Pregnancy and cyanotic congenital heart disease

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I have nothing to disclose.
Maternal mortality is increasing 70% of pregnancies with cardiac maternal disease.
Mortality and morbidity is from 8 to 100 folds higher between patients with CHD

Pregnant women with congenital heart disease

- Placental development
  - Low resistance

- Change in heart rate
  - Increase in systolic volume

- Increase in plasmatic volume

- Decrease in systemic vascular resistance (SVR)

- Increase in cardiac output

- Volume overload

- Increased right to left shunt

- PAH inability to increase pulmonary blood flow

- Precipitates heart failure
Pregnant women with congenital heart disease

Roos-Hessenlink et al; Heart 2009: 95: 680-86;

IVC COMPRESSION
CHAMBER AND AORTA DILATATION
THROMBOSIS RISK X 6 DURING PREGNANCY
Can we predict the risk in CHD

From prospective studies...

- FC > II o cyanosis
- LVOTO > 30 mmHG peak
- Previous event
- Systemic EF < 40%

From retrospective studies specific por CHD...

- Mechanical prosthesis
- Significant L/RAV regurgitation
- Severe PR
- Cardiac medication
- Complex CHD
- RV dysfunction

Khairy et al., Circulation 2006; 113:517-24
Drenthen et al., EHJ 2010;
Pregnant women with congenital heart disease

**Risks for the mother**
- Arrhythmia
- Heart Failure
- Mortality

**Risks for the fetus and neonate**
- Transmission CHD
- Preterm SGA
- Mortality

**Long term implications**

**pregnancy**

**delivery**

**postpartum**
Pregnancy and cyanotic congenital heart disease

Heart failure 20%
Increasing cyanosis
Cardiovascular event including stroke and mortality
Less than 5% in cyanotic without PAH vs 35% in Eisenmenger

Drenthen W.et.al. JACC 2007;49: 2303-11
Cyanotic congenital lesions without PAH

**SPECIFIC SERIES ON CYANOTIC PATIENTS WITHOUT PAH**

**Presbitero** N= 96 pregnancies in 44 patients  
Cardiac complications 32% maternal mortality 2%

*Presbitero P, Circulation 1994 :89*

<table>
<thead>
<tr>
<th></th>
<th>N of patients</th>
<th>N of cyanotic patients</th>
<th>Cardiac complications cyanotic</th>
<th>N of Eisenmeger</th>
<th>Mortality on Eisenmenger</th>
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</thead>
<tbody>
<tr>
<td><strong>Avila</strong></td>
<td>191</td>
<td>49</td>
<td>20%</td>
<td>7</td>
<td>50%</td>
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<td><strong>Manso</strong></td>
<td>83</td>
<td>5</td>
<td>60%</td>
<td>2</td>
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<td><strong>Fesslova</strong></td>
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<td><strong>Aggarwal</strong></td>
<td>196</td>
<td>24</td>
<td>33.3%</td>
<td>8</td>
<td>25%</td>
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</table>

*Avila Clin Cardiol 2003; 26:135-142  
Manso, Rev Esp Cardiol 2008;61:(3) 236-43  
Fesslova V, IJC 2009 131:257-264  
Aggarwal , ANZOG 2009:49:376*
SPECIFIC SERIES ON EISENMENGER

Avila n= 12 patients Cardiac complications: mortality 25%; increased cyanosis and functional class in all

Weiss n=73 retrospective review 1976-1996
Maternal mortality 36% 88.5% during postpartum period

Bédard n=73 retrospective review comparing Weiss series vs 1997-2007
Maternal mortality decreases from 36 to 28%

Has there been any progress made on pregnancy outcomes among women with pulmonary arterial hypertension?

Elisabeth Bédard¹,², Konstantinos Dimopoulos¹,², and Michael A. Gatzoulis¹,²*
New advanced therapies can help?

- Sildenafil use during pregnancy reported
- Prostaclicin inhaled or i.v.
- Bosentan not used due to teratogenic effect in animals

Recent reports on pregnancy and PAH, all patients were on medication (sildenafil +/- ilprost inhaled)
Pregnant women with congenital heart disease

WHO III

Significantly increased risk of maternal mortality or severe morbidity. Expert counselling required.

WHO IV

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.
As soon as pregnancy is confirmed
Counseling prior to pregnancy—Eisenmenger offer interruption

Complete cardiac assessment
02 Saturation
Medication
Clinic and echocardiographic

Drenthen W.et.al. JACC 2007;49: 2303-11
Regular fup in cardiology including Sat
Echo: increased velocity and gradients
Weight control and hypertension

Early admission when functional class deteriorates
or O2 saturation drops
Bed rest and O2 supplementation

Low molecular heparin thromboprophylaxis
Indicated if admission and peripartum period
No consensus during all pregnancy
Recurrence risk

- Global risk of transmission of 3.7%

  AVSD 7-12%
  Tetralogy of Fallot 5%
  TGA 0.5%

NT thickness and normal cariotip 25% CHD

Autosomal dominant

22q11 Deletion

“De novo” mutations Risk transmission 50%

Burn J Lancet 1998;351: 311-316
Oyen N Circulation 2009; 120: 295-301
Obstetric care during pregnancy

- Risks for the mother
- Risks for the fetus

**Obstetrics**
- ev 3-4 w until 26
- 2-3 weeks de 26-36
- Once w 36-partum

**Anesthesiology**
- 32-34w

**IUGR**
- 28 w-monthly
- 28-34 FHM
- Atosiban if PROM

**SGA**
- Preterm delivery

**Offspring mortality**

*Drenthen W.et.al. JACC 2007;49: 2303-11*
Pregnant women with congenital heart disease

- **Delivery**
  - Careful plan management
  - **Cardiology**
  - **Obstetrics**
  - **Anaesthesiology**

- **Minimize pain and anxiety**
- **Monitoring, fluid balance**
- **Left lateral position**

- **Spontaneous onset of labour**
- **Limit 2nd stage delivery**

**Epidural**

**Cardiac C-section:**
- Heart failure
- Warfarine
- “Aortic patients”

Caesarean delivery should be considered for obstetric indications or for patients with dilatation of the ascending aorta >45 mm, severe aortic stenosis, pre-term labour while on oral anticoagulants, Eisenmenger syndrome, or severe heart failure.
"Autotransfusion"
Increase in CVP

Heart failure

Thrombosis risk

Postpartum haemorrhage

Steer P, Gatzoulis MA "Pregnancy And Heart Disease"
Postpartum care of women with CHD

Delivery

Careful plan management

Cardiac environment
CCU

LMWH thromboprophylaxis

Oxitocine perfusion

Monitoring ECG, BP, CVP

- If breastfeeding inhibition: caution with hemodynamic changes

Weiss et al; JACC 1998
25% with high risk pregnancy were not aware of it
Ask actively in the outpatient clinic

Kovacs et al J Am Coll Cardiol 2008; 52:577-86
EHJ 2011 :32, 3147
Preconceptional assessment

Information to the patient including mortality and morbidity

In cyanotic patients:
• O2 saturation at rest
• Exercise test and decrease in saturation
• 22q genetic test in pulmonary atresia

WHO 4: Risk of combined hormonal contraceptives for different cardiac conditions

• Mechanical valves: Starr Edwards; BjorkShiley; any tricuspid valve
• Ischaemic heart disease
• PAH of any cause
• LVEF 30%
• Fontan circulation
• Cyanotic heart disease
• Previous arteritis

• Progesterone only pill
• Subdermal implant
• Intrauterine device impregnated

EHJ 2011;32, 3147
Thorne et al;Heart 2006;92:1520
Repaired cyanotic congenital heart disease

Tetralogy of Fallot

- More frequent cyanotic congenital heart disease repaired
- High percentage reach the adult life in a good condition
- Considered generally WHO II for complications

RV dysfunction

Severe PR

Pulmonary regurgitation considered risk of complications

Controversy regarding PVR prior to pregnancy

Kbairy et al., Circulation 2006; 113:517-24
Repaired cyanotic congenital heart disease

**Tetralogy of Fallot**

- PVR indicated prior to pregnancy if severe RV dilatation or symptoms
- Complications described during pregnancy are managed medically
- 10% CV complications: HF/Arrhythmia

BPS, reduced RVEF
RVH, restrictive physiology?
Previous arrhythmia, cardiac medication

<table>
<thead>
<tr>
<th>SERIES IN TETRALOGY</th>
<th>Pedersen</th>
<th>Meijer</th>
<th>Greutman</th>
<th>Balci</th>
<th>Khairy</th>
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<tbody>
<tr>
<td>N pregnancies</td>
<td>54</td>
<td>63</td>
<td>76</td>
<td>127</td>
<td>18</td>
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<tr>
<td>% Severe PR</td>
<td>?</td>
<td>28%</td>
<td>40%</td>
<td>56%</td>
<td>100%</td>
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<tr>
<td>Cardiac complications</td>
<td>0%</td>
<td>12%</td>
<td>9.6%</td>
<td>8%</td>
<td>27%HF 16%Arr</td>
</tr>
</tbody>
</table>

Meijer JM. et al Heart 2005;
Pedersen 2008 Cardiol Young 2009;
Greutmann M EHJ 2010;
Balci AJC 2010
Pregnancy and CHD

Pregnancy and heart disease program 2007 n=185; 70% CHD

Pregnancy and CHD patients
1994-2006 n=83
2007 n=127

2 pregnancies with PAH and CHD in early times
1 maternal mortality in peripartum period: premature partum 28 weeks
1 urgent C-section 32w mother and baby ok still alive

Cyanotic and congenital heart disease 5 pregnancies
2 spontaneous abortion
3 pregnancies: univentricular heart with pulmonary stenosis well balanced Sat 88-90%
  No maternal complications
  IUGR weight above 2000 no CHD transmission
LMHW 3T and peripartum
Repaired cyanotic congenital heart disease

**Ebstein’s anomaly**

- 56 women 111 pregnancies no mortality
- Functional class and presence of cyanosis
- Previous arrhythmia
- Prematurity and IUGR related to cyanosis and limited CO

**Transposition of great arteries**

- Arrhythmia
- De novo “leaks”
- RV dilatation and HF
- Pregnancy induced HT

**Systemic RV**

- Drenthen W, EHJ 2005;
- Guedes A et al., JACC 2004;
- Connolly HM JACC 1994;
Repaired cyanotic congenital heart disease

Small ASD or PFO

Severe right ventricular dysfunction

ESC 2011 FOCUS Session
Session title: Pregnancy and heart disease
• Inform regarding risks for the mother

  Complications during pregnancy/postpartum
  HF/Increasing cyanosis
  Including reference to mortality 30% in PAH

• Inform risk for the fetus

  High risk of miscarriage/Transmission/Prematurity/SGA
Conclusions

Pregnancy

- Care at tertiary center multidisciplinary approach
  - Exercise restriction
  - Supplementary 02 if admission
  - LMWH
  - advanced therapies in PAH?

- Fetal echo
- Plan of Delivery: vaginal partum with shortening 2nd stage in cyanotics and C-section for Eisenmenger
- Plan of Postpartum: cardiac environment
THANK YOU!!!