Congenital Heart Patients Reaching Reproductive Age

- As the result of the medical successes of the last 50 years, the most rapid growth in CHD is in patients in the late teens to 40.
- Issues of reproduction and pregnancy are therefore in the forefront
- Large cause of mortality

Marelli, Circulation 2007

Most Do Well in Adulthood

Englelfriet EHJ 2005
Repaired Tetralogy of Fallot

- Resection of pulmonic obstruction almost invariable results in pulmonary regurgitation
- Pulmonary regurgitation is generally well tolerated true childhood and early adulthood
- In 30’s and 40’s most patients begin to have symptoms, Most have complications by 50’s
- Mirrors most repairs of other complex lesions

Late Complications

Bouzas, EHJ, 2005
Pregnancy Concerns

• Occurs at time of late complications
• Pregnancy may present two patients with conflicting interest with the best interests of the mother and fetus often quite different
• Need to balance and advocate for each patient
• Because of these needs prenatal counseling essential preferable before even considering pregnancy

Prenatal Counseling

• Maternal Risk
• Fetal risk from maternal hemodynamics and medications
• Prospects for future surgery and ability to care for child (important for father as well)
• Potential of shortened life expectancy
• Recurrence risk
Recurrence Risk

- Overall population risk of CHD is around 0.8%. In parents with CHD, ranges 2-12% by diagnosis

Gatzoulis, BMJ 2006

Maternal Risk

- Considerable hemodynamic and hematologic changes occur with pregnancy and peripartum
- I tell patients pregnancy is low level exercise for several months and therefore need to make sure they are ready for it
- Understanding the circulatory changes allows to predict how an individual patients lesion will behave

Head PMJ 2005
Hemodynamic Changes

Box 1 Cardiovascular changes in pregnancy
- Decreased systemic and pulmonary vascular resistance
- Increased heart rate
- Increased plasma volume therefore decreased packed cell volume/raised net systemic pressure
- Increased end diastolic volume and cardiac output
- Third heart sound and ejection systolic murmur

Head PMJ 2005

Gatzoulis BMJ 2006

Net Change

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Average change during pregnancy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Blood volume</td>
<td>+ 35%</td>
</tr>
<tr>
<td>Cardiac output</td>
<td>+ 40 – 43%</td>
</tr>
<tr>
<td>Stroke volume</td>
<td>+ 30%</td>
</tr>
<tr>
<td>Heart rate</td>
<td>+ 15 – 17%</td>
</tr>
<tr>
<td>Systemic vascular resistance</td>
<td>+ 15 – 21%</td>
</tr>
<tr>
<td>Mean arterial pressure</td>
<td>No significant change</td>
</tr>
<tr>
<td>Systolic blood pressure</td>
<td>+3 – 5 mm Hg</td>
</tr>
<tr>
<td>Diastolic blood pressure</td>
<td>+5 – 10 mm Hg</td>
</tr>
<tr>
<td>Central venous pressure</td>
<td>No significant change</td>
</tr>
<tr>
<td>Serum colloid osmotic pressure</td>
<td>+10%</td>
</tr>
<tr>
<td>Hemoglobin</td>
<td>+23.1 g/dl</td>
</tr>
</tbody>
</table>

Connolly, IJC 2005
Understanding These Changes Predicts Outcome

- Lesion that do worse with increased cardiac output, increased heart rate, volume and lower SVR will do the worse.
- In general then stenotic lesions, pulmonary hypertension, Fontan, advanced heart failure are at higher risk, right to left shunt may increase.
- Regurgitant lesions and left to right shunts if PA pressure normal probably ok.

Markers of Risk

- Multi-center study of 562 consecutive women with pregnancies looked for risk factors.
- 13% of all pregnancies had pulmonary edema/CHF, arrhythmia, stroke, or cardiac death.

<table>
<thead>
<tr>
<th>Table 3: Multivariate Analysis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Condition</td>
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<tr>
<td>----------</td>
</tr>
<tr>
<td>Pulmonary hypertension</td>
</tr>
<tr>
<td>Left heart failure</td>
</tr>
<tr>
<td>Pulmonary edema</td>
</tr>
<tr>
<td>Arrhythmia</td>
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</tbody>
</table>

Siu Circulation 2001)
Risk Score

- Assigned point each of the following: 1) Prior event-CHF, CVA or TIA, arrhythmia, or cyanosis 2) > NYHA class II 3) Obstructive lesion (MVA<2.0cm², AVA<1.5cm², or aortic gradient >30 mmHg) 4) EF<40%
- Predicted risk for arrhythmia, pulmonary edema or CHF, CVA, arrest, or death

Lesion Specific Risk

- Functional status is a powerful integrative predictor, but anatomy matters too
- Certain anatomy just isn’t as well adapted to increased CO, thrombotic risk, and pregnancy
Fontan Pregnancy

- 10-15 years ago generally thought too high risk to recommend
- But patients still got pregnant and did ok
- Largest series still published only as an abstract Canobbio et al at ACC 2013 of retrospective analysis from 12 North American Centers with 71 pregnancies in 45 woman with 51 (73%) resulting in live birth—12 SAB, 3 therapeutic, 2 fetal demise
- Mean gestation 34 wks, 2169 g
- 33% (17) maternal complications—5 CHF, 12 arrhythmia-5 atrial, 2 ventricular, no deaths but one resuscitated arrest at delivery
- 1 neonatal death, 2 children CHD, 52% obstetric complications; 5 late deaths in f/u of (67%) with death at mean 12 years
- Dutch and Belgian registry 10 pregnancies—50% mischarage and 1 ectopic, 4 delivery with all complicaitons AF, CHF, premature delivery, neonatal death, growth retardation
Approach

- Pregnancy needs to be planned, so stress contraception
- Screen out medicines unsafe in pregnancy
- Echo plus complementary CT or MRI when needed
- Exercise testing to formally evaluate functional status
- Consideration of pre-pregnancy surgery is delaying, e.g., TOF PVR
- Discussion of social, genetic, and long-term risk

Exercise Intolerance—Perhaps Best Single Indicator of Success

- Exercise performance is decreased in congenital heart disease with more severe lesion showing more pronounced reduction in VO2
- Even “asymptomatic” patients have decreased VO2
- VO2 predicts outcome
  Diller, Circulation 2005
Anticoagulation

- While Coumadin presents a fetal danger particularly of skeletal malformation, heparin, either UFH or LMWH, may not provide as good anti-coagulation
- Ideal management in high risk patients is unclear and controversial

* Box 3: Safety profiles of cardiac drugs in pregnancy

1. Anticoagulation
   - While Coumadin presents a fetal danger particularly of skeletal malformation, heparin, either UFH or LMWH, may not provide as good anti-coagulation
   - Ideal management in high risk patients is unclear and controversial
Contraception

Box 4: Contraception and termination of pregnancy for women with congenital heart disease

Contraception

None of the methods available today is optimal for women with congenital heart disease.

- "Natural methods" (abstinence, withdrawal, safe period) and "barrier methods" (condom, diaphragm) are not recommended high-risk group because of the potential for unexpected pregnancy and the increased risk of thromboembolism.
- Oral contraceptives should be avoided in women at risk of thromboembolism with major forms of congenital heart disease (transannular patch, Fontan circulation, andFontan), because of the thrombotic properties of estrogens.
- Progestin-only oral contraceptives (the "mini pill") are not recommended for women with severe mitral valve regurgitation or right-sided congenital heart disease because of the continued risk of thromboembolism.
- In general, women should be informed of the risk of thromboembolism and be given the option of other contraceptive methods.

Termination of pregnancy

- The risk of teratogenicity increases with increasing gestational age and should be performed as soon as the decision has been made, preferably in the first trimester.
- Surgical abortion under local anesthesia is the preferred method.
- Medical abortion with mifepristone (misoprostol) is an alternative for women who are not suitable for surgical abortion.

Obstetric Management

- Labor is associated with increased risk as there is a further increase in heart rate, cardiac output to about 80% basal pre-pregnancy level, and hypertension.
- Uterine contraction also results in autotransfusion of 300-500cc blood.
- In general, vaginal delivery is preferred as less rapid hemodynamic shifts.
- Left lateral to limit IVC compression.
- Pain control.
- Assisted second stage as needed.
- Avoid induction.
Post-Partum

- Blood loss of delivery
- Auto-transfusion from uterine contraction increased intra-vascular volume
- Oxytocin--associated vasodilation
- Ergometrine-hypertension
- Thrombophilia-6 fold increase of thromboembolic event during pregnancy increases to 11 fold peri-partum
- Coumadin ok in breast feeding

Connolly IJC 2005

MUSC Experience-First 6 Months

Distribution of Diagnoses

High Patient Acuity

Diagnoses Made in Adulthood

Presentation of Late Diagnoses
Case Tetralogy

• 35 year TOF post suspected trans-annular patch presented with palpitations and pregnancy.

• ECG IART
MRI

- LV EF 55%
- LVEDV 192 ml
- LVESV 87 ml
- SV 105
- RVEF 42%
- RVEDV 384 (~160 ml/m2)
- RVESV 224 (~105 ml/m2)
- SV 160
- Regurgitant fraction 65% by velocity encoded imaging
Case

- 35 year old presented with new pregnancy
- MRI RVEDV 166 ml/m2
- RVESV 66 ml/m2
- VO2 36.1 (123% predicted)
- Uncomplicated pregnancy 2 1/2 years ago
Case

- 37 year old with HTN but otherwise well woman with one prior pregnancy 6 years ago transferred to MUSC after developed and acute onset of palpitations at 36 weeks while driving
- EMS found normal vitals but O2 saturations were in the mid-80’s
- Transferred to MUSC where exam notable for a 2-3/6 murmur at the LUSB consistent with outflow tract murmur (?: Increased CO of pregnancy and shunt vs.. PS)

ECHO
Management

- R to L atrial level shunting but why
- Thought due to streaming of flow similar to orthodeoxia and SVR and as well as increased CO stimulating R to L flow
- Did not appear to have pulmonary hypertension but would dramatically increased risk at delivery
- Noted orthodeoxia-Platypnea with sat falling 5% with standing
Cath

- Elected to due RHC to confirm PA pressures and confirm anatomy
- Bidirectional shunt-QP/QS 1.1; large secundum ASD, atrial membrane directing SVC blood across septum
- Saline infusion raised LA pressure but RA remained constant and fully saturated

Outcome

- With onset of labor developed hypertension to 160-180 and began fully saturating
- Elected c-section which was well tolerated
- Post-delivery sats remained in high 80’s to low 90’s
Likely Physiology

- Right atrial membrane act as a baffle encouraging r to l streaming
- This is enhanced with increased cardiac output during pregnancy
- Pulmonary gradient increases with increased flow creating mild RV hypertension
- Decreased SVR reduces relative LV compliance

Follow-up

- Post pregnancy sats increased but did not normalize
- Continues anticoagulation post-delivery in highest thromboembolic risk period
- Inadequate rim for percutaneous closure so referred for surgery