

**Anaesthetic tutorial of the week 111:
Cardiac disease in pregnancy: tutorial 1**

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The first 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:

- Other congenital heart disease
- Pulmonary hypertension
- Cardiomyopathy
- Ischaemic heart disease in pregnancy
- Approaches to management

Multiple-choice questions:

Answer true or false to each part

1. In severe aortic stenosis recommended management includes:

- a) Invasive blood pressure monitoring
- b) Oxytocin bolus after delivery
- c) Single shot spinal
- d) Regional anaesthesia may be suitable for delivery
- e) Management in a centre providing cardiac surgery

2. Risk factors for cardiac events in pregnancy are:

- a) Cyanotic heart disease
- b) Previous arrhythmia
- c) NYHA class 2 symptoms
- d) Previous corrective cardiac surgery
- e) Left ventricular dysfunction

3 Clinical signs that should prompt more investigations are:

- a) Systolic murmur grade 2/6
- b) Diastolic murmur grade 2/6
- c) 3rd heart sound
- d) 4th heart sound
- e) Fixed split 2nd heart sound

Key points

- **Cardiac disease is a leading cause of maternal mortality in the UK**
- **Maternal death due to ischaemic heart disease is increasing in the developed world**
- **Rheumatic heart disease is the leading cause of maternal heart disease in the developing world**
- **Some conditions still have such a high maternal mortality rate that foetal termination is advised**
- **Low dose epidural infusion for labour with assisted second stage is suitable management in many cases**
- **Oxytocin can cause haemodynamic instability and should be used with care in mothers with cardiac disease**

Background

The incidence of cardiac disease in pregnancy is increasing in the developed world. Cardiac disease was the commonest cause of maternal death in the most recent Confidential Enquiry into Maternal and Child Health (CEMACH) which reports the deaths of UK women while pregnant or within 42 days of the end of pregnancy. The leading cause of death was myocardial infarction, mainly secondary to ischaemic heart disease, followed by cardiomyopathy, sudden adult death syndrome and dissection of the thoracic aorta. Deaths related to pulmonary hypertension and congenital heart disease have fallen but deaths due to rheumatic heart disease have increased due to immigration of women from areas of high prevalence. Pregnant immigrant women are at highest risk of morbidity and mortality in the UK. The maternal mortality rate for cardiac disease was 2.27 per 100 000 maternities in 2003-05. The prevalence of cardiac disease in pregnancy in the developed world is 0.2-3%.

Introduction

More women with congenital heart disease now survive to adulthood due to improved surgical techniques and drug therapy. Many of these women have a good quality of life and may not appreciate the risks that pregnancy and delivery can impact upon their heart.

The incidence of ischaemic heart disease has risen due to the later age at which women are conceiving and an increase in those with risk factors for ischaemic heart disease including diabetes and obesity.

Risk factors for a cardiac event in pregnancy for a woman with pre-existing heart disease are:

1. Prior cardiac event
2. Poor New York Heart Association (NYHA) functional class (class 3/4) or cyanosis (see below)
3. Left heart obstruction
4. Systemic ventricular dysfunction

Cardiac event is defined as: pulmonary oedema, arrhythmia, cerebrovascular event, cardiac arrest or death.

If no risk factors are present then risk of cardiac event is 5%, 1 factor 27% and 2 factors 75%.

NYHA 1: Known cardiac disease with no limitation of physical activity and no objective evidence of cardiovascular disease

NYHA 2: Slight limitation of normal physical activity and objective evidence of minimal disease

NYHA 3: Marked limitation of physical activity and objective evidence of moderate disease

NYHA 4: Severe limitation of activity including symptoms at rest and objective evidence of severe disease

Rheumatic fever is still prevalent in the developing world so severe uncorrected valvular heart disease is more common in women reaching child-bearing age. Rheumatic heart disease most commonly affects the mitral valve causing stenosis or regurgitation but can also affect the aortic valve and less commonly the pulmonary valve.

Mothers with known cardiac disease need to be seen early in pregnancy or preferably preconception so they can be counselled on potential complications and mortality to themselves and their foetus. For some conditions including pulmonary hypertension and Eisenmenger syndrome, termination of pregnancy may be recommended due to the risk of maternal death which may be as high as 60%.

Pregnant women with cardiac disease should ideally be managed in specialist centres by a multi-professional team of cardiologists, obstetricians and anaesthetists who will make a plan for delivery and monitor cardiac function throughout pregnancy by regular clinical examination and investigations including ECG, echocardiogram and in some cases cardiopulmonary exercise testing.

Antenatal care should focus on preoptimisation of cardiovascular function with appropriate medication. In some cases medication will need to be altered to avoid potential teratogenicity.

Ideally the same team should review the mother throughout pregnancy to identify subtle changes in function. There should be a low threshold for admission and investigation as minimal activity (bed rest) can improve symptoms markedly.

Mothers who develop incipient cardiac disease in late pregnancy are difficult to diagnose as symptoms are similar to the later stages of pregnancy i.e. shortness of breath, swollen ankles, fatigue, reduction in exercise capacity, murmurs, third heart sound and arrhythmias. Loud 4th heart sound, diastolic murmur, grade 3/6 systolic murmur, fixed splitting of second heart sound or opening snap should raise suspicion and appropriate investigations including ECG and echocardiography should be performed.

Physiological Changes in Pregnancy

Physiological changes of pregnancy have been discussed in a previous tutorial. By the 20th week of gestation, cardiac output increases by 40% due to increased stroke volume and heart rate. Blood volume increases by up to 50% while systemic vascular resistance (SVR) falls. This will place additional strain on a failing heart and in those with a significant shunt. As SVR falls there will be increased right to left shunting which may reduce pulmonary perfusion. Coupled with the increased oxygen demand of pregnancy this can lead to increasing hypoxia.

Labour and delivery

Labour and delivery can further compromise cardiac function. Pain may lead to a tachycardia which reduces diastolic coronary filling as well as increasing circulatory catecholamines which increase SVR. Cardiac output increases during each contraction due to autotransfusion of up to 500ml of blood into the circulating volume. In some cases, pushing during the 2nd stage may cause haemodynamic compromise due to the effects of the Valsalva manoeuvre.

Potential problems at delivery include:

- Blood loss may be poorly tolerated since many women are unable to compensate by the usual response of increasing heart rate and stroke volume and may rely on venous return to maintain cardiac output.
- Bleeding may be increased due to concurrent anticoagulation.
- Pulmonary oedema is more likely to occur peripartum due to fluid shifts and as a consequence of additional IV fluids to replace losses. Careful fluid balance is essential and this may be aided by running drug infusion in smaller volumes of increased concentration.
- Arrhythmias and tachycardia are poorly tolerated. Drugs causing tachycardia should be avoided or limited (e.g. oxytocin, ephedrine). Alpha-adrenergic agonists (e.g. phenylephrine and metaraminol) are the vasopressors of choice.
- Reduction in SVR by regional techniques or other drugs may cause problems if cardiac output is fixed (e.g. severe aortic stenosis) or in the presence of a shunt. Increased right to left shunting reduces pulmonary blood flow and increases hypoxia.
- Women may be at very high risk of venous thromboembolism and measures including TED stockings and prophylactic heparin should be administered.
- Women with a right to left shunt may be at risk of air embolism so meticulous care should be taken with IV lines and drugs.
- Acute pulmonary hypertension at the time of delivery can cause right ventricular failure and cardiac ischaemia. The risk of death is high in those with severe pulmonary hypertension and severe morbidity can occur in those with relatively mild pulmonary hypertension.

Postpartum

- Women must be closely monitored post-partum for at least 72 hours. The majority of deaths in women with pulmonary hypertension occur after delivery.
- Fluid balance must be carefully monitored, anticoagulation re-established if necessary and good post-delivery analgesia maintained to prevent tachycardia and catecholamine increase.
- It takes 6-8 weeks for the cardiovascular system to return to pre-pregnancy function.

Risk stratification of different cardiac lesions

- Low risk; mortality (0.1-1%):
 - Most repaired lesions, uncomplicated left to right shunts, mitral valve prolapse, bicuspid aortic valve, aortic regurgitation, mitral regurgitation, pulmonary stenosis and regurgitation.
- Intermediate risk; mortality (1-5%):
 - Metal valves, single ventricles, systemic right ventricles, switch procedures, unrepaired cyanotic lesions, mitral stenosis, mild/mod aortic stenosis and severe pulmonary stenosis.
- High risk; mortality (5-50%):
 - Symptomatic at preconception with heart failure (NYHA 3/4), severe ventricular dysfunction, severe aortic stenosis, Marfans with aortic valve lesion or aortic dilation and pulmonary hypertension.

Congenital and Acquired Valvular Heart Disease

- Women may often have asymptomatic valvular disease until compromised by the physiological changes of pregnancy.
- Pregnancy is better tolerated in regurgitant lesions than in stenotic lesions as reduced SVR promotes forward blood flow. Labour can be managed with a carefully titrated epidural.

Aortic stenosis (AS)

- If asymptomatic before conception then pregnancy is usually well tolerated.
- In severe stenosis (valve area $<0.5\text{cm}^2$ (best guide) or gradient > 60 mmHg) mortality is up to 5%.
- Fall in SVR poorly tolerated in severe aortic stenosis due to fixed cardiac output.
- Tachycardia, bradycardia, hypovolaemia and venocaval compression poorly tolerated.
- Coronary artery perfusion dependent on aortic diastolic pressure so hypotension can lead to acute left ventricular failure secondary to ischaemia.
- Blood pressure should be closely monitored and carefully managed with alpha-adrenergic agonists and appropriate fluid to replace losses.

- Avoid ephedrine due to the risk of tachycardia as a result of beta 1 receptor stimulation.
- Invasive arterial blood pressure monitoring should be used in cases of severe AS.
- General anaesthesia may be indicated in severe aortic stenosis; however there are many reports of carefully titrated regional anaesthesia with a vasopressor infusion and close observation of blood pressure. Gradual epidural top-up, combined spinal epidural with low dose spinal block and careful epidural top-up and cautious spinal block with a spinal catheter have all been successfully used to manage women with moderate and severe AS.
- Acute aortic valve replacement may be required in cases of severe symptomatic stenosis. Women at risk must be managed in a hospital with cardiac surgery facilities.

Mitral stenosis

- Defined as severe if valve area on echocardiography is $<1\text{cm}^2$
- Problems due to large gradient between left ventricle and left atrium which may cause pulmonary oedema, pulmonary hypertension and right ventricular failure.
- Dyspnoea at rest is an important marker of severity.
- If pulmonary hypertension is already present then risk to the mother is significant and termination of pregnancy maybe the best option.
- Tachycardia and arrhythmias will reduce cardiac output by reducing diastolic filling.
- Rate control is most important and tachyarrhythmias should be controlled with β -blockers or digoxin.
- If the valve stenosis is severe a balloon valvotomy may be considered during pregnancy.
- Vaginal delivery with carefully titrated epidural analgesia and invasive arterial blood pressure monitoring is preferable in those who are symptomatic or with severe stenosis.

Pulmonary stenosis

- Pulmonary stenosis in isolation constitutes 10% of congenital heart disease.
- Also part of the Tetralogy of Fallots and may be present following its repair.
- Isolated pulmonary stenosis is often asymptomatic but may lead to increased right ventricular work and right ventricular failure.
- Balloon valvuloplasty may be considered in women developing right heart failure.
- Goals during labour and delivery are to maintain right ventricular preload, left ventricular afterload and right ventricular contraction.
- Vaginal delivery with epidural analgesia is probably the most optimal management.

Further Reading

Will be listed in: Cardiac Disease and Pregnancy: Tutorial 2.

MCQ answers.

1 TFFTT

Oxytocin and single shot spinal both cause large decreases in SVR which is very poorly tolerated in those with severe aortic stenosis. Cautious regional anaesthesia may be appropriate for delivery with close monitoring of BP and therapeutic measures to maintain SVR.

2 TTFFT

Risk factors for cardiac events in pregnancy include prior cardiac event (heart failure, TIA or CVA, arrhythmia, pulmonary oedema, cardiac arrest), poor NYHA functional class (class 3/4) or cyanosis, left heart outflow obstruction, systemic ventricular dysfunction.

3 FTFTT

Signs that should prompt further cardiac investigation are loud 4th heart sound, diastolic murmur, grade 3/6 systolic murmur, fixed split 2nd heart sound and an opening snap.