

ASSESSMENT OF PREGNANCY RISK IN WOMEN WITH CONGENITAL HEART DISEASE

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Congenital Heart Disease

- most common form of heart disease complicating pregnancy in Western countries (~70%)
- presence of maternal CHD associated with significant increase in
 - heart failure
 - arrhythmias
 - cerebrovascular events
 - embolic events
 - death

Physiological Adaptation of Pregnancy

Antepartum

- ↑ blood volume ~ 40% (peak ~ 32 wks)
- ↑ cardiac output 30-50% (peak ~ 24 wks)
- ↑ heart rate 10-20 bpm (peak ~ 28 wks)
- ↑ pulm blood flow ~ 47%
- ↓ PVR ~ 24% (peak wk 8 onwards)
- ↓ SVR ~ 30% (peak ~ 24 wks rising to near normal towards term)

Preconception

- discussion regarding pregnancy and fetal risks
- consider additional diagnostic evaluation
- consider repair of residual defects pre pregnancy
- consider genetic referral
- discontinue teratogenic drugs
- planned management if anti coagulation required

Prepregnancy Interventions

- ascending aorta conduit
- PVR
- AVR
- Mitral valvuloplasty
- branch PA balloon ± stent

Genetic Risk

~ 18% CHD specific defined genetic abnormality

Autosomal dominant

- Marfan
- Holt-Oram
- Noonan
- Alagille
- CHARGE
- 22q 11.2 microdeletion
- Williams syndromes

Overall risk

- 3-8%
- 10% (if 1 sibling CHD)

Higher risk

- Heterotaxy
- AVSD
- Obstructive lesions LVOT (~ 20%)

Common Maternal Complications during Pregnancy

- Congestive cardiac failure
- Arrhythmias
- Embolic events

Rare

- Maternal death
- Irreversible ventricular dysfunction

Fetal Risk

Higher frequency of

- spontaneous abortions ~ 15-25%
- CHD (fetal echo ~ 20-24 wks)
- Small for gestational age; IUGR
- interventricular haemorrhage
- ↑ risk of pre term births

Modified WHO classification of maternal cardiovascular risk

Risk Class	Risk of Pregnancy by Medical Condition
I	No detectable increase of maternal mortality and no/mild increase in morbidity
II	Small increase risk of maternal mortality or moderate increase in morbidity
III	Significantly increased risk of maternal mortality or severe morbidity
IV	Extremely high risk of maternal mortality or severe morbidity. Pregnancy contraindicated. Termination should be discussed.

Modified WHO Classification of Maternal Cardiovascular Risk (2014)

WHO Pregnancy Risk Category	Risk Description	Maternal Risk Factors
1	No detectable increase in maternal mortality and no/mild increase in morbidity risk	Mild pulmonary stenosis PDA, MVP Post ASD/VSD/PDA/ anomalous PVD repair
2	Small increase in maternal mortality and moderate increase in morbidity risk	Unoperated ASD, VSD Repaired TOF Most arrhythmias
3	Significantly increased maternal mortality or severe morbidity risk. Specialist cardiac and obstetric monitoring required throughout pregnancy, childbirth and the puerperim	<ul style="list-style-type: none"> Mechanical valve Systemic RV Fontan circulation Unrepaired cyanotic heart disease Other complex CHD Aortic dilatation 40-45 Marfan syndrome Aortic dilatation 45-50 in bicuspid aortic valve aortopathy
4	Extremely high maternal mortality or severe morbidity risk. Pregnancy is C/I. Termination should be considered.	<ul style="list-style-type: none"> Pulmonary arterial hypertension Severe systemic ventricular dysfunction LVEF <30%, NYHA Class III-IV Previous peripartum cardiomyopathy with residual LV impairment Severe MS/AS Native severe coarctation Aortic dilatation >45mm Marfan Aortic dilatation >50mm in BAV

Consider Termination if

- severe pulmonary hypertension
- severe systemic ventricular dysfunction
- severe aortic dilatation
- mechanical mitral valve prosthesis

Medications During Pregnancy

		FDA pregnancy Category	Teratogenic risks	Other	Lactation
B-Blockers	Metoprolol Propranolol Carvedilol Atenolol	C C C D	None	Possible association with fetal growth restriction (atenolol, propranolol)	Probably safe, except for atenolol
Combined α -B blockers	Labetalol	C	None	No	No
ACEI	Captopril Enalapril Lisinopril	C (first trimester) D (2 nd and 3 rd trimester)	Fetal renal dysplasia Oligohydramnios Fetal death		Yes (captopril and enalapril) Lisinopril – unknown
Vasodilators	Sildenafil	B	None	No	Unknown
Diuretics	Furosemide	C	None	Possible association with neonatal PDA and sensorinical hearing loss	No
	Spironlactone (not recommended)	C	None	Antiandrogenic effects of male fetuses	Yes
Antiarrhythmics	Sotalol Amiodarone Adenosine	B D C	None None	Thyroid dysfunction and IUGR	No No Unknown
Endothelium receptor antagonists	Bosentan Macitentan	X X			

Maternal Risk with Congenital Heart Disease and Pregnancy

Low Risk

- left to right shunts (ASD – risk of paradoxical embolus low)
- repaired AVSD
- Ebstein's anomaly (if no cyanosis)
- pulmonary/mitral/aortic/tricuspid regurgitation

Medium Risk

- moderate to severe aortic stenosis
- mild to moderate systemic ventricular dysfunction
- repaired coarctation of the aorta
- post TOF repair with severe PR
- post arterial switch repair for DTGA

Maternal Risk with Congenital Heart Disease and Pregnancy

High Risk

- post Fontan repair
- cyanotic
- post Mustard/Senning repair for DTGA
- Marfan's with Ao root > 40mm

Extreme risk

- Eisenmenger syndrome
- severe pulmonary hypertension
- symptomatic aortic stenosis
- Marfan's with Ao root > 45mm
- symptomatic severe systemic ventricular dysfunction

Aortic Stenosis and Pregnancy

Low risk

- mild aortic stenosis
- NYHA class I
- exercise test satisfactory
- normal LV function

Medium

- asymptomatic moderate-severe aortic stenosis

Very high

- symptomatic aortic stenosis
- AS with LV dysfunction
- bicuspid aortic valve with ascending aorta >45mm
- consider aortic balloon valvuloplasty

Moderate to Severe Asymptomatic Aortic Stenosis and Pregnant

Risks of

- congestive cardiac failure ~10%
- arrhythmias ~ 10%
- mortality rare

Fetal risks (25%)

- prematurity
- IUGR
- low birth weight

Pregnancy & delivery

- vaginal delivery usually preferred, early epidural \pm planned induction
- Caesarean section may be necessary if early delivery required (ie 34-36 weeks)

Pregnancy post atrial switch repair (Mustard/Senning)

Very high risk if

- NYHA >II; poor exercise tolerance
- RV dysfunction EF <40%
- >mild TR

Pregnancy and Marfan Syndrome

Contra-indicated if

- aortic root $\geq 45\text{mm}$ (ECS + Canadian guidelines)
- previous aortic dissection

Pre-pregnancy

- recent CTA/MRA
- if identified mutation, preimplantation genetic diagnosis possible
- stop ACEI/ARB
- continue BB

Pregnancy

- echo every 4-6 weeks

Gene(s) most commonly identified for the clinical aortopathy diagnosis (CSANZ 2016)

Condition	Gene
Marfan Syndrome (AD)	FBN1
Loeys-Dietz Syndrome (AD)	TGFBR2 TGFBR1 SMAD3 TGFB2
Enhlers Danlos Syndrome (Type IV)	COL3A1
Arterial Tortuosity Syndrome (AR)	SLC2A10
Non-systemic TAAD	ACTA2 MYH11 PRKGI MYLK
Bicuspid Aortic Valve	NOTCHI

Turner Syndrome

Aortic surgery pre pregnancy if

- Aortic root $> 2.5\text{cm}/\text{m}^2$ (otherwise $> 2.7\text{cm}/\text{m}^2$)

High risk

- Aortic root $2.0\text{cm}/\text{m}^2 - 2.4\text{cm}/\text{m}^2$

Fontan and Pregnancy

Very high risk (maternal and fetal) if -

- past atrial arrhythmias
- past pulmonary emboli
- ↓ exercise tolerance
- NYHA >II
- cyanosis with arterial O₂ satⁿ <85% at rest or on exercise
- systemic ventricular dysfunction ↓ EF

High Risk

- arterial O₂ satⁿ > 85% (rest and exercise)
- normal systemic ventricular EF
- good exercise tolerance (>9min Bruce protocol)
- ≤ mild atrioventricular regurgitation
- NYHA Grade 1-2
- sinus rhythm

Pregnancy and Cyanotic Congenital Heart Disease (excluding Eisenmenger's, severe PHT)

- mixed group (Ebstein's + PFO; CCTGA/VSD/PS; univentricular hearts; unrepaired TOF; PA + VSD)

Risks

- congestive cardiac failure (? Permanent deterioration)
- arrhythmias mainly atrial
- cardioembolic events (esp post partum)

Prepregnancy

- repair if possible
- exercise test helpful
- fetal risks very high if arterial O₂ saturation <85%

Eisenmenger Syndrome

- termination advised
- maternal mortality improved but still ~25%
- cease ERA – (teratogenic in animals)
- consider sildenafil/tadalafil ± prostacyclin derivatives
- consider diuretics, anticoagulants, restrict exercise
- early planned delivery around 32-34 weeks
- stay in hospital at least 1 week post delivery

Mode of Delivery

- generally vaginal delivery preferred
- consider induction/early epidural
- haemodynamic monitoring during labour and delivery

Caesarean section usually preferred/necessary if

- delivery before 36 weeks
- Marfan syndrome/dilated aorta

CONCLUSION

- **Management of pregnancy includes**
 - preconception care
 - pregnancy management
 - planning of labour and mode of delivery
 - postpartum care

- **Very high risk**
 - systemic ventricular dysfunction
 - cyanosis (arterial O₂ satⁿ <85%)
 - NYHA >II, poor exercise tolerance
 - pulmonary hypertension

CONCLUSION

Pregnancy in Women with Congenital Heart Disease

- multidisciplinary team required
- medical management/interventions frequently required
- significant risks to both mother and baby

However,

- outcomes generally good except in the very high risk groups

Predictors of maternal CVS events and risk score from CARPREG study

- Prior cardiac event (CCF, TIA, CVA, arrhythmias)
- NYHA class >II or cyanosis
- mitral stenosis, aortic stenosis
- systemic ventricular EF <40%

0 point	5%
1 point	27%
>1 point	75%

ZAHARA predictors

- past arrhythmia event
- NYHA class >II
- severe aortic stenosis (PG >50mmHg)
- mechanical valve prosthesis
- moderate/severe atrioventricular valve regurgitation
- use of cardiac medication pre-pregnancy
- cyanosis