M. Canadian Consensus Conference on Adult Congenital Heart Disease. Canadian Journal of Cardiology 1996; 14:395-452.
Modes of Delivery

• Vaginal deliveries result in smaller shifts in blood volume and less hemorrhage
• Fewer overall complications, including infection and thromboembolism
• Increased cardiac workload of labor can be significantly reduced with epidural coverage
**Modes of Delivery**

<table>
<thead>
<tr>
<th></th>
<th>Cases</th>
<th>Control</th>
<th>P-value</th>
</tr>
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<tbody>
<tr>
<td><strong>Induction of Labor</strong>*</td>
<td>232 (50%)</td>
<td>267 (28%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td><strong>Mean GA</strong></td>
<td>38 +/- 2 weeks</td>
<td>38 +/- 3 weeks</td>
<td>0.91</td>
</tr>
<tr>
<td><strong>Cesarean Section</strong></td>
<td>182 (33%)</td>
<td>352 (31%)</td>
<td>0.66</td>
</tr>
<tr>
<td><strong>Primary</strong></td>
<td>119 (21%)</td>
<td>266 (24%)</td>
<td>0.02</td>
</tr>
<tr>
<td><strong>Repeat</strong></td>
<td>63 (11%)</td>
<td>86 (8%)</td>
<td>0.02</td>
</tr>
<tr>
<td><strong>Cardiac Indic</strong></td>
<td>2 (0.4%)</td>
<td>0</td>
<td>0.11</td>
</tr>
<tr>
<td><strong>Vaginal Deliveries</strong></td>
<td>377 (67%)</td>
<td>766 (69%)</td>
<td>0.5</td>
</tr>
<tr>
<td><strong>Assisted Vaginal</strong></td>
<td>164 (29%)</td>
<td>120 (11%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td><strong>Forceps</strong></td>
<td>66 (12%)</td>
<td>27 (2%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td><strong>Vacuum</strong></td>
<td>98 (18%)</td>
<td>93 (8%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td><strong>Cardiac Indic</strong></td>
<td>90 (16%)</td>
<td>0</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

*Includes rates in all vaginal and non-elective CS deliveries; 92% (151/164) after 37 weeks
Modes of Delivery

• Despite increased induction rates, C-section rates no different in two groups
• Assisted delivery is frequent in CHD, but over 50% of women with CHD were able to be delivered spontaneously
  – many multiparous and precipitous
  – others with stable heart disease with no history of cardiac event
• No differences in lacerations despite increased assist rate
Second Stage of Labor

• No significant difference in time of second stage between cases and controls
  – 55 vs 50 mins, p = 0.18
  – Regardless of parity

• If assisted delivery for cardiac indication
  – Mean duration of second stage was 80 mins (p<0.001)
  – 20% with second stage >190 mins (>2SD above control group mean)
Cohort >190mins

- No postpartum hemorrhage requiring transfusion
- No cardiac events
  - No correlation between length of second stage and adverse cardiac event
Second Stage, Reconsidered

• “Shortened” ≠ “Assisted”
• In some patients, goal is to decrease expulsive effort, not time
• Time for passive descent of presenting part prior to assist
• No differences in cases and controls
  – Lacerations
  – Fetal trauma
## Obstetric Complications

<table>
<thead>
<tr>
<th></th>
<th>Cases</th>
<th>Controls</th>
<th>P-values</th>
</tr>
</thead>
<tbody>
<tr>
<td>Postpartum Hemorrhage*</td>
<td>29 (5%)</td>
<td>8 (1%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>3rd or 4th degree Laceration; Vaginal **</td>
<td>6/292 (3%)</td>
<td>11/748 (2%)</td>
<td>0.50</td>
</tr>
<tr>
<td>3rd or 4th degree Laceration; Assisted **</td>
<td>4/90 (4%)</td>
<td>7/49 (14%)</td>
<td>0.04</td>
</tr>
</tbody>
</table>

* Hemorrhage >500ml vaginal or 1L C-section
**Lacerations exclude episiotomies
Postpartum Hemorrhage

- 11 women with cardiac disease (2%) required transfusion
- 9 on anticoagulant or were cyanosed
- 2 non-cyanotic women not on anticoagulants
  - 1 C-section for placenta previa
  - 1 with assisted vaginal delivery for heart disease
    - Not in group with >2SD second stage
Cardiac Events

- During labor and delivery and up to 48 hours postpartum
- None in control group
- Occurred in 2% (8/559) of cardiac cases
  - CHF (pulmonary edema on CXR or exam), n=5*
  - arrhythmia, n=3*
  - aortic dissection, n=1
  - No cardiac arrest or cardiac death
  - One event in a vacuum-assisted delivery for cardiac reasons (second stage = 105 mins)
  (*one woman with both a-fib and CHF)
Final Thoughts

- Majority of women with cardiac disease can have successful pregnancy
- Diagnostics prior to pregnancy help in cardiac management during pregnancy (preconception counseling)
- High rate of post-pregnancy interventions
  - Many pulmonary valve replacements in TOF
Final Thoughts

• Increase in number and severity of CHD patients to be expected
• Interplay of systems will need interdisciplinary approach
  – Often to include anesthesia, hematology, genetics
• Understanding of individual anatomy and hemodynamics is essential
• Risk models applied and adjusted to the individual can serve as a guide
Questions?