



# Cardiac Lecture #5: Congenital Cardiac Lesions

February 7, 2023

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# Updates



- Next Maternal Webinar March 7<sup>th</sup>  
Topic: Valvular Heart Disease | Speaker: Marissa Platner, MD
- Q3 & Q4 HTN Data Submission via Survey 123
- GaPQC Annual Meeting - April 13<sup>th</sup> & 14<sup>th</sup>
- 28-Day Anti-Racism Challenge  
<https://www.perinatalqi.org/page/Anti-RacismChallengev2>



## Lectures 2022



- March 1, 2022 - GaPQC Kick-Off Cardiac Education Webinar
- September 6, 2022 - Intro Lecture: Building a Cardio Ob Team
- October 4, 2022 - Lecture 1: Cardiac Physiology
- November 1, 2022 - Lecture 2: Cardiac Warning Signs
- December 6, 2022 - Lecture 3: Cardiomyopathy

[Open GaPQC Maternal Youtube Playlist](#)

## Lectures 2023



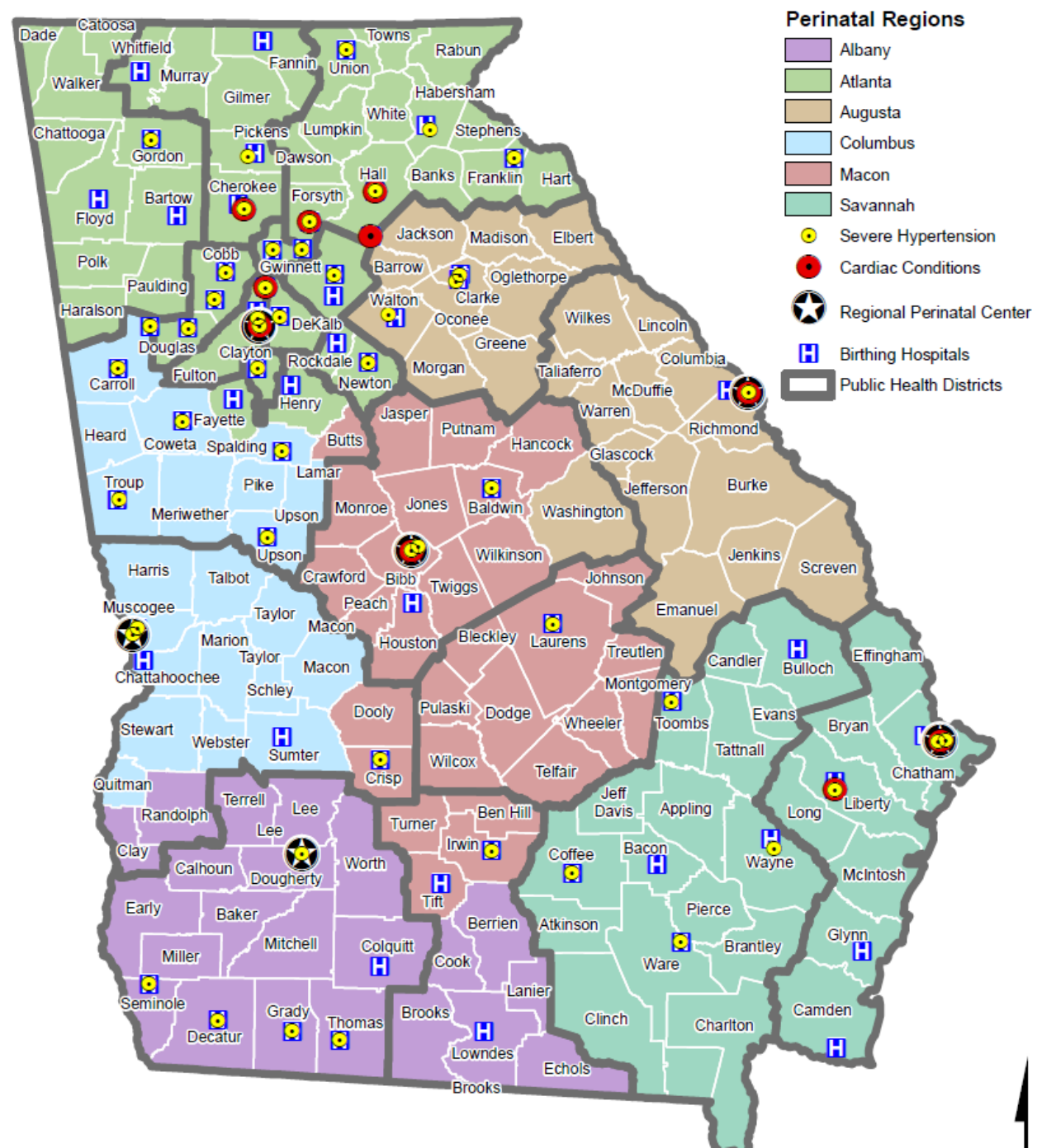
- January 3, 2023 – Lecture 4: Acute MI/Arrhythmias
- February 7, 2023 – Lecture 5: Congenital Cardiac Lesions
- March 7, 2023 – Lecture 6: Valvular Heart Disease
- May 2, 2023 – Lecture 7: Pulmonary Hypertension
- June 6, 2023 – Lecture 8: Ob Anesthesia and L&D Considerations

[Open GaPQC Maternal Youtube Playlist](#)

# Lecture Presentations

March 1, 2022 - GaPQC Kick-Off Cardiac Education Webinar (pdf)	<a href="#">Download</a>
September 6, 2022 - Intro Lecture: Building a Cardio-Ob Team (pdf)	<a href="#">Download</a>
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# Birthing Hospitals Participating in GaPQC's Maternal Patient Safety Bundles



# Key Driver Diagram: Maternal Cardiac Conditions

## GOAL:

To reduce severe morbidity & mortality related to maternal cardiac conditions in Georgia.

## SMART AIM:

By 02/6/2026, **National Wear Red Day**, to reduce harm related to existing and pregnancy related cardiac conditions through the 4<sup>th</sup> trimester by **20%**.

## Key Drivers

**Readiness:** EVERY UNIT - Implementation of standard processes for optimal care of cardiac conditions in pregnancy and post-partum.

**Recognition & Prevention:** EVERY PATIENT - Screening and early diagnosis of cardiac conditions in pregnancy and post-partum.

**Response:** EVERY UNIT - Care management for every pregnant or postpartum woman with cardiac conditions in pregnancy and post-partum.

**Reporting/System Learning:** EVERY UNIT - Foster a culture of safety and improvement for care of women with cardiac conditions in pregnancy and post-partum.

**Respectful, Equitable, and Supportive Care** — EVERY UNIT/PROVIDER/TEAM MEMBER - Inclusion of the patient as part of the multidisciplinary care team.

## INTERVENTIONS

- Train all obstetric care providers to perform a basic Cardiac Conditions Screen.
- Establish a protocol for rapid identification of potential pregnancy-related cardiac conditions in all practice settings to which pregnant and postpartum people may present.
- Develop a patient education plan based on the pregnant and postpartum person's risk of cardiac conditions.
- Establish a multidisciplinary "Pregnancy Heart Team" or consultants appropriate to their facility's designated Maternal Level of Care to design coordinated clinical pathways for people experiencing cardiac conditions in pregnancy and the postpartum period. S1**
- Establish coordination of appropriate consultation, co-management and/or transfer to appropriate level of maternal or newborn care.
- Develop trauma-informed protocols and training to address health care team member biases to enhance quality of care
- Develop and maintain a set of referral resources and communication pathways between obstetric providers, community-based organizations, and state and public health agencies to enhance quality of care. \*

- Obtain a focused pregnancy and cardiac history in all care settings, including emergency department, urgent care, and primary care.
- In all care environments assess and document if a patient presenting is pregnant or has been pregnant within the past year. S2**
- Assess if escalating warning signs for an imminent cardiac event are present.
- Utilize standardized cardiac risk assessment tools to identify and stratify risk.
- Conduct a risk-appropriate work-up for cardiac conditions to establish diagnosis and implement the initial management plan.

- Facility-wide standard protocols with checklists and escalation policies for management of **cardiac symptoms**.
- Facility-wide standard protocols with checklists and escalation policies for management of people **with known or suspected cardiac conditions**.
- Coordinate transitions of care including the discharge from the birthing facility to home and transition from postpartum care to ongoing primary and specialty care.
- Offer reproductive life planning discussions and resources, including access to a full range of contraceptive options in accordance with safe therapeutic regimens. \*
- Provide patient education focused on general life-threatening postpartum complications and early warning signs, including instructions of who to notify if they have concerns, and time and date of a scheduled postpartum visit.**

- For pregnant and postpartum people at high risk for a cardiac event, establish a culture of multidisciplinary planning, admission huddles and post-event debriefs.
- Perform multidisciplinary reviews of serious complications (e.g. ICU admissions for other than observation) to identify systems issues. S4**
- Monitor outcomes and process data related to cardiac conditions, with disaggregation by race and ethnicity due to known disparities in rates of cardiac conditions experienced by Black and Indigenous pregnant and postpartum people. **Process Measures – 1-5**

- Screen for structural and social drivers of health that might impact clinical recommendations or treatment plans and provide linkage to resources that align with the pregnant or postpartum person's health literacy, cultural needs, and language proficiency.
- Engage in open, transparent, and empathetic communication with pregnant and postpartum people and their identified support network to understand diagnoses, options, and treatment plans.
- Include each pregnant or postpartum person and their identified support network as respected members of and contributors to the multidisciplinary care team. \*S5**



# CCOC Process Measures: Reporting and Systems Learning



**P1:** Standardized  
Pregnancy Risk  
Assessments for People  
with Cardiac Conditions



**P2:** Multidisciplinary  
Care Plan for Pregnant  
People with Cardiac  
Conditions

**P3:** OB Provider and  
Nursing Education –  
**Cardiac Conditions**

**P4:** OB Provider &  
Nursing Education–  
**Respectful and  
Equitable Care**

**P5:** ED Provider and  
Nursing Education –  
Cardiac Conditions

**OP1:** Cardiovascular  
Disease (CVD)  
Assessment Among  
Pregnant and  
Postpartum Women



Clayton Allen Smith Jr., MD  
Emory Adult Congenital Heart Center  
Assistant Professor, Department of Cardiology  
Emory School of Medicine



EMORY  
UNIVERSITY  
SCHOOL OF  
MEDICINE

EMORY  
ADULT CONGENITAL  
HEART CENTER

# Pregnancy In Congenital Heart Disease

Clayton (Tony) Smith MD

Assistant Professor of Internal Medicine and Pediatrics

Emory Adult Congenital Heart Center

Children's Healthcare of Atlanta Cardiology



# Disclosures

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None

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# Objectives

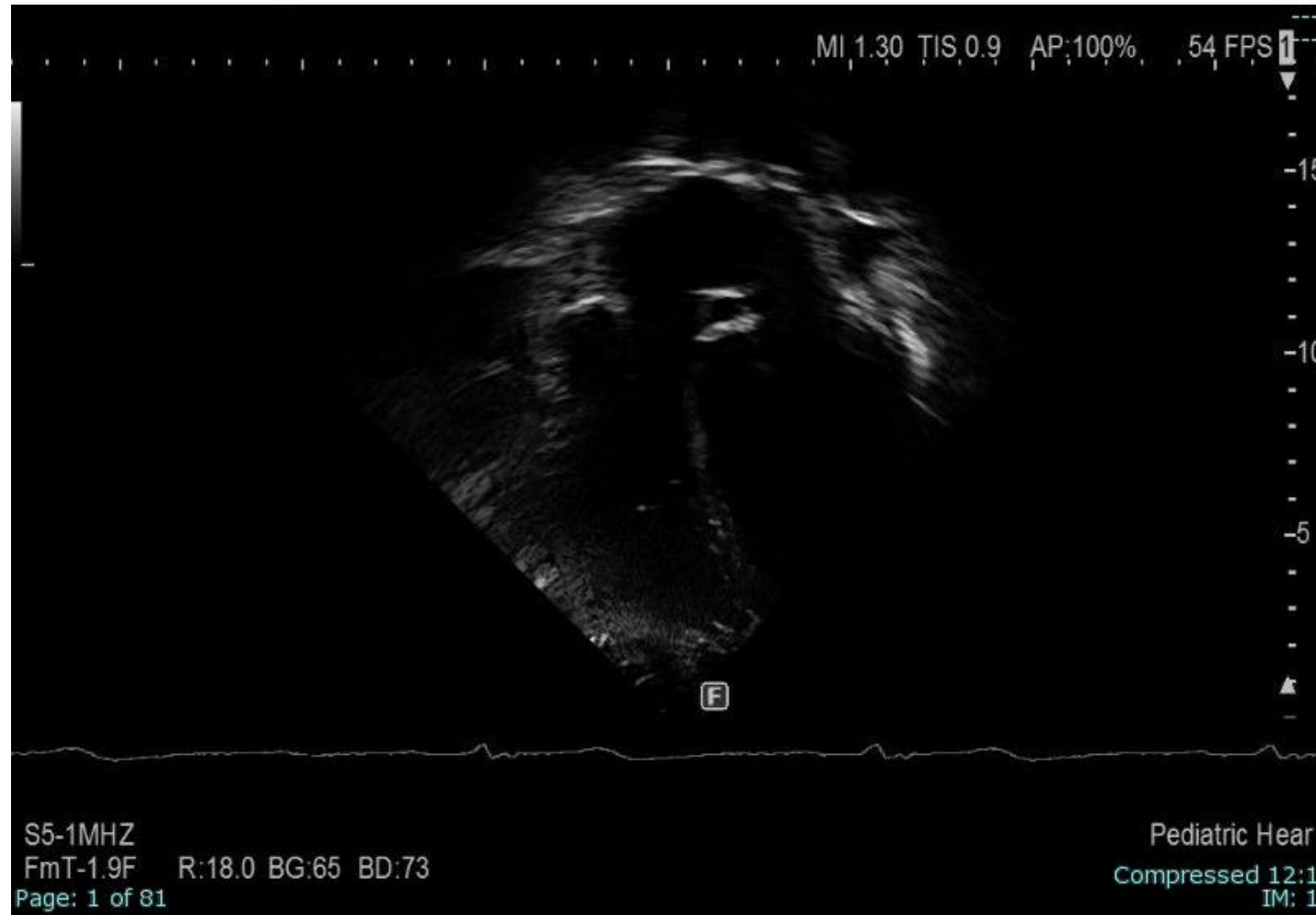
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- Review the physiologic changes that occur during normal pregnancy and how this impact cardiovascular function
  - Understand essential role of preconception counseling in women with congenital heart disease (CHD)
  - Be familiar with pregnancy issues related to selected Congenital Heart Defects
    - Left Heart obstructive lesions
    - Tetralogy of Fallot/Conotruncal lesions
    - Pulmonary Hypertension/Eisenmenger syndrome
    - Single Ventricle/Fontan
  - We will not cover:
    - Connective tissue disorders
    - Isolated valvular disease
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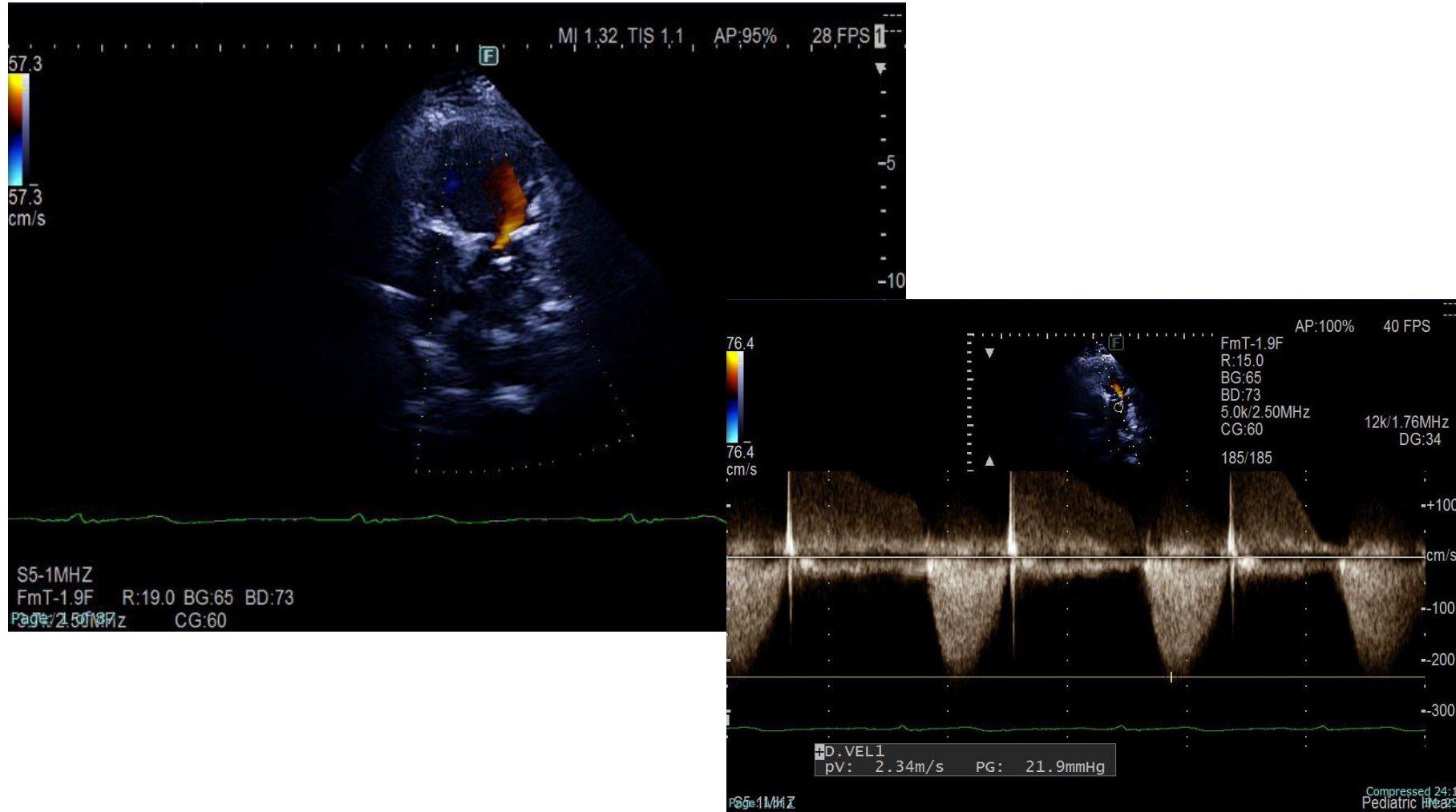
21 yo G1P0 with history of TOF  
Initial repair in first 3 months of  
life  
Residual VSD closure and  
enlargement of RVOT and LPA at  
10 months  
Surgical pulmonary valve  
replacement at 15 yo due to  
progressive RV enlargement  
Presented to her pediatric cardiologist  
office at 10 weeks gestation



# AE- Baseline Echo

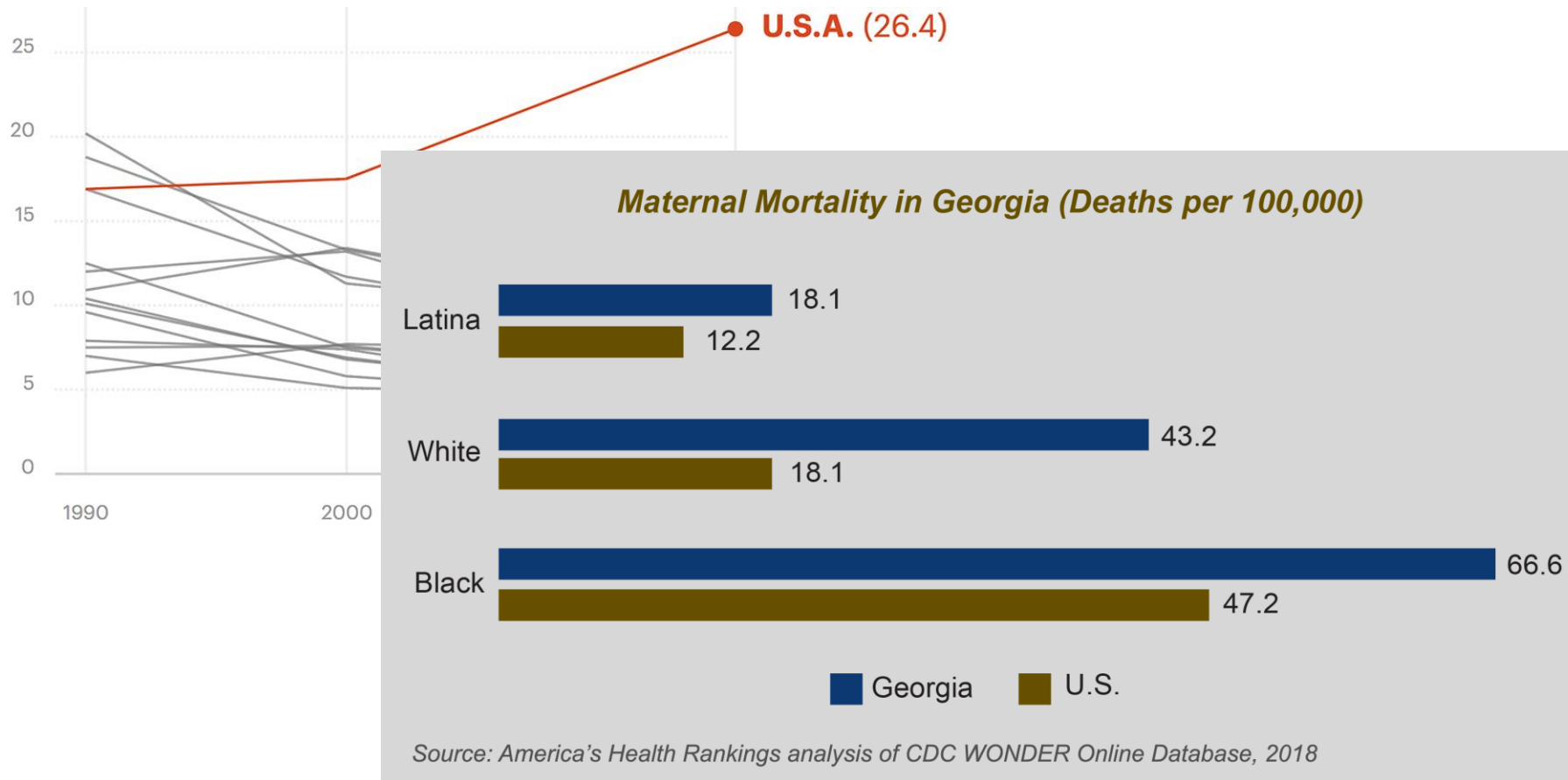


# AE- Baseline Echo

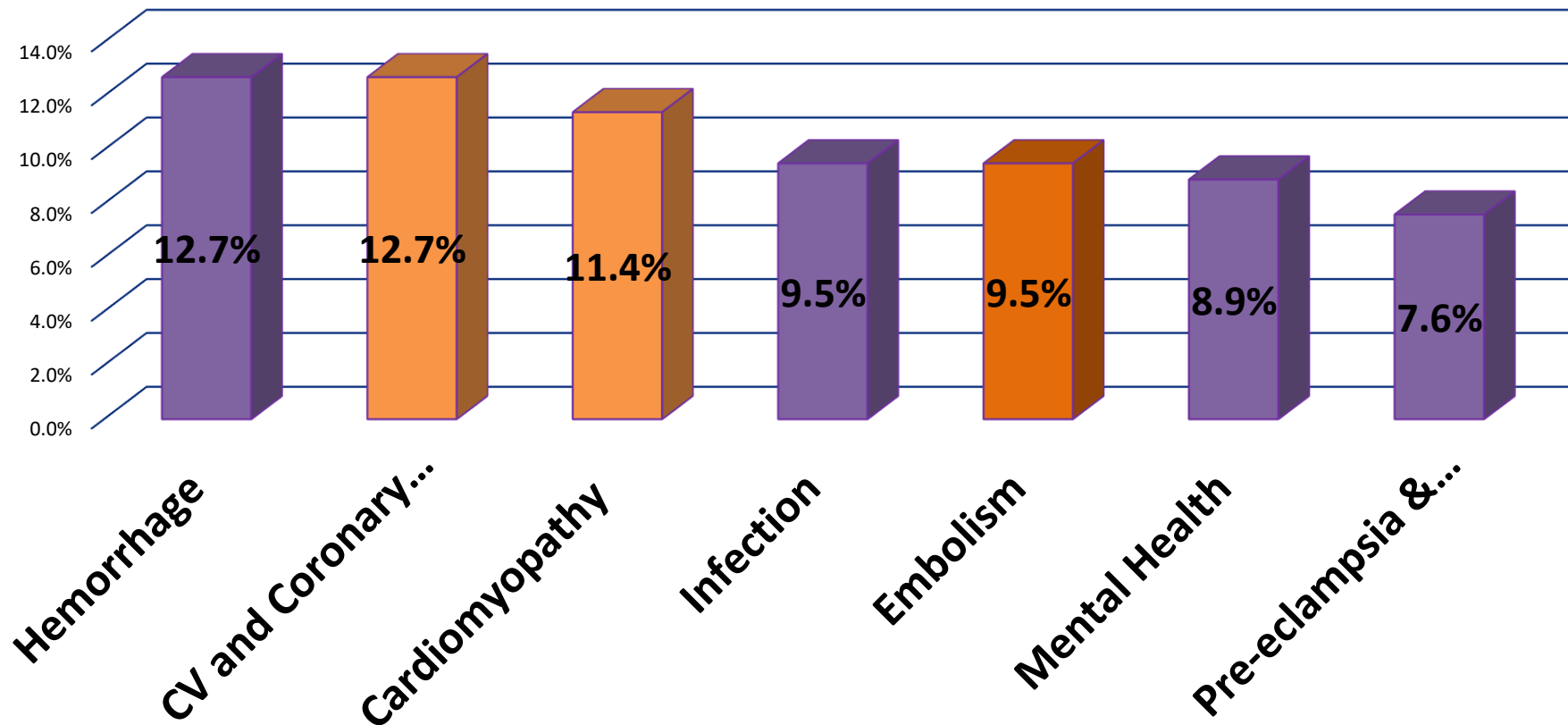


# Scope of the Problem

Maternal Mortality Rate- Deaths per 100,000 births



# CV Disease is the #1 cause of maternal mortality in the US Cardiovascular Causes account for ~25% of Maternal Mortality



# Physiologic Changes in Pregnancy

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Now that I'm in the  
third trimester, I  
sleep sitting up so  
I can breathe.

#pregnancyproblems

somee cards  
user card





**HIGH-FLOW,  
LOW RESISTANCE CIRCULATION**

## Circulation

Volume 141, Issue 23, 9 June 2020;, Pages e884-e903  
<https://doi.org/10.1161/CIR.0000000000000772>



### AHA SCIENTIFIC STATEMENT

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# Cardiovascular Considerations in Caring for Pregnant Patients: A Scientific Statement From the American Heart Association

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**Laxmi S. Mehta, MD, FAHA, Chair, Carole A. Warnes, MD, FAHA, Vice Chair, Elisa Bradley, MD, Tina Burton, MD, Katherine Economy, MD, Roxana Mehran, MD, Basmah Safdar, MD, Garima Sharma, MD, Malissa Wood, MD, Anne Marie Valente, MD, Annabelle Santos Volgman, MD, FAHA, and On behalf of the American Heart Association Council on Clinical Cardiology; Council on Arteriosclerosis, Thrombosis and Vascular Biology; Council on Cardiovascular and Stroke Nursing; and Stroke Council**

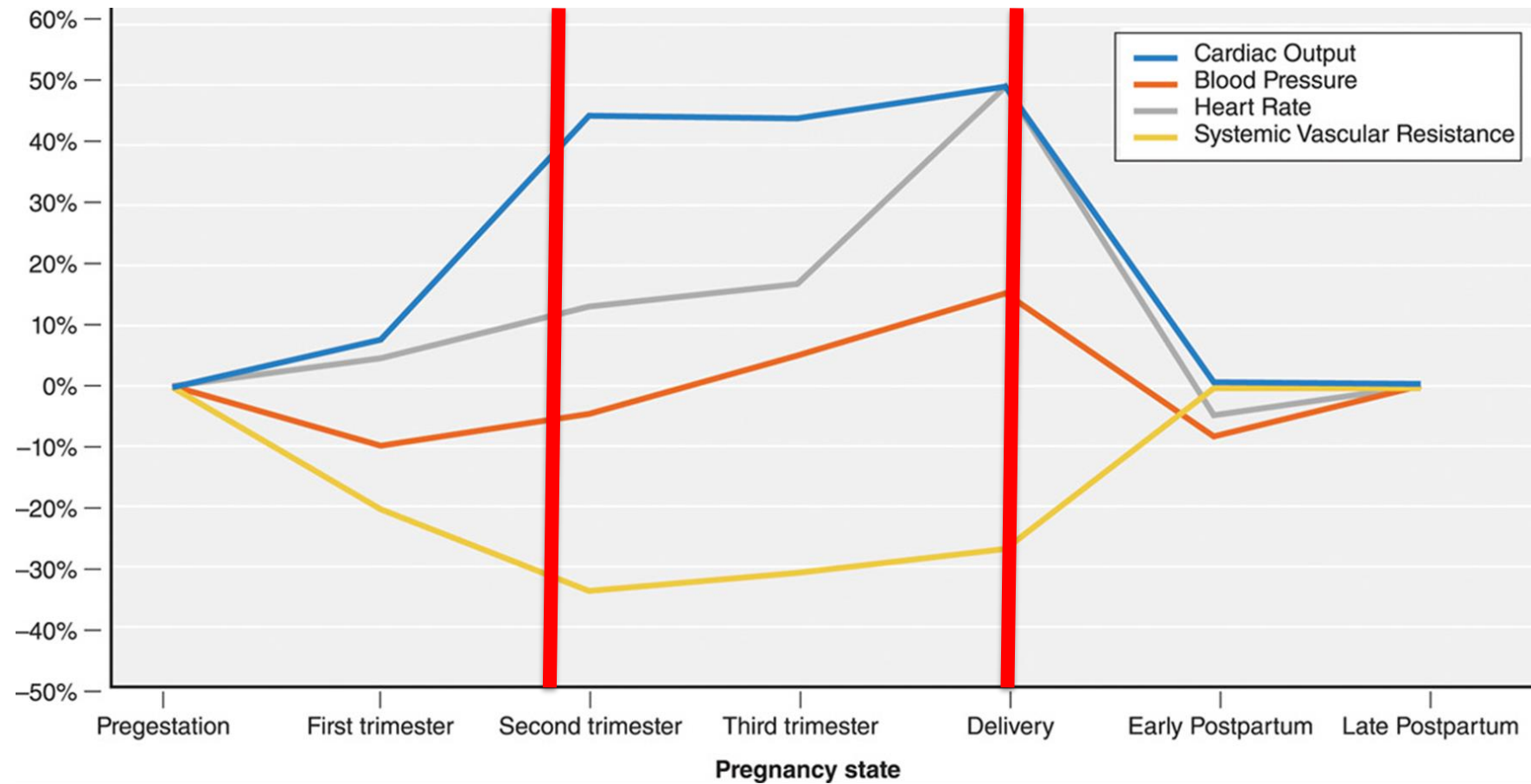
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# Pregnancy: Hemodynamic Changes

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- Cardiac Output increases 30-50% above baseline
    - Preload increased (increased blood volume)
    - Afterload decreased (decreased SVR, PVR)
    - Heart rate increases by 15-20 bpm
  - Resistance decreases to accommodate increased flow
-

# Relative Physiologic changes throughout pregnancy











Circ 2020; 141(23):e884-903

	1 <sup>st</sup> Trimester	2 <sup>nd</sup> Trimester	3 <sup>rd</sup> Trimester	During Labor	Early Postpartum (<3 Months)	Late Postpartum (3-6 Months)
Cardiac Output	↑	↑	↑	↑	↔	↔
Blood Pressure	↓	↓	↑	↑	↓	↔
Heart Rate	↑	↑	↑	↑	↓	↔
Systemic Vascular Resistance	↓	↓	↓	↓	↑	↔

Circ 2020; 141(23):e884-903

# Labor & Delivery

	Sympathetic nervous system	Systemic vascular resistance	Cardiac output/ stroke volume	Preload
L&D		 relative increase from pregnancy	 20%	 300-500 cc
Post-Partum		 relative increase from pregnancy	 50%	

# Objectives

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  - Understand essential role of preconception counseling in women with congenital heart disease (CHD)
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-

CLINICAL STATEMENTS AND GUIDELINES - AHA SCIENTIFIC  
STATEMENT AHA SCIENTIFIC STATEMENT

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## Management of Pregnancy in Patients With Complex Congenital Heart Disease: A Scientific Statement for Healthcare Professionals From the American Heart Association

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Mary M. Canobbio, RN, MN, FAHA, Chair, Carole A. Warnes, MD, FRCP, Co-Chair, Jamil Aboulhosn, MD, Heidi M. Connolly, MD, Amber Khanna, MD, Brian J. Koos, MD, DPhil, Seema Mital, MD, FAHA, FRCPC, Carl Rose, MD, Candice Silversides, MD, FRCPC, and Karen Stout, MD, FAHA

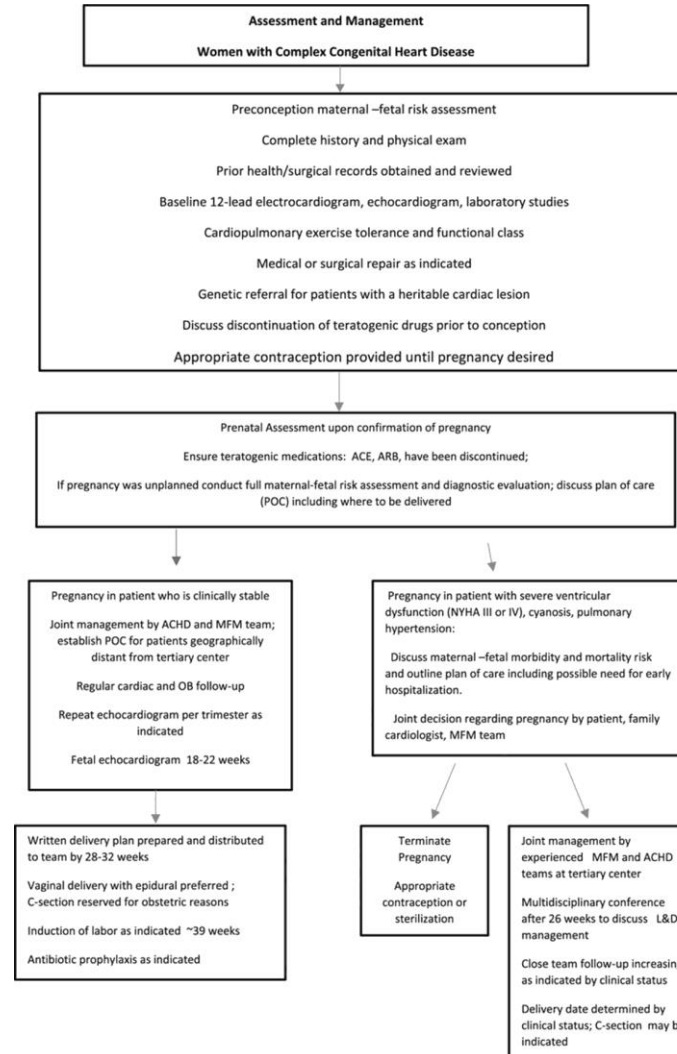
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**ABSTRACT:** Today, most female children born with congenital heart disease will reach childbearing age. For many women with complex congenital heart disease, carrying a pregnancy carries a moderate to high risk for both the mother and her fetus. Many such women, however, do not have access to adult congenital heart disease tertiary centers with experienced reproductive programs. Therefore, it is important that all practitioners who will be managing these women have current information not only on preconception counseling and diagnostic evaluation to determine maternal and fetal risk but also on how to manage them once they are pregnant and when to refer them to a regional center with expertise in pregnancy management.

**Key Words:** AHA Scientific Statements ■ heart defects, congenital ■ heart diseases ■ pregnancy



# AHA Guideline



The first (and arguably MOST IMPORTANT) step:

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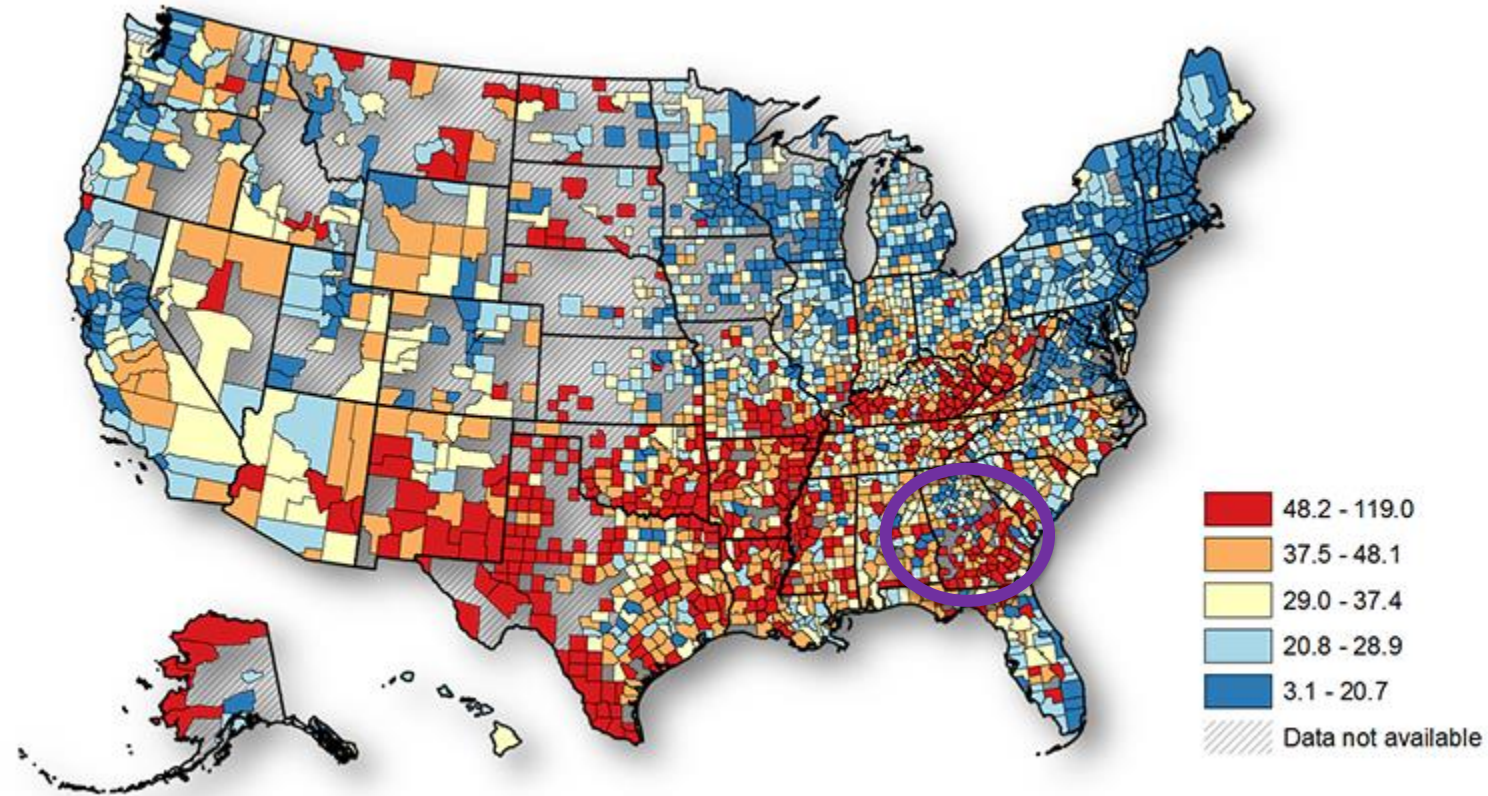
Assessment and Management  
Women with Complex Congenital Heart Disease

## Preconception maternal –fetal risk assessment

Prior health/surgical records obtained and reviewed  
Baseline 12-lead electrocardiogram, echocardiogram, laboratory studies  
Cardiopulmonary exercise tolerance and functional class  
Medical or surgical repair as indicated  
Genetic referral for patients with a heritable cardiac lesion  
Discuss discontinuation of teratogenic drugs prior to conception  
Appropriate contraception provided until pregnancy desired

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# Teenage Birth Rates by Quintile



That is a lot of red!!!

cdc.gov

- Pre-existing chronic conditions contribute to more than HALF of maternal cardiac deaths
  - More than 90% of female children born with a heart defect will reach child-bearing age
  - Many choose (or accidentally become) pregnant with SIGNIFICANT risk to themselves and the baby
  - Preconception counseling MUST begin during pediatric care as part of the transition process to adult care (within reason)
  - This includes discussion (and prescription) of appropriate and effective contraception!
-

# Risk Assessment: ZAHARA\*

Predictor	Points	Total Points	Risk
History of dysrhythmia	1.5	<b>0</b>	<b>2.9%</b>
Cardiac medication before pregnancy	1.5		
NYHA Class III or IV before pregnancy	0.75	<b>0.5-1.5</b>	<b>7.5%</b>
Left-sided heart obstruction Peak gradient > 50 mm Hg <i>or</i> Aortic valve area < 1 cm <sup>2</sup>	2.5		
Moderate/severe AV valve regurgitation (left-sided)	0.75	<b>1.51-2.50</b>	<b>17.5%</b>
Moderate/severe AV valve regurgitation (right-sided)	0.75		
Mechanical valve prosthesis	4.25	<b>2.51-3.50</b>	<b>43.1%</b>
Cyanotic heart disease	1		
<b>Total points<sup>a</sup></b>	<b>0-13</b>	<b>&gt;3.51</b>	<b>70%</b>

Abbreviations: AV, atrioventricular; NYHA, New York Heart Association.  
<sup>a</sup> An increasing ZAHARA risk score correlates with an increasing risk for adverse maternal cardiac events during pregnancy.

\*Zwangerschap bij Aangeboren HARTafwijkingen

# Risk Assessment: CARPREG II (Canadian Cardiac Disease In Pregnancy Score)

Predictor	Points
Prior cardiac events or arrhythmias	3
Baseline NYHA 3–4 or cyanosis	3
Mechanical valve	3
Systemic ventricular dysfunction LVEF < 55 %	2
High-risk valve disease or left ventricular outflow tract obstruction (aortic valve area < 1.5 cm <sup>2</sup> , subaortic gradient > 30, or moderate to severe mitral regurgitation, mitral stenosis < 2.0 cm <sup>2</sup> )	2
Pulmonary hypertension, RVSP > 49 mmHg	2
High-risk aortopathy	2
Coronary artery disease	2
No prior cardiac intervention	1
Late pregnancy assessment	1

*Primary cardiac event risk: score = 1, 5 % risk, score = 2, 10 % risk, score = 3, 15 % risk, score = 4, 22 % risk and 41 % risk if score greater than 4. NYHA = New York Heart Association Functional Classification; LVEF = left ventricular ejection fraction; RVSP = right ventricular systolic pressure. Source: Silversides et al., 2018, with permission from Elsevier.<sup>14</sup>*

- Published 2006
  - Integrates all known maternal cardiovascular risk factors, with emphasis on the specific type of congenital heart defect
  - Analyzed both maternal and offspring risk (mortality and morbidity)
-

# Modified WHO Classification

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<b>Risk Category</b>	<b>Risk Description</b>
I	No detectable increase in maternal mortality and no/mild increase in morbidity risk
II	Small increase in maternal mortality and moderate increase in morbidity risk
II-III	Moderate increase in maternal mortality and morbidity risk
III	Significantly increased maternal mortality or severe morbidity risk. Expert counseling required. In the event of pregnancy, intensive cardiovascular and obstetric monitoring needed.
IV	Extremely high maternal mortality or severe morbidity risk. Pregnancy is contraindicated. In the event of pregnancy, termination should be discussed.

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# Risk Category I- minimally increased risk\*

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<b>Risk Category</b>	<b>Risk Description</b>	<b>Maternal Risk Factors</b>
I	No detectable increase in maternal mortality and no/mild increase in morbidity risk	<ul style="list-style-type: none"><li>• Mild PS, small PDA, mitral valve prolapse</li><li>• Successfully repaired simple lesions (ASD, VSD, PDA, anomalous veins)</li><li>• Isolated PACs or PVCs</li></ul>

\*2.5-5%

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## Risk Category II- Small increased risk\*

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<b>Risk Category</b>	<b>Risk Description</b>	<b>Maternal Risk Factors</b>
II	Small increase in maternal mortality and moderate increase in morbidity risk	<ul style="list-style-type: none"><li>• Unoperated ASD/VSD</li><li>• Repaired TOF</li><li>• Most arrhythmias</li></ul>

\*5.7-10.5%

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## Risk Category II-III- Moderate increased risk\*

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<b>Risk Category</b>	<b>Risk Description</b>	<b>Maternal Risk Factors</b>
II-III	Moderate increase in maternal mortality and morbidity risk	<ul style="list-style-type: none"><li>• Mild LV impairment</li><li>• HCM</li><li>• Native or tissue valvular disease (that do not fall into I or IV)</li><li>• Marfan Syndrome without aortic dilation</li><li>• Repaired coarctation</li></ul>

\*10-19%

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## Risk Category III- Significantly Increased Risk\*

Risk Category	Risk Description	Maternal Risk Factors
III	Significantly increased maternal mortality or severe morbidity risk. Expert counseling required. In the event of pregnancy, intensive cardiovascular and obstetric monitoring needed.	<ul style="list-style-type: none"><li>• Mechanical valve</li><li>• Systemic RV</li><li>• Fontan Circulation</li><li>• Cyanotic heart disease (unrepaired)</li><li>• Other complex CHD</li><li>• Marfan Syndrome with aortic dilation 40-45 mm</li><li>• Bicuspid aortic valve with aortic dilation 45-50 mm</li></ul>

\*19-27%

# Risk Category IV- Extremely increased risk\*

Risk Category	Risk Description	Maternal Risk Factors
IV	Extremely high maternal mortality or severe morbidity risk. Pregnancy is contraindicated. In the event of pregnancy, termination should be discussed/advised. If pregnancy continues, follow level III recommendatios.	<ul style="list-style-type: none"><li>• Pulmonary Arterial Hypertension (of any cause)</li><li>• Severe systemic ventricular dysfunction (EF &lt; 30%, NYHA 3-4)</li><li>• Previous peripartum cardiomyopathy with ANY residual LV impairment</li><li>• Severe mitral stenosis</li><li>• Severe SYMPTOMATIC aortic stenosis</li><li>• Marfan Syndrome with aortic dilation &gt;45 mm</li><li>• Bicuspid aortic valve with aortic dilation &gt;50 mm</li><li>• Native severe coarctation</li></ul>

\*- Includes Fontan with complications

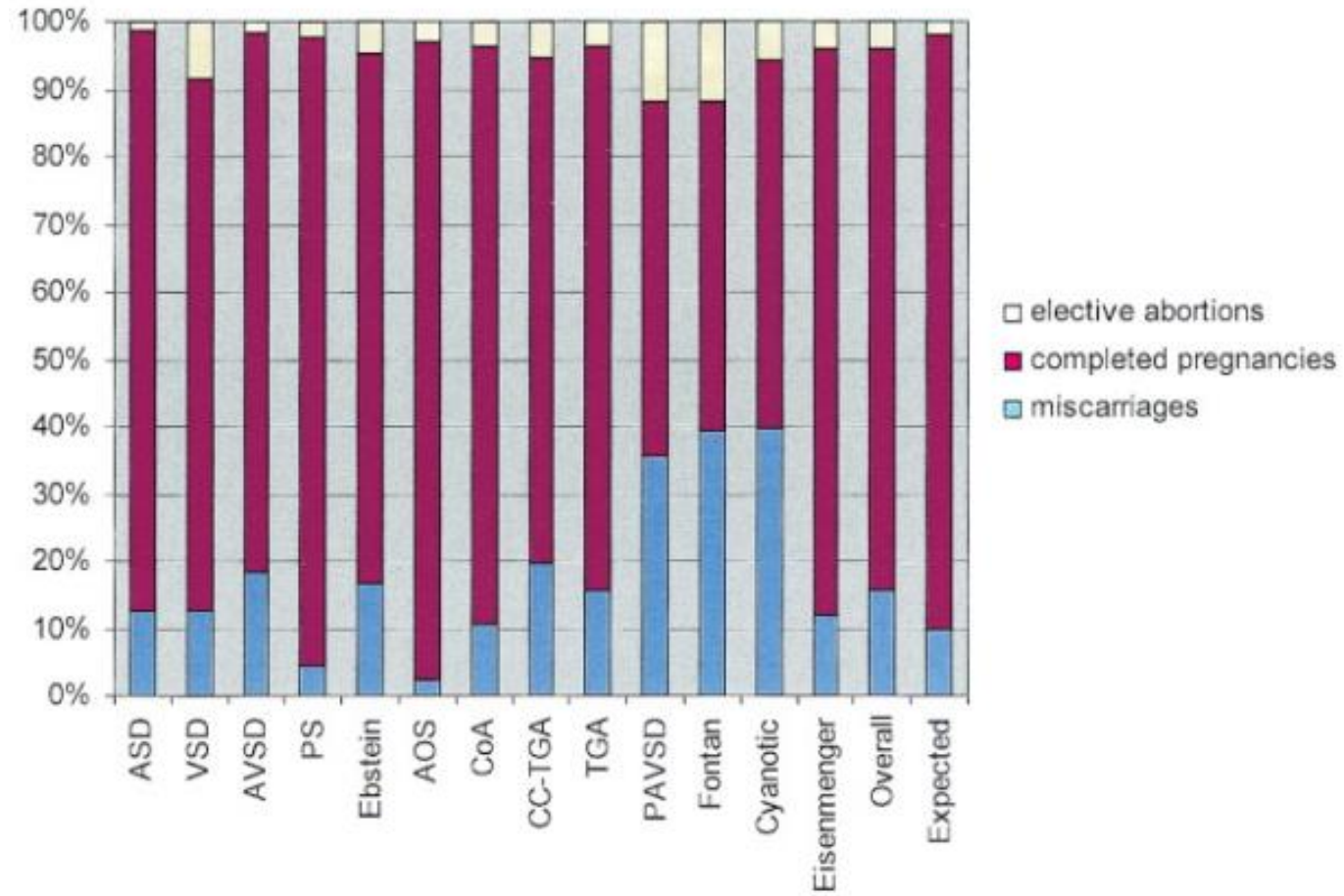
\*40-100%

# Fetal Risk

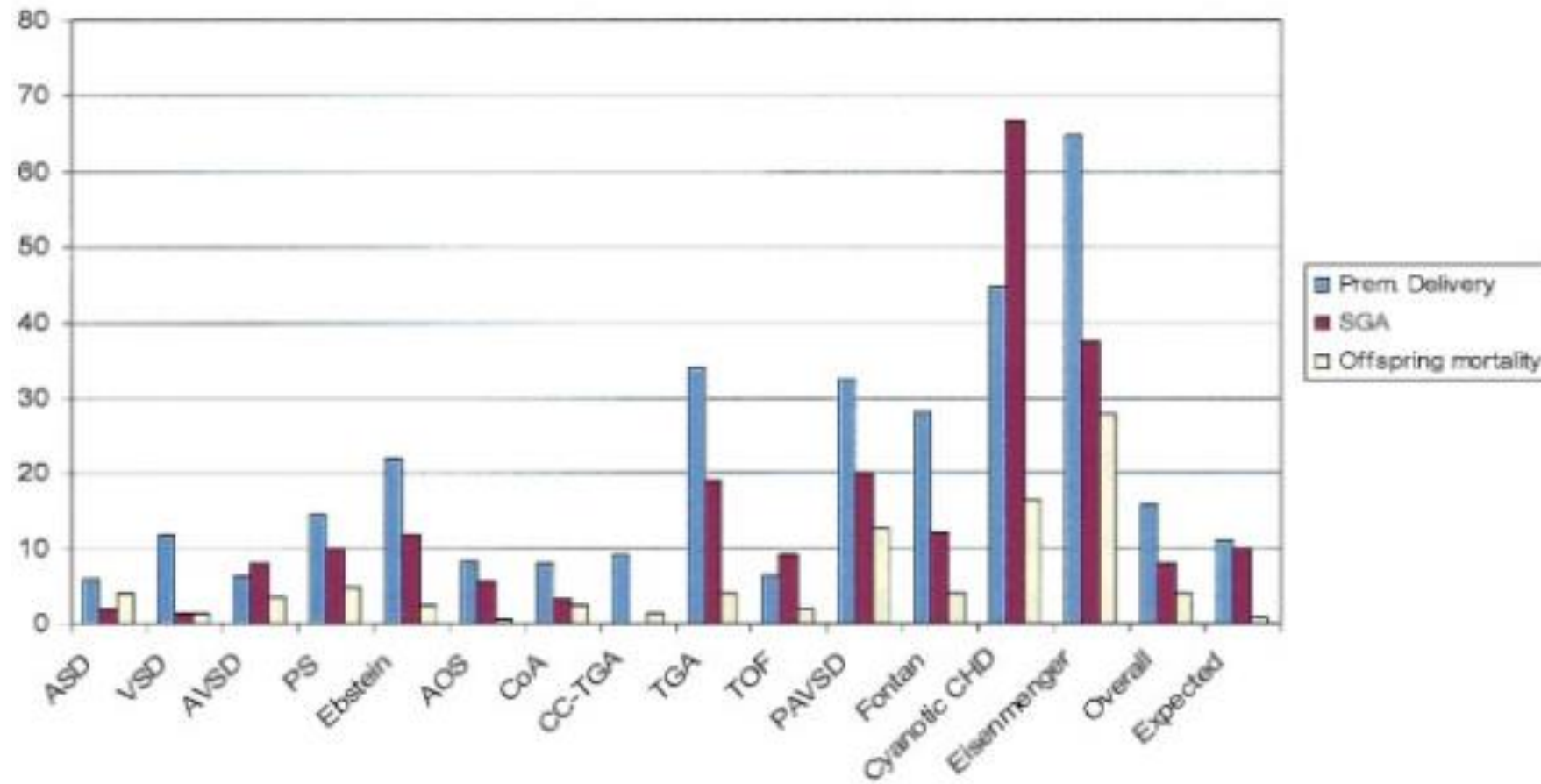
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- Highest risk is pre-term birth (22-65% of pregnancies)
  - Increased risk of pregnancy loss
  - IUGR, SGA, IVH
  - Increased risk of fetal CHD
  - Increased perinatal mortality (as high as 400%)
-

# Fetal Risk



# Fetal Risk





# My Approach to preconception counseling:

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- Assess patient's functional status by history and exam
  - Evaluate risk category using a combination of ZAHARA, CARPREG-II, and mWHO stratifiers
  - +/- CPET testing (especially for mWHO II, II/III, and III)
  - Discuss the degree of risk in detail with patient
  - Refer to MFM for obstetric preconception counseling
  - Genetics?
  - Ultimately, it is the patient's decision- all we can do is give our recommendations
  - "The worst pregnancy in CHD is the OOPSIE pregnancy!"
    - T. Smith, supposedly
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# Objectives

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- Review the physiologic changes that occur during normal pregnancy and how this impact cardiovascular function
  - Understand essential role of preconception counseling in women with congenital heart disease (CHD)
  - **Be familiar with pregnancy issues related to selected Congenital Heart Defects**
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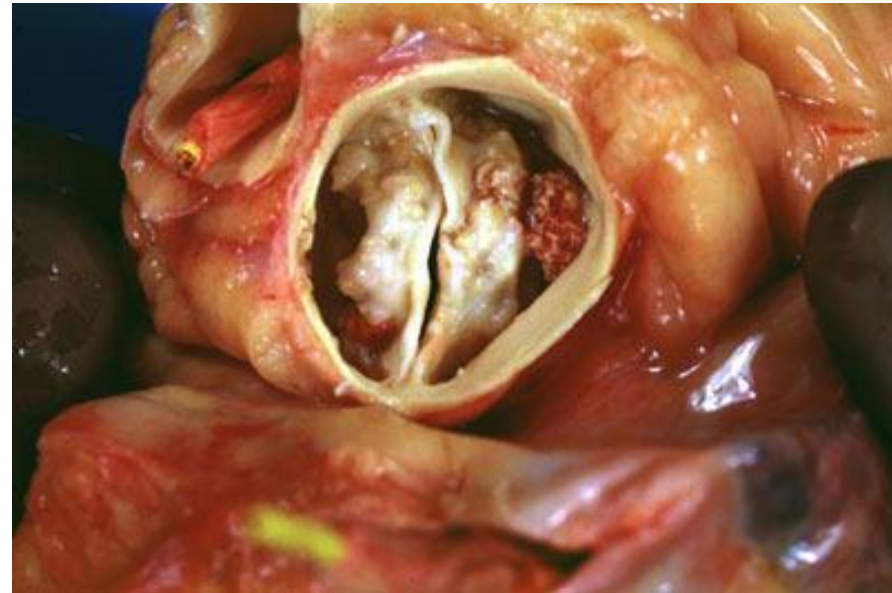
# Bicuspid Aortic Valve

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1-2% of the total population

Most common cause of isolated AS in adults

Thickening noted as early as 2<sup>nd</sup> decade



# Bicuspid Aortic Valve

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- Lesion progresses to significant AS in 75% of patients requiring eventual surgery
- Aortic valve regurgitation
- Associated aortic dilatation



# Coarctation of the Aorta

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Congenital condition whereby the aorta narrows in the area where the ductus arteriosus inserts

Blood flow to the aorta that is distal to the narrowing is dependent on the ductus arteriosus

Ductal coarctation - Usually appears when the ductus arteriosus closes

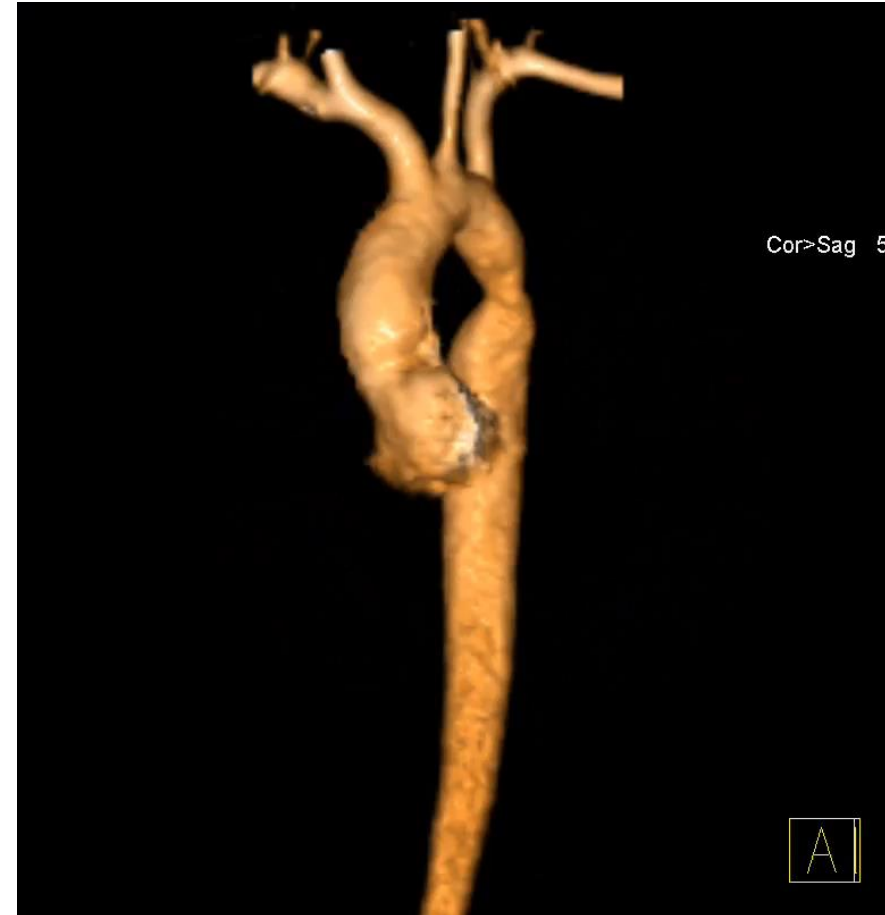
Postductal coarctation - Narrowing is distal to the insertion of the ductus arteriosus; most common



# Coarctation of the Aorta

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- Initially regarded as a simple lesion
- Well described late complications:
  - Early Systemic HTN
  - Premature atherosclerosis
  - Cerebrovascular events
  - Complication of BAV disease
  - Recurrence/residual coarctation



# Recurrent Coarctation and Coarctation Stenting

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# Left Heart Obstruction and Pregnancy

- Degree of Risk depends on degree of obstruction
- Mild-Mod usually ok, most mod-severe are ok
  - 60 patients: 8% heart failure, 10% low birth weight
  - 49 pregnancies, ½ severe stenosis: no mortality, 10% with complications of pulm edema and atrial arrhythmia, one intervention
- Severe stenosis- pregnancy is not recommended
  - Arrhythmia
  - Myocardial ischemia
  - Worsening LVH with diastolic or systolic dysfunction/heart failure

Risk Category	Risk Description	Maternal Risk Factors
II-III	Moderate increase in maternal mortality and morbidity risk	<ul style="list-style-type: none"> <li>• Mild LV impairment</li> <li>• HCM</li> <li>• Native or tissue valvular disease (that do not fall into I or IV)</li> <li>• Marfan Syndrome without aortic dilation</li> <li>• Repaired coarctation</li> </ul>

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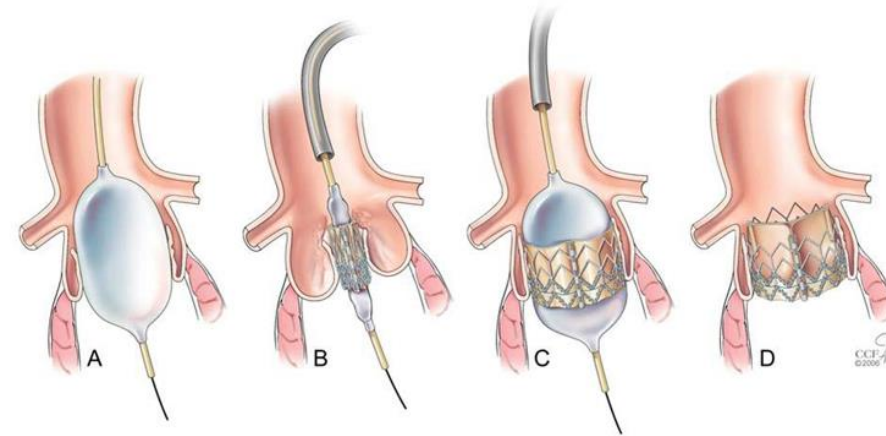
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# Valve/Aortic interventions during pregnancy

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- Ideally, interventions should occur prior to pregnancy
- Peak velocity  $>4.0$  m/s, mean gradient  $>40$  mmHG)
- Growing literature on interventions during pregnancy



# Valve/Aortic interventions during pregnancy

**TABLE 1 Published Cases of PBAV During Pregnancy**

First Author, Year	Age (y)	GA (weeks)	Etiology of Aortic Disease	Gradient Preprocedure (mmHg)	Gradient Postprocedure (mmHg)	Fluoroscopy Time (min)	Post-Procedure AR	Maternal Complications	Fetal Complications
Angel, 1988 <sup>25</sup>	17	19	Congenital	150 (peak)	68 (peak)	20.1	NA	None	None
Mchvor, 1991 <sup>26</sup>	19	14	Congenital	64 (peak)	32 (peak)	29	NA	None	None
Savas, 1991 <sup>27</sup>	22	22	Rheumatic	45 (peak)	22 (peak)	46 <sup>a</sup>	NA	None	None
Banning, 1993 <sup>28</sup>	26	14	Congenital	128 (peak)	50 (peak)	NA	Moderate	None	None
Banning, 1993 <sup>28</sup>	19	16	Congenital	123 (peak)	60 (peak)	NA	NA	None	None
Lao, 1993 <sup>29</sup>	26	16	Congenital	70 (mean)	30 (mean)	NA	NA	Transient seizure following 48 s of hypotension during the second inflation	None
Perloff, 1994 <sup>30</sup>	26	36	Congenital	100 (peak)	30 (peak)	NA	NA	None	None
Bhargava, 1998 <sup>41</sup>	27	26	Rheumatic	132 (peak)	41 (peak)	4.1	Trivial	None	None
Tumelero, 2004 <sup>42</sup>	16	27	Congenital	105 (peak)	20 (peak)	NA	Mild-moderate	None	Emergency CS secondary to oligohydramnios and placental insufficiency
Radford, 2004 <sup>43</sup>	36	13	Congenital	40 (mean)	11 (mean)	53	Moderate	Pulmonary edema post-delivery	CS at 39 wks due to fetal heart rate decelerations
Yap, 2006 <sup>44</sup>	25	16	Rheumatic	65 (mean)	28 (mean)	N/A	Moderate	None	None
Dawson, 2012 <sup>45</sup>	43	28	Rheumatic	40 (mean)	18 (mean)	N/A	Moderate	None	None
Dawson, 2012 <sup>45</sup>	32	26	Congenital	70 (mean)	25 (mean)	N/A	N/A	N/A	N/A
Vinotha, 2012 <sup>46</sup>	27	19	Congenital, endocarditis	118 (peak)	N/A	N/A	Mild	Sinus tachycardia (120 s) after PBAV, managed medically	Emergency CS at 32 wks secondary to fetal compromise with IUGR

<sup>a</sup>Fluoroscopy time includes triple valve intervention.  
AR = aortic regurgitation; CS = cesarean section; GA = gestational age; IUGR = intrauterine growth restriction; mmHg = millimeters of mercury; N/A = information not available; PBAV = percutaneous balloon aortic valvuloplasty.

*Long term impact of radiation dose on fetus/child???*

Elkayam et al. JACC. 2022.

**TABLE 2 Published Cases of TAVR During Pregnancy**

First author	Hodson et al <sup>53</sup>	Gandhi et al <sup>54</sup>	Maluenda et al <sup>55</sup>	Berry et al <sup>56</sup>	Chengode et al <sup>57</sup>	Herbert et al <sup>58</sup>	Zhong et al <sup>59</sup>
Patient age (y)	22	29	39	33	34	30	29
GA at procedure (wk)	22	14	23	22	22	19	12
Type of the valve	Native BAV	23-mm CE Perimount Magna	21-mm Freestyle Medtronic	21-mm CE Magna	21-mm CE Perimount Magna (M-27 mm)	19-mm Magna Ease	27-mm Freestyle Medtronic
Age of prior intervention (y)	9 (PBAV)	24	23	N/A	26	25	16
Symptoms	Dizziness, DOE, chest pain	NYHA class III HF, CCS class 3	NYHA class III HF	Progressive DOE	NYHA class III HF	NYHA class III HF	NYHA class III HF
Peak/mean aortic gradient before TAVR	110/56	149/98	98/51	102/61	148/66	153/92	104/65
AVA before procedure (cm <sup>2</sup> )	1	0.8	N/A	0.66	0.7	0.8	0.63
Degree of AR	Moderate	Mild	Severe	Moderate	Mild-to-moderate	None	None
PA pressure (mmHg)	N/A	Normal	Normal	N/A	72	52	N/A
LV function	Normal	Normal	Normal	Normal	Normal	Normal	Normal
Imaging modality	IVUS, 3D-TEE, Fluoro	TEE, Fluoro	CTA, Cine	CTA	TEE, Fluoro	TEE, Fluoro, Cine	FA US, CT chest, TEE, Fluoro
Type of valve	Core Valve	Sapien XT	Sapien XT	Sapien 3	Sapien XT	Sapien 3	Core Valve
Size of TAVR valve (mm)	26	23	23	20	23	20	26
Fluoroscopy time (min) + radiation dose (mGy)	10.03 (AK 101 mGy)	16.03 (AK 298 mGy)	NA	N/A	3:06 (AK 16 mGy)	18	30 mGy (fetal radiation dose estimate for all procedures)
Peak/mean aortic gradient post-TAVR (mmHg)	N/A	47/23	NA	61/23	68/24	52/27	24/14
AVA post-procedure (cm <sup>2</sup> )	N/A	N/A	NA	1.1	N/A	N/A	N/A
Procedural complications	LBBB/mild PVL	No PVL	No PVL	Trace PVL	No PVL	No PVL	Trace PVL
Meds post-procedure	ASA 81 mg every day	Dalteparin 12,500 units SQ/d x 1 mo; ASA 81 mg/d indefinitely	Clopidogrel 75 mg every day (ASA allergy)	N/A	ASA 81 mg/d; LMWH 80 mg SQ/d until admission for delivery	N/A	LMWH during pregnancy. ASA post-delivery
Delivery mode	Vaginal	Vaginal	NA	CS	Vaginal	Vaginal	Planned vaginal converted to CS
GA at delivery (wk)	38	39	NA	37	Full term	33	36
Maternal complications	Persistent LBBB	None	None	None	None	None	Premature rupture of the membrane
Fetal complications	None	None	None	None	None	None	None

<sup>a</sup>Transcatheter aortic and mitral double valve-in-valve implantation through left ventricular apical approach.  
3D = 3-dimensional; AK = air kerma; AR = aortic regurgitation; ASA = aspirin; AVA = aortic valve area; BAV = bicuspid aortic valve; CCS = Canadian Cardiovascular Society; cine = cineangiography; CS = Cesarean section; CTA = computed tomography angiography; DOE = dyspnea on exertion; FA = femoral artery; Fluoro = Fluoroscopy; GA = gestational age; IVUS = intravascular ultrasound; LBBB = left bundle branch block; LMWH = low-molecular-weight heparin; LV = left ventricle; meds = medications; NA = information not available; NYHA = New York Heart Association; PA = pulmonary artery; PBAV = percutaneous balloon aortic valvuloplasty; PVL = paravalvular leak; SQ = subcutaneous; TEE = transesophageal echocardiography; US = ultrasound.

# Coarctation Stenting During Pregnancy

Patient number/ pregn. weeks or time after delivery	Age (years)	Diagnosis	S/D blood pressure mmHg	PG (TEE), mmHg	Invasive PG, mm Hg	Type of stent
1/19 week (w).	19	Mid-aortic s	290/110	90	140	CP covered 8Z34
2/22 w.	33	CoA	160/80	40	60	Pulmaz 3910
3/23 w.	19	CoA with hypo- plastic aortic arch	150/80	35	55	CP covered 28 mm
4/15 w.	28	ReCoA	160/60	25	50	Andrastent 26XL
5/2 m.	29	CoA	130/70	25	45	Andrastent 30XL
6/48 hr.	28	CoA+PDA, Ao dissection	150/80	30	55	CP covered 34 mm
7/6 m	25	CoA	160/120	25	40	Andrastent 26XL
8/5 yrs.	31	CoA	160/80	30	50	Andrastent 30XL
9/2 yrs.	22	CoA	140/90	20	35	Andrastent 26XL
10/1 yr.	26	CoA	130/75	35	60	Andrastent 30XL

S/D pressure – systolic/diastolic pressure in the left arm (mean in pressure Holter); PG – pressure gradient; Mid-aortic S. – mid-aortic syndrome (suprarenal); CoA – native coarctation of the aorta; ReCoA – aortic coarctation after surgical repair 24 y/ago; Hypoplastic aortic arch (B); CoA+PDA, Ao dissection – Aortic aneurysm dissection type II DeBakey after 24 hours after delivery; PDA – patent ductus arteriosus.

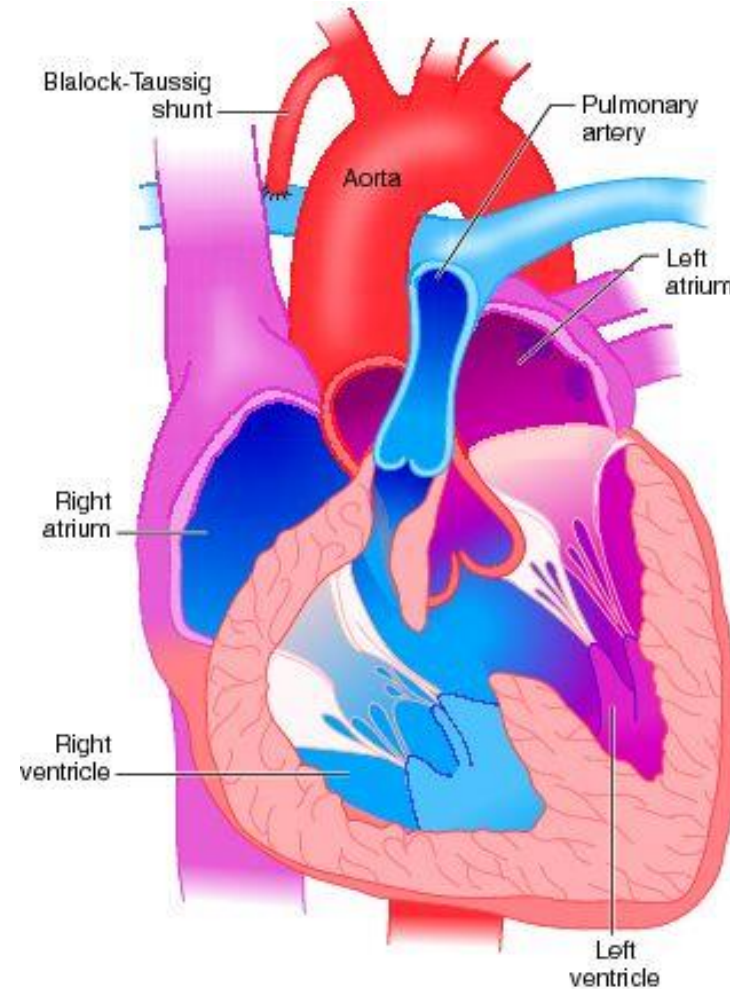
# Objectives

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- Review the physiologic changes that occur during normal pregnancy and how this impact cardiovascular function
  - Understand essential role of preconception counseling in women with congenital heart disease (CHD)
  - **Be familiar with pregnancy issues related to selected Congenital Heart Defects**
    - Left Heart obstructive lesions
    - **Tetralogy of Fallot/Conotruncal lesions**
    - Pulmonary Hypertension/Eisenmenger syndrome
    - Single Ventricle/Fontan
-

# Tetralogy of Fallot

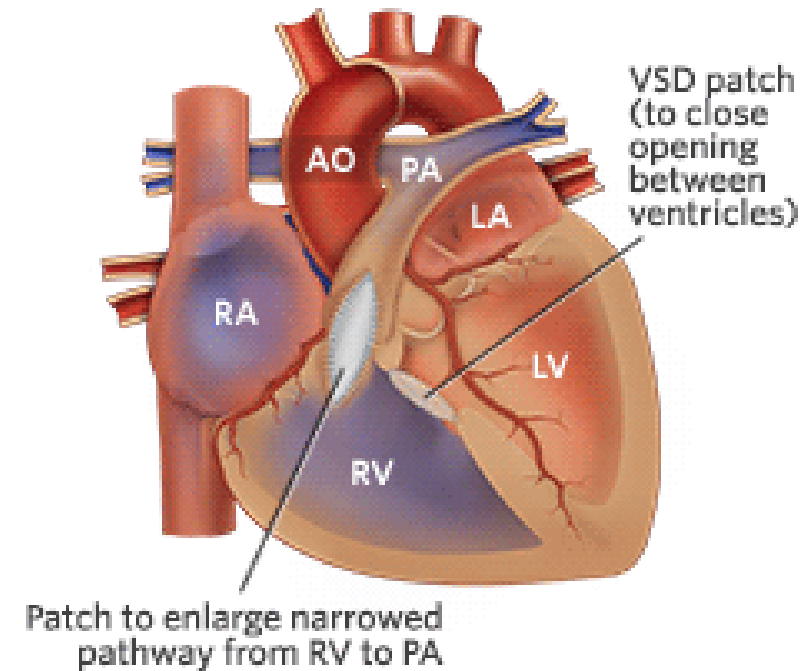
- Ventricular Septal Defect
  - Anterior malalignment of the conal septum\*
- Overriding aorta
- RV outflow tract obstruction
- RV hypertrophy



# Tetralogy of Fallot- Repair

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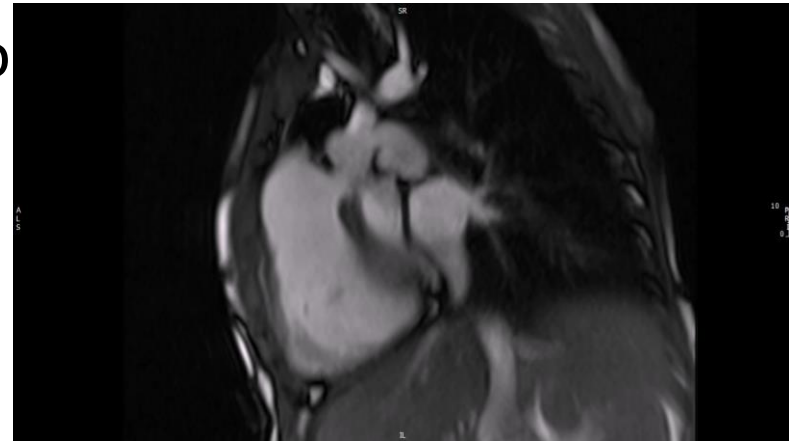
- Closure of VSD
- Relief of obstruction in the RV outflow
- +/- disruption of the pulmonary valve (depending on size)



# Tetralogy of Fallot- Complications after Repair

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- Chronic Pulmonary valve leak leading to RV enlargement
- Residual RV outflow obstruction
- RV progressive dysfunction
- SVT and VT
- Risk for Sudden cardiac death
- Similar issues are seen in other forms of CHD
  - Ie. Truncus arteriosus, double outlet right ventricle, AS with Ross-Konno operation, etc.



# Tetralogy of Fallot- Pregnancy

- Risk depends largely on degree of valve dysfunction
- ? Those with a well functioning native PV (valve sparing repair) or bioprosthetic valve are likely at less risk
- ? Stenotic lesions at higher risk than regurgitant lesions

Risk Category	Risk Description	Maternal Risk Factors
II	Small increase in maternal mortality and moderate increase in morbidity risk	<ul style="list-style-type: none"><li>• Unoperated ASD/VSD</li><li>• Repaired TOF</li><li>• Most arrhythmias</li></ul>

Risk Category	Risk Description	Maternal Risk Factors
II-III	Moderate increase in maternal mortality and morbidity risk	<ul style="list-style-type: none"><li>• Mild LV impairment</li><li>• HCM</li><li>• Native or tissue valvular disease (that do not fall into I or IV)</li><li>• Marfan Syndrome without aortic dilation</li><li>• Repaired coarctation</li></ul>



# Tetralogy of Fallot- Pregnancy

**Table I.** Patient characteristics for the total cohort and for patients with and without completed pregnancies

	All patients (N = 204)	Patients without completed pregnancies* (n = 135)	Patients with completed pregnancies* (n = 69)	P value†
Mean age at inclusion, y (±SD)	31 ± 7.5	28 ± 6.7	36 ± 5.8	<.001
Mean age at repair, y (±SD)	5.1 ± 2.7	4.7 ± 2.5	6.1 ± 2.8	<.001
Surgeries before or after repair (%)				
Blalock-Taussig shunt	22 (10.8)	15 (11.1)	7 (10.1)	NS
Waterston shunt	29 (14.2)	14 (10.4)	15 (21.7)	<.05
Potts shunt	2 (1.0)	0 (0.0)	2 (2.9)	NS
RVOT procedure	42 (20.6)	24 (17.8)	18 (26.1)	NS
PVR	44 (21.6)	26 (19.3)	18 (26.1)	NS
Valvular dysfunction (%)				
Tricuspid regurgitation‡	27 (18.5)	15 (18.1)	12 (19.0)	NS
RVOT obstruction	46 (31.5)	21 (25.3)	25 (39.7)	NS
Pulmonary valve regurgitation‡	77 (52.7)	39 (47.0)	38 (60.3)	NS
Ventricular septal defect (%)	13 (8.9)	6 (4.4)	7 (11.1)	NS
Mental disability (%)	12 (5.9)	12 (8.9)	0 (0.0)	<.01

RVOT, Right ventricular outflow tract; NS, not significant ( $P > .05$ ).

\* Completed pregnancy, lasting >20 weeks.

† Comparison of women without completed pregnancies versus women with completed pregnancies.

‡ Moderate or severe.<sup>1,5</sup>

- 157 pregnancies in 74 women (out of 204 total rTOF patients)
- 30 miscarriages (19%); 4 elective abortions (2.5%)

Balci et al. American Heart Journal. 2011.

	n = 123
Mean age at inclusion, y (±SD)	36.4 ± 5.6
Mean age at repair, y (±SD)	6.5 ± 4.2
Surgery before/after repair, n (%)	
Blalock-Taussig shunt	12 (9.8)
Waterston shunt	22 (17.9)
Potts shunt	2 (1.6)
RVOT procedure	47 (38.2)
PVR	33 (26.8)
Valvular dysfunction, n (%)	
Tricuspid regurgitation (moderate/severe) <sup>1,5</sup>	25 (20.3)
RVOT obstruction	54 (43.9)
Pulmonary valve regurgitation (moderate/severe) <sup>1,5</sup>	69 (56.1)
Ventricular septal defect, n (%)	20 (16.3)
History of arrhythmias, n (%)	5 (4.1)
History of heart failure, n (%)	3 (2.4)
NYHA class ≥II pre-pregnancy, n (%)	8 (6.5)
Cardiac medication used pre-pregnancy, n (%)	7 (5.7)
β-Blockers†	1 (0.8)
Calcium-channel blockers	1 (0.8)
Digoxin	3 (2.4)
Amiodarone	2 (1.6)
Vitamin K antagonists, n (%)	2 (1.6)
Mean age at pregnancy, y (±SD)	26.8 ± 4.1
Primipara, n (%)	32 (26.0)
Multipara, n (%)	91 (74.0)
Mean pregnancy duration, wk (±SD)	37.8 ± 4.5

\* Completed pregnancy, lasting >20 weeks.

† Stopped using because of pregnancy.

# Tetralogy of Fallot- Pregnancy

	n = 123	
	ZAHARA n (%)	Healthy population <sup>§</sup> (%)
Cardiovascular events	10 (8.1)	
Arrhythmias	8 (6.5)	<1
Heart failure	2 (1.6)	<1
Thromboembolic events	1 (0.8)	<.3
Obstetric events	73 (58.9)	
Cesarean delivery	25 (20.3)	6.5
Assisted <sup>†</sup> vaginal delivery	16 (13.0)	17
PPH	12 (9.7)	2.9
Prolongation of 2nd stage of delivery	10 (8.1)	<2.7
PPROM	8 (6.5)	1.5
PIH	6 (4.8)	10
Prolongation of cervix ripening	5 (4.1)	–
Preeclampsia	4 (3.2)	1.4
Hyperemesis	2 (1.6)	0.6
Soluto placenta	1 (0.8)	<1
Uterus rupture	1 (0.8)	<0.001
Offspring events	42 (33.9)	
SGA	23 (18.5)	10
PB	22 (17.7)	10
OM	8 (6.4)	0.9
CHD	3 (2.4)	0.6
Other offspring events <sup>‡</sup>	3 (2.4)	–

PB, Preterm birth; OM, offspring mortality.

\* Completed pregnancy, lasting >20 weeks.

† Delivery assisted using forceps or vacuum extraction.

‡ Other offspring events were fetal asphyxia, trisomy 13, and hydrocephalus.

§ Normal population, based on literature.<sup>4,24,25,27,29,32,33</sup>

**Table IV.** Results of univariable and multivariable logistic regression models and corresponding risk scores for cardiac, obstetric, and offspring events

Univariable analysis	Cardiovascular events <sup>*</sup>	Obstetric events <sup>*</sup>	Offspring events <sup>*</sup>
Palliative surgery	1.3 (0.3-5.1)	0.8 (0.4-1.7)	2.7 (1.1-6.2) <sup>‡</sup>
History of arrhythmias	9.3 (1.8-46.9) <sup>‡</sup>	5.2 (0.6-43.6)	2.0 (0.5-8.5)
Prior PVR	3.1 (0.8-11.4) <sup>†</sup>	2.2 (0.9-5.3) <sup>†</sup>	1.4 (0.6-3.2)
RVOT obstruction	0.9 (0.2-3.2)	1.2 (0.6-2.4)	1.3 (0.6-2.7)
Pulmonary valve regurgitation <sup>§</sup>	0.5 (0.1-1.9)	0.5 (0.23-0.99) <sup>‡</sup>	1.3 (0.6-2.7)
Pulmonary AV valve regurgitation	0.6 (0.1-2.3)	1.4 (0.6-3.4)	2.1 (0.9-5.2) <sup>†</sup>
Patent shunt	0.8 (0.1-3.8)	2.5 (0.9-6.7)	1.1 (0.4-2.9)
Smoking during pregnancy	0.8 (0.1-3.8)	0.9 (0.4-2.5)	1.4 (0.5-3.7)
Use of cardiac medication pre pregnancy	11.8 (2.2-63.3) <sup>‡</sup>	–	5.5 (1.0-29.2) <sup>‡</sup>
NYHA class >II	0.6 (0.1-5.3)	–	0.3 (0.0-2.7)

Multivariable analysis for endpoints cardiovascular, obstetric and offspring events		Odds ratio (95% CI)
<b>Cardiovascular events</b>		
Use of cardiac medication pre pregnancy		11.7 (2.2 – 62.7) <sup>‡</sup>
<b>Obstetric events</b>		
Pulmonary valve regurgitation		0.5 (0.2 – 0.99) <sup>‡</sup>
<b>Offspring events</b>		
Palliative surgery		3.3 (1.3 – 8.2) <sup>‡</sup>
Use of cardiac medication pre pregnancy		8.1 (1.4 – 48.6) <sup>‡</sup>

AV, Atrioventricular.

\* Expressed as OR (95% CI).

† P < .1.

‡ P < .05.

§ Moderate or severe.<sup>14,15</sup>

# Pulmonary Valve interventions during pregnancy

**TABLE 5 Reports of PBPV During Pregnancy**

First author	Oylumlu et al <sup>112</sup>	Johny et al <sup>113</sup>	Sener et al <sup>114</sup>
Patient age (y)	32	34	23
GA at procedure (wk)	28	31	34
Type of valve	Congenital	Congenital	Congenital
Age at prior intervention (y)	None	PBPV at the age of 26 y during the first pregnancy (second trimester)	None
Reason for intervention	Exertional chest pain, mild dyspnea	NYHA class III HF	NYHA class II HF
Peak gradient before valvuloplasty (mmHg)	126 (TTE)	192 (invasive)	122 (TTE)
Degree of regurgitation	None	N/A	None
RV function	Normal	Normal	RV dilatation, RV hypertrophy
Imaging modality	Fluoroscopy	Fluoroscopy	Fluoroscopy
Fluoroscopy time (min)	N/A	N/A	N/A
Peak gradient post valvuloplasty (mmHg)	37 (peak instantaneous)	120 (peak-to-peak)	48 (peak instantaneous)
Procedure complications	None	Mild hypotension with uterine contractions for 30 mins	None
Delivery mode	N/A	CS	CS
GA at delivery (wk)	N/A	36	Full term
Maternal complications	None	None	None
Fetal complications	None	None	None

CS = cesarean section; GA = gestational age; HF = heart failure; min = minute; N/A = information not available; NYHA = New York heart association; PBPV = percutaneous balloon pulmonary valvuloplasty; RV = right ventricle; TTE = transthoracic echocardiography.

**Valve Implantation During Pregnancy**

	Detzner et al <sup>115</sup>	Ormerod et al <sup>116</sup>
Patient age (y)	20	21
GA at procedure (wk)	13	23
Type of valve	22-mm Contegra conduit RV-PA	19-mm homograft conduit RV-PA
Imaging modality	12	3
Reason for intervention	NYHA class II HF, with concern for worsening symptoms during pregnancy	Asymptomatic but fetal IUGR
Peak gradient before procedure (mmHg)	42	23
Peak gradient post procedure (mmHg)	N/A	N/A
Valve area (cm <sup>2</sup> )	Moderate	Severe
RV pressure (mmHg)	70/18	72/15
RV function	Normal	Normal
Imaging modality	Low frame rate fluoro	MRI, Fluoro, Cine
Type of valve	22-mm Melody	22-mm Melody
Fluoroscopy time (min)	N/A	N/A
RV pressure/PV gradient post-procedure (mmHg)	RV 44/13; PV 7	RV 76/12; PV 27 (trivial PR)
VA post-procedure (cm <sup>2</sup> )	N/A	N/A
Procedural complications	None	None
Meds post-procedure	N/A	N/A
Delivery mode	N/A	CS
GA at delivery (wk)	30 + 6	32
Maternal complications	None	Pre-eclampsia
Fetal complications	Dichorionic twins	SGA (1,500 gm)

Cine = cineangiography; CS = cesarean section; Fluoro = fluoroscopy; GA = gestational age; HF = heart failure; IUGR = intrauterine growth restriction; meds = medications; MRI = magnetic resonance imaging; N/A = information not available; NYHA = New York Heart Association; PA = pulmonary artery; PR = pulmonic regurgitation; PV = pulmonary valve; RV = right ventricle; SGA = small for gestational age; VA = valve area.

# Tetralogy of Fallot- Pregnancy

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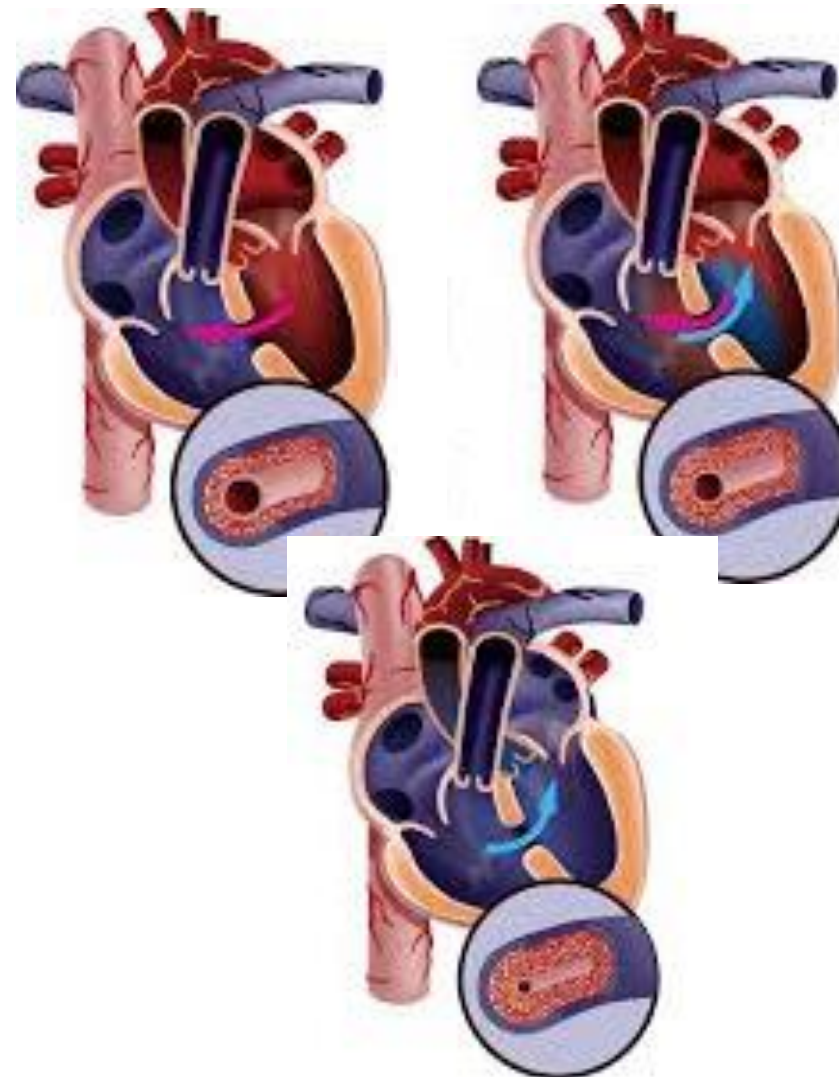
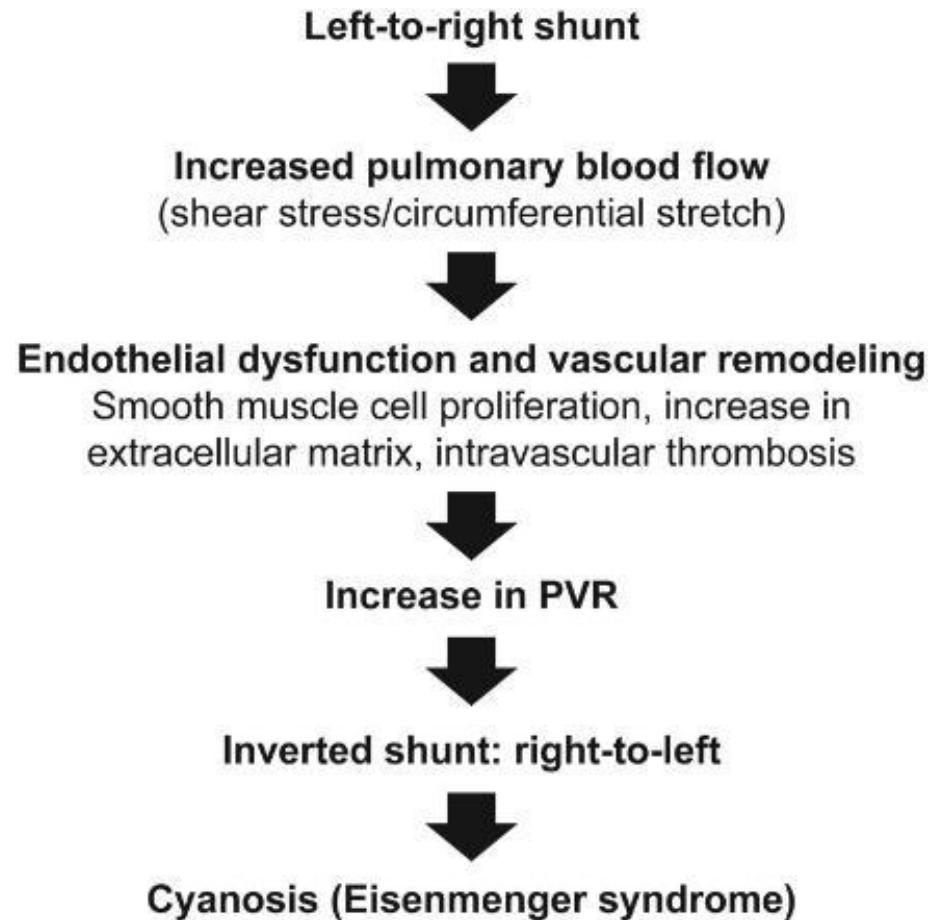
- Pregnancy is increased risk, but highly dependent on each patient
  - Best results probably come from more aggressive optimization by cardiology BEFORE pregnancy
  - C-section rarely indicated (aortic dilatation, arrhythmias, CHF)
  - Like most patients- need regular cardiac and MFM monitoring.
-

# Objectives

---

- Review the physiologic changes that occur during normal pregnancy and how this impact cardiovascular function
  - Understand essential role of preconception counseling in women with congenital heart disease (CHD)
  - **Be familiar with pregnancy issues related to selected Congenital Heart Defects**
    - Left Heart obstructive lesions
    - Tetralogy of Fallot/Conotruncal lesions
    - **Pulmonary Hypertension/Eisenmenger syndrome**
    - Single Ventricle/Fontan
-

# Eisenmenger Syndrome



- Polycythemia
- Coagulopathy
- Gout
- Renal dysfunction
- Stroke (coagulopathic, R→L shunt; IV lines)
  - Increased with microcytosis
  - Atrial fibrillation
- High risk pregnancy (30-50% mortality)



# Eisenmenger Syndrome- Pregnancy

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- Maternal O<sub>2</sub> saturation <85%
  - 12% live birth rate
- No phlebotomy
- Possible benefit of supplemental oxygen
- Air Filters on all Ivs
- No cardiac indications for C/s
- Monitor for ~48 hours in ICU





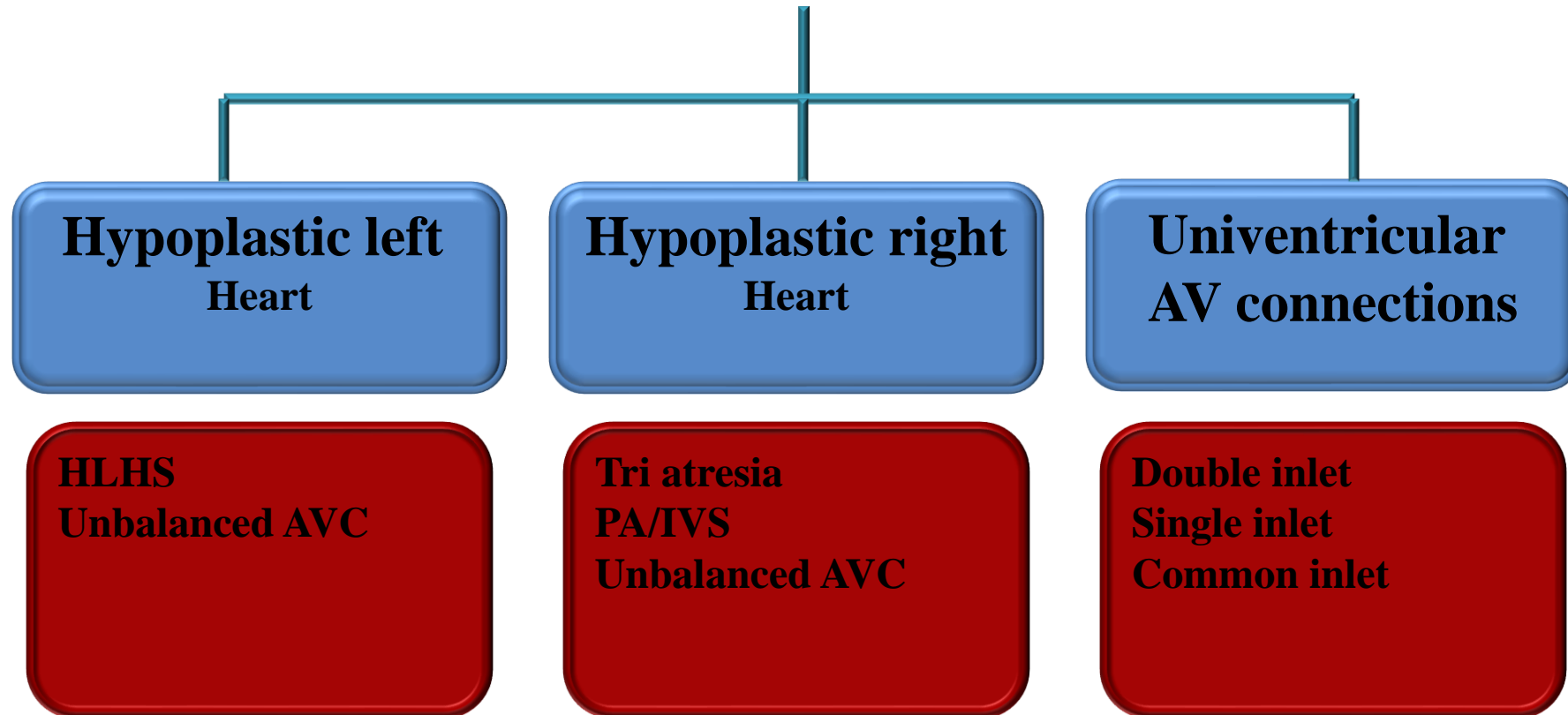
# Objectives

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- Review the physiologic changes that occur during normal pregnancy and how this impact cardiovascular function
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  - **Be familiar with pregnancy issues related to selected Congenital Heart Defects**
    - Left Heart obstructive lesions
    - Tetralogy of Fallot/Conotruncal lesions
    - Pulmonary Hypertension/Eisenmenger syndrome
    - **Single Ventricle/Fontan**
-

# Single Ventricle Heart Disease

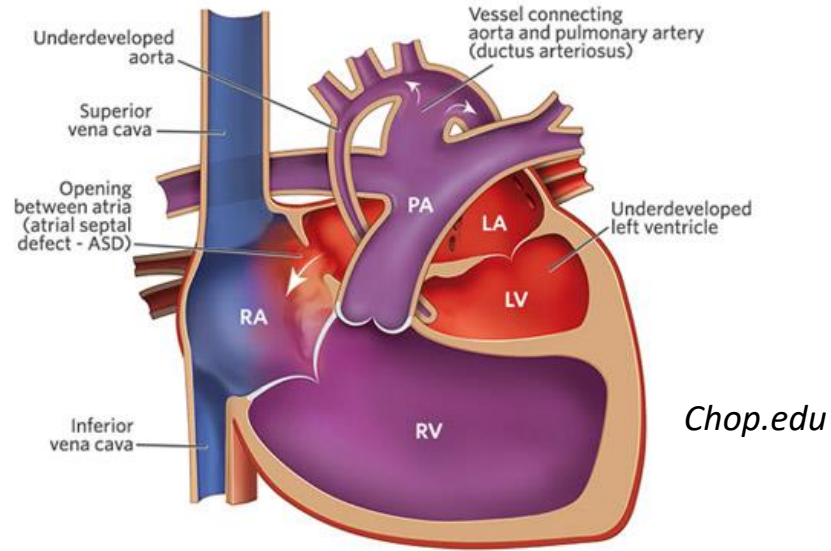
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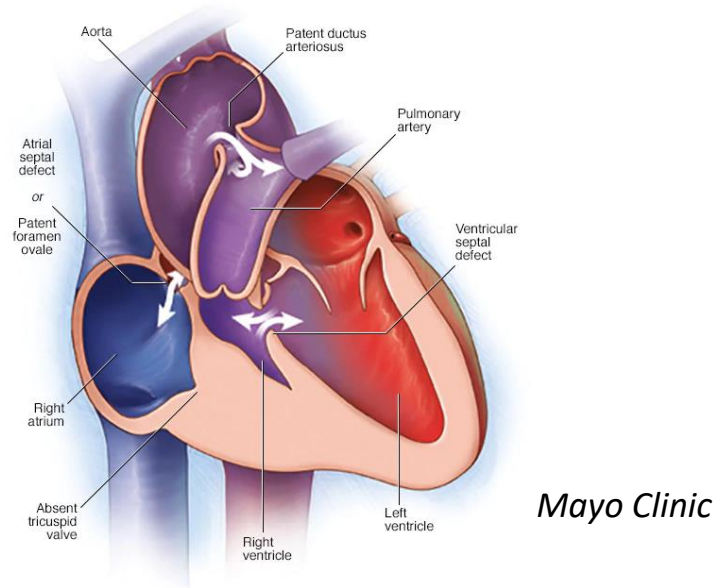
**Systemic or pulmonary outflow obstruction**

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# Single Ventricle Heart Disease

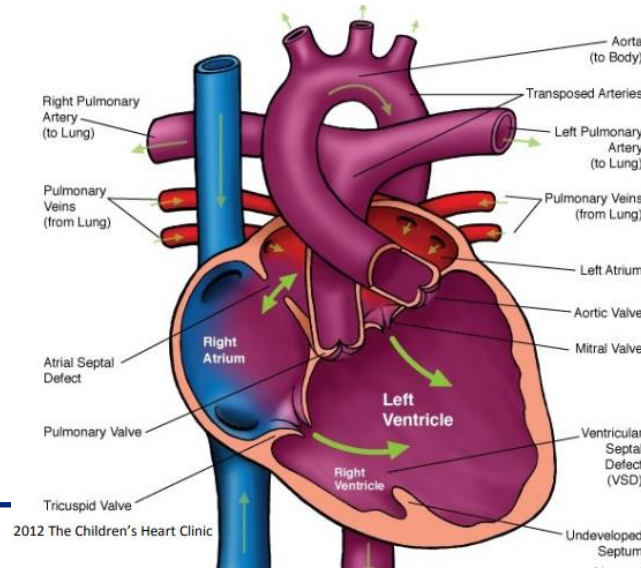


*Chop.edu*



*Mayo Clinic*

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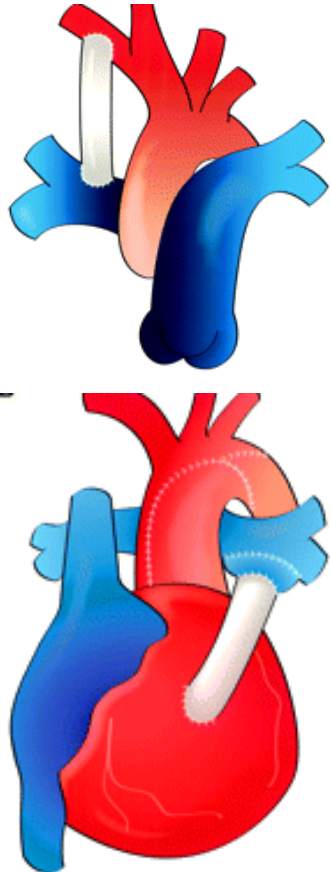


2012 The Children's Heart Clinic

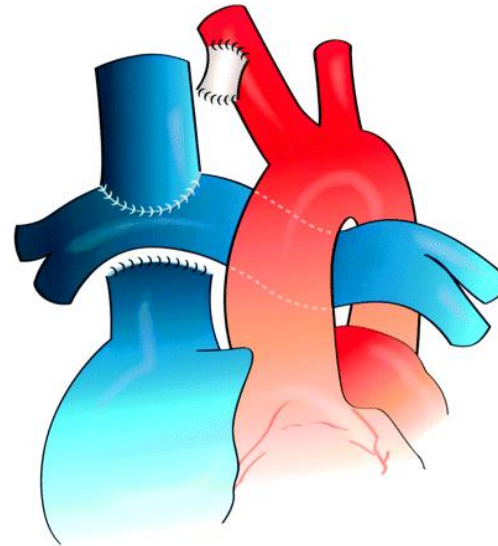
*Children's of Minnesota*

# Fontan Palliation

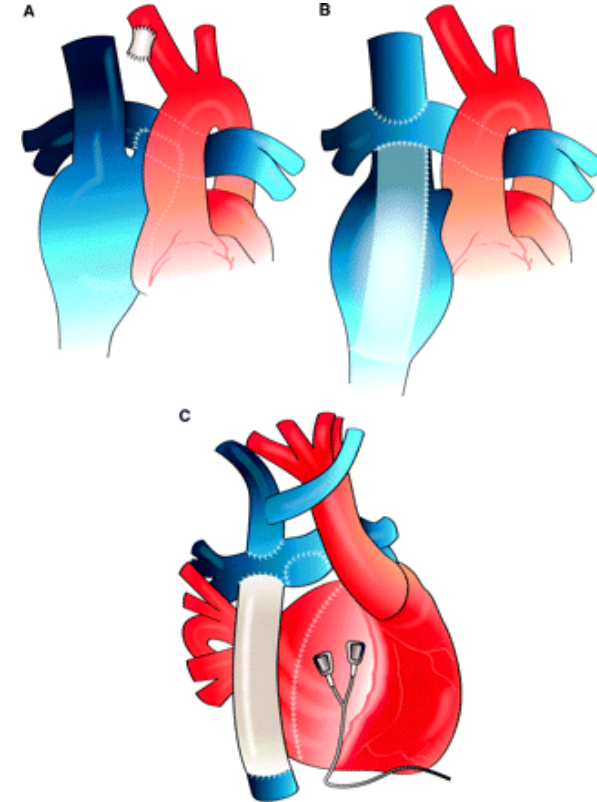
## Stage I Norwood or BT



## Stage II Glenn

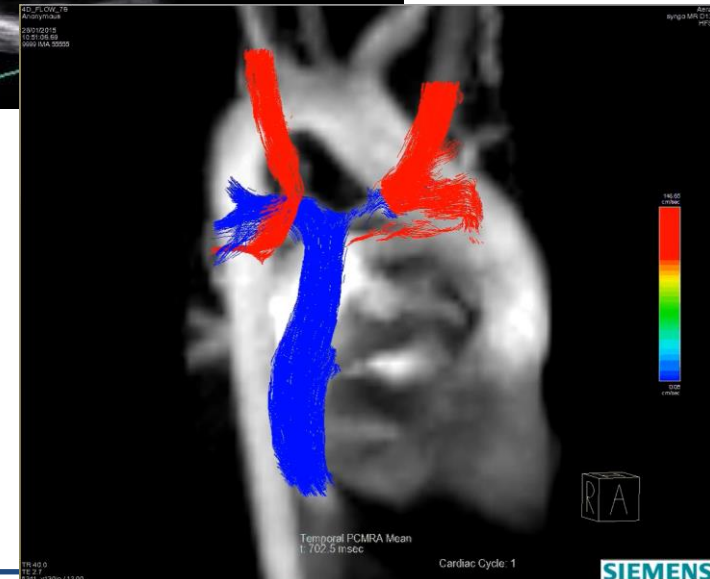
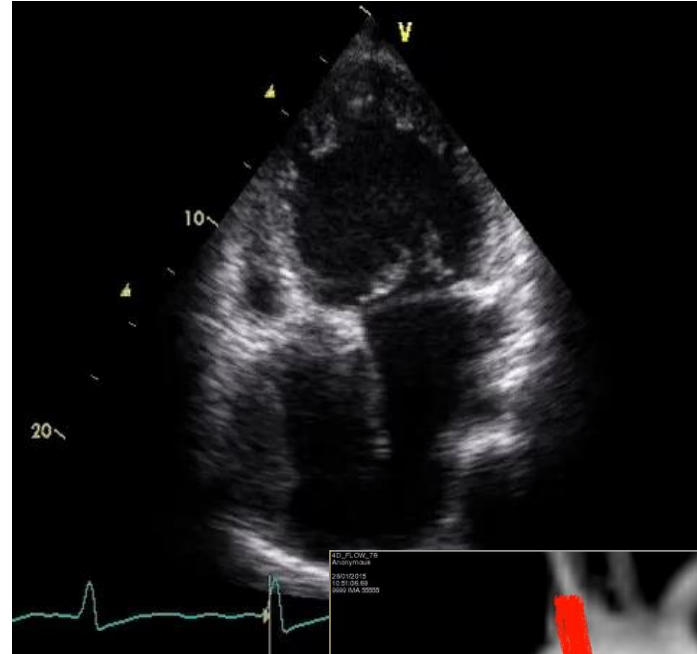


## Stage III Fontan



# Fontan Physiology

- Reliant on low systemic filling pressure/thoracic pressure to promote flow through Fontan pathway
- Develop progressive cardiac dysfunction
- Cyanosis
- Frequent arrhythmia
- Liver congestion and dysfunction



# Fontan Pregnancy

- Difficult to increase cardiac output (single RV > LV)
- Arrhythmia risk
- Increased thromboembolism risk
- IVC obstruction/increased abdominal/chest pressure

Risk Category	Risk Description	Maternal Risk Factors
III	Significantly increased maternal mortality or severe morbidity risk. Expert counseling required. In the event of pregnancy, intensive cardiovascular and obstetric monitoring needed.	<ul style="list-style-type: none"><li>• Mechanical valve</li><li>• Systemic RV</li><li>• Fontan Circulation</li><li>• Cyanotic heart disease (unrepaired)</li><li>• Other complex CHD</li><li>• Marfan Syndrome with aortic dilation 40-45 mm</li><li>• Bicuspid aortic valve with aortic dilation 45-50 mm</li></ul>

Risk Category	Risk Description	Maternal Risk Factors
IV	Extremely high maternal mortality or severe morbidity risk. Pregnancy is contraindicated. In the event of pregnancy, termination should be discussed/advised. If pregnancy continues, follow level III recommendations.	<ul style="list-style-type: none"><li>• Pulmonary Arterial Hypertension (of any cause)</li><li>• Severe systemic ventricular dysfunction (EF &lt; 30%, NYHA 3-4)</li><li>• Previous peripartum cardiomyopathy with ANY residual LV impairment</li><li>• Severe mitral stenosis</li><li>• Severe SYMPTOMATIC aortic stenosis</li><li>• Marfan Syndrome with aortic dilation &gt;45 mm</li><li>• Bicuspid aortic valve with aortic dilation &gt;50 mm</li><li>• Native severe coarctation</li><li>• Fontan with complications!</li></ul>

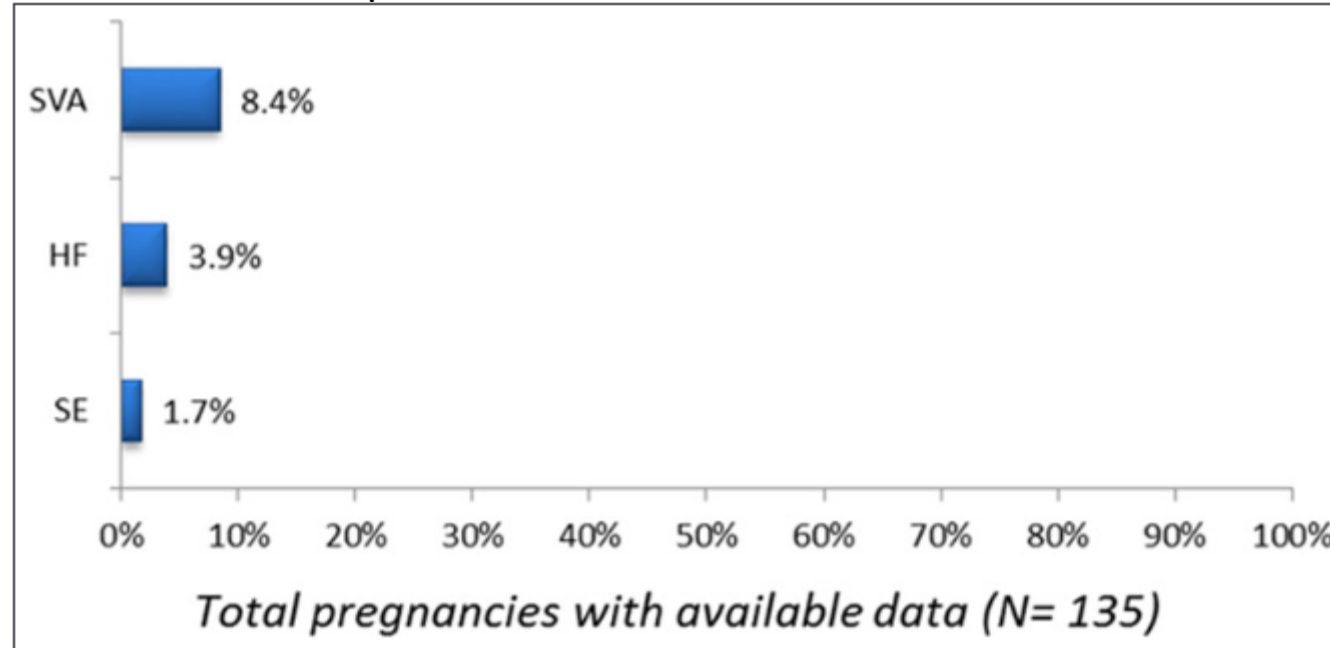
- Baseline evaluation including cardiopulmonary exercise testing and liver assessment
  - Use of Lovenox/LMWH?
    - Should be used in those with thromboembolic event or atrial arrhythmias; possible benefit in others
    - Continue aspirin in those already on this
  - IVC obstruction late in pregnancy may cause decompensation
-

- Avoid general anesthesia
    - PPV results in reduced cardiac output
  - Fluid balance
    - Requires preload to maintain Fontan/Glenn pressure; too much can exacerbate CHF
  - Consider c-section if reduced function; assisted second-stage
  - Telemetry due to risk of arrhythmia
  - Monitor in ICU for 24-48 hours
-



# Fontan Pregnancy Outcomes

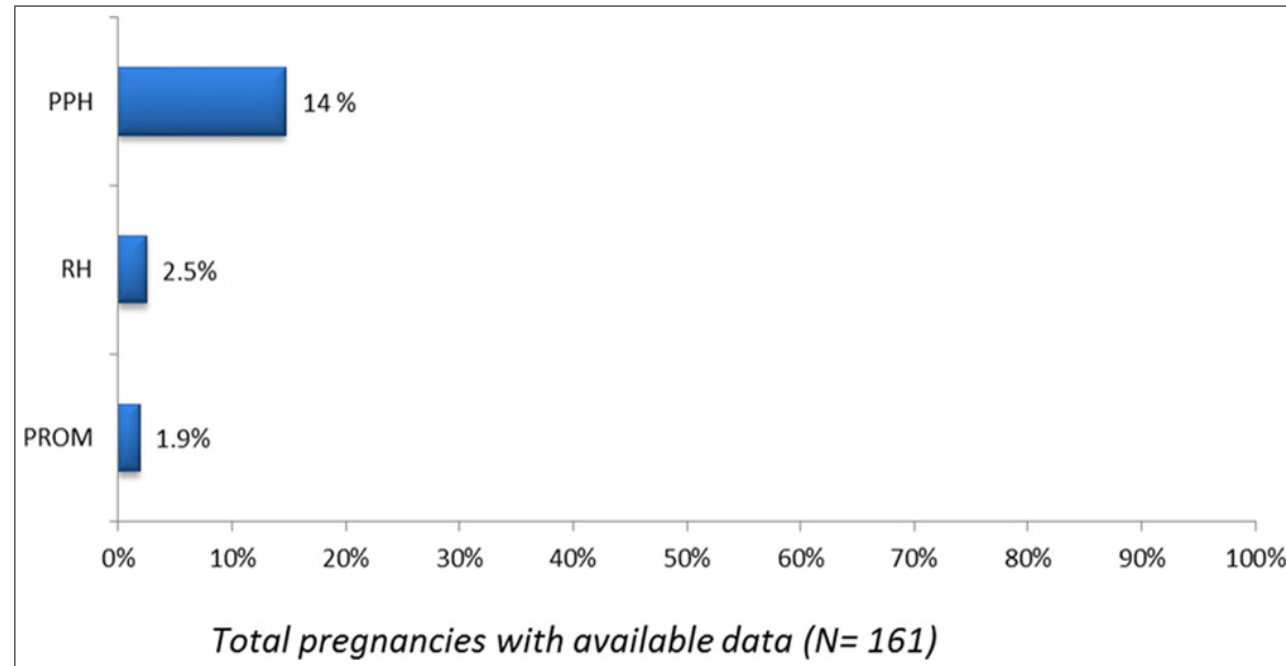
## Maternal CV Complications



**Figure 2. Cardiovascular complications.** HF indicates heart failure; SE, systemic embolism; and SVA, supraventricular arrhythmia.

# Fontan Pregnancy Outcomes

## Obstetric Complications



137 pregnancy losses amount 255 pregnancies  
(54%)

115 spontaneous miscarriages

19 elective terminations

1 ectopic pregnancy

115 Live births

68 premature deliveries (59%)

6 neonatal deaths

# Return to AE- Pregnancy Course

Seen at 12 weeks by ACHD/MFM

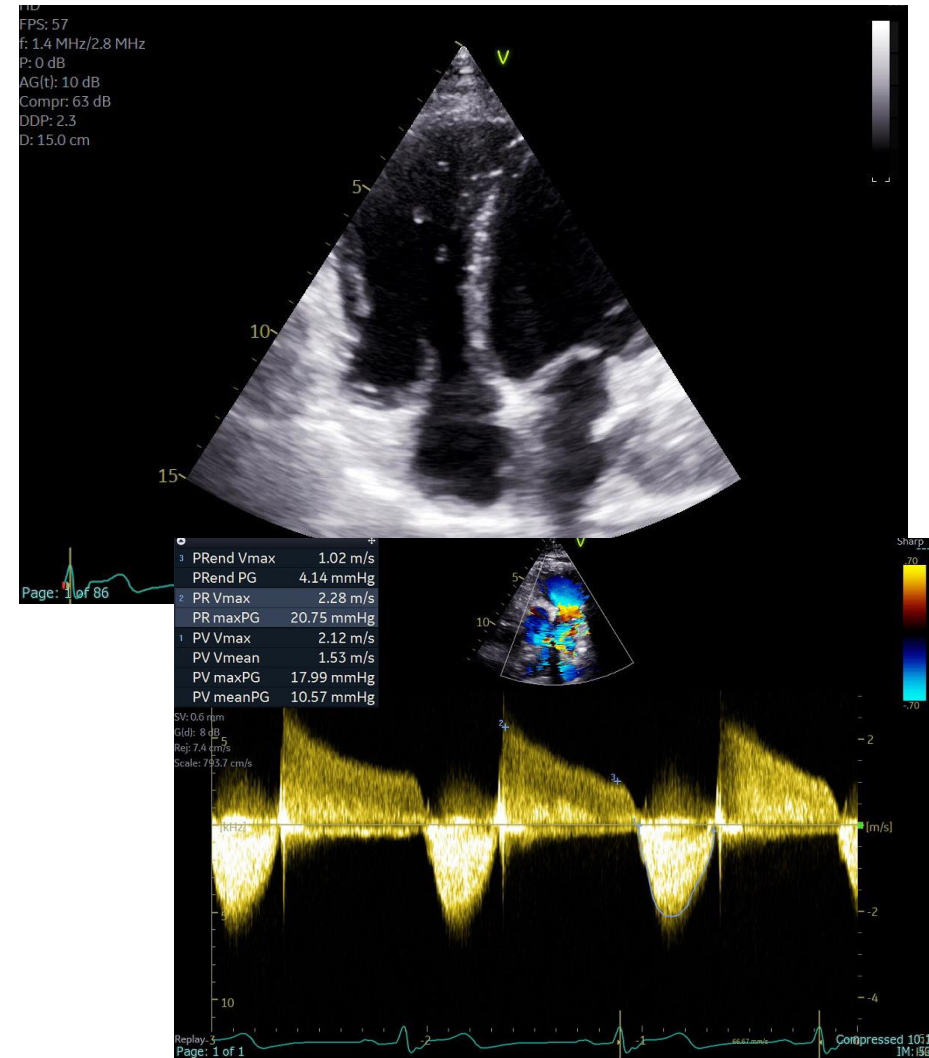
Developed increased palpitations  
into the mid portion of her  
pregnancy- monitor with isolated  
PVCs

Third Trimester:

BP 146/94, increased lower extremity  
edema and JVD

UA positive for protein

Admitted for suspected pre-eclampsia  
and decompensated right heart failure



# AE- Delivery

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Diagnosis of pre-eclampsia was confirmed

BP control and diuresis started

After multidisciplinary discussion, planned induced delivery @ 31.2 weeks gestation

Hemodynamics improved

Infant required short course in the NICU, but doing well!

Mother being evaluated for valve-in-valve PVR



# Take Home Points

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- Pregnancy is a high-flow, low resistance circulation which impacts the cardiac system in multiple ways
  - Preconception counseling is a critical part of the management of a woman with CHD
    - Nothing worse than an unplanned pregnancy!
  - Management of pregnancy in the congenital heart patient is complex and dependent on a number of factors including original diagnosis, surgical procedures, presence of residual disease, or other complicating factors
    - Requires a team-based effort between MFM, ACHD, anesthesia, surgery, etc. for optimal outcomes
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# Thank you!

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## Question #1

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Which of these conditions is considered mWHO IV and pregnancy is contraindicated?

- A. Tetralogy of Fallot with free pulmonary regurgitation
  - B. Uncorrected VSD with evidence of bidirectional shunting and SpO<sub>2</sub> of 88%
  - C. Hypoplastic left heart syndrome s/p Extracardiac Fontan
  - D. Bicuspid aortic valve with aortic stenosis, peak velocity 3.1 m/s, moderate stenosis
-

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- D. Bicuspid aortic valve with aortic stenosis, peak velocity 3.1 m/s, moderate stenosis, aortic dimension 3.7 cm.

Of the presented scenarios, the second patient has Eisenmenger syndrome which is a contraindication to pregnancy. Fontan palliated patients are considered mWHO III. The patient with repaired TOF with severe PR and the BAV patient would be considered mWHO II-III.

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## Question #2

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A patient with Tricuspid atresia s/p lateral tunnel Fontan palliation presents to the ER at 33 weeks gestation. She has respiratory distress and is intubated in the ER. On arrival to the ICU, pH on ABG is 7.1 with PCO<sub>2</sub> of 43 and base deficit of -10 with elevated lactate. Vitals are HR 110, BP 90/55, SpO<sub>2</sub> 95%. Ventilator settings are tidal volume 400 ml, RR 15, FiO<sub>2</sub> 60%, PEEP 12. What is the next best step in management?

- A. Immediate reduction of PEEP followed by immediate prep for delivery
- B. Increase in FiO<sub>2</sub> to maintain SpO<sub>2</sub> >97%.
- C. Prepare for Ecmo cannulation
- D. Start systemic heparinization

The patient shows evidence of cardiogenic shock with metabolic acidosis. This is likely being caused by excessively high intrathoracic pressure in a patient with Fontan palliation. High intrathoracic pressure (such as in PPV) can result in poor Fontan flow, decreased filling to the systemic ventricle, and worsening shock. First step should be reduction of PEEP to improve forward flow. Given her late-preterm pregnancy and presentation with heart failure, preparation for emergent delivery would be reasonable.

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## Question #3

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A patient with Tetralogy of Fallot repaired at 6 months of life with a valve-sparing technique presents for preconceptual counseling. Review of cardiac studies shows that she has normal biventricular function with normal chamber sizes. There is mild pulmonary valve stenosis and moderate pulmonary valve regurgitation. She reports no symptoms and has no history of arrhythmia. What is the best next step?

- A. Referral for surgical pulmonary valve replacement
- B. Cardiopulmonary exercise testing
- C. Recommend against ever becoming pregnant
- D. Proceed with pregnancy without additional testing

CPET would provide additional risk stratification information in this moderate risk patient. She would likely be considered either mWHO II or II-III. CPET testing has been shown to help predict pregnancy risk in such patients, primarily looking at total peak VO<sub>2</sub> and HR responsiveness in mothers. Some practices may consider recommending proceeding with pregnancy without additional testing.

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## Question #4

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A patient with a history of bicuspid aortic valve with moderate-severe aortic stenosis (peak velocity 3.7 m/s, mean gradient 37 mmHg) with preserved LV function presents at 24 weeks gestation to OB triage with new onset dyspnea with exertion. Urgent echocardiography shows severe AS with peak velocity 4.2 m/s, mean gradient of 45 mmHg, moderate aortic regurgitation, and an LVEF of 45%. What is the next best step in management?

- A. Immediate induction of labor and delivery
- B. ECMO cannulation
- C. Surgical aortic valve replacement (AVR)
- D. Transcatheter aortic valve replacement (TAVR)

TAVR would be the most reasonable option in this woman with new onset LV dysfunction in the setting of severe aortic valve stenosis. TAVR has been used safely in pregnancy to rescue patients with aortic valve disease with low maternal and fetal complication rates. Surgical AVR would likely be very high risk for the fetus and likely to end in fetal loss.

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