Έγκυος και συγγενής καρδιοπάθεια: Τι πρέπει να γνωρίζει ο Καρδιολόγος Ενηλίκων

Αντώνιος Π. Βλάχος
Αναπληρωτής Καθηγητής Παιδοκαρδιολογίας
Πανεπιστήμιο Ιωαννίνων
• No conflict of interest
Pregnancy and CHD

- Hemodynamic changes: fall in systemic vascular resistance and increase in cardiac output
  - Systemic BP ↓ early in gestation usually 10 mmHg below baseline in the second trimester to a mean of 105/60 mmHg
  - A 30 to 50 percent increase in intravascular volume in normal pregnancy beginning at 12 to 14 weeks gestation and peaking early to mid-third trimester
  - Marked fluctuations in cardiac output occur during normal labor and delivery

- Risk of thromboembolism
  - Women at risk for thrombosis: prosthetic heart valves, atrial arrhythmias, Fontan connection, or previous thromboembolic events
Pregnancy and CHD

• ESC registry CHD the most prevalent form of structural heart disease (66 percent) affecting pregnancy outcomes worldwide

• Cardiac complications: 11% of completed pregnancies, with HF 5% and arrhythmias 4.5% the most common

• Cardiovascular events reported primarily in Eisenmenger patients and in those with palliated or unrepaired cyanotic heart disease

• Increases in the frequencies of specific congenital heart diseases, cardiac dysrhythmias, cardiomyopathy, and heart failure were observed with severe complications during hospitalizations for delivery among women with chronic heart disease more common in 2004 to 2006 than in 1995 to 1997
Risk scores

• WHO
• CARPREG
• ZAHARA
Table 6
Modified WHO classification of maternal cardiovascular risk: principles

<table>
<thead>
<tr>
<th>Risk class</th>
<th>Risk of pregnancy by medical condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>No detectable increased risk of maternal mortality and no/mild increase in morbidity.</td>
</tr>
<tr>
<td>II</td>
<td>Small increased risk of maternal mortality or moderate increase in morbidity.</td>
</tr>
<tr>
<td>III</td>
<td>Significantly increased risk of maternal mortality or severe morbidity. Expert counselling required. If pregnancy is decided upon, intensive specialist cardiac and obstetric monitoring needed throughout pregnancy, childbirth, and the puerperium.</td>
</tr>
<tr>
<td>IV</td>
<td>Extremely high risk of maternal mortality or severe morbidity; pregnancy contraindicated. If pregnancy occurs termination should be discussed. If pregnancy continues, care as for class III.</td>
</tr>
</tbody>
</table>

Modified from Thorne et al. 72

WHO = World Health Organization
### Conditions in which pregnancy risk is WHO I

- 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

### Conditions in which pregnancy risk is WHO II or III

**WHO II** *(if otherwise well and uncomplicated)*

- Unoperated atrial or ventricular septal defect
- Repaired tetralogy of Fallot
- Most arrhythmias

**WHO II–III** *(depending on individual)*

- Mild left ventricular impairment
- Hypertrophic cardiomyopathy
- Native or tissue valvular heart disease not considered WHO I or IV
- Marfan syndrome without aortic dilatation
- Aorta <45 mm in aortic disease associated with bicuspid aortic valve
- Repaired coarctation

### WHO III

- Mechanical valve
- Systemic right ventricle
- Fontan circulation
- Cyanotic heart disease (unrepaired)
- Other complex congenital heart disease
- Aortic dilatation 40–45 mm in Marfan syndrome
- Aortic dilatation 45–50 mm in aortic disease associated with bicuspid aortic valve

### Conditions in which pregnancy risk is WHO IV *(pregnancy contraindicated)*

- Pulmonary arterial hypertension of any cause
- Severe systemic ventricular dysfunction (LVEF <30%, NYHA III–IV)
- Previous peripartum cardiomyopathy with any residual impairment of left ventricular function
- Severe mitral stenosis, severe symptomatic aortic stenosis
- Marfan syndrome with aorta dilated >45 mm
- Aortic dilatation >50 mm in aortic disease associated with bicuspid aortic valve
- Native severe coarctation
Risk factors

• Pulmonary hypertension (pulmonary vascular disease)
• Maternal cyanosis
• Poor maternal functional class
• History of arrhythmia
• Maternal anticoagulants
• NT-proBNP value >128 pg/mL → ZAHARA II
Obstetrical issues

• Miscarriage
• Pregnancy induced hypertension, preeclampsia, eclampsia
• Premature labor
• Postpartum hemorrhage
• Thromboembolic events
• Endocarditis prophylaxis
ASD

unrepaired

- supraventricular arrhythmias
  - atrial fibrillation or atrial flutter
  - right ventricular failure

- risk of paradoxical embolization
  - leg or pelvic veins

- acute blood loss
  - augments the left-to-right shunt

repaired

- low risk pregnancy

atrial arrhythmias
VSD

small VSDs
repaired VSD - no significant residua
No risk

Qp:Qs <1.7
normal PAp
preserved functional aerobic capacity
↓ risk

larger VSD shunts
arrhythmias
ventricular dysfunction
pulmonary hypertension
↑ risk

VSD and
moderate or more pulmonic stenosis
DCRV
LVEF<40%
↑ risk

Eisenmenger
contraindicated
Eisenmenger syndrome pregnancy is **contraindicated**

Eisenmenger syndrome
- maternal mortality from 30 to 50 percent
- has not changed significantly in the past 50 years
- mortality risk cannot be predicted in an individual patient

The majority of maternal deaths occur during or in the first week after delivery

Most deaths are due to
- thromboembolism
- volume depletion -> ↑ R→L shunt
  - intense cyanosis
  - preeclampsia
- a sudden increase in systemic vascular resistance may fatally reduce cerebral blood flow
Cyanotic CHD

- Unrepaired TOF
- TOF/PA with AP collaterals,
- Some single-V conditions
- TA
- Ebstein's anomaly with RtoL via an ASD,
- ccTGA with VSD or ASD

- Arrhythmias, HF, CVD
- Fetal issues
  - Prematurity
  - Fetal mortality
  - Fetal CHD
- maternal hemoglobin ≥20 g/dL: live birth rate of 8%
- maternal arterial oxygen saturation ≤85 percent: live birth rate of 12%
Specific LVOT lesions

• BAV high risk if Ao root >45mm
• Marfan extremely high risk if Ao root >40mm
  • Acceleration of Ao root dilation after pregnancy
• Beta – blocker administration
Congenital AS

• Bicuspid severe AS: delay conception until the AS has been addressed
• Valve intervention prior to pregnancy for asymptomatic patients with severe AS
  • Intervention for PG cath >50 mmHg
  • ↓ risk for repaired BAV/AS and mean gradient < 25mmHg
• If BAV/AS early in pregnancy with severe AS and/or a LVEF < 40% consider termination of pregnancy followed by reparative surgery before another attempt at pregnancy.
• If severe AS and no or only mild symptoms then conservatively with bed rest, beta blockers, and oxygen. Intervention during pregnancy is recommended only for refractory New York Heart Association (NYHA) class III or IV
CoA

• Unrepaired coarctation during pregnancy
  • If normotensive: pregnancy to term
  • repair of the coarctation a few weeks after delivery
  • if ↑ blood pressure during pregnancy: intervention is recommended
  • surgery and stent placement during pregnancy should be considered by a multidisciplinary team
  • For stent placement: procedure preferably after the second trimester with abdominal shielding

• Repaired coarctation repair usually uncomplicated pregnancy
  • rate of miscarriage and preeclampsia are higher than in the general population
Repaired TOF

WHO II

Degree of PR

RV volume
RVESV 80-85 ml/m²

RV function

RVEDV 160-170 ml/m²
D-TGA

- Mustard or Senning
  - morphologic RV → systemic
  - PAP level
  - conduction and rhythm abnormalities

Jatene after 1982
- successful pregnancies

L-TGA
- AVVR
- development of HF
- risk of heart block → spontaneous

Data not yet available: degree of RV dysfunction that imposes ↑ pregnancy risk
PS or ↑ PVR:
balanced i.e. adequate, but not excessive pulmonary blood flow

↑ PVR: ↑↑ risk

Single-V unrepaird

Pregnancies reported in TA

Single-V repaired

Satisfactory systemic ventricular function

Normal sinus rhythm

High rates of spontaneous abortion and preterm deliveries observed

Could tolerate pregnancy but ↑ complication rate

? Alteration in placental blood flow caused by Fontan physiology

PS: successful reported
Evaluation

• Preconception
  • detailed history, prior interventions
  • physical exam
  • 12-lead electrocardiogram
  • ECHO
  • functional status (which may include exercise testing)

• ECHO
  • detailed anatomy
  • ventricular size and function
  • assessment of valve function
  • pulmonary pressures
Pregnancy in Patients With Congenital Heart Disease: A Contemporary Challenge
Despina Ntiloudi, MD, MSc, Thomas Zegkos, MD, MSc, Athanasios Koutsakis, MD, George Giannakoulas, MD, and Haralambos Karvounis, MD

Clinical factors:
- NYHA class
- Cyanosis

Medical History
- Heart failure
- Thromboembolism
- Arrhythmia
- Smoking
- Mechanical valve

CHD subtype:
- Pulmonary arterial hypertension
- Mitral stenosis: Severe?
- Aortic stenosis: Severe?
- Symptomatic?
- Aortic Coarctation: Native? Severe?
- Marfan syndrome: aorta dilatation?
- BAV: aortic dilatation?
- Cyanotic disease

Echocardiographic factors:
- Systemic ventricular EF
- Mitral valve area
- Aortic valve area
- Peak LVOT Gradient
- Subpulmonary ventricular dysfunction
- Atrioventricular valve regurgitation

Blood Test/ Other :
- NT-pro BNP
- CPET

Cardiology in Review 2017;25: 326–330
TRANSITION TO ACHD