Multidisciplinary approach to management of High Risk Patient with Congenital Heart Disease

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Growing Population of Adults With Congenital Heart Disease

- Medical and surgical advances have resulted:
  - 85% of children with complex lesions are expected to survive to adulthood

- Most females with CHD reach childbearing age and majority wish to become pregnant

2006 NHLBI Working Group – ACHD
Williams R, et.al 2006 JACC
Pregnancy risk for Females With Heart Disease

- In western countries maternal heart disease is the major cause of maternal mortality during pregnancy.
- Patients with CHD have a 10% risk of adverse maternal outcomes and 4% risk of adverse outcomes for neonates.
- Women with complex CHD have twice the risk of adverse outcomes than women with simple CHD.

Normal Physiologic changes in pregnancy

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Post-partum</th>
<th>Percentage of change</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cardiac output</td>
<td>Within 1 h</td>
<td>30% above pre-labour values</td>
</tr>
<tr>
<td></td>
<td>24-48 h</td>
<td>Just below pre-labour values</td>
</tr>
<tr>
<td></td>
<td>2 weeks</td>
<td>10% above pre-pregnant values</td>
</tr>
<tr>
<td></td>
<td>12-24 weeks</td>
<td>Baseline pre-pregnancy values</td>
</tr>
<tr>
<td>Heart rate</td>
<td>Immediate</td>
<td>Decrease</td>
</tr>
<tr>
<td></td>
<td>2 weeks</td>
<td>Pre-pregnant values</td>
</tr>
<tr>
<td>Stroke volume</td>
<td>48 h</td>
<td>Remains above pre-labour values</td>
</tr>
<tr>
<td></td>
<td>24 weeks</td>
<td>10% above pre-pregnant values</td>
</tr>
</tbody>
</table>
**Adverse maternal –fetal events**

**Facing CHD patient at high risk**

<table>
<thead>
<tr>
<th>Maternal</th>
<th>Fetal</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventricular decompensation</td>
<td>Fetal wastage</td>
</tr>
<tr>
<td>Arrhythmias:</td>
<td>Prematurity</td>
</tr>
<tr>
<td>Atrial fib/flutter</td>
<td>Small for gestational age</td>
</tr>
<tr>
<td>Ventricular fib/tachycardia</td>
<td>Congenital malformation</td>
</tr>
<tr>
<td>Sudden death</td>
<td></td>
</tr>
<tr>
<td>Thromboembolism : PE, Stroke</td>
<td></td>
</tr>
</tbody>
</table>
Pregnancy Management for High risk Congenital Heart Patient
Pregnancy Management for High risk CHD patient

- Bethesda Conference (1990,2000) Care of the Adult with Congenital Heart Disease
- Guidelines
  - ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease
  - ESC 2011 Guidelines on the Management of Cardiovascular Diseases during Pregnancy
- Canadian Cardiovascular Society 2009 Consensus Conference on the management of adults with congenital heart disease: Complex congenital cardiac lesions
- The Cardiac Society of Australia and New Zealand Adult Congenital Heart Disease (ACHD): Recommendations for Standards of Care
Defining the high risk obstetrical patient?

Modified WHO classification of maternal cardiovascular risk: principles

<table>
<thead>
<tr>
<th>Risk class</th>
<th>Risk of pregnancy by medical condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>No detectable increased risk of maternal mortality and no/mild increase in morbidity.</td>
</tr>
<tr>
<td>II</td>
<td>Small increased risk of maternal mortality or moderate increase in morbidity.</td>
</tr>
<tr>
<td>III</td>
<td>Significantly increased risk of maternal mortality or severe morbidity. Expert counselling required. If pregnancy is decided upon, intensive specialist cardiac and obstetric monitoring needed throughout pregnancy, childbirth, and the puerperium.</td>
</tr>
<tr>
<td>IV</td>
<td>Extremely high risk of maternal mortality or severe morbidity; pregnancy contraindicated. If pregnancy occurs termination should be discussed. If pregnancy continues, care as for class III.</td>
</tr>
</tbody>
</table>

Risk stratification models

- **CARPREG** “Predictors of Risk for Pregnancy-Related Complications in Women With Heart Disease”
- Siu SC. et al, Circulation 1997

- **ZAHARA I,II** “Pre-pregnancy risk assessment and counselling of the cardiac patient” Pieper, P. G. Netherlands Heart Journal, 2011
- **ROPAC Registry**

Modified from Thorne et al 2006, WHO = World Health Organization
HIGH RISK CONDITIONS IN PREGNANCY

- Decreased ventricular function (EF<35%) – LV /RV
- Uncorrected cyanotic CHD,
- Severe left & right heart obstruction/ stenosis
  (e.g. Coarctation of Aorta, Aortic/Mitral Pulmonic stenosis)
- Prosthetic valves (Anticoagulation)
- Aorthopathies with dilated aorta (>40mm)
- Pulmonary HTN (PAP >3/4 systemic)
  (e.g. Primary PHTN, Eisenmenger’s syndrome)
The Main Aims of Management for the High Risk CHD Patient

- Early risk assessment,
- Optimization of care through the organization of a multidisciplinary team for patients considered moderate to high risk,
- Regular monitoring for deterioration throughout pregnancy,
- Organizing a plan of delivery, and surveillance for deterioration
  - Labor
  - Delivery
  - Immediate post-partum period.
Recommendations for moderate to high risk patients with CHD

- Specialized (regional/tertiary) center where a *multidisciplinary team* with knowledge and experience in adult CHD is available.

This includes:
- Cardiologist,
- High risk obstetrician, anesthesiologist and neonatologist
- Geneticist
- Access to cardiac surgical support

- A coordinated “written” care pathway outlining delivery and postpartum care
Who is the high risk obstetrical patient?

- We know who by diagnosis who is considered high risk;
- We know who are low risk;
- But the range of moderate to intermediate risk is wide;
- Haven’t defined who among moderate or intermediate have the potential to develop complication during pregnancy and become high risk;
How to manage the high risk patient with CHD?

There are no models of care on “how to manage these patients.”
Ahmanson/UCLA ACHD (AACHD)  
UCLA Department of Obstetrics:  

A Model for Obstetrical Management of the High risk patient with ACHD

- AACHD program established 1981
- Reproductive services began 1984-85
- High risk obstetrics begun 1992
- Reorganized into multi-disciplinary team approach – 2003
  lead by AACHD and UCLA Department of Maternal Fetal Medicine.
UCLA AACHD 4-Category Risk* Model for Pregnancy Management

- **Low risk (Class A):** No residual effects; carries no additional risk; may be managed as general population; may deliver in community

- **Intermediate (Class B):** Clinically stable at time of conception; poses a potential risk for embolization, arrhythmias, hypertension; ventricular dysfunction;
  - antepartum and delivery plan determined by clinical status

*Definition of risk is based upon published reports*
UCLA AACHD 4-Category Risk* Model for Pregnancy Management

- **Unknown (Class C)** Reports data is limited; risk mild to moderate if clinically stable at time of conception, but data indicates a high risk for potential complications;
  - should be managed/delivered in high risk regional care center

- **High: (Class D)** High maternal and fetal morbidity and mortality; Pregnancy is contra-indicated; always managed and delivered in high risk center
# UCLA AACHD Pregnancy Risk Management Model

<table>
<thead>
<tr>
<th>CATEGORY A LOW</th>
<th>CATEGORY B INTERMEDIATE</th>
<th>CATEGORY C UNKNOWN</th>
<th>CATEGORY D HIGH</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Surgically-repaired</strong></td>
<td><strong>Unoperated</strong></td>
<td><strong>Repaired</strong></td>
<td></td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>ASD, VSD</td>
<td>Fontan for SV,TA</td>
<td></td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>Coarctation of aorta</td>
<td>Atrial repair for TGA</td>
<td></td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>PS (moderate)</td>
<td>Rastelli for PA</td>
<td></td>
</tr>
<tr>
<td>Pulmonic stenosis</td>
<td>Ebstein’s anomaly (without cyanosis);</td>
<td>Prosthetic valves (anticoagulation);</td>
<td></td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>Congenitally correct TGA</td>
<td>CyaHotic CD</td>
<td></td>
</tr>
<tr>
<td>Congenital complete heart block</td>
<td></td>
<td>P. hypertension</td>
<td></td>
</tr>
<tr>
<td>Aortic valve stenosis (gradient &lt;25 mm Hg)</td>
<td></td>
<td>Severe aortic/mital steosis</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Unoperated</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bicuspid AV valve with no obstruction</td>
</tr>
<tr>
<td>ASD/VSD (small left to right shunts)</td>
</tr>
</tbody>
</table>

* Operated
- Coarctation of aorta
- Tetralogy of Fallot
- Ebstein’s Anomaly

* Presence of residual effects after operation; should be managed in high-risk tertiary care center.
Obstetrical Management of the High risk CHD patient

- **UCLA Team approach:**
  - Cardiologist team: ACHD faculty and fellows, APN
    - Consultants: Electrophysiology, pulmonary medicine
    - Coordinator (Nurse) of high risk patient
  - Obstetrical team: perinatologist (MFM), anesthesiologist;
    - MFM fellows, perinatal nurse manager; labor and delivery team (lead RNs, resident), neonatologist
  - Additional: ICU nurse manager, cardiac surgery team; social workers ethics and legal consultants
  - Written delivery plan developed at 32 wks, reviewed by MDT and entered into electronic medical record by week 34 (updated until admission)
  - For high risk patients: multi-disciplinary team conference once fetal viability is established
  - For geographically distant patient, a collaborative plan for OB and cardiology “shared care”
Multidisciplinary Team

- MFM Obstetrician
- Cardiac Anesthesiologist
- Social Worker
- Ethicist
- Cardiac Surgical Team (VAD, ECMO)
- Geneticist
- Neonatologist (NICU Team)
- Labor & Delivery Teams
  - RN, Fellows, Residents
- ACHD Cardiologist
  - ACHD team: APN, Fellows, Consultants: EP, Pulmonary, Hem
Obstetrical Management of the High risk CHD patient

- UCLA Team approach:
  - Designation of high risk is determined by cardiology or obstetrical team
  - If stable sees cardiology beginning of each trimester and has echo
  - Nurse coordinator is notified and meets with patient/spouse (and parents) and follow patient with OB team
  - Delivery plan is drafted at beginning of 3rd trimester; For Category D patients begin after 28 wks
  - Draft of delivery plan is sent to entire team for review and question about management, then uploaded and available till patient is admitted for IOL or if patient comes in earlier
Clinical Management for “high risk” mother
Development of Delivery Plan at 32 wks.

CATEGORY C (Class B)
Antepartum Care

- Repeat echocardiogram at beginning of each trimester
- Fetal echocardiogram at 18-22 wks
- Address lifestyle issues: work, physical activity, sexual activity
- Plan of care for geographically distant patient

LABOR

- Admission date for induction of labor (goal 39 wks)
- Monitoring
  - Cardiac: Remote tele-monitor; monitor
  - O2 Saturation
  - Hemodynamic: arterial line, central line
  - TBD
- Fluid limits/allowances

- Medications e.g. anticoagulation
- Labs: BNP on admission/discharge
- Prevent thromboembolism: IV air bubble filters if have shunts

Delivery

- Vaginal; directions regarding pushing 2nd stage vs assisted deliveries
- Anesthesia: Epidural
- Pacemaker/ICD management: magnet on standby for C-section

Post-partum Care

- Clinically stable patient: Transfer to PP floor w/ remote telemonitoring for 24-48 hrs.
**UCLA DELIVERY PROTOCOL FOR PATIENT with D-TGA s/p atrial Repair**

**Patient:** MM  
**MRN:**  
**DOB:** 5/12/88  
**EDC:** 9/17/17

- **DX:** D-Transposition of Great Arteries

**BACKGROUND**
- **Primary diagnosis:** Transposition of the great arteries;
- **Surgical History:** Senning atrial switch “early in life” dates unknown
- **Medical History since surgery**
- **Medications:** Prenatal
- **Allergies:** Latex
- **Social History:** LVN; Lives in Palmdale with husband, Kevin.
- **Insurance:** PPO

**Medical Team for L&D**
- **MFM OB:** Tina Nguyen, MD
- **MFM Fellows:** Yalda Afshar MD, Emily Scarbetta MD, Ilina Datkhaeva, MD
- **Anesthesia:** Richard Hong, MD; Jason Hirsch, MD OB Anesthesia Fellow
- **ACHD Cardiology Team:** Jamil Aboulhosn # 20550  
Leigh Reardon, # 23665  
Jeannette Lin, MD #23665  
Gentian Luri, MD #28059
- **Electrophysiology:** Jeremy Moore MD  
Kevin Shannon, MD #10732
- **ACHD Fellows:** Ian Lindsey, MD Dr.

- **Nurse practitioners:** Linda Houser and Pam Miner
- **Clinical specialist** for OB: Mary Canobbio x 53091

**Risk Category:** Moderate risk (Category C)

- **Pregnancy course:** G1 P0
- **Cardiac follow-up**
  
History of NSVT in early pregnancy; Throughout 2nd 3rd trimester pregnancy e has been mostly unremarkable. Except for c/o mild LE edema and mild DOE from cardiac side has been asymptomatic. She denies chest pain or pressure, shortness of breath at rest or with daily activity, palpitations, lightheadedness or syncope.

A Holter monitor obtained here did showed 6 runs of SVT, the longest 12 beats and the fastest 200 bpm.

- **Last echo:** 7/20/17  
**CONCLUSIONS:** 1. D-TGA s/tatus post atrial switch (Senning) surgery. Ventricular EF 55-60%. 4. **Right ventricle is the systemic ventricle.** Moderately enlarged right ventricular size and low normal systolic function. 5. Severely increased RV wall thickness. 6. Mild to moderate tricuspid (systemic AV) valve regurgitation. 7. Systemic venous return and pulmonary venous return baffles appear to be patent with no flow acceleration or obstruction noted.

- **Fetal echo** -Normal
- **Antepartum Follow-up (obstetrical complications)**
- **Labor – Delivery:** Induction: Admission date: 9/7/17
- **Anesthesia:** Epidural; limited pushing to avoid decrease in vascular resistance and tachycardia.

- **Infective endocarditis prophylaxis:** x None required
**Potential problems**

1. Potential decrease in systemic right ventricular function.

Avoid fluid overload; observe for S&S of DOE, SOB, and peripheral edema

Measure BNP post-delivery

Potential for arrhythmias: monitor for complaints of palpitations, dizziness, syncope

**Monitoring** : ECG: Remote telemonitoring during active labor; NOTE: Call ACHD attending/fellow for reported runs of SVT; or PACs. PVC that persistent for 5 minutes or if it’s associated with symptoms (lightheadedness, dizziness, presyncope, syncope)

Pulse oximetry during active labor and delivery and for 12to 24 hrs. post-partum

**Cardiac Tests/Diagnostic**:

- ECG: TBD per ACHD Cardiology team (last done: 7/20/17: NSR)
  - Echocardiogram TBD per ACHD Cardiology Team
  - LABS: BNP: last: 6/27/17: 59 7/20/20: 77; Repeat on admission and prior to discharge

**Pain Management**:

- IV narcotics for pain control

**Special precautions**:

- Anti-embolic stockings (TED hose)
- Alternating leg—SCDs
- Bubble-air filters

**Delivery Plan**

- **Admit**
  - Thursday, September 14, 2017

Scheduled to arrive at:
- Induction per protocol
- Analgesia—IV narcotics prn

Intravenous fluids: once NPO begin IV infusion at 120 ml/hrs.; monitor closely for signs of fluid overload
- Vaginal with Minimal push
- No pushing during

**Recovery/Postpartum**

- Routine post-partum recovery: telemonitoring to be determined by cardiology
  - Transfer to postpartum recovery room for observation w/ ECG monitoring
  - Transfer to CCU post-delivery per ACHD team
  - Restart ACE inhibitor:

**Breastfeeding**:

- YES
- NO

**Nursing Care and Discharge Planning**

- Routine nursing care
- Discuss Contraception: considering Mirena
- Review meds:
- Instruct to call Cardiology (310 794-9629) for follow-up visit:
- Instruct to call Obstetrics for follow-up visit.
These special deliveries
Take Special Team Work!
These special deliveries take special Team Work! “The UCLA Team”
Summary

- Despite the well known high risk CHD lesion, the spectrum of who is high risk is wider because cardiac lesions and their residual effects can be negatively impacted by pregnancy.
- Pre-conception counseling and evaluation is essential in defining potential and actual high risk patient.
- In order to ensure safety to mother and fetus, it takes a coordinated multidisciplinary team approach to manage the women with CHD.
- Identifying the challenges will guide us in ensuring....
Health Happy Babies,
Healthy moms to care for them!