MULTIDISCIPLINARY APPROACH TO PERIPARTUM CARE

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GROWING POPULATION OF ADULTS WITH CONGENTITAL HEART DISEASE

- Medical and surgical advances have resulted
 - 80% -95% of babies born with congenital cardiac anomalies expected to reach adulthood;
 - 85% of children w/ complex lesions are expected to survive to adulthood

- 2006 NHLBI Working Group ACHD
- In the past it was thought that congenital heart defects seen in adults represented the mild and simple portion of the spectrum,

But now the number of adults with condition of moderate or severe complexity outweighs that of children

- Continuing medical breakthroughs result in new populations of adult survivors with ever more complex disease including recipients .
- For the number of those living with congenital heart defect first time, adults with single-ventricle physiolgy represent a significant number of CHD patients

Williams R, et.al 2006 JACC

GROWING POPULATION OF FEMALES WITH HEART DISEASE





- Most women with CHD reach childbearing age and majority wish to become pregnant.
- Congenital heart disease is now the most common form of heart disease encountered during pregnancy in the U.S
- Pregnancy is the most common non-cardiac issue, cardiology providers see in practice

NORMAL PHYSIOLOGIC CHANGES IN PREGNANCY



Parameter	Post-partum	Percentage of change
Cardiac output	Within 1 h	30% above pre-labour values
	24–48 h	Just below pre-labour values
	2 weeks	10% above pre-pregnant values
	12-24 weeks	Baseline pre-pregnancy values
Heart rate	Immediate	Decrease
	2 weeks	Pre-pregnant values
Stroke volume	48 h	Remains above pre-labour values
	24 weeks	10% above pre-pregnant values

PREGNANCY RISK: ADVERSE MATERNAL – FETAL EVENTS

Maternal

Fetal

Primary cardiac events

Ventricular decompensation

Arrhythmias:

Atrial fib/flutter

Ventricular fib/tachycardia

Sudden death

Thromboembolism : PE, Stroke **Fetal wastage**

Prematurity

Small for gestational age

Congenital malformation

MANAGEMENT PRINCIPLES

• PRE-CONCEPTUAL COUNSELING AND DIAGNOSTIC EVALUATION

• STRATIFICATION OF MATERNAL RISK

• OBSTETRICAL MANAGEMENT OF PREGNANCY

DETERMINE ORGANIZED PLAN OF DELIVERY AND SURVEILLANCE OF DETERIORATION

Preconception Counseling and Diagnostic Evaluation

- Counseling "critical" for moderate -high risk CHD
 - Understand potential risk to mother and fetus
 - Genetic counseling & evaluation referral
 - Review of medications ; change teratogenic drugs
 - Assess need for correction of clinical problems prior to conception e.g. arrhythmias, residual shunts
 - Control non-cardiac disease –e.g.- thyroid disorders

- Diagnostic evaluation
 - H & P
 - Echocardiogram (resting and stress CPX)
 - Electrocardiogram
 - Additional diagnostics
 - Catheterization
 - EP studies
 - MRI/CT
 - Laboratory work
 - Full evaluation including clotting function (platelets, PT, PTT)
 - BNP
 - Chromosomal studies

WHO IS THE HIGH RISK OBSTETRICAL PATIENT?

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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 "<u>Pre-pregnancy risk assessment and</u> <u>counselling of the cardiac patient</u>" Pieper, P. G. Netherlands Heart Journal, 2011

• ROPAC " Registry Of Pregnancy And Cardiac disease . Roos-Hesselink

MODIFIED WHO CLASSIFICATION OF MATERNAL CARDIOVASCULAR RISK: PRINCIPLES

Risk class	Risk of pregnancy by medical condition
	No detectable increased risk of maternal
	mortality and no/mild increase in morbidity.
She Et 1	Small increased risk of maternal mortality or
Ш	moderate increase in morbidity.
	Significantly increased risk of maternal
Ш	mortality or severe morbidity. Expert
	counselling required.
	If pregnancy is decided upon, intensive specialist
Christian S	cardiac and obstetric monitoring needed
	throughout pregnancy, childbirth, and the
	puerperium.
29. A. S. M.	Extremely high risk of maternal mortality or
IV	severe morbidity; pregnancy contraindicated. If
	pregnancy occurs termination should be
3 82.12	discussed. If pregnancy continues, care as for
	class III.

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)

Based on Sui 2002

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

MULTIDISCIPLINARY TEAM

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



• A coordinated "written" care pathway outlining delivery and postpartum care

MULTIDISCIPLINARY TEAM



ADULT CONGENITAL HEART ASSOCIATION (ACHA)

ACHD ACCREDITATION PROGRAM (2016)

Specialized (regional/tertiary) center

• Comprehensive Care Center



Cardiac Care Center



- Availability and experienced ACHD cardiology and high-risk obstetric care
- Availability of high-risk obstetric care

Recognize when to refer to an ACHD Comprehensive Care Center

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 WITHCHD

- 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.

• Awarded ACHA Accreditation "Comprehensive Care Center" 2017

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

DEFINITION OF PREGNANCY RISK IS BASED UPON PUBLISHED REPORTS

- Unknown (Class C) Reported 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 contraindicated; always managed and delivered in high risk center

UCLA CHD PREGNANCY RISK MANAGEMENT MODEL

CATEGORY A	CATEGORY B	CATEGORY C	CATEGORY D
LOW	INTERMEDIATE	UNKNOWN	HIGH
Surgically-repaired	Unoperated	Repaired	Cyanotic CHD
Atrial septal defect	ASD, VSD	Fontan for SV,TA	P. hypertension
Ventricular septal	Coarctation of aorta	Atrial repair for TGA	Severe aortic/mital
Defect	PS (moderate)	Rastelli for PA	stenosis
Patent ductus arteriosus	Ebstein's anomaly	Prosthetic valves	Eisenmenger Synd
Pulmonic stenosis	(without cyanosis);	(anticoagularion)	Vent. dysfunction
Unoperated Bicuspid AV valve with no obstruction ASD/VSD (small left to right shunts) Mild PS	 Congenital complete Aortic valve stenosis (g * Operated Coarctation of aor Tetralogy of Fallot Ebstein's Anomaly 	heart block gradient <25 mm Hg) ta	

presence of residual effects after operation; Should be managed in high-risk tertiary care center

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UCLA CHD PREGNANCY RISK MANAGEMENT MODEL

CATEGORY A LOW	CATEGORY B INTERMEDIATE	CATEGORY C UNKNOWN	CATEGORY D HIGH
Surgically-repaired Atrial septal defect Ventricular septal Defect Patent ductus arterious Pulmonic stenosis	Unoperated ASD, VSD Coarctation of aorta PS (moderate) Ebstein's anomaly (without cyanosis);	Repaired Fontan for SV,TA Atrial repair for TGA Rastelli for PA Prosthetic valves anticoagularion)	Cyanotic CHD P. hypertension Severe aortic/mital stenosis Eisenmenger Synd
Tetralogy of Fallot	Vent. dysfunction Congenitally correct TGA Congenital complete heart block Aortic valve stenosis (gradient <25 mm Hg)		
Unoperated Bicuspid AV valve with resolution ASD/VSD (small left to right shunts) Mild PS	* Operated Coarctation of aor Tetralogy of Fallot Ebstein's Anomary	t I	

presence of residual effects after operation; Should be managed in high-risk tertiary care center

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OBSTETRICAL MANAGEMENT OF THE HIGH RISK CHD PATIENT

• UCLA Team approach:

- Cardiologist team: ACHD faculty and fellows, APN (NP/CNS)
 - Consultants: Electrophysiology, pulmonary medicine
- Obstetrical team: perinatologist (MFM), anesthesiologist knowledgeable in cardiac anesthesia; MFM fellows, perinatal nurse manager; labor and delivery team (lead RNs, resident), neonatologist
- Additional: ICU nurse manager, cardiac surgery team ; ethics and legal consultants
- Written delivery plan developed at 32 wks, reviewed by MDT and entered into system 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".

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 life style 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
 - 02 Saturation
 - Hemodynamic : arterial line, central line TBD
- Fluid limits/allowances
 - Leg vein care

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

Delivery

• Vaginal;

Permit pushing 2nd stage

- Anesthesia : Epidural preferred
- Pacemaker/ICD management: magnet on standby for Csection
- Post-partum Care
- Clinically stable patient: Transfer to PP floor w/ remote telemonitoring for 24-48 hrs.

PRINCIPLES OF MANAGEMENT: LABOR AND DELIVERY: CATEGORY D

- Management dictated by cardiac status and pregnancy course; planning begins after week 26
- Multi-disciplinary conference (26wks)
- Early admission dictated by clinical status;
 - Location:
 - L&D w/ cardiac RN ;
 - ICU : fetal monitoring required
- MD-Team notified of patient pending admission (including CTS support)

- Monitoring
 - 02 Saturation
 - EKG
 - Hemodynamic g : arterial line, central line
- Prevent thromboembolism:IV air bubble filters if have shunts, leg vein care

Delivery

- Induction: 37 wk. or earlier
- Mode of delivery : TBD by maternal status; often C-section
- Anesthesia : TBD
- Notification of standby teams: CTS (e.g VAD)

Post Partum

- ICU admission
- Close follow-up after discharge

UCLA DELIVERY PROTOCOL FOR PATIENT WITH D-TGA S/P ATRIAL REPAIR

- Patient: MM MRN: DOE DX: D-Transposition of Great Arteries
 - DOB: 5/12/88

EDC: 9/17/17

- BACKGROUND
- Primary diagnosis: Transposition of the great arteries;
- Surgical History: Senning atrial switch "early in life" dates unknown
- Medical History : History of NSVT but controlled with Metoprolol
- Pregnancy: developed volume overload and runs of NSVTpost delivery
- Medications:, Prenatal, Metoprolol
- 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 Jeannette Lin, MD #23665 ,Gentian Luri, MD #28059
- Electrophysiology: Jeremy Moore MD # 22261 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: <u>Moderate risk (Category C)</u>
- Pregnancy course: G1 P0
- <u>Cardiac follow-up</u>

• Pregnancy course has been mostly unremarkable. Did have run of NSVT at beginning of second trimester but no further event and has been asymptomatic. She denies chest pain or pressure, shortness of breath at rest or with daily activity, palpitations, lightheadedness or syncope.

She was referred here for higher level of care in setting of cardiac diagnosis

• 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. LVentricular 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

THESE SPECIAL DELIVERIES TAKE SPECIAL TEAM WORK



THESE SPECIAL DELIVERIES TAKE SPECIAL TEAM WORK! "THE UCLA TEAM"





SUMMARY

- Understand the impact normal pregnancy can have cardiac lesions and their residual effects following operative repair.
- Pre-conception counseling and evaluation is essential in defining potential and actual pregnancy risk.
- 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!











THANK YOU