pregnancy in tetralogy of fallot

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Fallot is the commonest cyanotic congenital heart condition.
branch PA stenosis

right aortic arch

anomalous coronaries

ASD

VSD
‘total correction’, Lillehei, 1954
branch pulmonary artery stenosis
residual VSD
PS/PR
LV dysfunction
endocarditis
atrial arrhythmias
ventricular arrhythmias
RV dilatation and dysfunction
aortic root dilatation/AR/dissection
residual VSD
RV dilatation and dysfunction
When should you replace the pulmonary valve?

Severe PR with

- Symptoms
- Deteriorating CPET
- RVEDV >150-170 mls/m²
- RVESV >82-90 mls/m²
- Decreasing RV function

Oosterhof T Circulation 2007
Ferraz Calvacanti PE JACC 2013
pulmonary valve replacement
heart disease is the **leading** cause of death in pregnancy.....
MBRRACE-UK Mothers and Babies: Reducing Risk through Audits and Confidential Enquiries across the UK
Table 9.2. Causes of maternal death from cardiac disease; UK: 1994–2008

<table>
<thead>
<tr>
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<tbody>
<tr>
<td><strong>Acquired</strong></td>
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<tr>
<td>Aortic dissection</td>
<td>7</td>
<td>5</td>
<td>7</td>
<td>9</td>
<td>7</td>
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<tr>
<td>Myocardial infarction (MI)</td>
<td>6</td>
<td>5</td>
<td>8</td>
<td>12</td>
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<td>Ischaemic heart disease (no MI)</td>
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<td>0</td>
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<td>Sudden adult death syndrome (SADS)</td>
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<td>3</td>
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<tr>
<td>Peripartum cardiomyopathy</td>
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<td>7</td>
<td>4</td>
<td>0*</td>
<td>9**</td>
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<td>Other cardiomyopathy</td>
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<td>3</td>
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<td>4</td>
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<td>Myocarditis or myocardial fibrosis</td>
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<td>Mitral stenosis or valve disease</td>
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<td>Thrombosed aortic or tricuspid valve</td>
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<tr>
<td>Infective endocarditis</td>
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<td>1</td>
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<tr>
<td>Right or left ventricular hypertrophy or</td>
<td>1</td>
<td>2</td>
<td>2</td>
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<tr>
<td>hypertensive heart disease</td>
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<td></td>
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<tr>
<td><strong>Congenital</strong></td>
<td></td>
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<td></td>
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<tr>
<td>Pulmonary hypertension (PHT)</td>
<td>7</td>
<td>7</td>
<td>4</td>
<td>3</td>
<td>2</td>
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<tr>
<td>Congenital heart disease (not PHT or thrombosed aortic valve)</td>
<td>3</td>
<td>2</td>
<td>2</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td><strong>Other</strong></td>
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<td>2</td>
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<td>0</td>
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<tr>
<td><strong>Total</strong></td>
<td>39</td>
<td>35</td>
<td>44</td>
<td>48***</td>
<td>53</td>
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*Twelve Late deaths reported in 2003–05.
**Two Late deaths reported in 2006–08.
***Includes one woman for whom information on cause was not available.
2/3 patients have congenital heart disease
lots of women with CHD get pregnant
predictors of maternal events in pregnancy

NYHA Class > II cyanosis
previous cardiac event
left heart obstruction (mitral/aortic stenosis)
impaired systemic ventricular function (EF < 40%)
severe pulmonary regurgitation and poor RV mechanical valve
dilated aorta

Drenthen W et al, ZAHARA. Eur Heart J. 2010 Sep;31(17):2124-32
Khairy P et al. Circ 2006;113(4):517-24
and in fallot?

cardiac events 7-12%

arrhythmias (SVT)
RV impairment
heart failure

previous arrhythmia x9 risk

use of cardiac meds x12 risk

Veldtman GR et al. JACC 2004, 44 (1)
predictors of fetal events in pregnancy

- NYHA Class > II
- cyanosis
- left heart obstruction (mitral/aortic stenosis)
- smoking
- anticoagulants
- severe pulmonary regurgitation and poor RV
- mechanical valve
- dilated aorta

Khairy P et al. Circ 2006;113(4):517-24
and in Fallot?

- mortality ≈6%
- prematurity ≈6%
- SGA ≈9%
- recurrence 2-10%

unrepaired and ≥mod PR predict low birthweight

use of cardiac meds x8 risk

Veldtman GR et al. JACC 2004, 44 (1)
think 22q11
digeorge syndrome/catch 22/velocardiofacial syndrome/schprintzen syndrome

Cardiac abnormalities (congenital heart defects - 75%)

Abnormal facies

Thymic aplasia

Cleft palate (70%)

Hypocalcemia

22q11.2 microdeletion

uncorrected fallot is **high risk**

<table>
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<tr>
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<th>uncorrected</th>
<th>corrected</th>
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<tbody>
<tr>
<td>cardiac complications</td>
<td>40%</td>
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<tr>
<td>obstetric complications</td>
<td>70%</td>
<td>40%</td>
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<tr>
<td>spontaneous abortion</td>
<td>37.5%</td>
<td>0</td>
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<tr>
<td>preterm birth</td>
<td>25%</td>
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</table>

unrepaired previous event RV dysfunction ≥ moderate PR cardiac medication
fetal echo
see once per trimester (more if RV dysfunction)
local delivery if possible
low threshold for epidural
short active 2\textsuperscript{nd} stage
epidural anaesthesia higher shorter active 2\textsuperscript{nd} stage

is the **heart** affected by the pregnancy?

**NO**

19 repaired TOF v 38 controls

**no effects** on RV volumes, aortic dimensions, or exercise data

**YES**

13 repaired TOF v 26 controls

increase of RVEDV higher v controls (4.1 vs. 1.6 ml/m(2)/year, p=0.07).

no change in EF

Egidy Assenza G et al.. Int J Cardiol. 2013;168(3):1847-52

Cauldwell M et al. JAHA 2017, 6 (7)
Regional referral pathway for pregnant women with known cardiac disease

- All women with known cardiac disease should have received pre-conception counselling by a cardiologist (ACHD Cardiologist in the case of congenital cardiac disease).
- For further information on specific management, please refer to the ESC Pregnancy Guidelines

Congenital Cardiac Disease

**Low risk**
- Patent Ductus Arteriosus (isolated without pulmonary hypertension)
- Moderate mitral/aortic regurgitation
- Mild mitral/aortic stenosis
- Total anomalous pulmonary venous drainage repair
- Bicuspid aortic valve, no aortopathy
- Repaired ASD/VSD

**Do not usually require tertiary assessment.**
- Assessment by consultant cardiologist in local centre may wish to discuss with ACHD cardiologist
- Will require fetal echocardiogram (except PDA)

- **Moderate risk**
  - Repaired coarctation with no sequelae
  - Repaired Fallot’s
  - Severe pulmonary stenosis/regurgitation
  - Bicuspid aortic valve aortopathy < 40mm

- Initial assessment by local consultant cardiologist. Referral or discussion with ACHD cardiologist recommended. Likely to receive antenatal and intrapartum care locally.
- A locally devised multidisciplinary plan should be available in the patient’s maternity
involve **senior clinicians** from the **multidisciplinary team**
symptoms? medication? family history?

echo CPET MRI

do they need a PVR pre-pregnancy?

maternal and fetal risks?
progesterone methods are safe
WHO II
small increased risk of maternal mortality
or
moderate increase in morbidity
Overview

The Heart Disease and Pregnancy website provides information for patients and physicians about the risks associated with pregnancy and management strategies for pregnant women with heart disease trying to have a baby. Included are educational materials on heart conditions.