

PREGNANCY & CONGENITAL HEART DISEASE

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**If you have heart disease
Don't fall in love**

**If you fall in love
Don't make love**

**If you make love
Don't get pregnant**

**If you get pregnant
Don't have a child**





**“ I’m never having kids. I hear they take
9 months to download “**

HEART DISEASE & PREGNANCY



What do we know ?

PREGNANCY AND HEART DISEASE

- **2 % pregnancies involve maternal CV disease**
- **Most women with CV disease can have a pregnancy with proper care, but there is an increased risk to mother and fetus.**
- **Pre - pregnancy evaluation mandatory**

PREGNANCY & ADULT CHD



Getting there first
Pre - pregnancy counseling

PRE – CONCEPTION COUNSELING

Issues to be discussed

- **Effects of pregnancy on mother's cardiac situation : risks to mother**
- **Whether risks will change with time or Rx**
- **Long- term outlook for the mother**
- **Risks to baby**
- **Alternative options : adoption, surrogacy**

PRE – CONCEPTION COUNSELING

- Visit to regional center where expertise in ACHD **and** high risk pregnancy Rx
- History, physical exam, ECG, CXR
- Echocardiogram : assess ventricular function, valves, vessels, PAp and shunts
- Alternative imaging if arterial assessment suspect. If PAp in doubt : cardiac cath
- Exercise testing : functional status

GENETIC COUNSELING

Risk of congenital abnormality

- **Family history**
- **Fetal risks average 4- 6%. Up to 50% in single gene defects (Marfan syndrome, 22q11 deletion)**
- **Consider genetic analysis in some situations (e.g. tetralogy of Fallot).**
- **Offer appointment with clinical geneticist**
- **Discussion of partner's genetic history**

PRE – CONCEPTION COUNSELING

Ante partum care

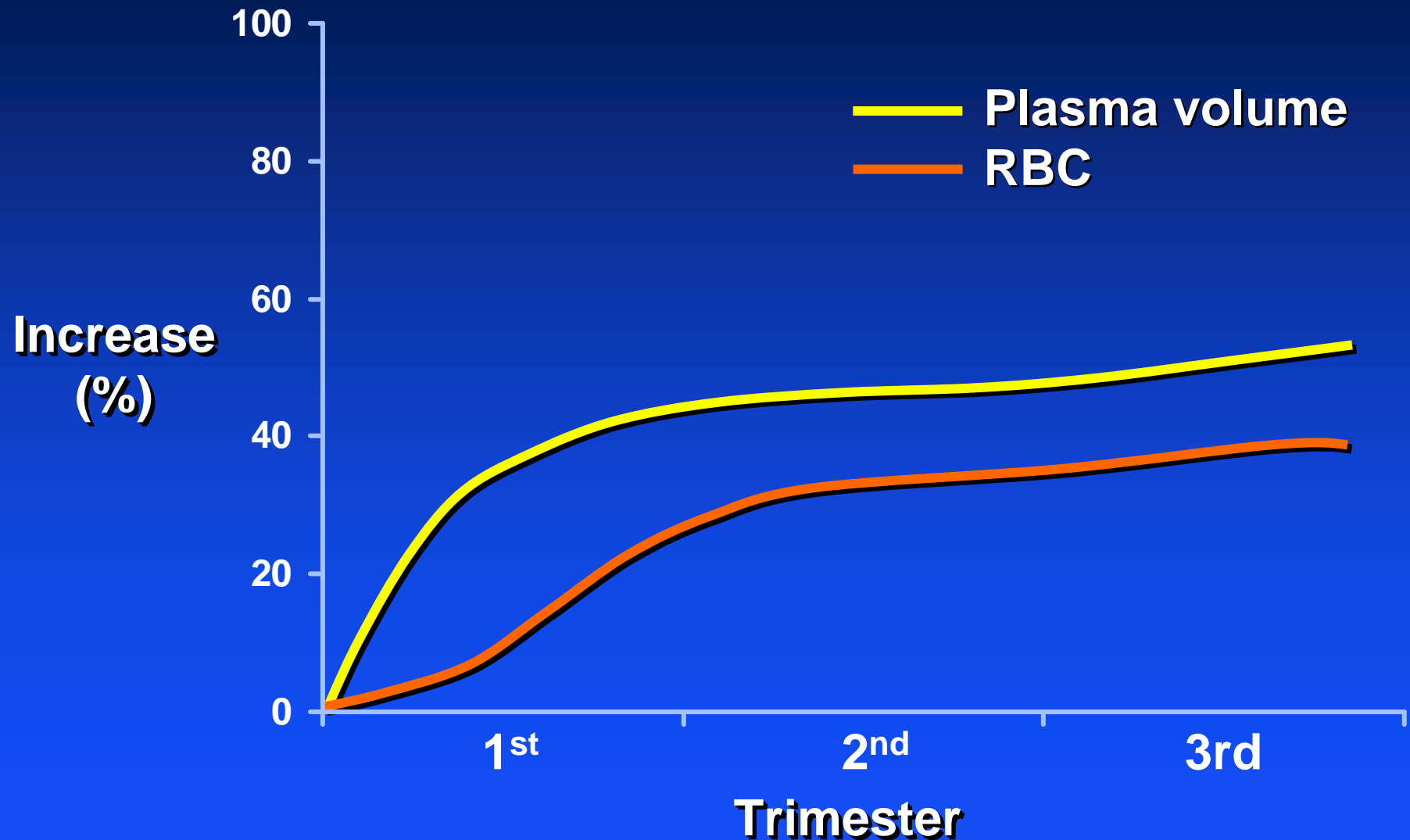
- Plan CV visits in event of pregnancy :
“ Call me immediately / x weeks.. etc ”
- Outline principles of CV follow up and plan
- Introduction to high risk obstetrician and discuss obstetric visits, location, timing and mode of delivery

AREAS OF CONCERN IN CONGENITAL HEART DISEASE AND PREGNANCY

- Genetic counseling
- Avoidance of drugs harmful to baby
- Antepartum care – c-v and obstetric
- Fetal growth and development
- Labor – heart and obstetric considerations
- Antibiotic prophylaxis
- Postpartum care
- Contraception

PREGNANCY AND THE HEART

Volume Changes



PREGNANCY AND THE HEART

Hemodynamics During Pregnancy

Peripheral resistance ↓
↑ uterine blood flow

Blood volume ↑ 40- 45%

Heart rate ↑ 10- 20%

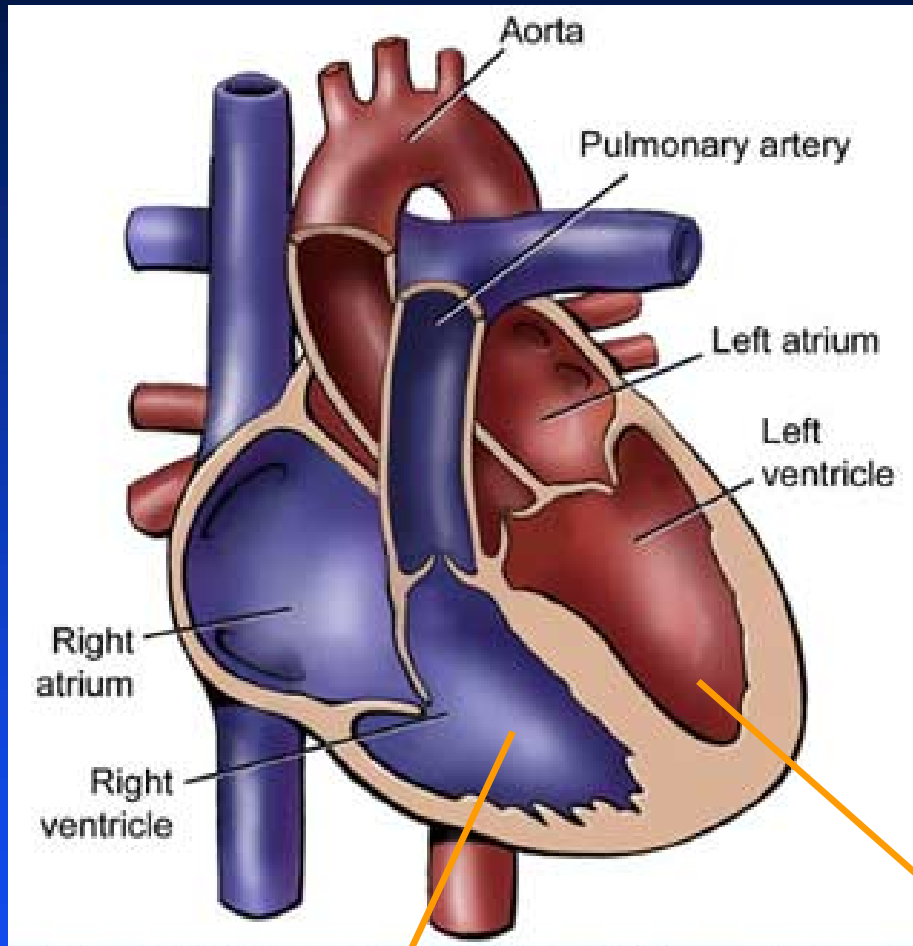
**Cardiac
output ↑ 30%**

Blood pressure → or ↓

Pulmonary vascular resistance ↓

Venous pressure in legs ↑

NORMAL HEART



25 mmHg

120 mmHg

PREGNANCY AND HEART DISEASE

Before pregnancy

- High-risk pregnancy unit, clinical exam, ECG, CXR, Echo, exercise test
Discuss maternal and fetal risks

During pregnancy

- Team approach, fetal Echo

Labor and delivery

- Obstetrician, cardiologist, anesthesiologist
Tailor management to specific needs

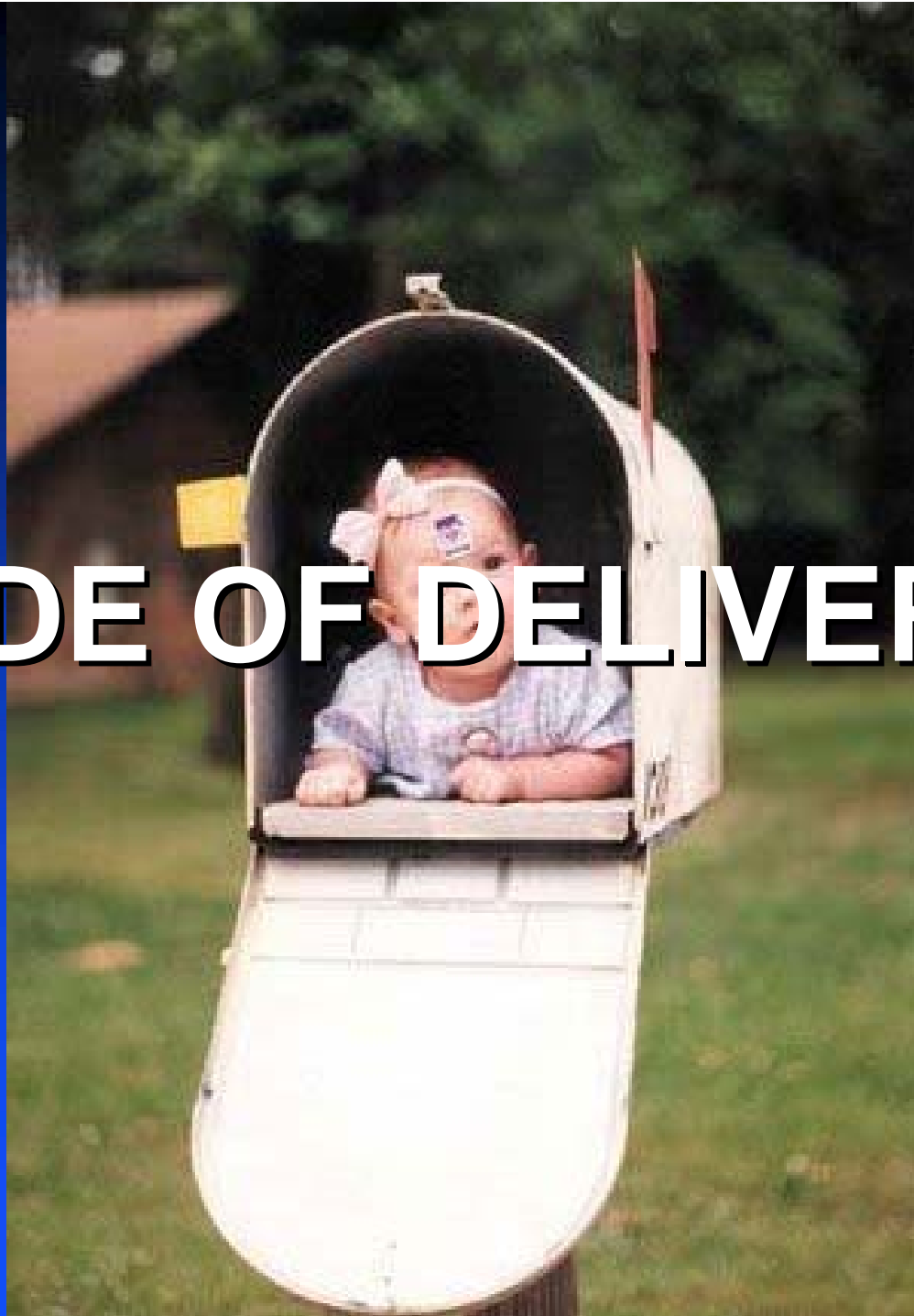
CHANGES DURING LABOR & DELIVERY

Uterine contractions – 300 - 500 cc blood
into circulation

CO ↑ 25% 1st stage
50% 2nd stage
80% delivery (vaginal)

**Loss of 300 - 400 cc in normal
vaginal delivery**

MODE OF DELIVERY ?



HEMODYNAMICS DURING LABOR AND DELIVERY

Volume Loss

- **Vaginal delivery 300 - 400 cc**
- **Cesarean section 500 - 800 cc**
 - **↑ venous return**

**Hemodynamics return to baseline
at \approx 4 - 6 weeks**

PREGNANCY : MEDICATIONS



PREGNANCY & HEART DISEASE

Drugs

Relatively safe

Digoxin

Quinidine

Procainamide

Ca channel blockers

β blockers

Lasix

Heparin

Not safe

Phenytoin

ACE inhibitors

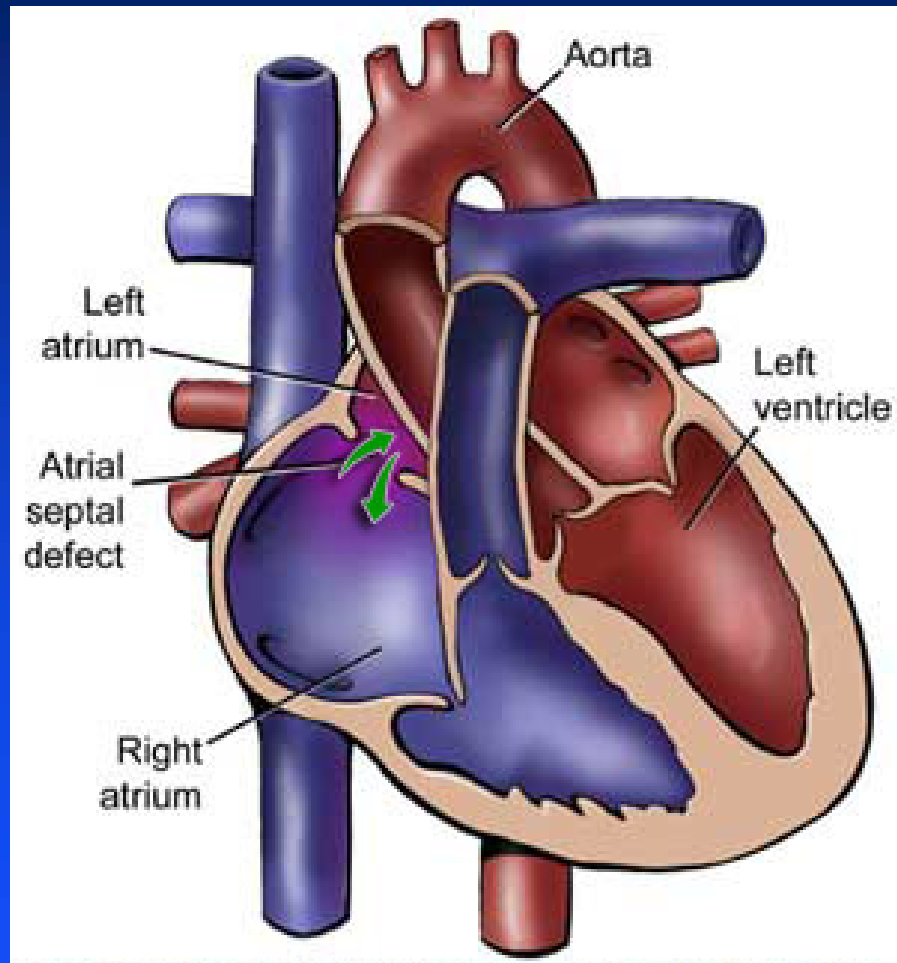
Coumadin

Statins

PREGNANCY & HEART DISEASE

Simple lesions

ATRIAL SEPTAL DEFECT

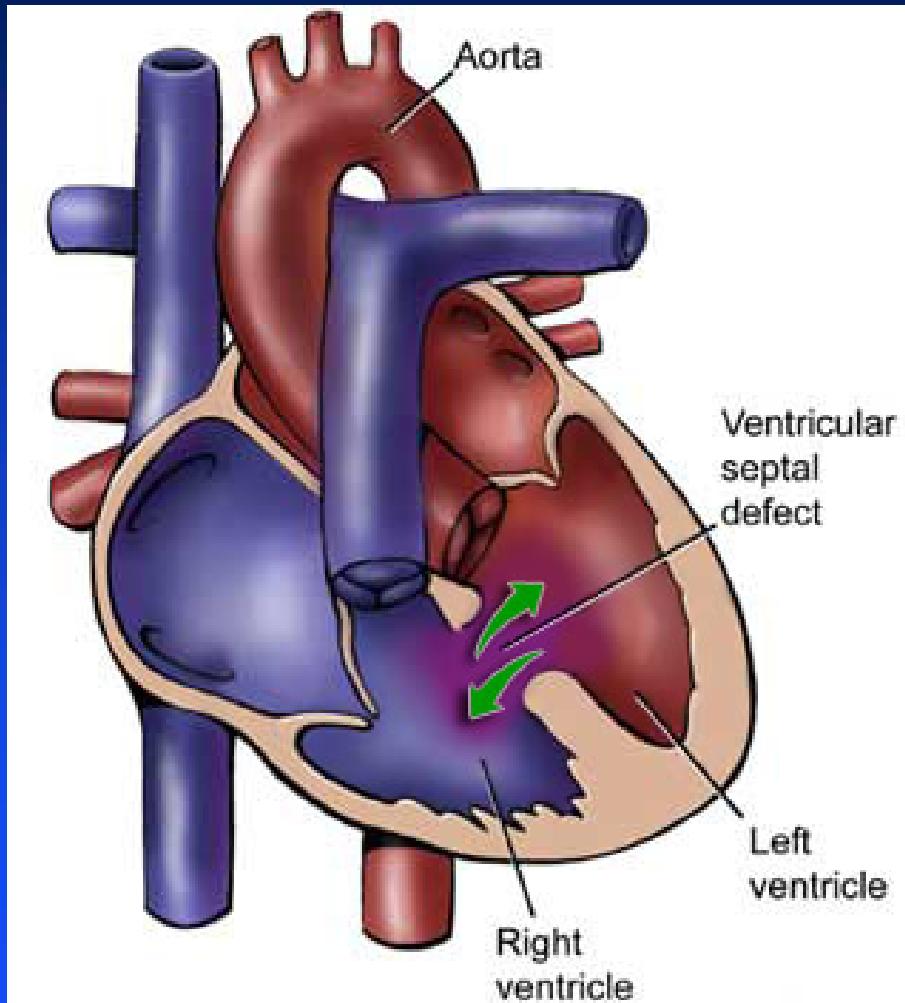


- Left to right shunt
- RV enlarged
- May be tricuspid regurg
- Be sure no pulmonary hypertension
- Concern about blood clots from legs passing through hole

ASD AND PREGNANCY

- Usually tolerate pregnancy well
- ↑ risk for atrial arrhythmias & paradoxical embolus
- Baby ASA after 1st trimester
- Early postpartum ambulation
- Elective closure before pregnancy is preferable

VENTRICULAR SEPTAL DEFECT



- Left to right shunt
- Be sure ventricles have normal size & function
- Be sure no pulmonary hypertension

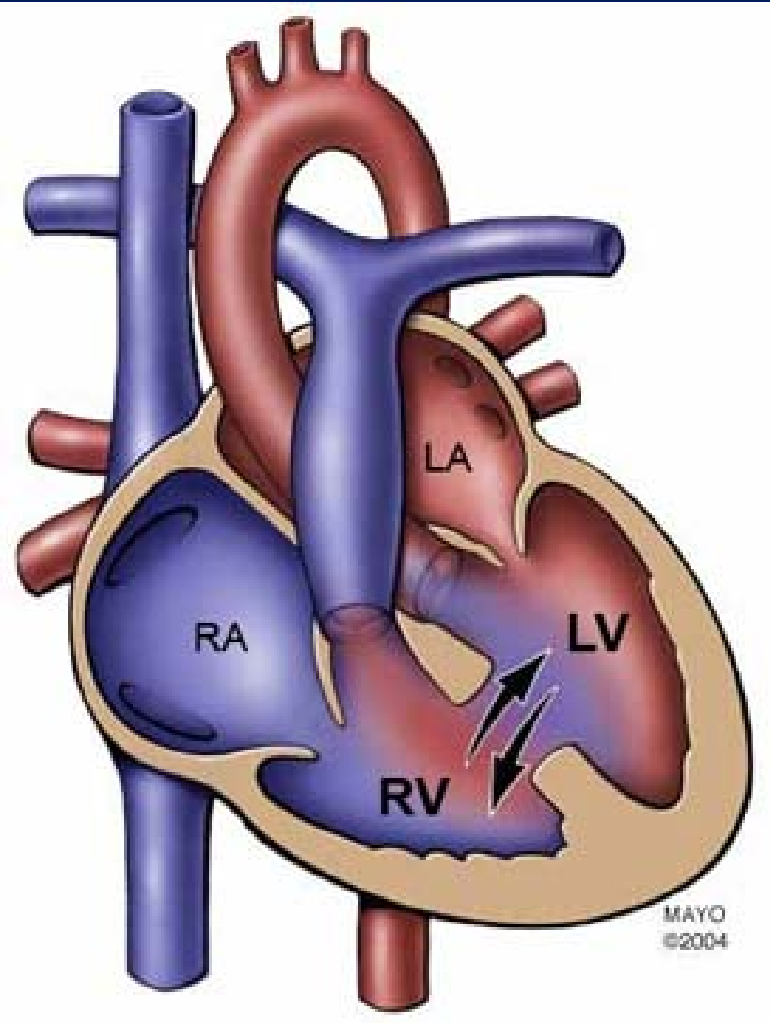
CARDIAC DISEASE AND PREGNANCY

High Risk

Avoidance or interruption of pregnancy

- **Pulmonary hypertension**
- **Cyanotic heart disease**
- **Reduced ventricular function $EF \leq 40\%$**
- **Symptomatic obstructive lesions AS, PS, Coarctation**
- **Marfan syndrome with big aortic root**

EISENMENGER SYNDROME



Large shunt (usually VSD) produces pulmonary hypertension and irreversible pulmonary vascular disease.

Shunt reverses $R \rightarrow L$ causing cyanosis

EISENMENGER SYNDROME

44 cases with 70 pregnancies

- **Maternal death in 30% pregnancies**
 - 52% pt died in connection with a pregnancy**
- **Post - partum refractory hypoxia**
- **Blood clot in lung (Pulmonary infarction)**
- **C section high risk – ? bias**

PULMONARY HTN AND PREGNANCY

Newer treatments ?

- **Intravenous prostacyclin (epoprostenol)**
- **Sildenafil**
- **Inhaled nitric oxide**
- **Case reports : successful outcomes
with both vaginal delivery and C. section**

CONGENITAL HEART DISEASE : PH

Progress 1997- 2007 ? Published data

- While maternal mortality lower in last decade, mortality rates remain prohibitively high
- Pregnancy should still be discouraged

CARDIAC DISEASE AND PREGNANCY

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CYANOTIC CHD: PREGNANCY

Maternal Risk

- Hemoglobin high due to low O₂
 - Anemia exaggerates hypoxia
 - R to L shunt worsens
 - ↑ platelets stickier
↓ fibrinolysis
 - DVT & paradoxical embolus
- > Risk of thrombus

CYANOTIC CHD: PREGNANCY Fetal Risk

- **↑ spontaneous abortion**
- **Poor fetal growth**
- **Risk of inheriting CHD**

FETAL OUTCOME AND MATERNAL CYANOSIS

Arterial O₂ sat (%)	≤ 85	85 - 89	≥ 90
Pregnancies (no.)	17	22	13
Live births (no.)	2	10	12
Live born (%)	12	45	92

Presbitero P, Circ 1994



CARDIAC DISEASE AND PREGNANCY

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ACHD & PREGNANCY

Heart muscle problem

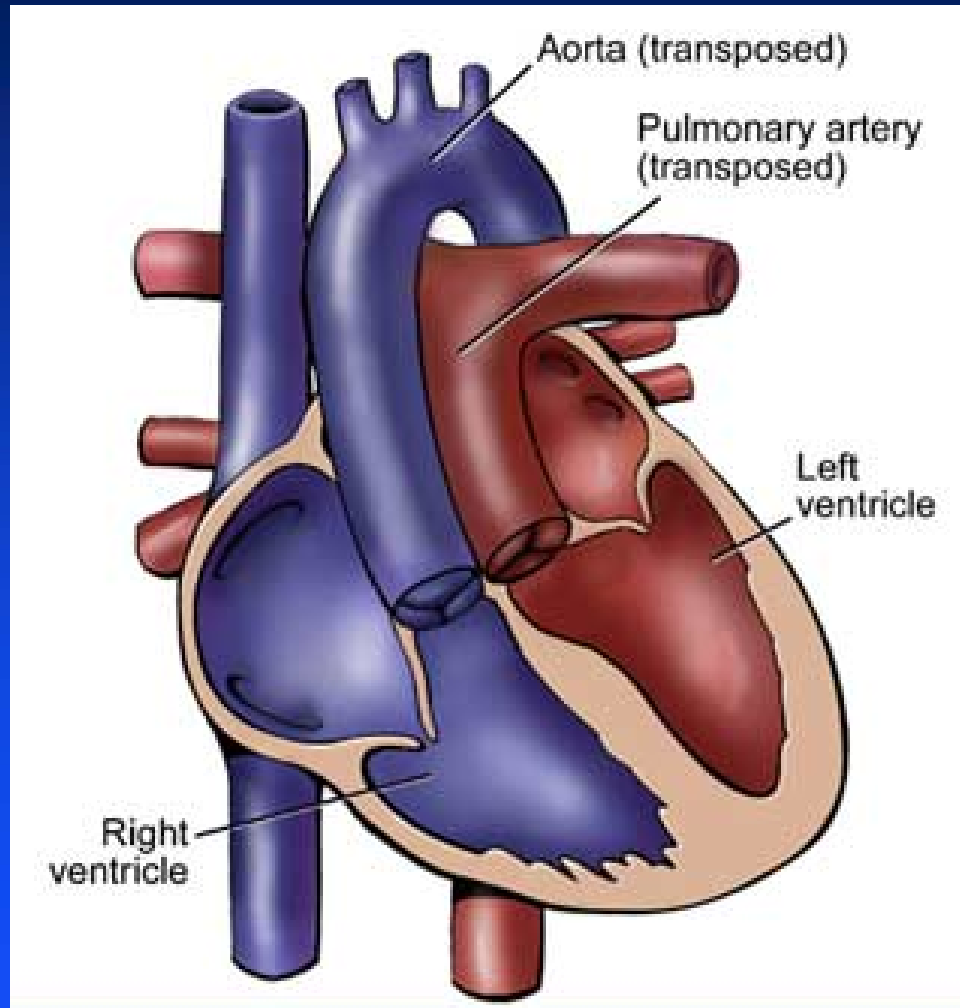
- Plasma volume ↑ 50 %
- If ejection fraction less than 40% -
NO PREGNANCY
- (Normal 55 - 70%)

COMPLEX CHD : LONG-TERM OUTCOME

Ventricular function

- Underlying cardiac anatomy abnormal
- Borderline baseline heart function
- Mild A-V valve regurgitation
- Pregnancy changes might be more profound
- Valve ring dilates, minor changes in ventricular shape : A-V valve coaptation
- Marked when systemic ventricle is RV

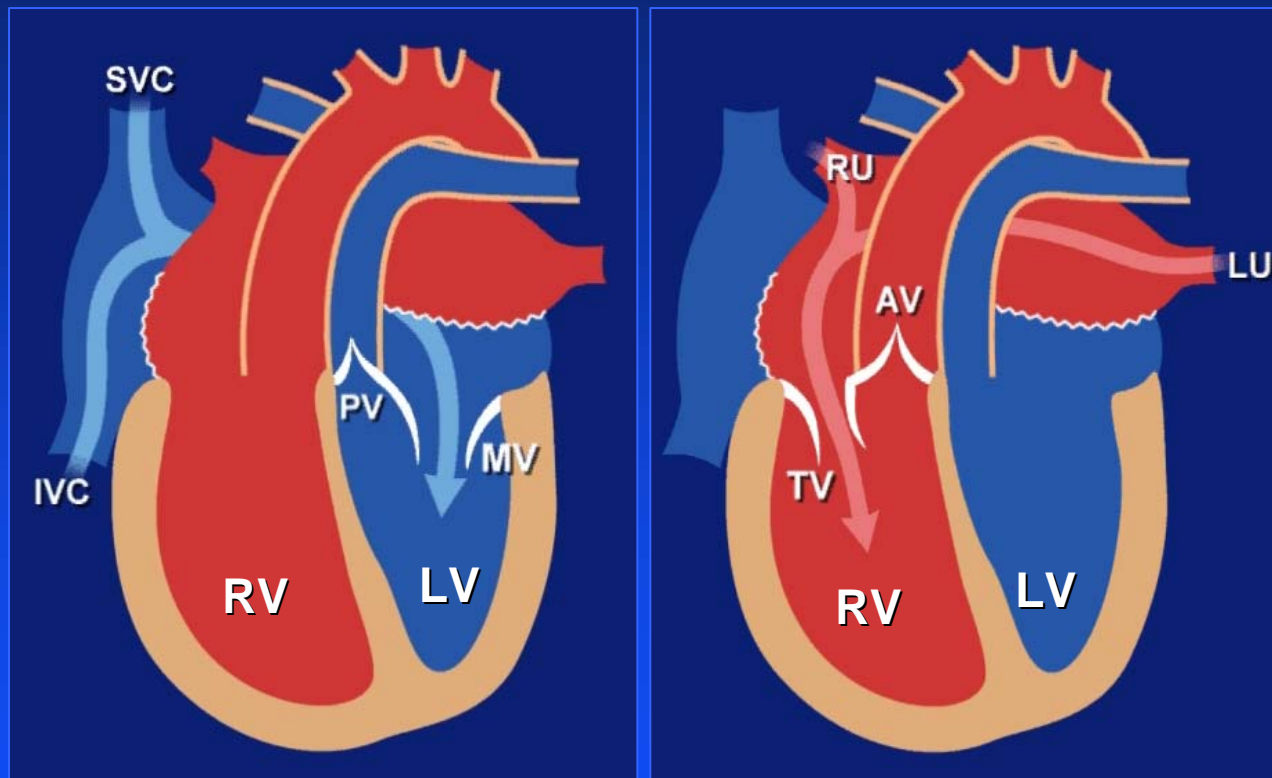
TRANSPOSITION OF THE GREAT ARTERIES



Aorta arises from RV
PA arises from LV

PREGNANCY & d- TGA

Mustard & Senning operations



Tricuspid regurg
RV vulnerable to
failure

RV is the systemic ventricle

SYSTEMIC RIGHT VENTRICLE

Mustard operation

- 16 women with 28 pregnancies
- **RV dilatation (n=18)**

absent = 4

mild/moderate = 12

severe = 2

**Progressed
in 5 (31%) :
no recovery**

SYSTEMIC RIGHT VENTRICLE

Mustard operation

- 16 women with 28 pregnancies

- **RV systolic dysfunction (n=21)**

absent = 16

mild/moderate = 4

severe = 1

**Progressed
in 4 (25%) :
no recovery in 3**

SYSTEMIC RIGHT VENTRICLE

Mustard operation

- 16 women with 28 pregnancies
- **Tricuspid regurgitation (n=20)**

absent = 8

mild = 9

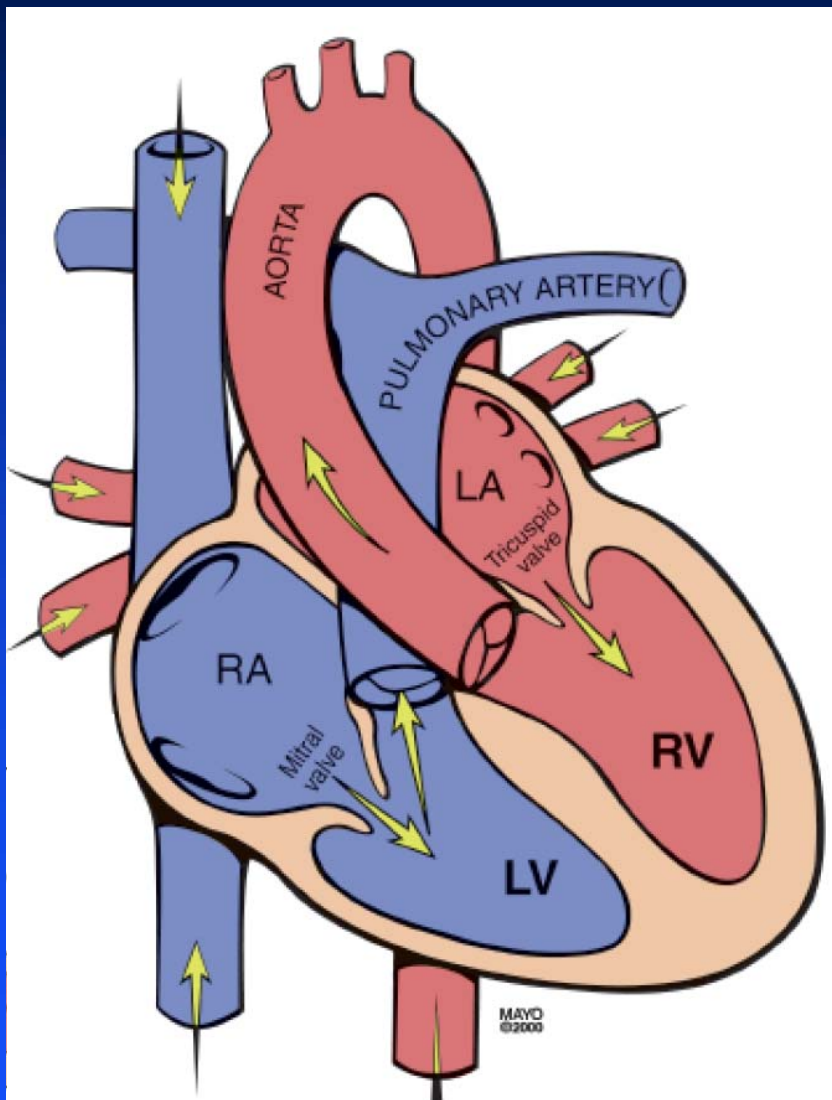
moderate = 3

**Progressed
in 8 (50%) :
no recovery in 3**

RECOMMENDATIONS : Systemic RV

- **Pre- conceptual counselling should include the possibility that pregnancy might cause a deterioration in cardiac status and impact long- term outcome**
- **Physicians should be alerted to the possibility of deterioration peri- and post-partum and should follow patients closely.**

CONGENITALLY CORRECTED TGA



- RV is systemic ventricle
- Systemic AV valve regurg common

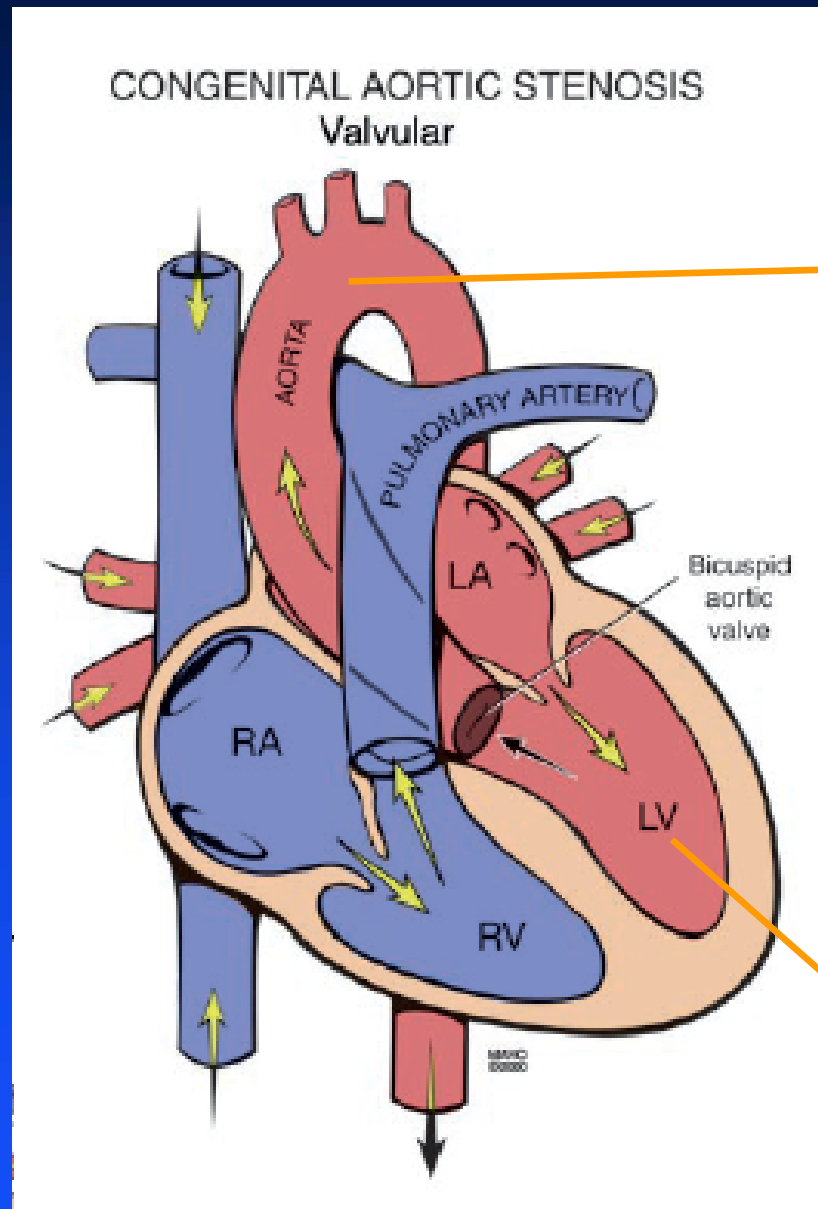
CARDIAC DISEASE AND PREGNANCY

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AORTIC STENOSIS



100 mmHg

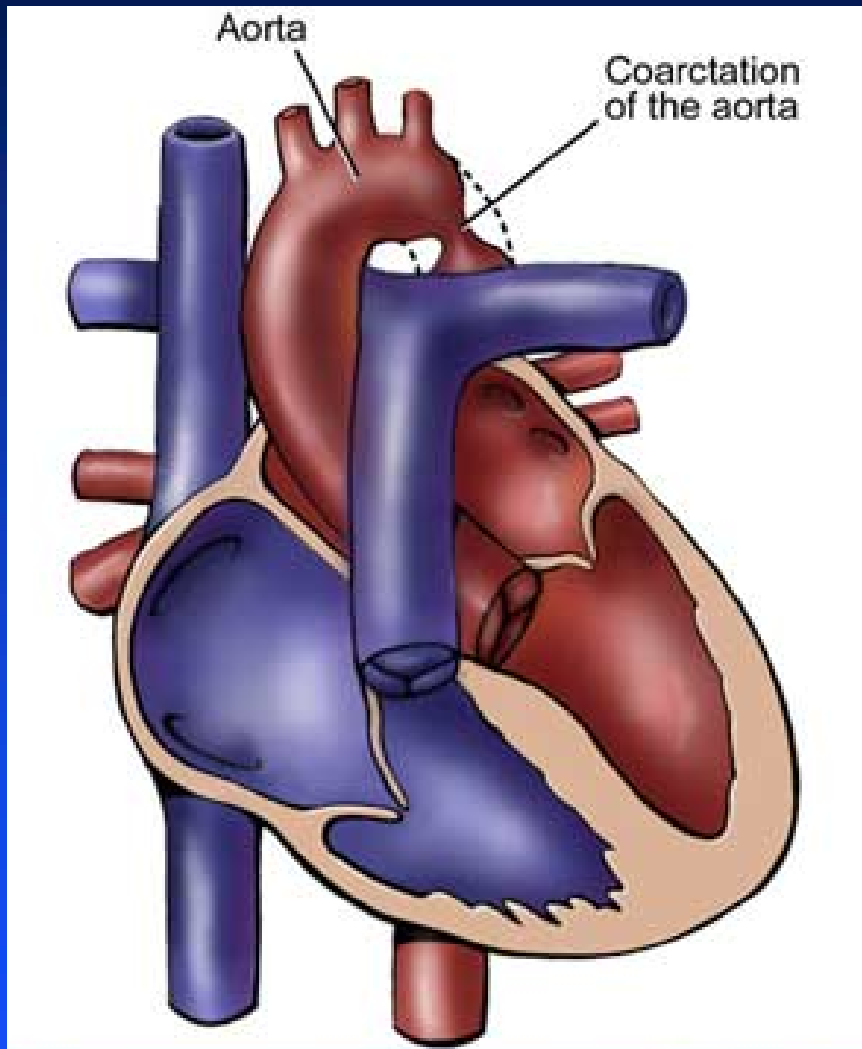
200 mmHg

ACHD & PREGNANCY

Obstructed valve problem

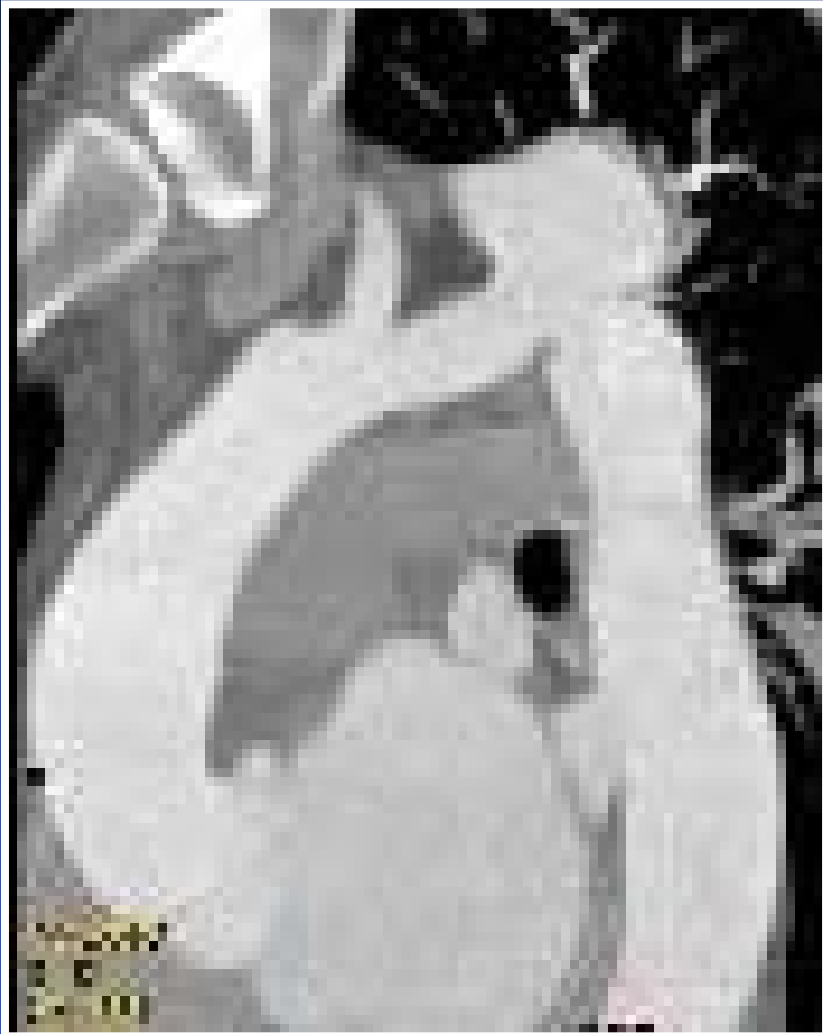
- If severe : **NO PREGNANCY**
- Have valve repaired or replaced first

COARCTATION OF THE AORTA



- **Narrowing of aorta usually beyond left Subclavian A.**
- **High BP above, low BP in legs**
- **Often associated with bicuspid aortic valve**

ACHD & PREGNANCY



**Coarctation repair
Aneurysm**

ACHD & PREGNANCY

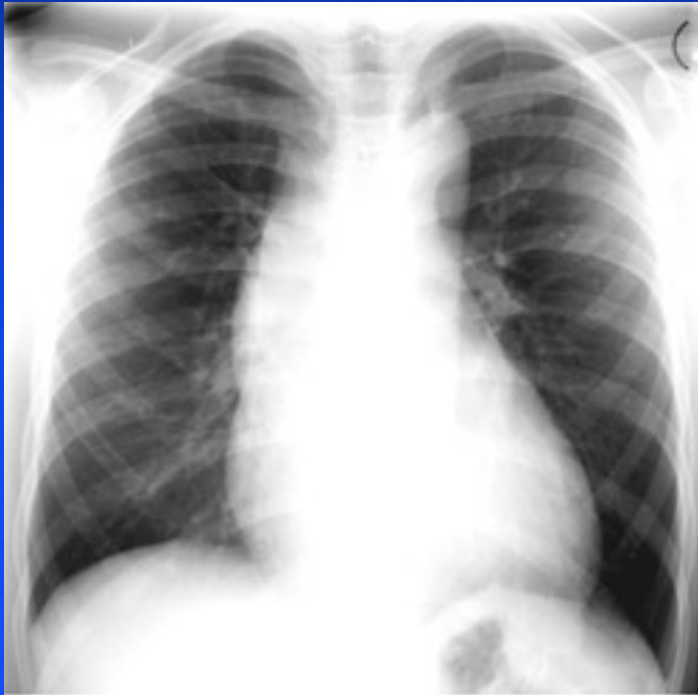
Connective tissue problem

- If aorta dilated more than 40mm wide -
NO PREGNANCY
- All tissues get softer during pregnancy
aorta may dissect, tear or rupture

MARFAN SYNDROME

Fetal risk

- Spontaneous abortion
- Inheritance 50%



Maternal risk

? Pre-existing CV disease
Hemodynamically
significant AR, MR
Aortic root dimension

MARFAN SYNDROME



If aortic root > 40 mm

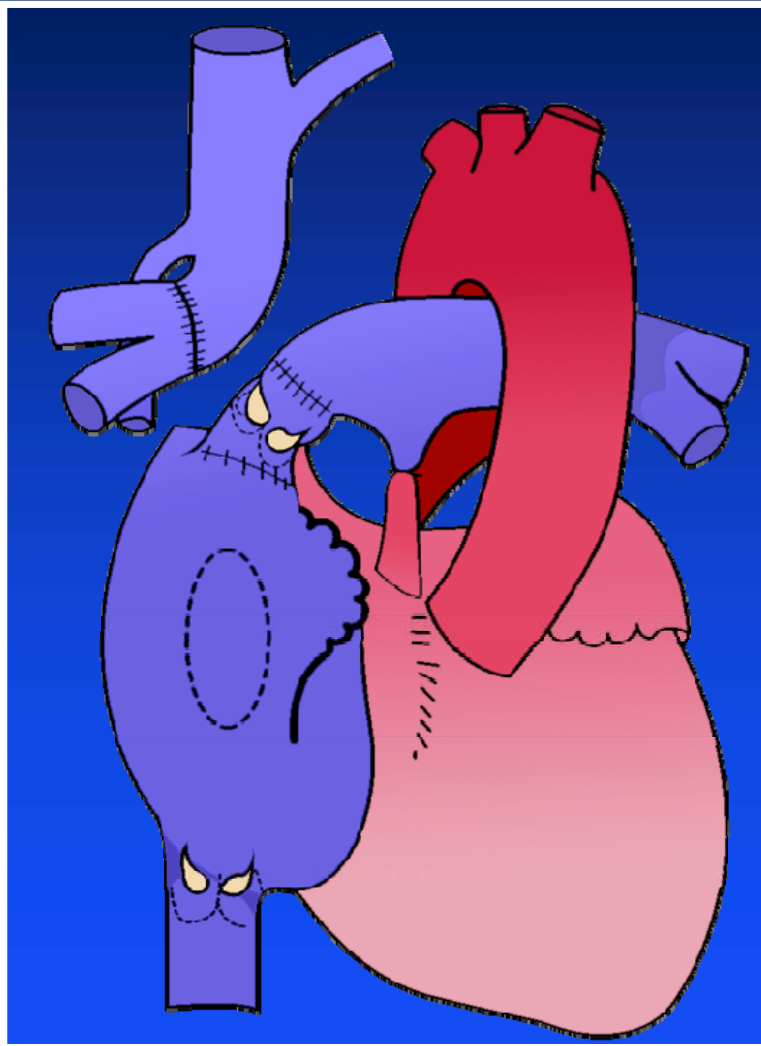
Advise against pregnancy

If aortic root < 40 mm

small risk of dissection,
rupture BUT unpredictable

Beta blockade appropriate

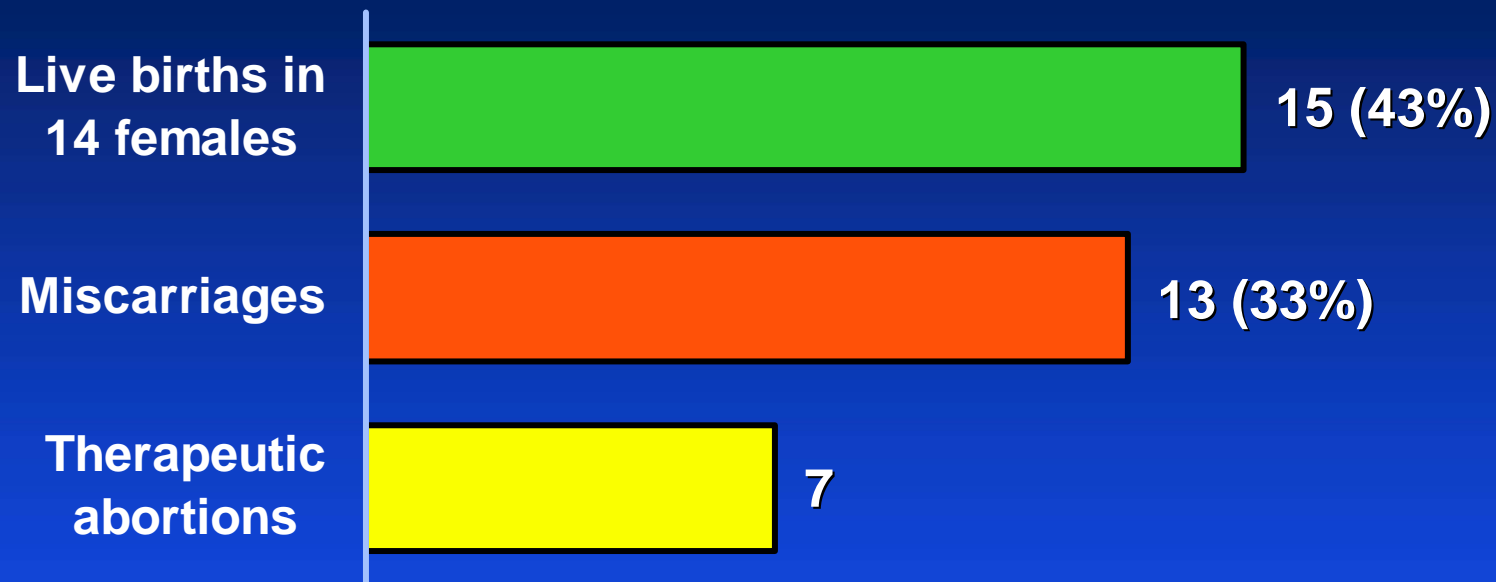
PREGNANCY AND FONTAN



**Pre - pregnancy assessment
mandatory**

- **Clinical evaluation**
History arrhythmias
Ex capacity
- **Echo**
EF, AV valve regurg
- **Drugs**
ACE inhibitors
Warfarin

FONTAN – PREGNANCY : 33 Pregnancies



- Mean 4 years between Fontan and pregnancy
- Maternal complications: 1 SVT, 1 CHF postpartum
- One infant has CHD (2^o ASD)

PREGNANCY AND FONTAN

Literature review: 39 completed pregnancies

- **All reported pregnancies: delivery premature in 15/39 (39%) : range 25-36 weeks (normal pre-term birth rate = 10-12%)**
- **Pre-term delivery important cause of infant morbidity and mortality**
- **Spontaneous miscarriage rate high: 34% (higher than usual 10-15%)**

COMPLEX CHD AND PREGNANCY

Conclusions

- Detailed pre- pregnancy evaluation mandatory
- Discussion of prognosis, maternal risks including potential for CV deterioration
- Development of multidisciplinary Rx plan
- Special vigilance in 3rd trimester and delivery plan with CV /anesthesia /OB team collaboration at time of delivery.

TISSUE HEART VALVE



MECHANICAL HEART VALVES



PREGNANCY & PROSTHETIC VALVES

Warfarin (coumadin) best blood thinner BUT crosses placenta and can affect baby 1st trim

Warfarin embryopathy



- Abnormal bones on X ray, optic atrophy, blindness, CNS dysfunction
- Risk approximately 6%
- Risk is dose - related

PREGNANCY & PROSTHETIC VALVES

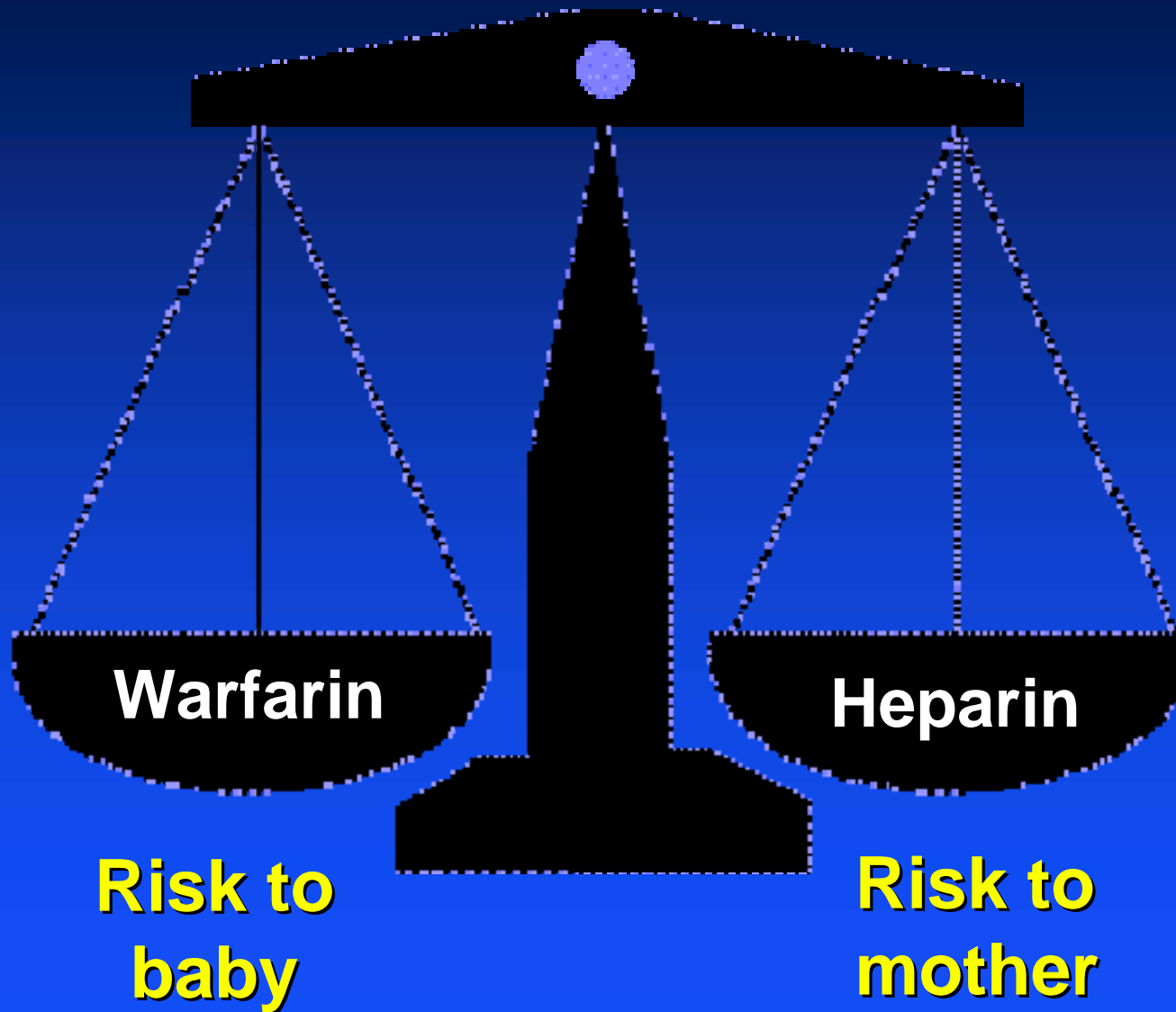
Heparin : i.v. or sub q
does not cross placenta
does not harm baby

BUT

Not a good blood thinner
in pregnancy



MECHANICAL VALVES & PREGNANCY



An intraoperative photograph showing a mitral valve prosthesis. The valve is a tilting-disc type, with a dark, circular disc visible in the center of the valve opening. The surrounding tissue is pinkish-red and appears moist. A blue surgical drape is visible at the bottom of the frame.

**Most complications occur with
tilting- disc mitral prostheses
and unfractionated heparin**

PREGNANCY & PROSTHETIC VALVES

40 articles : 976 ♀ 1234 pgy, 1966 - 97

- Warfarin throughout : embryopathy 6 % and lowest risk of valve thrombosis (3.9%)
- Heparin 6-12 wk eliminates this risk **BUT** increases risk of valve thrombosis (9 %)
- Fetal loss similar in those Rx with warfarin vs. those Rx heparin 1st trimester

PROSTHETIC VALVES AND PREGNANCY

43 women (58 pregnancies), Rx warfarin

31 mitral, 7 aortic, 5 mitral & aortic

INR 2.5 - 3.5, C section at 38 weeks

Fetal complications

Warfarin ≤ 5 mg

5 / 33 (15%)

No embryopathy

Warfarin > 5 mg

22 / 25 (88%)

9% embryopathy

ACHD & PREGNANCY

High risk

- Mechanical valve on heparin
- Worst valve : mitral valve

Risks of mechanical valve clot

Need for urgent surgery

Stroke

Death

PREGNANCY AND VALVE DISEASE

Low molecular weight heparin

DATA ?

LOW MOLECULAR WEIGHT HEPARIN

- **Enoxaparin (Lovenox) - product label**
“ not recommended for thromboprophylaxis in patients with prosthetic heart valves ”
- **Resulted from randomized study S. Africa**
12 pt enrolled (out of planned 110) :
2 deaths in enoxaparin group due to valve thrombosis. Each R_x 80mg sc every 12 hr

LMWH AND PROSTHETIC VALVES

Meta analysis 1989-2004

- 10 reports, only 2 prospective studies
- 75 women with 81 pregnancies
 - 44 had MVR , 8 had AVR
 - 5 had both AVR and MVR,18 not known

LMWH AND PROSTHETIC VALVES

Meta analysis 1989-2004

- **10 reports, only 2 prospective studies**
- **75 women with 81 pregnancies**
 - 44 had MVR , 8 had AVR
 - 5 had both AVR and MVR,18 not known
- **60 pregnancies : LMWH throughout**
- **21 pregnancies : LMWH 2nd half 1st trim and term**

LMWH AND PROSTHETIC VALVES

Meta analysis 1989-2004 : 81 pregnancies

- 51 pregnancies : anti Xa levels monitored
- 30 pregnancies : fixed dose used

RESULTS

- Thromboemboli in 10/81 pregnancies (12%)
- All had mitral prostheses
- All were on LMWH throughout pregnancy

LMWH & MECHANICAL VALVES

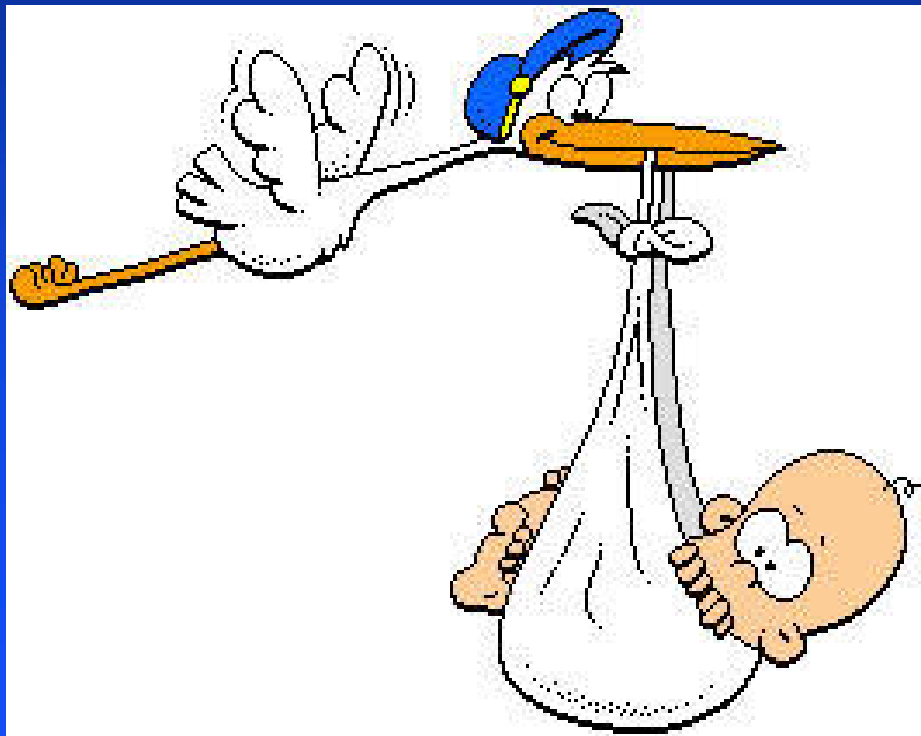
Thrombus

	Year	No. pregnancies	Valve thrombus
• Rowan J	2001	14	7%
• Oran B	2004	81	9%
• James AH	2006	76	17%
• Abildgaard U	2009	12	17%
• McLintock C	2009	34	15%
• Yinon Y	2009	23	4%
• Quinn J	2009	12	8%

PROSTHETIC VALVES & PREGNANCY

No perfect solution

Better choice



ACHD & PREGNANCY

Bottom line

- **ALWAYS** have an evaluation with a cardiologist who has expertise & training in ACHD and pregnancy care
- Ask : “ What is the risk to me ? ”
 - “ Is it possible pregnancy will make my heart worse ? “
 - “ What is my prognosis ? ”

ACHD & PREGNANCY

Bottom line

- Ask : “ What is the risk to my baby ? ”
- Ask : “Should I deliver at home or at a special center where the whole team is together ?”

**UNDERSTAND WHAT IS GOOD FOR YOU
AND YOUR PARTNER**

PREGNANCY AND HEART DISEASE

Conclusions

- **Most women with heart disease can have a pregnancy with proper care**
- **Pre-pregnancy evaluation vital**
- **High risk cases benefit from combined high-risk OB and cardiac care in the same center**

PREGNANCY AND HEART DISEASE

Most women with heart disease can have a pregnancy if properly managed



"It was all worth it"