Assessment and Management of Pregnancy in Patients with Congenital Heart Diseases

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Disclosure

No conflict of interest

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Physiology of Pregnancy

Risk stratification & Assessment

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Epidemiology

Heart diseases in women of childbearing age (16 – 45yo):

- Rheumatic heart disease
- Congenital heart disease
- Coronary artery disease
- Cardiac Arrhythmia
- Cardiomyopathy
- Inflammatory cardiac disease
- Pulmonary hypertension

Epidemiology

Pregnancy outcomes in women with cardiovascular disease: evolving trends over 10 years in the ESC Registry Of Pregnancy And Cardiac disease (ROPAC)

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European Heart Journal (2019) **00**, 1–8
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Pregnancy Outcomes in Women With Heart Disease

The CARPREG II Study

(J Am Coll Cardiol 2018;71:2419-30)

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妊娠合并心脏病患者心脏不良事件危险因素分析

周晓瑞　卢家凯　陈晓　李强　叶清　孙建萍　张京岚

【摘要】 **目的**　探讨心脏病孕产妇发生心脏不良事件的危险因素，为改善此类患者妊娠结局提供帮助。**方法**　回顾性分析2004—2012年在首都医科大学附属北京安贞医院综合外科监护病房住院的围产期心脏病孕产妇的临床资料。采用单因素和多因素logistic回归分析心脏病孕产妇发生心脏不良事件的危险因素。**结果**　190例孕龄≥20周心脏病孕产妇入选本研究，其中先天性心脏病134例(70.5%)，风湿性心脏病30例(15.8%)，心肌病10例(5.3%)，围生期心肌病2例(1.1%)，原发性高血压性心脏病14例(7.4%)。42例(22.1%)患者发生心脏不良事件。7例患者死亡，病死率3.7%。死亡原因为循环衰竭死亡4例；继发肺部感染，呼吸、循环衰竭死亡2例；风湿性心脏病二尖瓣置换术后患者，孕晚期瓣膜功能急性失调，急诊终止妊娠同时行二次换瓣术，术后因严重肺动脉高压、循环衰竭死亡1例。多因素logistic回归分析显示，孕前纽约心脏病协会心功能分级＞Ⅰ级、产前左心室射血分数＜50%、产前应用血管活性药、重度肺动脉高压[肺动脉收缩压(SPAP)＞80 mm Hg(1 mm Hg=0.133 kPa)]是心脏病孕产妇发生心脏不良事件的独立危险因素。**结论**　心脏病孕产妇心脏不良事件发病率高，重度肺动脉高压(SPAP ＞80 mm Hg)、孕前心功能差是导致心脏病孕产妇发生心脏不良事件的主要危险因素，临床工作中应加强对此类患者的密切监护及合理治疗。

An analysis of risk factors for postpartum cardiac events in pregnant women with heart disease
中华内科杂志 - Chinese Journal of Internal Medicine, 2013, Vol.52(11), pp.966-969

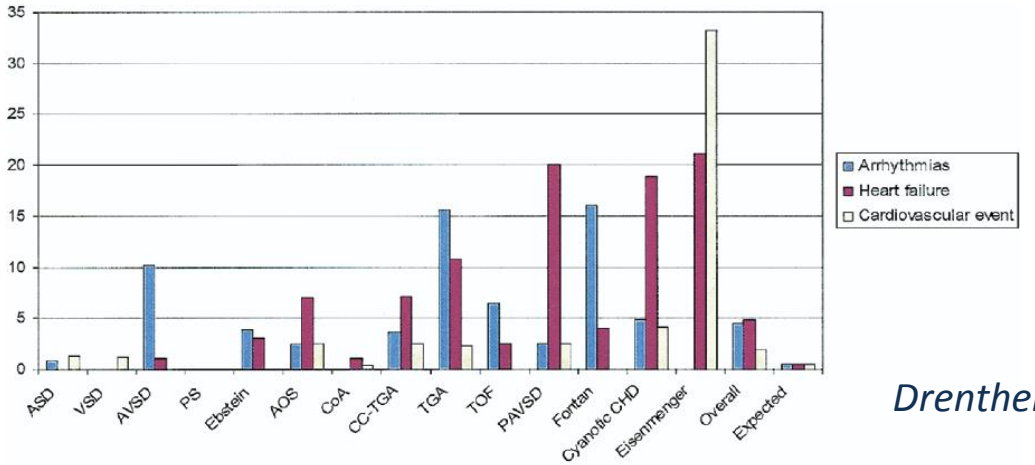
Epidemiology

	ROPAC 2019	CARPREG II, 2018	Beijing, 2013
Number	5739	1938	190
Type of Heart disease			
Congenital HD	3295 (57.4%)	1235 (63.7%)	134 (70.5%)
Valvular HD	1648 (28.7%)		RHD 30 (15.8%)
Cardiomyopathy	438 (7.6%)		12 (6.3%)
Ischaemic HD	95 (1.6%)		
Acquired HD		443 (22.9%)	
Pulmonary HT			14 (7.4%)
Isolated arrhythmia		260 (13.4%)	

Maternal Cardiac events

Maternal Cardiac Events		Reported	Expected
Heart failure	79 / 1663	**4.8%**	<0.5%
Arrhythmia	70 / 1562	**4.5%**	<0.5%
Cardiovascular events	33 / 1740	**1.9%**	<0.5%
Endocarditis	7 / 1372	**0.5%**	<0.01%

Overall cardiac events: 7.8%



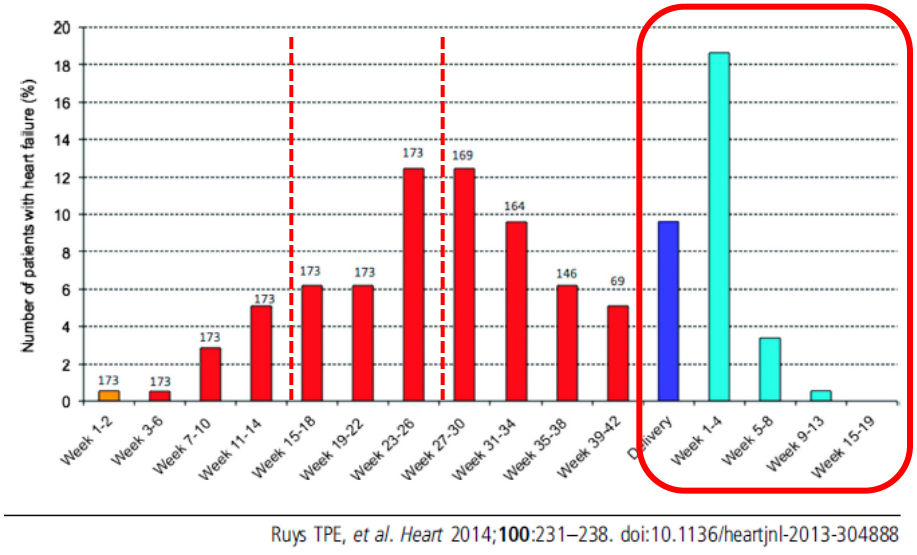
Drenthen et al. JACC 2007

Maternal Outcome

	ROPAC 2019	CARPREG II, 2018	Beijing, 2013
MACE		307 (15.8%)	42 (22.1%)
Heart failure	611 (10.6%)	120 (6.2%)	35 (18.4%)
Arrhythmia	185 (3.3%)	181 (9.3%)	10 (5.3%)
Maternal death	34 (0.6%)	6 (0.3%)	5 (2.6%)
Cardiac arrest		8 (0.4%)	
Stroke		13 (0.7%)	
Thromboembolism	87 (1.5%)	6 (0.3%)	
Myocardial infarction		8 (0.4%)	
Dissection	5 (0.1%)	7 (0.4%)	

Timing of maternal cardiac events

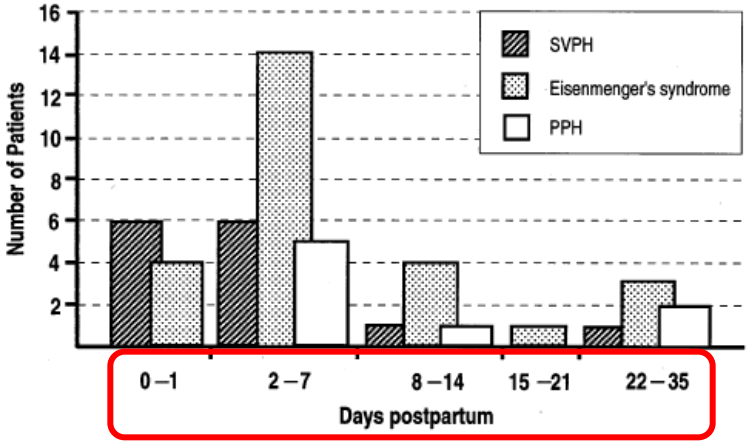
Heart failure



Ruys TPE, *et al*. *Heart* 2014;**100**:231–238. doi:10.1136/heartjnl-2013-304888

Pulmonary hypertension

JACC Vol. 31, No. 7
June 1998:1650–7



Obstetric Events

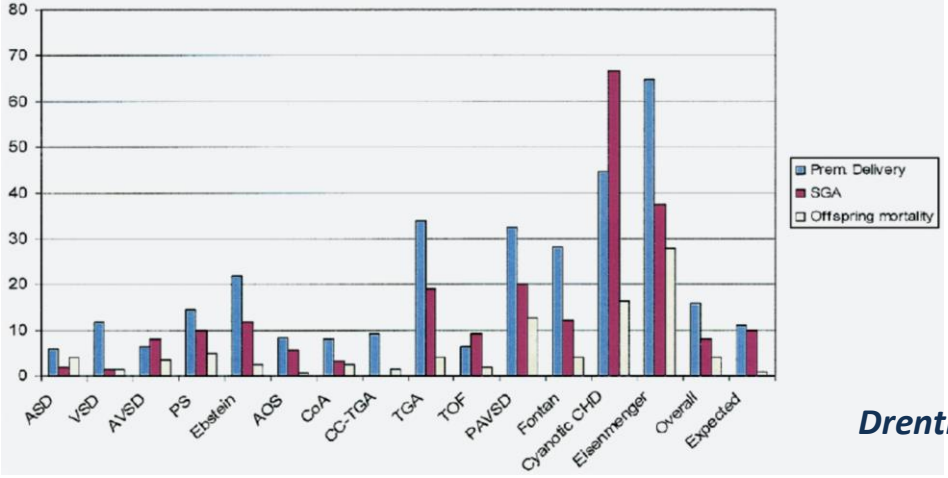
Obstetric Cx	Drenthen et al, 2007	CARPREG II, 2019	expected
PIH	54 / 989 (5.5%)	150/ 5739 (2.6%)	5.05%
Pre-eclampsia	35 / 1098 (3.2%)	159 /5739 (2.8%)	2 – 3%
Thromboembolism	**15 / 688 (2.2%)**		0.1%
PPH	44 / 552 (8.4%)	170/5739 (3.0%)	10 – 12%

Obstetric complication (at least 1 of the following)[a]	30 (32.6)
Pregnancy-induced hypertension or pre-eclampsia	3 (3.3)
Preterm delivery	19 (20.7)
<28 weeks	4 (21.1)
28–32 weeks	0 (0)
>32 weeks	15 (78.9)
Placental abruption	3 (3.3)
Preterm premature rupture of membranes	9 (9.8)
Post-partum hemorrhage	13 (14.1)
Intra-uterine fetal demise	2 (2.2)

Ouyang et al. Int J Cardiol 2010

Neonatal Events

Foetal / Neonatal Cx	Drenthen et al, 2007	CARPREG II, 2019	Expected
Premature delivery	**224/1413 (15.9%)**	**905/5739 (15.8%)**	10 – 12%
SGA / IUGR	110/1381 (8.0%)	254/5739 (4.4%)	10%
Foetal mortality	**31/1776 (1.7%)**	**72 /5739 (1.3%)**	<0.5%
Neonatal mortality	**41/1756 (2.3%)**	33/5739 (0.6%)	<0.5%
CHD recurrence	**56/1616 (3.5%)**		0.7-1%



Drenthen et al. JACC 2007

Cardiovascular Physiology of Pregnancy

Physiological Changes in Pregnancy

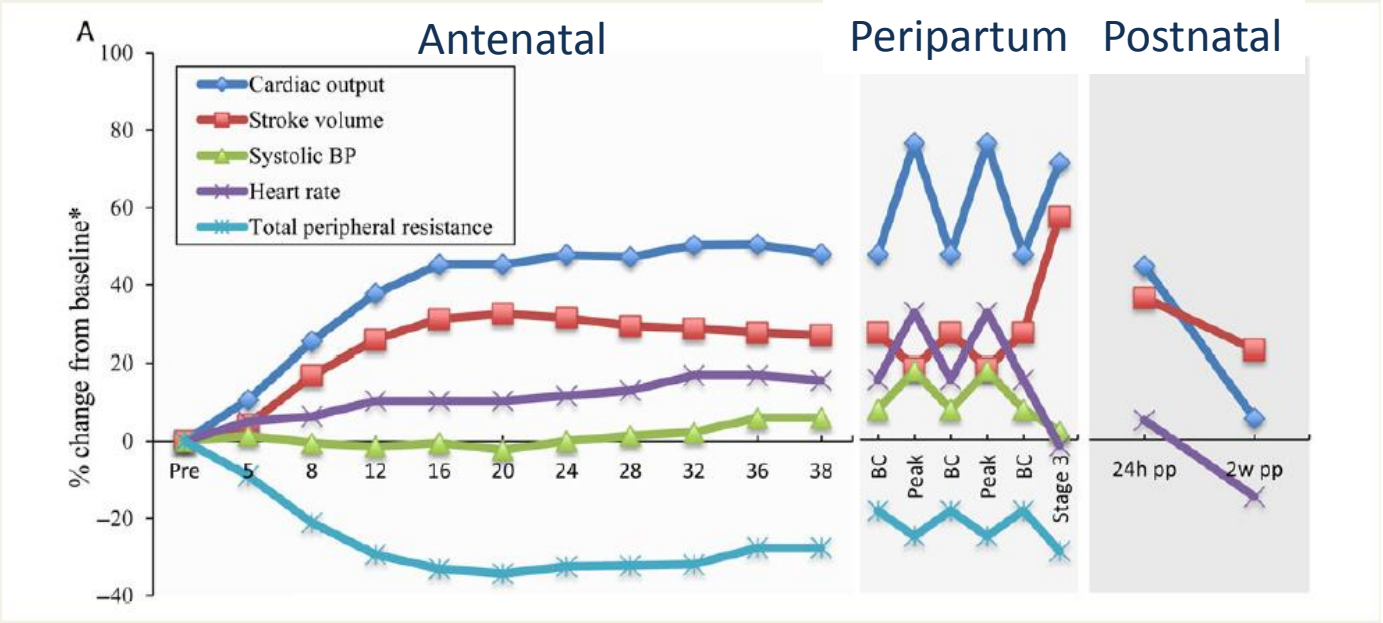


Figure 2 Haemodynamic changes during pregnancy, peripartum, and postpartum. (*A*) Pregnancy (weeks of gestation). Modified from Robson *et al.*[12] (*B*) Peripartum. Modified from: Adams *et al.*[13] BC, between contractions; Peak, at the peak of contraction; Stage 3, at the time of uterine contraction. (*C*) Postportum. Modified from Robson *et al.*[14,15] 24 h pp, 24 h postpartum; 2w pp, 2 weeks postpartum. *For cohorts in (*B*) and (*C*), relative changes from baseline were compared with the baseline values of the cohort from (*A*).

Greutmann & Pieper, EHJ 2015

Physiological Changes in Pregnancy

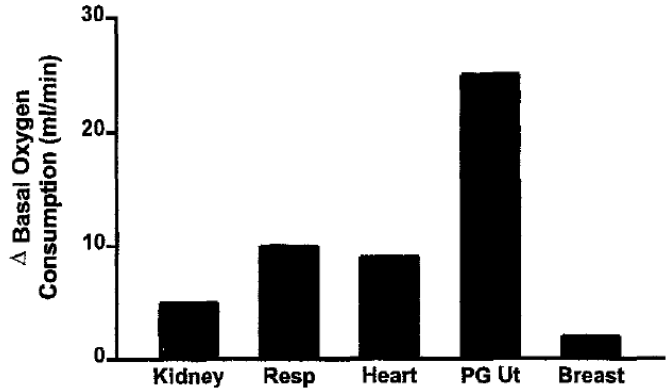


Figure 1. The maximal increase in basal oxygen consumption (mL/min) during pregnancy. (Data from ref 1.)

Metabolic demand of foetus & mother → ↑VO2 30%

Haematological:

Red cell mass ↑20-30%, Plasma volume ↑30 - 50% →↑ total blood volume, relative anaemia

Hypercoagulable state: ↑ factors VII, VIII, X, and fibrinogen

Renal:

↑ glomerular filtration rate (GFR) and renal plasma flow (RPF) early in pregnancy → ↑ creatinine clearance, BUN at ULN, lower creatinine. GFR and RPF return to normal in the third trimester

Alteration of pharmacokinetics:

↑ blood volume, ↓ plasma proteins, ↑ renal clearance, ↑ hepatic clearance, ↓ gastrointestinal absorption

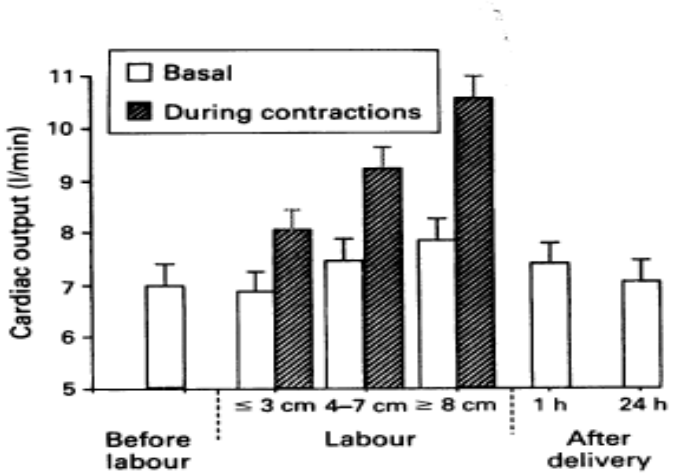
Cardiovascular Changes during Delivery

Pain / Anxiety / stress – further ↑ CO by 50-61%

Uterine contraction – 300-500 ml blood entering into circulation

Haemodynamic effects of uterine contraction:

Parameter	Change
Blood Volume	↑ 300-500ml
Cardiac Output	↑ 30 -60%
Heart Rate	↑ or ↓
Blood Pressure	↑
Peripheral Resistance	Unchanged
O_2 Consumption	↑ Up to 100%



Elkayam U et al. Cardiac Problems in Pregnancy. 1990.

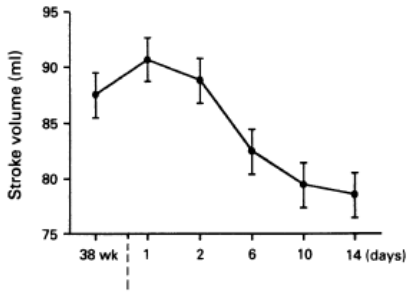
Post-partum Changes

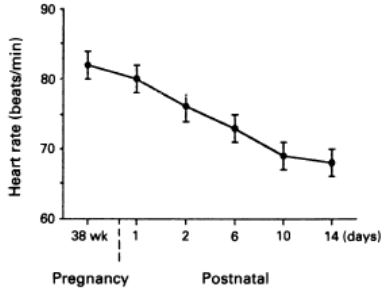
After delivery & postpartum:

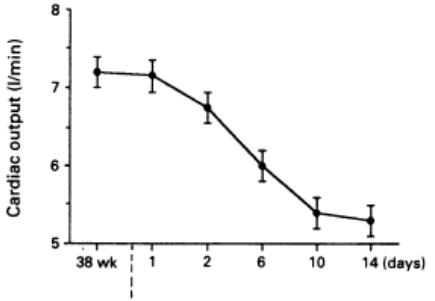
Venous return ↑ [redistribution of blood volume & relief venocaval compression] → ↑ CO up to 60 – 80%

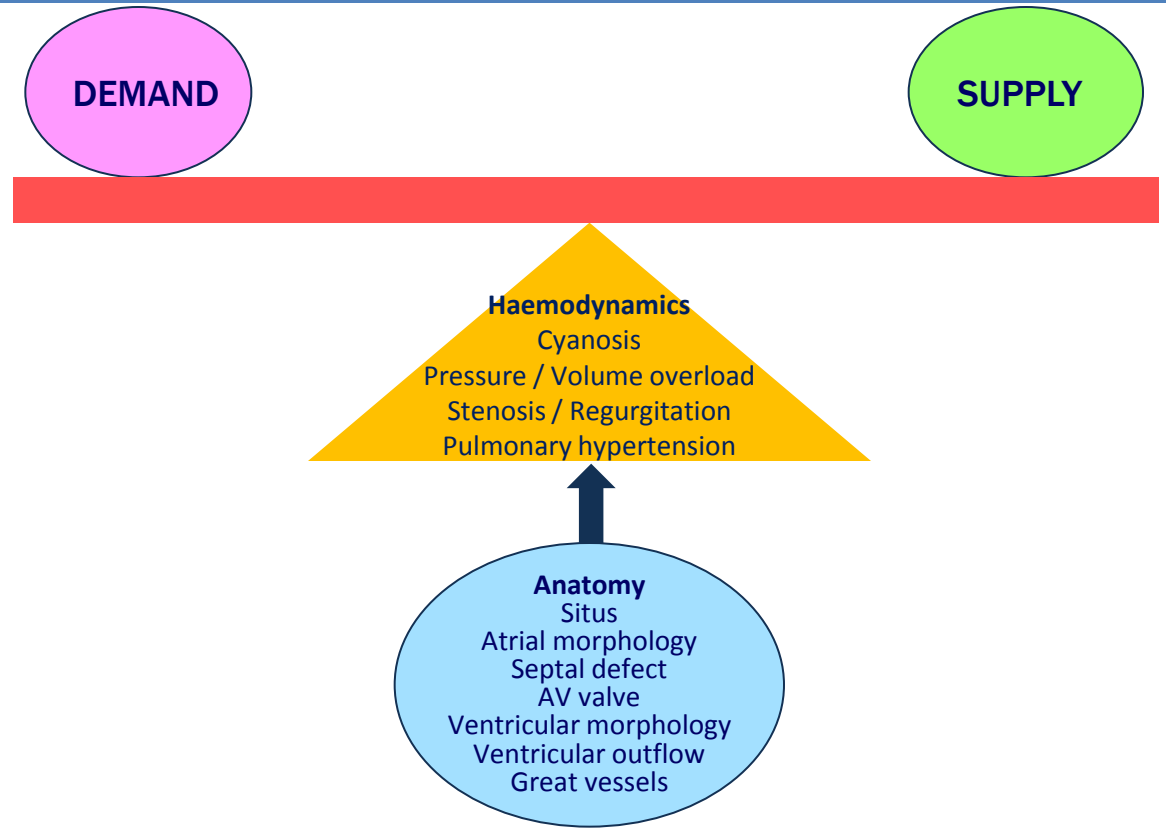
Spontaneous diuresis during first 24 - 48 hours

Haemodynamics return to baseline by 2 - 4 weeks








DEMAND
SUPPLY
Haemodynamics
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Situs
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Septal defect
AV valve
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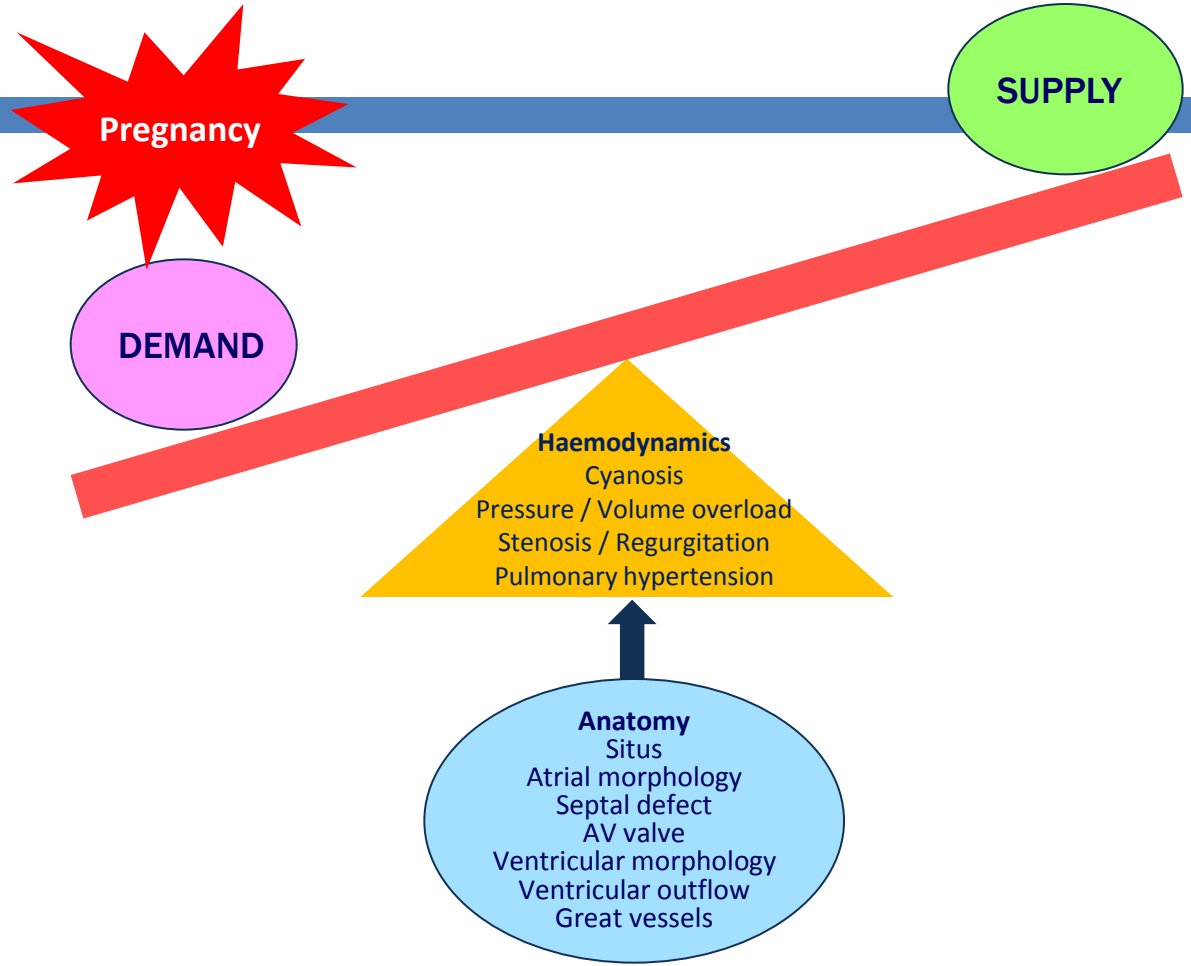
Ventricular outflow
Great vessels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Assessment & Care during Pregnancy

Management Guidelines

2018 ESC Guidelines for the management of cardiovascular diseases during pregnancy

The Task Force for the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology (ESC)

Endorsed by: the International Society of Gender Medicine (IGM), the German Institute of Gender in Medicine (DGesGM), the European Society of Anaesthesiology (ESA), and the European Society of Gynecology (ESG)

Authors/Task Force Members: Vera Regitz-Zagrosek* (Chairperson) (Germany), Jolien W. Roos-Hesselink* (Co-Chairperson) (The Netherlands), Johann Bauersachs (Germany), Carina Blomström-Lundqvist (Sweden), Renata Cífková (Czech Republic), Michele De Bonis (Italy), Bernard Iung (France), Mark Richard Johnson (UK), Ulrich Kintscher (Germany), Peter Kranke[1] (Germany), Irene Marthe Lang (Austria), Joao Morais (Portugal), Petronella G. Pieper (The Netherlands), Patrizia Presbitero (Italy), Susanna Price (UK), Giuseppe M. C. Rosano (UK/Italy), Ute Seeland (Germany), Tommaso Simoncini[2] (Italy), Lorna Swan (UK), Carole A. Warnes (USA)

The American College of Obstetricians and Gynecologists
WOMEN'S HEALTH CARE PHYSICIANS

ACOG PRACTICE BULLETIN

Clinical Management Guidelines for Obstetrician–Gynecologists

Number 212

Presidential Task Force on Pregnancy and Heart Disease
Committee on Practice Bulletins—Obstetrics. This Practice Bulletin was developed by the American College of Obstetricians and Gynecologists' Committee on Practice Bulletins–Obstetrics in collaboration with the Presidential Task Force on Pregnancy and Heart Disease members Lisa M. Hollier, MD, James N. Martin Jr., MD, Heidi Connolly, MD, Mark Turrentine, MD, Afshan Hameed, MD, Katherine W. Arendt, MD, Octavia Cannon, DO, Lastascia Coleman, ARNP, CNM, Uri Elkayam, MD, Anthony Gregg, MD, MBA, Alison Haddock, MD, Stacy M. Higgins, MD, FACP, Sue Kendig, JD, Robyn Liu, MD, MPH, FAAFP, Stephanie R. Martin, DO, Dennis McNamara, MD, Wanda Nicholson, MD, Patrick S. Ramsey, MD, MSPH, Laura Riley, MD, Elizabeth Rochin, PhD, RN, NE-BC, Stacey E. Rosen, MD, Rachel G. Sinkey, MD, Graeme Smith, MD, PhD, Calondra Tibbs, MPH, Eleni Z. Tsigas, Rachel Villanueva, MD, Janet Wei, MD, and Carolyn Zelop, MD.

Pregnancy and Heart Disease

Management of Pregnancy in Patients With Complex Congenital Heart Disease

A Scientific Statement for Healthcare Professionals From the American Heart Association

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.

Expanded diagnostic, medical, and surgical management options have improved the long-term survival of patients with congenital heart disease (CHD). Thus, most women born with CHD will reach reproductive age. The ability to bear children is a major point of care for this growing population. As a result, pregnancy counseling and management are among the major noncardiac issues facing pediatric and congenital cardiac providers.

For the majority of patients, the ability to conceive and carry a pregnancy to term will present little problem. However, for those with complex CHD, pregnancy may be associated with an increased risk compared with women with milder forms of CHD, regardless of whether they are clinically stable at the time of conception. This document provides an overview of the management of the patient with complex CHD who becomes pregnant.

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Candice Silversides, MD, FRCPC
Karen Stout, MD, FAHA
On behalf of the American Heart Association Council on Cardiovascular and Stroke Nursing; Council on Clinical Cardiology; Council on Cardiovascular Disease in the Young; Council on Functional Genomics and Translational Biology; and Council on Quality of Care and Outcomes Research

Assessment for Pregnancy

General concerns:

- Status of underlying cardiac disease
- Need for intervention in future
- Prognosis & life expectancy

Maternal concerns:

- Maternal morbidity and mortality associated with pregnancy
- Timing of pregnancy: 20's vs 30's

Foetal concerns:

- Foetal death and morbidity
- Prematurity, neonatal mortality and morbidity
- Risk of recurrence of CHD

Psychosocial concerns:

- Family completion
- Ability to provide child care
- Availability of support – unmarried vs married, other family members

Assessment of Pregnancy in Women with Heart Diseases

Timing:

- Pre-pregnancy
- Pregnancy
  - antepartum
  - intrapartum
  - post-partum

Modalities:

- Clinical (NYHA class, saturation, cardiac symptoms/signs)
- ECG +/-Holter
- Echocardiography
- Exercise testing (CPET)
- +/- CXR
- +/- MRI heart

Risk Stratification

Risk Stratification

Pregnancy Outcomes in Women With Heart Disease

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Predictors of pregnancy complications in women with congenital heart disease

Willem Drenthen[1]*, Eric Boersma[2], Ali Balci[1], Philip Moons[3], Jolien W. Roos-Hesselink[4], Barbara J.M. Mulder[5,6], Hubert W. Vliegen[7], Arie P.J. van Dijk[8], Adriaan A. Voors[1], Sing C. Yap[4], Dirk J. van Veldhuisen[1], and Petronella G. Pieper[1] On behalf of the ZAHARA Investigators

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Aims Data regarding pregnancy outcome in women with congenital heart disease (CHD) are limited.

Methods and results In 1802 women with CHD, 1302 completed pregnancies were observed. Independent predictors of cardiac, obstetric, and neonatal complications were calculated using logistic regression. The most prevalent cardiac complications during pregnancy were arrhythmias (4.7%) and heart failure (1.6%). Factors independently associated with maternal cardiac complications were the presence of cyanotic heart disease (corrected/uncorrected) ($P < 0.0001$), the use of cardiac medication before pregnancy ($P < 0.0001$), and left heart obstruction ($P < 0.0001$). New characteristics were mechanical valve replacement ($P = 0.0014$), and systemic ($P = 0.04$) or pulmonary atrioventricular valve regurgitation related with the underlying (moderately) complex CHD ($P = 0.03$). A new risk score for cardiac complications is proposed. The most prevalent obstetric complications were hypertensive complications (12.2%). No correlation of maternal characteristics with adverse obstetric outcome was found. The most prevalent neonatal complications were premature birth (12%), small for gestational age (14%), and mortality (4%). Cyanotic heart disease (corrected/uncorrected) ($P = 0.0003$), mechanical valve replacement ($P = 0.03$), maternal smoking ($P = 0.007$), multiple gestation ($P = 0.0014$), and the use of cardiac medication ($P = 0.0009$) correlated with adverse neonatal outcome.

Conclusion In our tertiary CHD cohort, cardiac, obstetric, and neonatal complications were frequently encountered, and (new) correlations of maternal baseline data with adverse outcome are reported. A new risk score for adverse cardiac complications is proposed, although prospective validation remains necessary.

ZAHARA

Predictors of pregnancy complications in women with congenital heart disease

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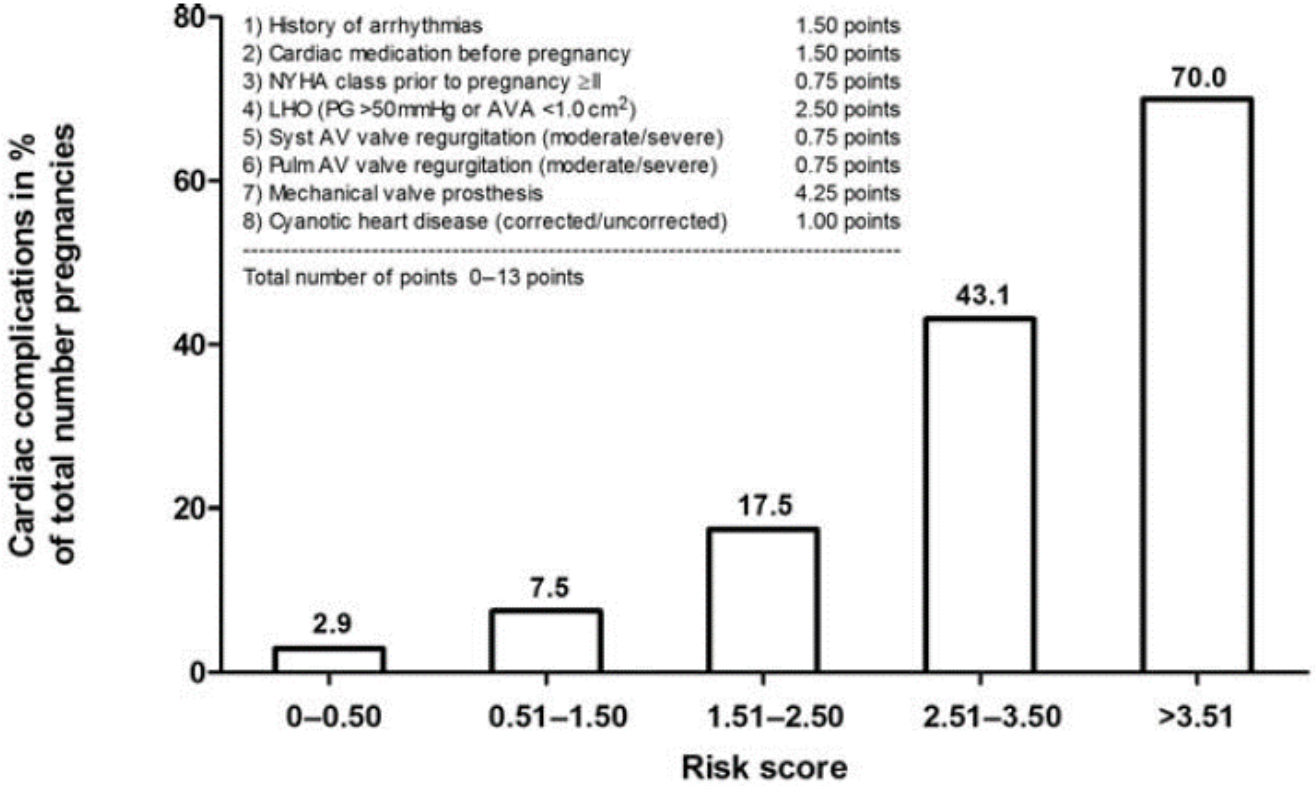


Eur Heart J 2010

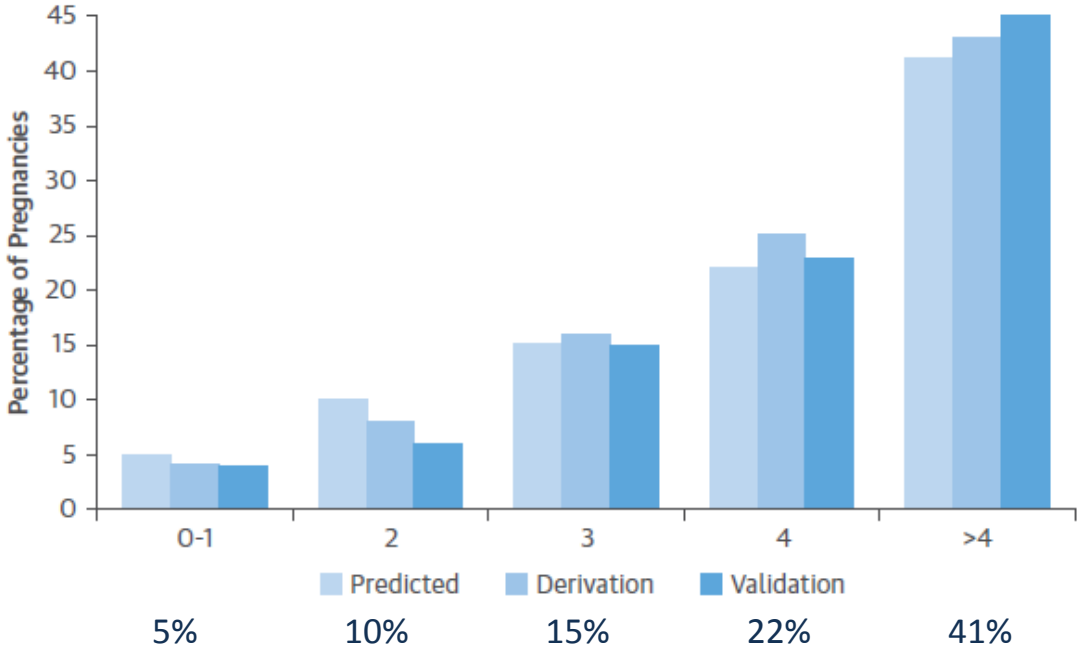
CARPREG II Risk Index

Pregnancy Outcomes in Women With Heart Disease

The CARPREG II Study (J Am Coll Cardiol 2018;71:2419–30)

Candice K. Silversides, MD, MS,[a,b] Jasmine Grewal, MD,[c] Jennifer Mason, RN,[a,b] Mathew Sermer, MD,[a,b] Marla Kiess, MD,[c] Valerie Rychel, MD,[d] Rachel M. Wald, MD,[a,b] Jack M. Colman, MD,[a,b] Samuel C. Siu, MD, SM, MBA[a,b,e]

FIGURE 4 CARPREG II Risk Prediction Index: Incidence of Adverse Cardiac Events Stratified According to CARPREG II Risk Scores



PREDICTOR	POINTS
Prior cardiac events or arrhythmias	3
Baseline NYHA III-IV or cyanosis	3
Mechanical valve	3
Ventricular dysfunction	2
High risk left-sided valve disease/ left ventricular outflow tract obstruction	2
Pulmonary hypertension	2
Coronary artery disease	2
High risk aortopathy	2
No prior cardiac intervention	1
Late pregnancy assessment	1

Predicted risks for primary cardiac events

modified WHO class – ESC 2018

	mWHO I	mWHO II	mWHO II–III	mWHO III	mWHO IV
Diagnosis (if otherwise well and uncomplicated)	Small or mild – pulmonary stenosis – patent ductus arteriosus – mitral valve prolapse Successfully repaired simple lesions (atrial or ventricular septal defect, patent ductus arteriosus, anomalous pulmonary venous drainage) Atrial or ventricular ectopic beats, isolated	Unoperated atrial or ventricular septal defect Repaired tetralogy of Fallot Most arrhythmias (supraventricular arrhythmias) Turner syndrome without aortic dilatation	Mild left ventricular impairment (EF >45%) Hypertrophic cardiomyopathy Native or tissue valve disease not considered WHO I or IV (mild mitral stenosis, moderate aortic stenosis) Marfan or other HTAD syndrome without aortic dilatation Aorta <45 mm in bicuspid aortic valve pathology Repaired coarctation Atrioventricular septal defect	Moderate left ventricular impairment (EF 30–45%) Previous peripartum cardiomyopathy without any residual left ventricular impairment Mechanical valve Systemic right ventricle with good or mildly decreased ventricular function Fontan circulation. If otherwise the patient is well and the cardiac condition uncomplicated Unrepaired cyanotic heart disease Other complex heart disease Moderate mitral stenosis Severe asymptomatic aortic stenosis Moderate aortic dilatation (40–45 mm in Marfan syndrome or other HTAD; 45–50 mm in bicuspid aortic valve, Turner syndrome ASI 20–25 mm/m^2, tetralogy of Fallot <50 mm) Ventricular tachycardia	Pulmonary arterial hypertension Severe systemic ventricular dysfunction (EF <30% or NYHA class III–IV) Previous peripartum cardiomyopathy with any residual left ventricular impairment Severe mitral stenosis Severe symptomatic aortic stenosis Systemic right ventricle with moderate or severely decreased ventricular function Severe aortic dilatation (>45 mm in Marfan syndrome or other HTAD, >50 mm in bicuspid aortic valve, Turner syndrome ASI >25 mm/m^2, tetralogy of Fallot >50 mm) Vascular Ehlers–Danlos Severe (re)coarctation Fontan with any complication

Modified WHO I & II

mWHO I
Small or mild - Pulmonary stenosis - PDA - Mitral valve prolapse
Repaired simple lesions (ASD, VSD, DA, anomalous pulmonary venous drainage)
Atrial or ventricular ectopics (isolated)

Maternal cardiac events: 2.5 – 5%

Cardiology follow-up: once every trimester

Care & Delivery: local hospital

mWHO II
Unoperated ASD or VSD
Repaired tetralogy of Fallot
Most arrhythmias (SVT)
Turner syndrome without aortic dilatation

Maternal cardiac events: 5.7 – 10.5%

Cardiology follow-up: once every trimester

Care & Delivery: local hospital

Modified WHO II - III

mWHO II - III
Mild LV impairment (EF > 45%)
Hypertrophic cardiomyopathy
Native or tissue valve disease not considered mWHO I or IV (mild MS, moderate AS)
Mild aortic dilation: Marfan < 40mm, BAV < 45mm
Repaired coarctation
Atrioventricular septal defect

Maternal cardiac events:
10 - 19%
Cardiology FU: bimonthly
A/N care & delivery:
referral centre

Modified WHO III

mWHO III
Moderate LV impairment (EF 30 – 45%)
Systemic RV with good or mildly decreased ventricular function
Fontan circulation – patient well and uncomplicated cardiac condition
Unrepaired cyanotic heart diseae Other complex heart disease
Mechanical valve
Moderate mitral stenosis Severe aortic stenosis
Moderate aortic dilation: Marfan 40-45mm, BAV 45-50mm, Turner syndrome ASI 20 - 25mm/m2, TOF <50mm
Ventricular tachycardia

Maternal cardiac events:
19 – 27%
FU: monthly or bimonthly
A/N care and Delivery:
expert centre

Modified WHO IV

mWHO IV
Pulmonary arterial hypertension of any cause
Severe systemic ventricular dysfunction (LVEF <30% or NYHA III-IV)
Systemic RV with moderate-to-severely decreased ventricular function
Previous peripartum cardiomyopathy with any residual LV impairment
Fontan with any complication
Severe mitral stenosis Severe symptomatic aortic stenosis
Severe aortic dilation: Marfan or other HTAD >45mm, BAV >50mm, Turner syndrome ASI > 25mm/m2, TOF >50mm
Vascular Ehlers-Danlos Syndrome
Severe (re) coarctation

Maternal cardiac events:
40 – 100%

Counselling: pregnancy contraindicated / TOP

FU: at least monthly

A/N care or Delivery:
Expert centre

Antenatal Care

Maternal:

- Level of surveillance (Risk level)
- Surveillance of cardiac adverse event
- Monitoring of cardiac function
- Need for treatment
- Maternal drugs adjustment
- Anticipated hospitalization during pregnancy

Foetal:

- Monitoring of foetal well being and growth
- Screening of ConHD – foetal echo by 18-22weeks

Antenatal Care

Signs and symptoms of normal pregnancy versus heart failure

May be present in normal pregnancy	Suggests cardiac pathology
Fatigue	Chest pain
Exertional dyspnoea	Severe breathlessness, orthopnoea, paroxysmal nocturnal dyspnoea, cough
Palpitation (re-entry tachycardia, atrial and ventricular premature beats)	Atrial flutter or fibrillation, ventricular tachycardia
Elevated jugular venous pressure	Systemic hypotension
Sinus tachycardia 10–15% above normal heart rate	Sinus tachycardia >15% above normal heart rate
Full volume pulse	
Third heart sound	Fourth heart sound
Systolic flow murmur	Pulmonary oedema
Pedal oedema	Pleural effusion

Thorne, Heart 2004

Antenatal Care

Chest X-ray:
Straightened left heart border
Increased cardiothoracic ratio
Increased pulmonary vascular markings
Small pleural effusion (early post-partum period)

ECG:
Sinus tachycardia
left ward shift of QRS axis
Small Q wave, inverted T wave in lead III
Increase R/S ratio in V1 and V2
ST segment and T waves changes
Atrial and ventricular ectopics

Echo during pregnancy

Left ventricular mass ↑ up to 50%, partially reversed by 12 weeks post-partum

Left atrial size ↑

Left ventricular vol ↑

Left ventricular FS ↔

Left ventricular filling (diastolic function) insignificant ↓:

- mitral E velocity : ↑14% (early), ↓4% (late)

- mitral A velocity : steadily ↑ 19%,

- E/A : ↑ 6% (early), ↓10% (late)

Gestation (wk)	Left atrium	Left atrium/aorta	Left ventricle filling*	L ventricle mass (g)	Left ventricle mass index (g/m^2)	Left ventricle systolic function (%)†
14–19	3.0 ± 0.2	1.18 ± 0.12	2.1 ± 0.6	102 ± 16 (−15)	63 ± 10 (−15)	32 ± 4
20–23	3.1 ± 0.6	1.34 ± 0.28	2.1 ± 0.4	127 ± 18 (6)	77 ± 11 (4)	34 ± 4
P	.391	.428	.495	.038‡	.099	.014‡
24–27	3.2 ± 0.5	1.38 ± 0.23	2.1 ± 0.5	124 ± 23 (3)	75 ± 12 (1)	34 ± 4
P	.088	.431	.303	.096	.089	.277
28–31	3.2 ± 0.5	1.35 ± 0.23	2.0 ± 0.6	131 ± 24 (9)	78 ± 11 (5)	34 ± 4
P	.171	.335	.067	.210	.376	.032‡
32–36	3.2 ± 0.5	1.37 ± 0.23	1.8 ± 0.3	139 ± 30(16)	81 ± 13 (9)	33 ± 4
P	.457	.244	.208	.052‡	.072	0.074
37–term	3.5 ± 0.4	1.46 ± 0.17	1.8 ± 0.3	151 ± 35(26)	87 ± 15 (18)	35 ± 4
P	.016‡	.006‡	.309	.027‡	.048‡	.369
Postpartum	3.0 ± 0.5	1.27 ± 0.21	2.0 ± 0.5	120 ± 31 (0)	74 ± 16 (0)	32 ± 4
P	.002‡	.005‡	.048‡	.001‡	.006‡	.130

Maternal Drugs Safety – FDA Categories

Category A
Adequate and well-controlled studies have failed to demonstrate a risk to the fetus in the first trimester of pregnancy (and there is no evidence of risk in later trimesters).

Category B
Animal reproduction studies have failed to demonstrate a risk to the fetus and there are no adequate and well-controlled studies in pregnant women.

Category C
Animal reproduction studies have shown an adverse effect on the fetus and there are no adequate and well-controlled studies in humans, but potential benefits may warrant use of the drug in pregnant women despite potential risks.

Category D
There is positive evidence of human fetal risk based on adverse reaction data from investigational or marketing experience or studies in humans, but potential benefits may warrant use of the drug in pregnant women despite potential risks.

Category X
Studies in animals or humans have demonstrated fetal abnormalities and/or there is positive evidence of human fetal risk based on adverse reaction data from investigational or marketing experience, and the risks involved in use of the drug in pregnant women clearly outweigh potential benefits.

FDA classification:

- Majority of drugs is class C; insufficient information; better to provide the scarce information that is available than give a classification that gives no guidance
- One classification is given for the pregnancy – no differentiation for effects in 1st, 2nd and 3rd trimester.
- Subjective – ACE-inibitors (D), but statins (X)?
- False sense of security: ABCDX → graded categories?

30th June 2015: US Food and Drug Administration (FDA) replaced former A to X categories to Pregnancy and Lactation Labelling Rule (PLLR): descriptive risk summary and detailed information on animal and clinical data. PLLR applies immediately for prescription drugs approved after 30 June 215

2018 ESC Guidelines: Maternal Medications

Recommendations	Class[a]	Level[b]
Before pharmacological treatment in pregnancy is started, it is recommended to check *Table 7* for clinical safety data.	I	C
In the absence of clinical safety data, it is recommended to check the electronic drug table (www.safefetus.com) for pre-clinical safety data.	I	C
In the absence of adequate human safety data, decision-making should be based on individual drug efficacy and safety profiles, and the available animal data, and the decision must be made together with the patient.	IIa	C
Decision-making based on former FDA categories alone is no longer recommended.[11]	III	C

www.safefetus.com
www.ema.europa.eu
www.accessdata.fda.gov
www.embryotox.de

Peri-partum Managment

Planning for Delivery

Timing of delivery

- spontaneous vs induced labour vs no labour

Intrapartum monitoring

- ECG, cuff / intra-arterial BP , CVP, Swan-Ganz catheter insertion

Positioning

- left lateral decubitus [↓ aorto-caval compression]

Intrapartum anagelsia

- ↓ pain & stress

Mode of delivery

- Vaginal vs Caesarean section

Second stage management

- Shortened 2nd stage, elective instrumental delivery

Intra-partum antibiotic prophylaxis

Peri-partum Management

Timing of delivery: spontaneous vs induced labour

Depends on:

- Cardiac status
- Bishop score
- Foetal well-being and lung maturity

Spontaneous onset of labour and vaginal delivery is preferred

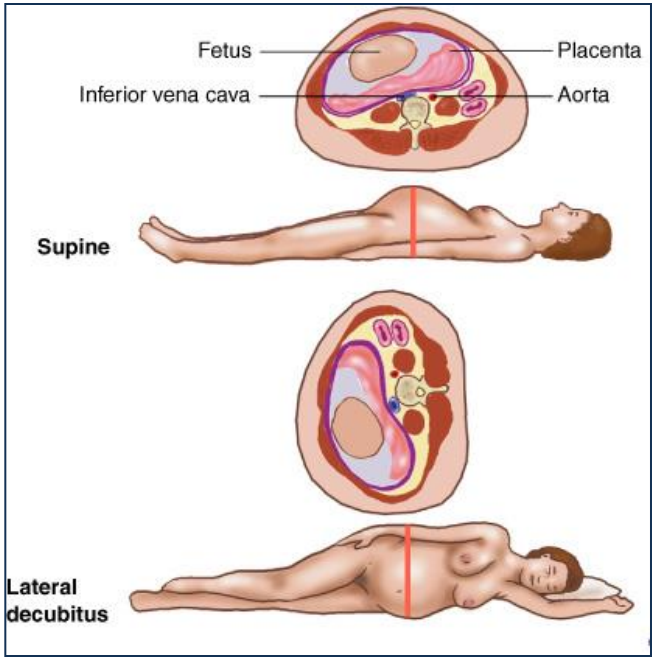
Induction of labour should be considered at 40 weeks of gestation in all women with cardiac disease.	IIa	C

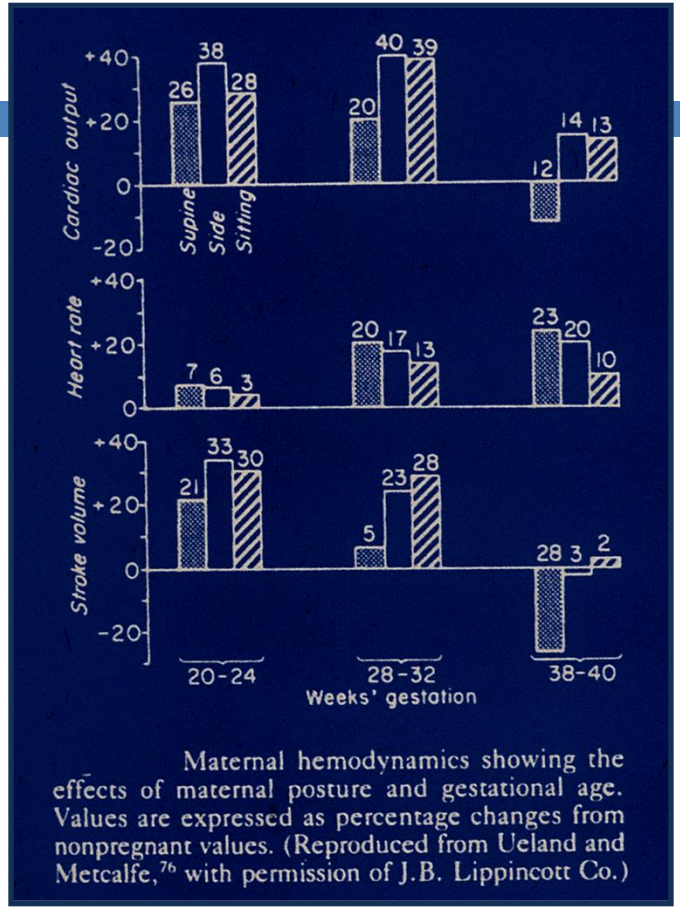
Induction of labour:

- Prostaglandins:
  - misoprostol (PGE1) 2mcg
  - dinoprostone (PGE2) 1 – 3 mg / 10mg (slow release)]: potential drop in SVR & hypotension
- Oxytocin
- AROM

Intrapartum Monitoring and Management

Maternal Posturing





Maternal hemodynamics showing the effects of maternal posture and gestational age. Values are expressed as percentage changes from nonpregnant values. (Reproduced from Ueland and Metcalfe,[76] with permission of J.B. Lippincott Co.)

Braunwald E et al. Heart Disease. 2001.

Planning for Delivery

Caesarean section recommended:

- Obstetrical reasons
- Oral Anticoagulation [Failure to switch from warfarin to heparin 2 weeks before labour] (Avoid forceps, use vacuum/suction devices)
- Heart failure
- Severe fixed obstructive cardiac lesions
  - Avoid vasodilation (reduced preload) with epidural anesthesia
- Marfan's with dilated aorta [>40mm] or aortic dissection
  - Avoid increased blood volume, aortic stress with contractions
- Severe pulmonary HTN

Second Stage Management

STAGES OF LABOUR		PHASES OF LABOUR			FUNCTIONAL DIVISION	MAIN EVENTS
STAGE	DESCRIPTION	PHASE	DESCRIPTION	SUB-CATEGORY		
First Stage	From onset of regular uterine contractions to full dilatation of the cervix	Latent	From onset of contractions until 3-5cm cervical dilatation		PREPARATORY DIVISION	(1) Uterine contractions of adequate frequency, duration and intensity (2) Cervical effacement
		Active	From 3-5cm to full (10cm) cervical dilatation	Acceleration phase		
				Phase of maximum slope	DILATATIONAL DIVISION	(1) Cervical Dilatation (2) Some fetal descent
				Deceleration phase		
Second Stage	From full cervical dilatation to delivery of the fetus	Passive	From full cervical dilatation until commencement of active pushing		PELVIC DIVISION	(1) Fetal Descent (2) Delivery of the fetus
		Active	From active pushing until delivery of the fetus			
Third Stage	From delivery of the fetus until delivery of the placenta					Delivery of the placenta

D'Souza & Sermer (unpublished)

Second Stage management:

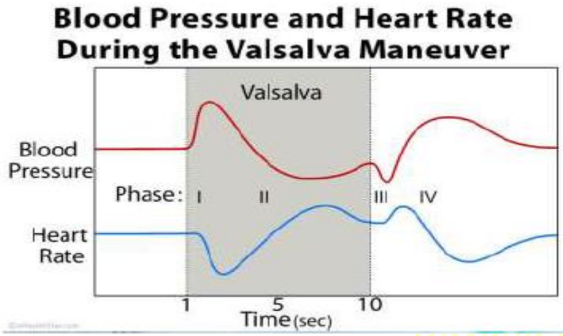
Active [pushing] vs Passive [no pushing] second stage

Prolonged Valsalva (active phase of second stage)

- Increase PA pressures, Increases R to L shunting
- Shunts: ASD, VSD, Tetralogy of Fallot, Eisenmenger's
- Increase heart rate, Hypotension

Chart 1. Valsalva Maneuver

Phase	1	2a	2b	3	4
Intrathoracic pressure	Increased	Increased	Increased	Normal	Normal
Mean arterial blood pressure	Increased	Decreased	Increase*	Decrease*	Increased
Heart rate	Decreased	Increased	Decrease*	Increase*	Decreased
Sympathetic activity	Decreased	Decreased	Increased	Increased	Increased
Parasympathetic (vagal) activity	Increased	Increased	Decreased	Decreased	Increased



Anaesthetic Management

	Single shot SA	Epidural anaesthesia	Continuous SA	Combined spinal-epidural anaesthesia	Saddle block
Left-to-right shunt	X	√	?	±	?
Aortic coarctation	X	±	?	?	?
Aortic stenosis	X	±	±	±	?
Idiopathic hypertrophic subarotic stenosis	X	±	±	±	±
Mitral stenosis	X	√	±	±	±
Aortic regurgitation	X	√	?	√	±
Mitral regurgitation	X	√	√	√	±
Unrepaired tetralogy of Fallot	X	±	±	±	?
Eisenmenger's syndrome	X	√	√	√	?
Primary pulmonary hypertension	X	±	±	±	±
Coronary artery disease	X	√	√	√	±

Antibiotic prophylaxis

Puerperal bacteremia :

per patient: 3.23%

per blood culture: 0.4 - 7%

asymptomatic

2.15% of 1116 patients

infective endocarditis after vaginal delivery:

0.09% (2 in 2165 patients) (manual removal of placenta)

Sugrue et al Br Heart J 1980

Antibiotic Prophylaxis

NICE guidelines 2008

Identifying cardiac risk factors

Regard patients with the following cardiac conditions as being at risk of developing infective endocarditis:

- Acquired valvular heart disease with stenosis or regurgitation
- Valve replacement
- Structural congenital heart disease (including surgically corrected or palliated structural conditions but excluding isolated atrial septal defect, fully repaired ventricular septal defect or fully repaired patent ductus arteriosus, and closure devices deemed to be endothelialised)
- Previous infective endocarditis
- Hypertrophic cardiomyopathy.

Prophylaxis for patients at risk of infective endocarditis

- Antibiotic prophylaxis against infective endocarditis is not recommended in the following circumstances:
  - For patients undergoing dental procedures
  - For people undergoing non-dental procedures at the following sites: upper and lower gastrointestinal tract; genitourinary tract (this includes urological, gynaecological, and obstetric procedures, and childbirth); upper and lower respiratory tract (this includes ear, nose, and throat procedures, and bronchoscopy).

AHA guidelines 2007

Table 3. Cardiac Conditions Associated With the Highest Risk of Adverse Outcome From Endocarditis for Which Prophylaxis With Dental Procedures Is Reasonable

Prosthetic cardiac valve or prosthetic material used for cardiac valve repair
Previous IE
Congenital heart disease (CHD)*
Unrepaired cyanotic CHD, including palliative shunts and conduits
Completely repaired congenital heart defect with prosthetic material or device, whether placed by surgery or by catheter intervention, during the first 6 months after the procedure†
Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device (which inhibit endothelialization)
Cardiac transplantation recipients who develop cardiac valvulopathy

*Except for the conditions listed above, antibiotic prophylaxis is no longer recommended for any other form of CHD.

†Prophylaxis is reasonable because endothelialization of prosthetic material occurs within 6 months after the procedure.

Antibiotic prophylaxis solely to prevent IE is not recommended for GU or GI tract procedures.

Although these guidelines recommend changes in indications for IE prophylaxis with regard to selected dental procedures (see text), the writing group reaffirms that those medical procedures listed as not requiring IE prophylaxis in the 1997 statement remain unchanged and extends this view to vaginal delivery, hysterectomy, and tattooing. Additionally, the committee advises against body piercing for patients with conditions listed in Table 3 because of the possibility of bacteremia, while recognizing that there are minimal published data regarding the risk of bacteremia or endocarditis associated with body piercing.

Post-partum Care

Early post-partum period [48-72 hours] is potentially dangerous

Blood loss / volume depletion:

- Uterine haemorrhage
- Uterine atony

Fluid shift and retention / volume loading:

- Auto-transfusion
- Overzealous IV fluid administration

Cardiac perturbation and arrhythmia

Acute pulmonary hypertension

Thromboembolism

Infective endocarditis

Post-partum Care

Prevention of post-partum haemorrhage

Use of uterotonic drugs

- oxytocin – improve uterine contraction, vasodilation and arterial hypotension, ↑PVR
- ergometrine – arterial hypertension
- continuous infusion at lowest effective rate [oxytocin 8-12mU/minutes over 4 hour]
- avoidance of bolus or high dose

Attention to fluid and hydration status

Post-partum monitoring for 48-72 hours, up to 7 days [14 days in PAH]

Prevention of thromboembolism: meticulous leg care, early ambulation

Surveillance for puerperial infection

Resumption of cardiac medications [caution: breast feeding]

Cardiac Pregnancy Management Algorithm – ACHD QMH

Period	Maternal	Foetal
First trimester	Initial assessment Review of medication Planning follow-up arrangement	Assessment of risk of CHD recurrence
Second trimester	Follow-up once Echo at 26 – 29wks [if necessary]	Foetal USG 16 – 20 wks Foetal growth
Third trimester	Follow-up +/- echo 4-8 weekly prn Pre-delivery echo 4 wks before EDC Refinement of delivery plan	Foetal growth
First stage	Labour: spontaneous vs induced Analgesia Duration of 1st stage	Foetal distress Foetal maturity
Second stage	Duration of 2nd stage Vaginal [+/- assisted] vs CS Fluid management Cardiovascular support	
Third stage	Syntocinon administration Post-partum haemorrhage	
Post-partum	Risk of PHT, ventricular dysfunction Follow-up 1 month after delivery Echo at 3-6 months after delivery	Baby's well being Baby's heart

Cardiac Pregnancy Care Plan

Cardiac Pregnancy Care Plan Version Apr-2019

Queen Mary Adult Congenital Heart Clinic: Cardiac Pregnancy Care Plan [QMACH_CPCP]

Date of Care Plan drafted								
Name: **ID No:**								
Drafted by								
Drafted at gestation								
Expected Date of Delivery								
Underlying cardiac condition								
Potential cardiac events during pregnancy								
Pregnancy Risk Class	Modified WHO risk class					Level of Risk		
	I	II	II-III	III	IV	Low	Moderate	High
Contra-indication against pregnancy	Pregnancy *is* / *is not* absolutely contra-indicated							
First Trimester	Expectant antenatal visit by obstetrician would be adequate							
Second Trimester	Cardiac follow-up at: _____ weeks [Date: _____] Monitoring of foetal growth Foetal ultrasound to assess if any foetal cardiac malformation at 16-20wk							
Third Trimester	Clinical follow-up: _____ weeks [Date: _____] Echo at 1 month before EDD: _____ [to be arranged]							

Cardiac Pregnancy Care Plan

Peripartum period	Cardiac monitoring recommended during peripartum period: - ECG / SaO2 / non-invasive BP / arterial line monitoring / CVP monitoring Mode of delivery : - [ ] Trial of vaginal delivery [if cardiac condition remained stable] - [ ] Elective assisted delivery only - [ ] Elective LSCS *First stage:* - Spontaneous onset of labour: *allowed / NOT allowed* - Duration of the first stage: *standard / ≤12 hours* - Adequate analgesia is recommended - Prophylactic antibiotics against endocarditis: [ ] Not routinely recommended for vaginal delivery [to be considered if instrumentation might be anticipated] [ ] Indicated preferably at onset of labour *Second stage:* - Duration of second stage (active phase) : [ ] Standard [ ] Shortened (≤ 30 minutes) [ ] Elective assisted delivery only - Other actions: *Third stage:* - Syntocinon infusion: [ ] Standard [ ] Reduced dose: [ ] Loading [ ] Maintenance dose [ ] Avoid - Other actions
Post-natal period	High-dependency unit is recommended: Yes [ ] No [ ] Post-natal stay in hospital for _____ days To monitor for ventricular dysfunction , cardiac arrhythmia, BP Cardiac assessment prior discharge : ECG [ ] Echo [ ] Cardiac follow-up __1__ months post-delivery

Pregnancy in Women with (Congenital) Heart Disease

Medical & Health Care Practitioners
Cardiologist, Obstetrician, Anaesthetist

Paturients

Risk Assessment and Stratification

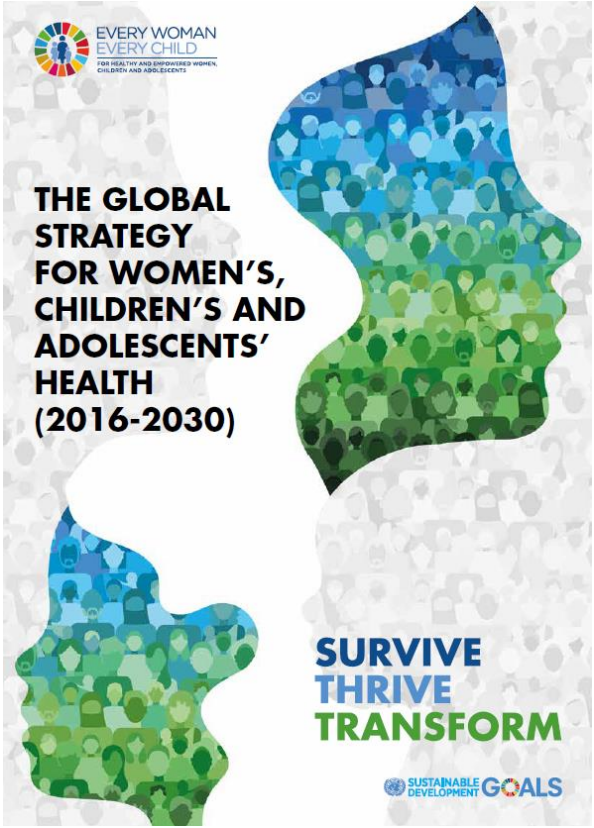
Risk Communication

Risk Perception

Treatment / Intervention

Counseling

Decision



LIFE COURSE	INTERVENTION PACKAGES	ENABLING ENVIRONMENT
Women's health	• sexual and reproductive health information and services; • nutrition; • management of communicable and non-communicable diseases; • screening and management of cervical and breast cancer; • gender-based violence prevention and response; • pre-pregnancy risk detection and management	**HEALTH SYSTEM ENABLERS** • policies for universal health coverage; sufficient and sustainable financing; • health workforce supported to provide good-quality care everywhere; • commodity supply; • health facility infrastructure; community engagement; • mainstreaming emergency preparedness; • human rights-, equity-and gender-based approaches in programming; • accountability at all levels
Pregnancy, childbirth and postnatal care	• antenatal care, • childbirth care; • safe abortion and post-abortion care; • prevention of mother-to-child transmission of HIV; • management of maternal and newborn complications; • postnatal care for mother and baby; • extra care for small and sick babies	
Child health and development	• breastfeeding; • infant and young child feeding; • responsive caregivingand stimulation; • immunization; • prevention and management of childhood illness and malnutrition; • treatment and rehabilitation of congenital abnormalities and disabilities	**MULTISECTOR ENABLERS** • policies and interventions in key sectors: finance and social protection; • education; • gender; • protection—registration, law and justice; water and sanitation; • agriculture and nutrition; • environment and energy; • labour and trade; • infrastructure, including facilities and roads; • information and communication technologies; • and transport
Adolescent health and development	• health education; • supportive parenting; • nutrition; • immunization; • psychosocial support; • prevention of injuries, violence, harmful practices and substance abuse; • sexual and reproductive health information and services; • management of communicable and non-communicable diseases	