CARDIAC DISEASE IN PREGNANCY: TUTORIAL 2

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The second of two tutorials exploring the presentation and management of pregnant women with heart disease.

Tutorial 1:

- Introduction and scope of the problem
- Physiological changes and challenges during labour and delivery
- Acquired and congenital valvular heart disease.

Tutorial 2:

- Non-valvular congenital heart disease
- Pulmonary hypertension
- Cardiomyopathy
- Ischaemic heart disease in pregnancy
- General approaches to management

NON-VALVULAR CONGENITAL HEART DISEASE:

Conditions with a left to right shunt

- Left to right shunt may be due to a ventricular septal defect (VSD), atrial septal defect (ASD) or patent ductus arteriosus.
- Small shunts do not usually cause problems.
- Moderate shunts may increase if SVR increases due to pain and catecholamine release. If there is a large drop in SVR (e.g. following spinal block) then the shunt may reverse in direction and may result in hypoxia.
- Large shunts (most likely from a VSD) can result in pulmonary hypertension.

Tetralogy of Fallot (ToF)

- Most common cyanotic congenital heart lesion.
- Large VSD, right ventricular outflow tract obstruction, right ventricular hypertrophy and overriding aorta.
- Risks dependant on the status of the repair.
- Pregnancy often well tolerated in those with repaired ToF but women should have their right ventricle fully assessed; deaths have occurred in recent years from arrhythmias secondary to unrecognised right heart failure.

Coarctation of the Aorta

Narrowing of the aorta around the site of the ductus arteriosus. Most women presenting with coarctation will have had a previous repair.

- Problems during labour and delivery are unlikely if successfully repaired, however late hypertension, re-coarctation and aneurysm formation at the site of previous repair may occur. All women with previous repairs should be closely monitored throughout pregnancy by serial

echocardiography and regular BP measurement (measure BP in both arms since left subclavian may have been used as part of previous repair).

- If present with un-repaired coarctation (native coarctation), risks to both mother and fetus are high due to hypertension refractory to medical treatment.
- Regional anaesthesia or analgesia must be carefully titrated with close monitoring of BP and drugs to maintain SVR (phenylephrine, metaraminol).
- In severe cases women are at risk of aortic rupture, dissection and left ventricular failure.

Pulmonary Hypertension (PH) and Eisenmenger Syndrome

- There is a very high risk of maternal mortality with PH and termination of pregnancy is often recommended.
- There is increased pulmonary vascular resistance resulting in an increased workload placed on the right heart.
- PH may be primary or secondary.
- The causes of secondary PH include:
    - cardiac, respiratory (chronic obstructive or parenchymal conditions, cystic fibrosis, obstructive sleep apnoea, thoracic cage abnormalities) venous thromboembolism, vasculitis, hyperviscosity syndrome, infection, portal hypertension, cirrhosis and drugs (oral contraceptive, crotalaria teas, appetite suppressants)
- PH is poorly tolerated due to insufficient adaptation of the right heart to the increased cardiac output and poor compliance of the pulmonary vasculature.
- Symptoms of right ventricular decompensation are: shortness of breath, fatigue, chronic cough, haemoptysis and syncope.
- Signs include; tachycardia, cyanosis, right ventricular heave, elevated JVP and hepatomegaly.
- Death occurs from irreversible right ventricular failure and arrhythmias.
- Therapies targeted at pulmonary arterial vasodilatation may be useful during pregnancy (e.g. prostaglandin analogues including Iloprost).
- Timing of delivery is dependant on the impact of PH on the mother as pregnancy progresses. If possible, aim to deliver at 32-34 weeks.
- Increases in pulmonary vascular resistance (PVR) must be prevented by avoiding rises in $PaCO_2$, falls in PaO_2 and pH, hypothermia, high ventilatory pressures and sympathetic agents.
- Right ventricular preload, left ventricular afterload and right ventricular contractility must be maintained.
- Vaginal delivery is probably the safest mode of delivery with a low dose epidural to reduce pain, stress and haemodynamic fluctuations while maximising oxygen consumption. Avoid pushing in second stage due to the reduction in venous return and right heart preload that results.
- Elective caesarean is sometimes performed especially when delivery is preterm. Regional anaesthesia may be appropriate but single shot spinals should be avoided due to the inability of the right ventricle to respond to hypotension.
- Pulmonary arterial pressure may rise significantly during intubation if general anaesthesia is provided. Measures to obtund the pressor response to laryngoscopy should be used. (See later.)
- Oxytocin should be used cautiously after delivery.
- Women should be monitored closely after delivery since most deaths occur 2-9 days post partum. Observation for at least 72 hours on a High Dependency Unit should be provided.

- Eisenmenger's syndrome is pulmonary hypertension at systemic values with reversal of bidirectional shunt either due to a large left to right communication or pulmonary artery occlusion.
- The shunt flow depends on the PVR: SVR ratio.
- Hypovolaemia will lead to shunt reversal, reduced cardiac output and increased cyanosis.
- Mortality in pregnancy and delivery is very high due to right ventricular failure.

CARDIAC DISEASE DEVELOPING IN PREGNANCY

Cardiomyopathy

- Peripartum cardiomyopathy is defined as onset of cardiac failure with no identifiable cause in the last month of pregnancy or within 5 months after delivery, in the absence of pre-existing heart disease.
- The incidence is 1:1500 to 1:4000 live births.
- Risk factors include previous peripartum cardiomyopathy, hypertension, pre-eclampsia, obesity, diabetes, Afro-Caribbean origin, increased parity, older maternal age and multiple gestations.
- The aetiology remains unclear but includes viral myocarditis, abnormal immune response to pregnancy or terbutaline tocolytic therapy.
- Diagnosis is difficult as many symptoms are similar to those you would expect in the last trimester (peripheral oedema, fatigue and shortness of breath).
- Suspected cases should be investigated by echocardiography as there are strict echo criteria for diagnosis.
- Treatment is supportive with medical stabilisation. As most cases present late in pregnancy, delivery of the foetus may significantly improve symptoms.
- Vaginal delivery may be best with low dose epidural and close monitoring of BP and fluid status.
- Patients should be monitored on a high dependency unit or cardiac care unit post-delivery.
- Medical treatment includes salt restriction, diuretics, vasodilators, digoxin for arrhythmias and inotropy and anticoagulation due to the high risk of thromboembolism.
- Mortality ranges from 18 to 56% and often occurs several months after delivery. In severe cases patients will be referred for heart transplantation after delivery.
- Idiopathic dilated cardiomyopathy can also develop which is similar to the above but does not fit the diagnostic criteria and has a worse long-term outcome.

- Pre-existing **hypertrophic cardiomyopathy** is generally well tolerated and most undergo successful vaginal delivery.
- Cardiac function depends on preload and afterload so if using regional anaesthesia it must be carefully titrated with invasive blood pressure monitoring.

- Pre-existing **dilated cardiomyopathy** may decompensate in pregnancy. Women with severe LV impairment secondary to dilated cardiomyopathy may be counselled against pregnancy due to the high risk of mortality.

Ischaemic heart disease and myocardial infarction

This is now the leading cause of cardiac maternal mortality in the UK.
In the last confidential enquiry of maternal deaths, all women who died from ischaemic heart disease had identifiable risk factors including:

- Obesity
- Advanced maternal age
- Higher parity
- Pre existing hypertension
- Smoking
- Family history of cardiac disease
- Type 2 diabetes mellitus

Previously undiagnosed ischaemic heart disease (IHD) usually manifests itself in the 3rd trimester, labour or post delivery at a time when maternal stress and cardiac demand are at their greatest. Most commonly it presents with chest pain, ischaemic changes on the ECG and elevated troponin but may sometimes present atypically with abdominal or epigastric pain. Any woman with chest pain suspicious of ischaemia, particularly in those with risk factors, should have an ECG.

Coronary angiography may be indicated in women with IHD to treat coronary artery occlusion and coronary artery dissection by stenting and angioplasty.

In the event of myorcardial infarction, primary percutaneous transluminal coronary angioplasty (PTCA) should be performed. If PTCA is not available, thombolysis should not be withheld in the pregnant or post-partum woman as the risk of bleeding is less than the risk of no treatment.

Beware the use of uterotonics. Ergometrine causes coronary artery vasospasm and should be avoided if there is a history or IHD.

Aortic dissection

- Associated with hypertension due to pre-eclampsia or coarctation of the aorta and connective tissue disorders including Marfan's and Ehlers-Danlos disease.
- Pregnancy related aortic dissection accounts for 50% of all aortic dissections in women under 40 years of age.
- Maternal mortality may be as high as 25%.
- Usually occurs in late pregnancy or post delivery.
- Presents with severe chest pain, interscapular pain, end-organ ischaemia or acute MI.
- Investigations include chest CT, MRI or Trans Oesophageal Echo (TOE).
- Management varies depending on the gestation of the foetus.
- If presents before 28 weeks then surgical repair with foetus in-situ is recommended as without surgery mortality may reach 80%.
- Cardiopulmonary bypass is associated with congenital malformations in the first trimester but is safer in second and third trimester.
- After 32 weeks delivery by caesarean section followed by corrective surgery.
- Between 28 and 32 weeks gestation, unless there is severe cardiovascular instability, medical management is provided to allow the foetus to mature
- Goals of anaesthetic management are: maintenance of cardiovascular stability with regional anaesthesia and labetalol infusion for control of BP.
- For caesarean section under GA, the hypertensive response to laryngoscopy must be avoided.

Management of women with transplanted heart

- Pregnancy is generally well tolerated if cardiac function is good.
- Problems due to the side effects of the immunosuppressant drugs.
- Spontaneous vaginal delivery is best management option.

GENERAL APPROACHES TO MANAGEMENT OF PREGNANT WOMEN WITH CARDIAC DISEASE

Monitoring

- Basic monitoring for mothers with heart disease during labour, delivery and in the immediate post partum period includes; blood pressure, pulse oximetry and continuous 3-lead ECG.
- Invasive blood pressure monitoring is very useful in higher risk cases and can be easily managed on labour ward with appropriate anaesthetic input.
- There is debate over the use of central venous pressure (CVP) and pulmonary artery catheter (PAC) monitoring. These interventions are not without risk and benefits may sometimes be limited.
- Long lines placed in the antecubital fossa can be used to measure CVP and infuse vasoactive drugs and may be a safer approach to central venous cannulation.

Delivery

- Consultant led delivery in a hospital used to dealing with cardiac disease with a high dependency area that can provide invasive monitoring is essential.
- Stress on the mother and her cardiovascular system must be minimised while maintaining placental and foetal circulation.
- Effective pain relief results in less tachycardia and catecholamine release. It also reduces the haemodymanic effects of pushing.
- Limited pushing in second stage i.e. assisted second stage may reduce cardiovascular instability.
- Low-dose epidural anaesthesia sited early in labour for effective pain control and reduced catecholamine release is highly beneficial in most cases.
- If caesarean section is required due to obstetric indications or decompensation of the underlying disease, then this can be done with either general or regional anaesthesia.
- If general anaesthesia is planned, measures to suppress the pressor response to laryngoscopy must be provided e.g. alfentanil 10-20 mcg/Kg.
- If regional anaesthesia is planned, single-shot spinals are best avoided. Alternative options include: careful titration of an epidural, combined spinal-epidural or incremental spinal anaesthesia (via spinal catheter).

There are many factors that influence the decision whether to proceed with regional or general anaesthesia and these include:

- Requirement for other procedures which may demand general anaesthesia e.g. DC cardioversion, post-delivery cardiac surgery, high inspired oxygen concentration (pulmonary hypertension), post operative ventilation, prolonged or complex surgery following previous surgery.
- Risk of reducing SVR with regional anaesthesia (left sided stenotic lesion and those with shunts) versus impairment of cardiac contractility with general anaesthesia.
- Impact of anticoagulation; risks of withholding it and risk of epidural haematoma.
- Risk of maternal or fetal death and how the mother feels about this.
- Airway abnormalities.
- Anaesthetic preference.
- Patient preference.

Prophylactic Antibiotics to prevent endocarditis

- American Heart Association (2007) guidelines and the UK National Institute for Health and Clinical Excellence (NICE) 2008 guidelines do **not** recommend administration of antibiotics solely to prevent endocarditis in patients who undergo a gynaecological or obstetric procedure since there is no beneficial evidence of this practice.

Anticoagulation

- Warfarin is teratogenic and not recommended during the first trimester of pregnancy. It is avoided in the third trimester since it crosses the placenta and can cause foetal haemorrhage. It also precludes regional anaesthesia and its effects may be difficult to rapidly reverse in an emergency.
- Low molecular weight heparin (LMWH) can be used instead of warfarin throughout the whole of pregnancy. Regional anaesthesia can be performed provided adequate time has elapsed since the last dose of LMWH.
- For women receiving prophylactic LMWH, regional anaesthesia or removal of epidural catheter can be performed 12 hours after last dose of LMWH. After insertion of epidural or spinal a dose can be given 4 hours later.
- In women receiving therapeutic doses of LMWH, 24 hours should elapse after the last dose of LMWH before regional anaesthesia or removal of epidural catheter. After insertion of epidural or spinal a dose can be given 4 hours later.

Uterine atony

Many oxytocics have severe consequences for those with cardiac disease but withholding them can lead to haemorrhage. A balanced individualised approach is best.

- Oxytocin can cause profound tachycardia, vasodilatation and hypotension when administered as an IV bolus so administer the bolus as an infusion (e.g. 5 units in 20mls over 5-10 mins). If at particular risk of cardiovascular effects (e.g. severe aortic stenosis) then it may be best omitted. A low dose infusion with 10 units per hour can be used post-delivery with careful monitoring.
- Ergometrine causes pulmonary vasoconstriction and hypertension so avoid in most cardiac cases especially pulmonary hypertension.
- Prostaglandin F2∝ (Carboprost) can cause severe bronchospasm, hypertension, cardiovascular collapse and pulmonary oedema making it unsuitable in most cases.
- Uterine massage can be used to provide temporary relief but may require adequate analgesia.
- Other surgical options in the event of refractory uterine atony include: an intrauterine balloon that can be left in 1-2 days after caesarean section or vaginal delivery, uterine compression sutures (e.g. B-lynch suture), internal iliac balloon catheterization/ligation and hysterectomy.

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