Cardiology Update:
Confidential Enquiry into Cardiac Maternal Deaths

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First detailed UK confidential enquiry review of cardiac maternal mortality since 2009
Global Maternal Mortality

Maternal mortality ratio (per 100,000 live births), 2013

UK 8.5 per 100,000 maternities

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Data Source: World Health Organization
Map Production: Health Statistics and Information Systems (HSI)
World Health Organization

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Causes of maternal death 2012-14

8.5 women per 100,000 died during pregnancy or up to six weeks after giving birth or the end of pregnancy in 2012 - 14

2 women per 100,000 died from heart disease
Causes of maternal death 2012-14

- Cardiac disease
- Sepsis*
- Neurological
- Other indirect
- Thrombois & thromboembolism
- Psychiatric**
- Amniotic fluid embolism
- Haemorrhage
- Early pregnancy deaths
- Indirect malignancies
- Anaesthesia
- Pre-eclampsia
Why is Heart Disease Dangerous in Pregnancy?

Can your heart do this?

Who is most at risk?

- Unable to increase cardiac output
- Impaired systolic ventricular function
- Severe mitral stenosis
- Severe aortic stenosis
- Coronary artery disease
- Pulmonary arterial hypertension

- Mechanical valves
- Aortopathy
• 77% of the women who died were not known to have heart disease before their pregnancy.

• Most of the women who died from heart disease did not get to see a cardiologist

So —

A&E and Acute Medicine services must make the life-saving diagnosis
Cardiac Causes of Maternal Death

Percent of Cardiac Deaths

50% - improved care may have made a difference to outcome

- 31% SADS MNH
- 22% Ischaemia
- 18% Myocardial disease
- 14% Aortic dissection
- 7% Valve disease
- 5% Pulmonary artery hypertension
- 4% Systemic hypertension
- 7% Congenital heart disease
Sudden Arrhythmic Death with Morphologically Normal Heart

- SADs MNH
  - Sudden unexplained death
  - Diagnosis of exclusion – cannot be made until autopsy
  - No cause found at autopsy, normal heart

- Incidence probably same as non-pregnant population

- Presume inherited arrhythmic condition e.g., channelopathy

- Autopsy tissue should be saved for genetic testing
- Family should be referred to inherited cardiac condition clinic
Deaths Due to Myocardial ischaemia

• ~50% atheroma, ~30% dissection

• Majority died suddenly at home, diagnosis made at autopsy

• Some had chest pain and sought medical help in the days before death

• Most with atheromatous disease had risk factors:

  Older age
  Smoking
  Obesity
  Diabetes
  Hypertension
  Family history of premature coronary disease
  Hypercholesterlaemia
Ischaemia - Vignette

- Older, hypertensive smoker, 34 weeks pregnant ➔ A&E
- Intermittent chest pain for 3 days, ➔ back and left arm
- No history for CVS risk factors taken
- Abnormal ECG
- No further Ix, obstetric team not contacted
- Discharged home
- Found dead next day
- Autopsy - extensive coronary artery atherosclerosis, thrombosed left anterior descending artery
Ischaemia – Messages For Care

• Severe chest pain requires a positive diagnosis, not simply exclusion of one cause
• Investigation for myocardial ischaemia is the same as in the non pregnant patient
  – History of pain
  – Serial ECGs
  – Serial troponin
  – Urgent cardiology review
Myocardial Disease

- Peripartum cardiomyopathy
- Dilated cardiomyopathy
- Left ventricular hypertrophy
- Myocarditis
- Obesity cardiomyopathy
- Hypertrophic cardiomyopathy
- Arrhythmogenic right ventricular hypertrophy

- Many causes of ventricular dysfunction
- All cause inability to ↑ cardiac output
  ➔ Arrhythmia & sudden death
  Pulmonary oedema & low cardiac output state
Myocardial Disease - Vignette

- A woman presented to A&E D3 post partum
- Breathless, palpitation, pink frothy sputum
- Anxious ++, tachycardic, tachypnoeic, unable to lie flat, dull bases, SaO2 85%
- Juniors’ ΔΔ chest infection
  - panic attack
  - pulmonary embolus
- Consultant review: cardiac?
- No echo available outside normal working hours
- Deteriorated, cardiac arrest
- Post resuscitation echo – severely impaired ventricular function
- Increasingly unstable; died following day
Myocardial Disease – Messages for Care

• Indicators of cardiac disease:
  – Orthopnea
  – Paroxysmal nocturnal dyspnoea
  – Pink frothy sputum = pulmonary oedema

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  *Cardiology review, chest XR and echo without delay*
Aortic Dissection

- 21 deaths (14%)
- All but one involved the ascending aorta
- Only 2 known to have high risk aortic disease pre pregnancy
- Timing of death
  - 1/3 3rd trimester
  - 2/3 post delivery

Missed opportunities

- 5 had antenatal FH of aortopathy or sudden death, not investigated
- 42% presented with symptoms indicative of severe aortic pathology in the days before their death
- 2 presented with neurological symptoms due to dissection of entire aorta
- 58% may have had a different outcome with improvement to care
Aortic Dissection - Vignette

- A woman presented to A&E in the middle of the night c/o sudden onset severe chest and neck pain
- Pregnancy was complicated by disabling symphysis pubis dysfunction
- Anxious, described severe pain starting in the neck and radiating in waves to the chest and back. She was clear that the pain was different to the ‘all over pain’ of her symphysis pubis dysfunction.
- Junior doctors’ ΔΔ pulmonary embolism anxiety attack
- Sent home, no investigations
- Sudden death at home 2 days later
- Autopsy: dissection of entire aorta, extensive cystic medial necrosis.
Aortic Dissection – Messages for Care

• Severe localising pain with anxiety is typical of aortic dissection

• Severe chest pain requires a positive diagnosis
  Any woman who leaves her baby to attend A&E at night requires a positive diagnosis

• Tired and emotional patients can have pathology

• Don’t withhold investigations or treatment because of pregnancy
Aortic Dissection - Vignette

- Woman found dead in bed in the third trimester

**Autopsy:**
- Ascending aortic dissection with haemopericardium and cystic medial necrosis and bicuspid aortic valve
- Non-Marfanoid dysmorphic features: clubbed feet, cleft palate, long fingers and unusual facies.
- Fetus had same dysmorphic features
- Her syndrome remained undiagnosed, tissue not saved for DNA analysis, family not referred for genetic screening.

Classical Loeys Dietz syndrome
Aggressive autosomal dominant familial aortopathy
Deaths from valvular disease

Predictors of poor outcome:
- Severe aortic or mitral stenosis
- Mechanical valve

2009-2014 - 11 deaths from valvular heart disease
Mechanical Valves in Pregnancy


All pregnancies in women with mechanical valves 2013-2015, N = 58

Maternal mortality 9%  valve thrombosis, CVA
Maternal morbidity 41%  31% haemorrhage, 10% valve thrombosis, CVA
Fetal demise 21%
Fetal morbidity 26%

**Risk factors**

Poor compliance
Poor monitoring and dose adjustment
Valve site/number

Poor maternal outcome: AV 39% vs MV 57% vs AV & MV 75%

- **NO IDEAL REGIME: MONITOR & TITRATE DOSE**
- **REFER FOR EXPERT CARE**
Mechanical Valve - Vignette

- Unplanned pregnancy in woman with mechanical valve
- Converted to LMWH, initially only once daily, too low dose
- Requested termination because she knew pregnancy high risk
- Presented to A&E the day before TOP
  - Breathless, unable to lie down, pink frothy sputum
  - Tachycardic, tachypnoeic, hypoxic
- Ventilated and treated for chest infection
- Echo thought to show right heart strain, Ix for PE negative
- Possibility of valve thrombosis not considered
- 4 days later, thrombosed mitral valve diagnosed
- Transferred to tertiary centre, but died
Mechanical Valves – Messages for Care

High maternal and fetal morbidity & mortality
  • Due to valve thrombosis, haemorrhage, stroke
  • Need specialist pre pregnancy counseling
  Warfarin vs LMWH
  • Need meticulous multidisciplinary specialist pregnancy care
    Ensure compliance
    Dose adjusted anticoagulation
Pulmonary Arterial Hypertension

• Highest risk cardiovascular condition in pregnancy
  - independent of underlying cause

• Risk of death
  40% without specialist care
  20-30% with specialist care

• With counselling, most women decide against pregnancy

Kiely 2010
Pulmonary Arterial Hypertension - Vignette

- First generation ethnic minority woman, first pregnancy
- Unexplained collapse during pregnancy, not investigated
- On day of delivery
  - SOB & ↓BP
    - continuous murmurs, ECG- RVH
- 2 day delay – cardiology SpR refused to review patient before echo performed
- Echo - severe pulmonary arterial hypertension secondary to patent arterial duct
- 2 week delay before transfer to regional pulmonary hypertension centre
  - selective pulmonary vasodilator therapy
  - continued to deteriorate and died.

Followed natural history of PAH in pregnancy
Pulmonary Arterial Hypertension – Messages For Care

- Unexplained collapse in pregnancy should be investigated
- Cardiology review should not be delayed in a sick patient
- Cyanosis more difficult to detect in non-white people
- Early access to specialist PAH care is likely to improve outcome
Congenital Heart Disease

Unlike the other groups, almost 2/3 were known to have congenital heart disease

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Opportunity for appropriate counselling and care
Increasing numbers of women with complex congenital heart disease have pregnancies, no increase in deaths since 1970s – is this a result of good pre pregnancy counselling and pregnancy care?

Pre pregnancy counselling and contraception should be offered to all teenagers and women with heart disease – both in the adult and paediatric transition services.
Key Messages

• Maternal mortality improving but -
  Deaths from heart disease continue to rise
  50% - improvements to care may have changed the outcome
  77% - no previously known heart disease
  Majority died without seeing a cardiologist

• Onus on obstetric and acute care doctors to **think heart disease**
  Take a good history.....
  breathlessness / chest pain / unexplained syncope
  Do the tests – don’t withhold because of pregnancy
  Make a diagnosis, don’t just exclude PE

• Get early, expert cardiology input