education

FROM THE JOURNALS Edited highlights of Richard Lehman's blog on http://bmj.co/Lehman

Collaborative care for heart failure

When Goethe published *The Sorrows of Young Werther* in 1774, it was a literary sensation and led to a new vogue in men's clothing and a spate of young suicides. When I get round to writing "The Sorrows of Old Lehman" I doubt that it will have quite the same effect on male fashion or the suicide rate. It will mostly be about how medicine goes around in the same circles and old lessons have to be learnt time and again. Take "heart failure." Properly speaking, this is a mode of death, not a diagnosis. If somebody is dying from a failing heart around the age of 80, then your expectations of any intervention need to be modest. If you can relieve symptoms of depression and fatigue, as they did in this US trial, that will be a very worthwhile success in its own right. The trajectory for general wellbeing is bound to be downwards whatever you do. The comparator here was usual care: let's try to improve this too. When I helped put together the first book on *Heart Failure and Palliative Care* in 2006, our subheading was "a team approach." Wherever you are in the system, do all you can to encourage humane care that supports the patient and carers through what is bound to be a sad and difficult journey. Count your success by what you can achieve, and don't be put off by what can't be achieved.

JAMA Intern Med
doi:10.1001/jamainternmed.2017.8667

Corticosteroids for septic shock

The first of two trials of steroids in septic shock is an exemplary study of 3800 patients in intensive care units across five countries. For once, the abstract of the paper underplays the results by declaring that the administration of hydrocortisone did not result in lower 90 day mortality than placebo among patients with septic shock. But in the main text the authors say, "This effect did not differ in any of the six prespecified subgroups. We observed a more rapid resolution of shock and a lower incidence of blood transfusion among patients who received hydrocortisone than among those who received placebo. Patients who had been assigned to receive hydrocortisone had a shorter time to intensive care unit discharge and earlier cessation of the initial episode of mechanical ventilation than did those who had been assigned to receive placebo." So, there is still a case for giving continuous intravenous hydrocortisone.

Now add a pinch of mineralocorticoid. When a French trial of steroids in septic shock was designed, there was a drug called drotrecogin alfa (activated) on the market, which was to be given in one of three arms of the study. However, it was subsequently withdrawn, leaving just two arms: placebo, or hydrocortisone plus fludrocortisone. Mortality in the hydrocortisone plus fludrocortisone group was lower than in the placebo group: 43% versus 49%. But it's impossible to compare this directly with the Australian study, because the patient characteristics were markedly different and the French mortality rate was very much higher: in fact, it's the same as in the original trials of steroids in sepsis done in the early 1960s. So, the message seems to be that hydrocortisone is of marginal benefit to a few patients with septic shock and that we don't really know whether added fludrocortisone makes a lot of difference.

N Engl J Med
doi:10.1056/NEJMoa1705835
N Engl J Med
doi:10.1056/NEJMoa1705716

Home lowering of blood pressure

At a population level, blood pressure is one factor that contributes to cardiovascular risk. That's about all that I would be willing to concede with the current state of knowledge. This trial shows that people with blood pressure above 140/90 mm Hg (labelled "hypertensive patients" in this paper: isn't it time we dropped these terms?) can monitor their blood pressure at home and bring it down more effectively by self titration than through the usual tedious process of repeated visits to a general practice surgery. Adding telemedicine contributes nothing extra. This is useful knowledge, but now we must start to get to grips with shared decision making and the individuation of treatment for total cardiovascular risk.

Lancet
doi:10.1016/S0140-6736(18)30309-X

Mesh best for small umbilical hernias

Last week you might have wondered if you should have simple suturing or mesh for the repair of your small umbilical hernia. This week you don't need to wonder, because a Dutch led trial gives the answer. It provides "high level evidence for mesh repair in patients with small hernias of diameter 1–4 cm. Hence we suggest mesh repair should be used for operations on all patients with an umbilical hernia of this size." Sometimes evidence actually settles a question.

Lancet
doi:10.1016/S0140-6736(18)30298-8

Pregnancy in women with congenital heart disease

0.5 HOURS

See http://learning.bmj.com for linked learning module

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One in 125 people is born with congenital heart disease.[1] For women with the condition, pregnancy induced cardiovascular stress can cause complications such as arrhythmia, heart failure, and thromboembolism.[2] The UK Confidential Enquiry into maternal deaths found that of 910 maternal deaths between 2009 and 2014,[3] 205 (22.5%) were caused by heart disease, and a minority from congenital heart disease. Clinicians in primary and emergency care increasingly encounter women with congenital heart disease who are planning pregnancy or who are pregnant at presentation. These women might seek information about the risks pregnancy poses to their own health, and to the health of the fetus. In this article, we highlight aspects of pre-conception, antenatal, and postpartum care for women with congenital heart disease.[3]

What are the common forms of congenital heart disease?

The European Registry on Heart Disease is the largest published cohort of women with pregnancy complicated by heart disease. In 2012, of 1321 pregnant women with heart disease, 66% had congenital heart disease.[4] Approximately one third of those women had simple shunt lesions, such as ventricular or atrial septal defects, and the rest had multiple lesions, including mitral and pulmonary valve abnormalities, aortic coarctation, transposition of the great arteries, and Marfan's syndrome. Very high risk conditions, such as univentricular circulation (Fontan), cyanotic heart disease, or inherited cardiomyopathies accounted for 2% to 5% of cases.[4] In a smaller Canadian study of 405 pregnancies in women with congenital heart disease, more than half had shunt lesions, repaired tetralogy of Fallot, or aortic coarctation.[5]

WHAT YOU NEED TO KNOW

- Women with congenital heart disease have increased risk of poor pregnancy outcomes
- Offer counselling and specialist multidisciplinary care before conception
- Refer women with congenital heart disease to a cardiologist during pregnancy for clinical assessment including cardiac function, and to review cardiac medications
- Offer a planned hospital birth
- Expert consensus suggests vaginal delivery with regional anaesthesia is preferred

What are the risks?

Risks for women with congenital heart disease in pregnancy

Maternal cardiac risk

Common complications of congenital heart disease
- Arrhythmias
- Heart failure
- Thromboembolic events

These complications can lead to need for major surgery, disability, premature death

Maternal obstetric risk

Higher incidence of
- Miscarriage
- Preterm pre-labour rupture of membranes
- Postpartum haemorrhage

For the woman

The overall risk of maternal death in women with congenital heart disease is approximately 1%, which is 100 times higher than the background risk for maternal mortality in the developed world.[2]

Information on the effects of pregnancy, both immediate and throughout life, on women with congenital heart disease is limited as most studies are retrospective,[12-14] and few include long term follow-up. Most women can be reassured that pregnancy is not associated with any apparent decline in cardiac function, even years later.[15 16]

Risks to the fetus of congenital heart disease in the mother

Higher incidence of
- Fetal growth restriction
- Preterm birth
- Intracranial haemorrhage
- Fetal and neonatal death

Congenital heart disease in infant:
Risk 3%-50% (background risk 0.8%)

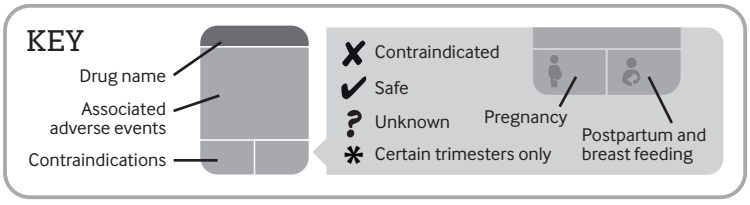
For the fetus

Reported miscarriage rates vary according to lesion; being highest (up to 50%) in women with cyanotic heart disease and women with a univentricular (Fontan) circulation.[17-19] The risk of congenital heart disease in the children of these women is higher than background, with 3% to 5% having the condition.[20 21] A UK single centre study (331 women) showed that preterm labour and pre-labour rupture of membranes were more common (12% and 14%, respectively) than in those without congenital heart disease, while the incidence of babies being small for gestational age (less than 10th centile) was 25% and the neonatal mortality rate was 4%.[22]

thebmj Visual summary

Using cardiovascular drugs during pregnancy

Many cardiac medicines appear safe for use during and after pregnancy, as summarised here. However, evidence is lacking for some drugs, and many of them carry risks of adverse events. Shared decision making, considering risks and benefits of medicines, is vital to help women weigh up potential risks to themselves and their unborn baby.



β blockers

These are commonly used by pregnant women. However, organ specific malformations are more prevalent in the offspring of women treated with β blockers, and fetal growth restriction has been reported

Labetalol

When used near term:
- Fetal hypoglycaemia
- Fetal bradycardia
- Fetal hypotension

Bisoprolol

- Fetal hypoglycaemia
- Fetal bradycardia

Atenolol

When used in second and third trimester:
- Fetal hypoglycaemia
- Fetal bradycardia
- Low birth weight

Methyldopa

Mild neonatal hypotension. Avoid post partum because of the risk of postnatal depression

Digoxin

Serum levels are unreliable

Calcium channel blockers

Not associated with an increased incidence of congenital anomalies in humans

Nifedipine

Potential synergism with magnesium sulphate can induce hypotension in mother and fetal hypoxia

Verapamil

⚠ Limited evidence

Well tolerated

Platelet aggregation inhibitors

Acetylsalicylic acid

Low dose aspirin safe in early pregnancy, but usually stopped at 34–36 weeks. No teratogenic effects reported

Clopidogrel

⚠ Limited evidence

Safe in animal studies, but caution is advised. Stop one week before anticipated delivery

Antiarrhythmic drugs

Adenosine

⚠ Limited evidence

No fetal adverse effects reported

Procainamide

⚠ Limited evidence

Unknown. Appears to be safe

Flecainide

⚠ Limited evidence

Unknown. Appears to be safe

Amiodarone

Might be used in special circumstances. Risk of hypothyroidism, goitre, bradycardia, fetal growth restriction, and preterm birth

Diuretics and aldosterone antagonists

Bumetanide, furosemide, and hydrochlorothiazide

Oligohydramnios and electrolyte imbalance in the fetus

Anticoagulants

Warfarin

Risk of fetal abnormalities if used in the first trimester. Can cause intracranial fetal bleeding in second trimester.

Heparin (low molecular weight)

Seldom osteoporosis and markedly less thrombocytopenia than unfractionated heparin.

Drugs contraindicated in pregnancy

ACE inhibitors†

High risk of fetal abnormalities. Captopril and lisinopril considered safe in breast feeding

Angiotensin receptor blockers

High risk of fetal abnormalities

Spironolactone

⚠ Limited evidence

Possible risk of anomalies of the external genitalia

† ACE inhibitors = Angiotensin converting enzyme inhibitors

Key considerations for counselling before conception for women with cardiovascular disease[8-29]

- Offer woman of childbearing age with cardiovascular disease counselling and risk stratification before conception
- Counselling is best made available within the paediatric cardiology transition service
- Offer the woman appropriate contraceptive advice
- In women contemplating pregnancy, change cardiovascular medications to those which can be used in pregnancy, and emphasise the importance of close monitoring
- In women not contemplating pregnancy, ensure effective discussion on contraception and early pregnancy termination

Preconception counselling

The UK Confidential Enquiry into maternal deaths highlighted the importance of preconception counselling.[3] Few studies examine the effect of counselling before conception on maternal and perinatal morbidity and mortality in the context of maternal heart disease.[4] Nevertheless, European Society of Cardiology and American Heart Association guidance emphasises that counselling before conception should be readily available at the transition from paediatric to adult cardiac care,[23-25] ideally by referral to a combined cardiology-obstetric clinic.

In the developed world, care is commonly undertaken by a multidisciplinary team, including an obstetrician and a cardiologist with access to a haematologist, geneticist, and/or anaesthetist.[26]

Women with heart disease who require IVF or medical therapies to become pregnant are advised to receive counselling before undergoing fertility treatment, as pregnancy might be contraindicated in some cases, or management of the fertility problem might need to be modified (such as single embryo transfer to reduce the risk of multi-fetal pregnancy).[24] In some conditions, such as pulmonary hypertension, women might be advised to avoid pregnancy.[23]

Women might have concerns about potential risks to themselves and their unborn baby,[27] and these can be discussed during preconception counselling. In women not contemplating pregnancy, ensure effective discussion on contraception and early pregnancy termination if necessary. Preconception counselling assessment (below) typically includes a clinical evaluation, imaging (notably an echocardiogram), risk stratification, and a review of current medications[30 31] to optimise cardiac status and/or avoid fetal exposure to known teratogens, such as angiotensin converting enzyme inhibitors. Data from the European Registry on Heart Disease showed that two thirds of women took cardiac medications at some point during their pregnancy.[32] Table 1 (see bmj.com) provides information on the use of common cardiac medications in pregnancy.

What assessment is needed

Urgently refer women who become pregnant and have not had preconception counselling, especially if they have not had a recent cardiac review. During assessment, women who have not previously had counselling usually undergo echocardiography and might require additional investigations, such as cardiac magnetic resonance imaging, for confirmation of the defect and to assess cardiovascular function. Such tests are best organised by a specialist able to evaluate the underlying condition fully and identify possible surgical interventions to improve cardiac function before pregnancy.

Several studies have evaluated risk scoring systems for women with congenital heart disease to better predict the likelihood of a cardiac complication.[11-48] Most scoring systems are limited, however, as they do not consider both fetal and maternal risks.[48 49] The modified World Health Organisation (mWHO) classification is the most widely adopted and simplest risk categorisation system (table 2, see bmj.com). This can assist in counselling women with congenital heart disease, and identify women who need referral to specialist services.

How should new cardiac symptoms be managed?

Although the physiological burden of normal pregnancy can cause breathlessness and palpitations, any new onset requires careful evaluation, often with referral to the cardiac and/or obstetric team.

Occasionally, women without a history of heart disease might present for the first time in pregnancy with a major cardiac event. The online article describes some important scenarios in women with known and previously occult congenital heart disease. Treatment is influenced by the gestation of the pregnancy, and usually requires referral to a specialist centre.[29]

How to deliver antenatal care

Antenatal care is usually based in a tertiary hospital, and is particularly recommended for women with risk categorised as mWHO 3 or 4. In the UK, there are 14 centres providing specialist care for women with congenital heart disease. Whether a woman needs antenatal care by a specialist team or can be safely cared for by a local hospital team with cardiology input is best discussed with the multidisciplinary team. The European Society of Cardiology recommends that all women with congenital heart disease should be reviewed by a cardiologist at least once before, and once during pregnancy, and should have a planned hospital birth. An integrated care record can help keep track of appointments and facilitate communication between different healthcare providers. Here we list measurements and observations undertaken at each antenatal visit.

Ultrasound screening of the fetus at 11-14 weeks' gestation is advised to detect abnormal nuchal translucency, which has a strong association with congenital heart disease in the fetus.[50] Fetal echocardiography at 18-20 weeks' gestation can detect major structural and functional abnormalities. The European Society of Cardiology recommends fetal growth monitoring using serial ultrasound biometry, particularly if a woman is taking β blockers, as maternal heart disease is associated with an increased risk of fetal growth restriction.[22]

Check and record the following at each antenatal visit

- Blood pressure and heart rate
- Heart rhythm
- Auscultation of heart sounds and lung bases
- Maternal oxygen saturations
- Proteinuria
- Fetal growth

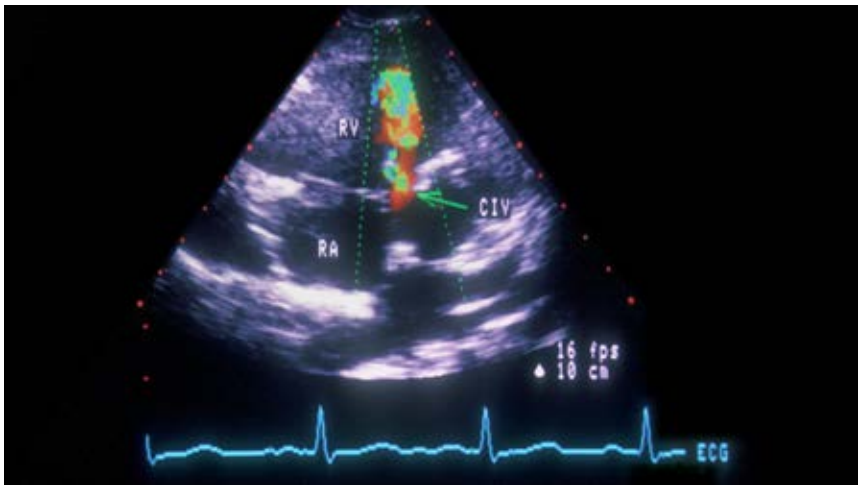
Management of labour

Most women with congenital heart disease can expect and are typically offered as normal a birth as possible, including spontaneous onset of labour. However, in general women are advised to have their birth in hospital and to attend as soon as in labour. A small number of women might require induction of labour for obstetric reasons (for example, fetal growth restriction) or cardiac reasons—for example needing to stop anticoagulants such as low molecular weight heparin before labour.[51] The usual induction methods, such as vaginal or oral administration of prostaglandins, are safe in women with congenital heart disease.

For pain, most experts recommend regional anaesthesia, as it blunts the cardiovascular response to pain,[23 26] thereby reducing strain on the heart. Maternal monitoring during labour is individualised and commonly includes continuous electrocardiographic monitoring, pulse oximetry, and non-invasive blood pressure measurement. Expert consensus is to offer antibiotic prophylaxis for infective endocarditis in women with high risk lesions, such as metallic heart valves or a history of endocarditis.[25]

No trials have evaluated the preferred mode of delivery in women with heart disease. Retrospective data from the European Registry on Heart Disease suggest that vaginal delivery should be encouraged, as caesarean section for cardiac indications alone does not confer any benefit to the mother or baby.[52] This view is supported by the European Society of Cardiology and the American Heart Association.[23 25] Nevertheless, a planned caesarean section might be preferable in some settings—for example, because access to specialist cardiac and anaesthetic services is limited, or because of the nature of the woman's condition. Despite the absence of robust clinical data, both the American Heart Association and European Society of Cardiology[23 25] recommend assisted delivery, either by vacuum extraction or forceps, in conditions such as Marfan syndrome or significant valvular stenosis, to minimise the duration of the active phase of the second stage of labour. However, this might not apply where disease is mild.[23 53]

After delivery, active management (cord clamping and administration of oxytocin) is recommended for all women with heart disease, as this reduces blood loss by up to 40%.[54] Oxytocin can cause profound hypotension and tachycardia as a bolus[55 56]; hence a slow infusion of 2 IU of oxytocin over 10 minutes is preferred.[57] Ergometrine is avoided as it causes hypertension and constriction of the coronary arteries, which can cause myocardial infarction.[58 59]



PIERRE BORIES, ISM/SPL

Doppler ultrasound scan of the heart, showing a ventricular septal defect

What to consider in the postnatal period

The puerperium is a high risk period as many haemodynamic changes occur concurrently. Intensity of maternal monitoring depends on the underlying congenital lesion, predisposition to arrhythmia, and the presence of symptoms of heart failure. Admission to an obstetric high dependency unit for continuous cardiac monitoring (telemetry) might be required, with multidisciplinary input in women at risk of arrhythmia or heart failure.

Some babies might benefit from referral to paediatric cardiology, as minor lesions might not have been detected earlier.

Medications are reviewed and adjusted at this time and further review in primary or secondary care can be discussed with the patient. Women can be reassured that breast feeding is safe with most cardiac medications (p 411, infographic), although advice should be individualised and the wishes of the woman considered. Contraception can be discussed with the woman and her partner before discharge, to assess whether they would like to avoid or space a future pregnancy.

Competing interests: None declared.

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EDUCATION INTO PRACTICE

- Describe your established referral pathway for women with cardiac disease who are pregnant or who are planning to conceive? Does this article offer you ideas on how to improve it?
- How many women of reproductive age in your practice have cardiac disease? Does this article offer you ideas on what standards of pre/ante/postnatal care you could audit?
- How would you explain the risks[3] associated with pregnancy and childbirth to a woman with a cardiac condition?

HOW PATIENTS WERE INVOLVED IN THE CREATION OF THIS ARTICLE

A woman with congenital heart disease who had recently given birth kindly reviewed this paper. Based on her experience of care under a multidisciplinary team during her pregnancy, she emphasised the importance of the team in caring for women with heart disease or other cardiac complications in pregnancy. She remarked that good communication was essential between members of the multidisciplinary team, and when discussing care plans with the patient.

Reduced fetal movements

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This is part of a series of occasional articles on common problems in primary care. *The BMJ* welcomes contributions from GPs.

CPD READING
0.5 HOURS

LEARNING MODULE
See http://learning.bmj.com for linked learning module

A 28 year old woman who is 32 weeks pregnant with her first baby tells you that, in the past 24 hours, she has not felt her baby move as much as usual.

A reduction or change in fetal movements can be a warning sign for adverse outcomes in pregnancy. This article aims to help healthcare professionals become more familiar with how to assess, counsel, and decide when to refer a woman presenting with reduced fetal movements.

The perception of fetal movements is subjective. Most women become aware of fetal movements by around 18-20 weeks' gestation, which may be felt as "a discrete kick, flutter, swish, or roll."[1] Nulliparous women may first become aware of fetal movements at a later gestation.[2] Fetal movements usually increase in number up to 32 weeks' gestation before reaching a plateau; fetal movements do not reduce in healthy late pregnancy and do not reduce before normal labour. They tend to become more frequent later in the day, and absent during fetal sleep cycles (lasting 20-40 minutes).[1 3] Women will come to learn the normal pattern of their baby's movements, which can vary considerably between women. A woman is said to have a reduction in fetal movements when there has been a decrease or change in her baby's normal pattern of movements. The Royal College of Obstetricians and Gynaecologists (RCOG) guidelines from 2011 did not recommend formal daily counting of fetal movements or "kick-charts" as they had not been shown to be beneficial and could increase maternal anxiety.[1]

The potential association of reduced fetal movements with adverse pregnancy outcomes are listed in the box. Up to 15% of pregnant women experience a change in fetal movements during their pregnancy.[4] Most women (about 70%) who perceive a reduction in fetal movements will have a normal outcome to their pregnancy.[1] However, 55% of women experiencing a stillbirth perceived a reduction in fetal movements before diagnosis.[5] Women who present with reduced fetal movements on two or more occasions are at increased risk of a poor perinatal outcome (including fetal growth restriction, preterm birth, or stillbirth) compared with those who attend only once.[6] Reduced fetal movements may represent an attempt by the fetus to conserve energy due to insufficient oxygen and nutrient transfer resulting from placental insufficiency.[7]

Associations with reduced fetal movements[1-7]

Factors that may decrease perception of fetal movement
- Early gestation
- Maternal stress
- Maternal obesity
- Oligohydramnios
- Anterior placenta

Maternal factors associated with reduced fetal movements
- Maternal alcohol consumption
- Medications (opioids and anxiolytics)

Fetal factors associated with reduced fetal movements
- Fetal sleep cycle (rarely exceeds 90 minutes)
- Anaemia
- Hydrops
- Congenital abnormalities
- Fetal growth restriction
- Stillbirth

HOW PATIENTS WERE INVOLVED IN THE CREATION OF THIS ARTICLE

The authors consulted Jane Brewin, chief executive of Tommy's Charity, and Elizabeth Hutton, chief executive of Kicks Count, who provided input to the articles based on their experiences of talking to women with reduced fetal movements. Tommy's Charity provided access to women with stories of care relating to reduced fetal movements. The lessons learnt from these representative stories (taking women's concerns seriously, being non-judgmental, and timely referral to the maternity unit) have been included in this article.

WHAT YOU NEED TO KNOW

- Reduced fetal movements are associated with adverse outcomes in pregnancy, including stillbirth, fetal growth restriction, placental insufficiency, and congenital malformations
- Perform clinical assessment of a woman with reduced fetal movements including symphysis-fundal height measurement and fetal heart auscultation
- Listen to the woman's concerns and refer her to the nearest maternity unit if she remains worried

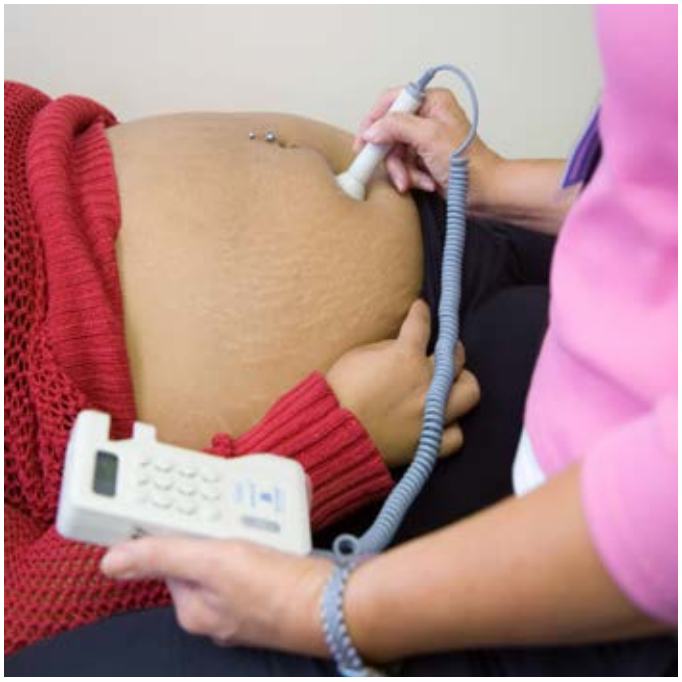
WHAT YOU SHOULD COVER

History

- *Time of onset and duration*—When did the patient first notice a reduction in fetal movements? Duration >24 hours is associated with poorer outcomes.
- *Previous episodes*—Is this the first presentation, or have there been recurrent episodes? Multiple consultations are a red flag and should prompt specialist review.
- *Gestation and parity*—Ask about the patient's estimated date of delivery and current gestation, and explore her previous obstetric history (fetal growth restriction, preterm birth, or stillbirth).
- *Maternal risk factors in current pregnancy which are red flags*—Smoking (active or passive), hypertension (chronic and gestational), pre-eclampsia, diabetes, advanced maternal age, and high maternal body mass index are red flags. Late booking is associated with poorer obstetric and neonatal outcomes.[8 9]
- *Antenatal progress to date*—Review previous ultrasound scans (indicating fetal or placental abnormalities), estimated fetal growth on ultrasound scan (growth <10th centile is a red flag[10]), and prenatal screening tests (indicating a higher risk of fetal chromosomal abnormalities).
- *Emotional wellbeing*—Recognise the emotional distress that some women may experience when presenting with reduced fetal movements and address these concerns directly.

Examination

- *Symphysis–fundal height*—Measure between the symphysis pubis and fundus of the uterus to screen for fetal growth restriction, and assess against gestation and previous measurements.
- *Customised centile chart*—Plot a customised centile (customised to correct for maternal height, weight, parity, and ethnic group) using computer software such as the one provided by the Perinatal Institute via the Gestation Network (www.gestation.net), referenced within the RCOG guideline.[10] A measurement below the 10th centile or serial measurements showing slow or static growth are clinically important[10] and need further specialised assessment (such as with ultrasound scan).
- *Fetal heart auscultation*—Listen for one minute using a handheld Doppler heart monitor to determine viability (normal range 110-160 beats/min). Measure the maternal pulse simultaneously to distinguish between maternal and fetal heart rates. If the fetal heart is not heard or is outside the normal range on Doppler auscultation, refer immediately to a maternity unit.



REX/SHUTTERSTOCK

Women's experiences

Theresa's story

"At around 34 weeks I felt my son wasn't moving as usual. I found the Kicks Count website, which explained how to monitor baby's movement, and a week later I was worried enough to go to hospital. A scan showed that I had an anterior placenta, which may have been making it difficult to feel my baby's kicks, and with that knowledge I felt reassured.

At a follow-up appointment I explained my worries about reduced fetal movement to a consultant, but she was pretty dismissive. I tried to put my fears to the back of my mind, but the advice from Kicks Count made me feel more and more certain that something was not right.

I went back to the hospital, where I felt that, as a first-time mum, I'd been labelled, my concerns not taken seriously. One midwife actually asked if I thought I was an anxious person. Forty minutes later my baby still hadn't moved, and the consultant explained I'd need an emergency caesarean section. I was angry that all my questions and fears, now justified, hadn't been investigated. I'd tell any pregnant woman to be aware of fetal movement and do not be afraid to keep asking questions."

Camilla's story

"My baby started moving at around 16 weeks and had a clear routine from the start. Then, one day I realised he wasn't moving as usual, his kicks less powerful and less frequent. I knew something was wrong, but when I arrived at maternity triage, predictably, I was asked; 'Is it your first?' and I knew I'd likely face that stereotype of an overanxious first-timer. The midwife told me our baby was fine, but I wasn't reassured.

Four times I reported reduced movements, yet, on each occasion, I'd be sent home, dismissed. I felt vulnerable, so scared that if I didn't push to have my concerns taken seriously, I could potentially lose my child.

The final time I raised the issue, I was reviewed by three consultants. The first and second tried to reassure me, before being bleeped to theatre. Thankfully, the third listened, read my notes thoroughly, and then booked a caesarean section for the following week. I can't explain the relief at finally being taken seriously. Nicolas arrived at 37^{+4} weeks and I fell in love instantly, relieved that he was here and safe. I'd tell any mum-to-be with concerns over fetal movement not to feel intimidated or worry about wasting anyone's time. It could save your baby's life."

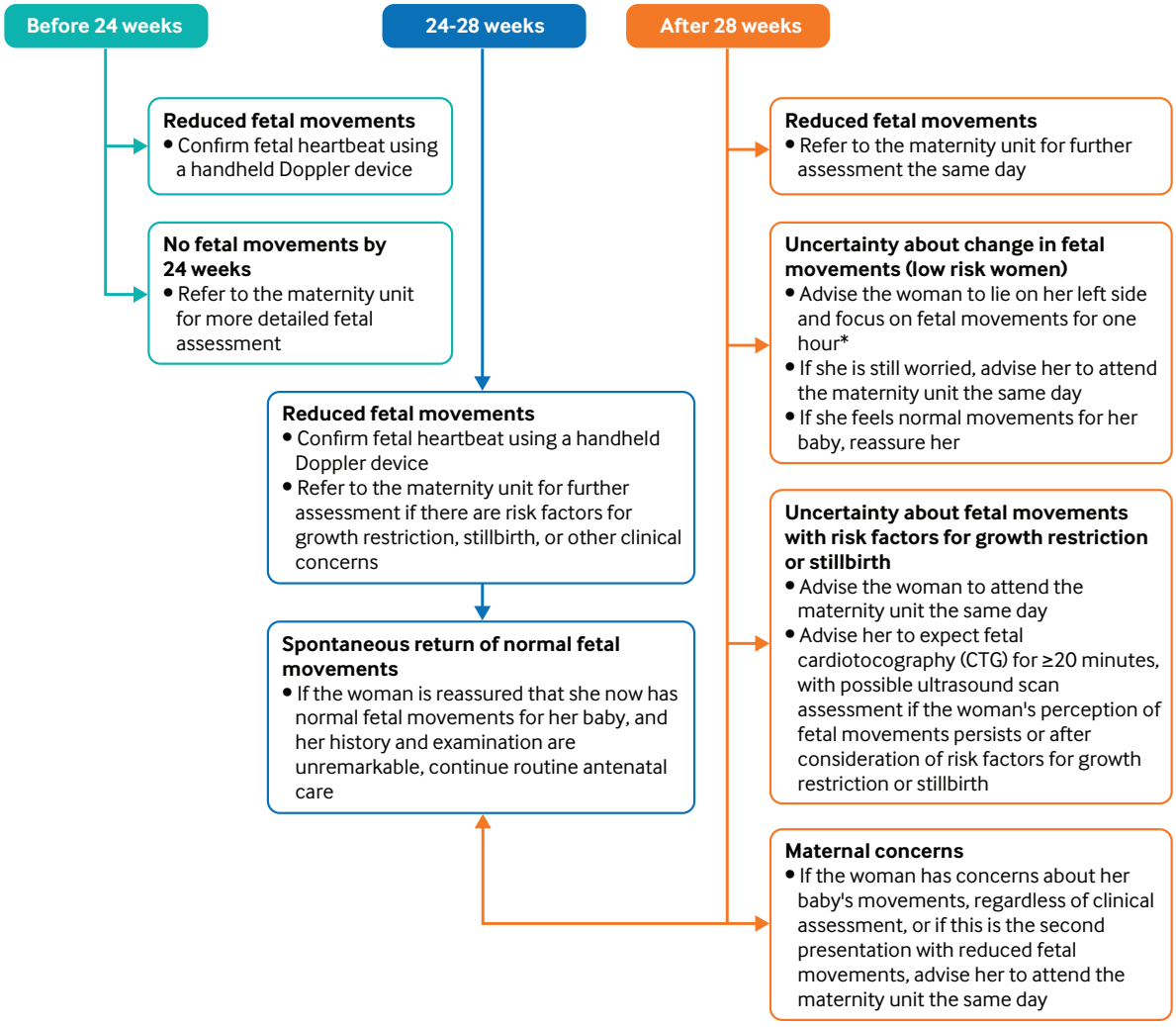


Fig 1 | Suggested approach to managing women in pregnancy with reduced fetal movements (from Royal College of Obstetricians and Gynaecologists guidelines[1])

WHAT YOU SHOULD DO

Figure 1 provides a suggested approach to managing women in pregnancy with reduced fetal movements.

Experiences of pregnant women can be read as part of the online supplement accompanying this article.

Competing interests: None declared.

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EDUCATION INTO PRACTICE

- Does your organisation or practice have a local guideline for reduced fetal movements?
- Think about the last pregnant woman you saw with reduced fetal movements. Did you feel confident assessing her clinically?
- How could you find out if pregnant women are aware that they should seek medical attention if there is a change or reduction in reduced fetal movements? Do they know who to contact?
- What proportion of women over 28^{+0} weeks' gestation with a history of reduced fetal movements have a cardiotocography performed to exclude fetal compromise?

FURTHER EDUCATIONAL RESOURCES

- Tommy's (www.tommys.org)
  —A UK registered charity that funds research into pregnancy problems and provides pregnancy health information to parents, including on reduced fetal movements (www.tommys.org/pregnancy-information/symptom-checker/my-babys-movements-have-slowed-down)
- Kicks Count (www.kickscount.org.uk)
  —A UK registered charity that aims to ensure that all pregnant women are aware of how important their babies' movements are, know the most up-to-date recommendations for monitoring movement, and have the confidence to speak to a medical professional if they have any concerns
- Each Baby Counts (www.rcog.org.uk/eachbabycounts)
  —A national quality improvement programme launched by the Royal College of Obstetricians and Gynaecologists to reduce the number of stillbirths, neonatal deaths, and brain injuries as a result of incidents occurring during term labour. Data are being analysed from all UK units to improve future care and make recommendations on how to improve practice at a national level
- Saving Babies' Lives (www.england.nhs.uk/mat-transformation/saving-babies/)
  —A care bundle for reducing stillbirth, developed by NHS England. It comprises four elements: reducing smoking in pregnancy, risk assessment and surveillance for fetal growth restriction, raising awareness of reduced fetal movements, and effective fetal monitoring during labour. An advice leaflet on reduced fetal movement should be provided to all pregnant women by the 24th week of pregnancy, and fetal movements discussed at every subsequent contact.

CASE REVIEW

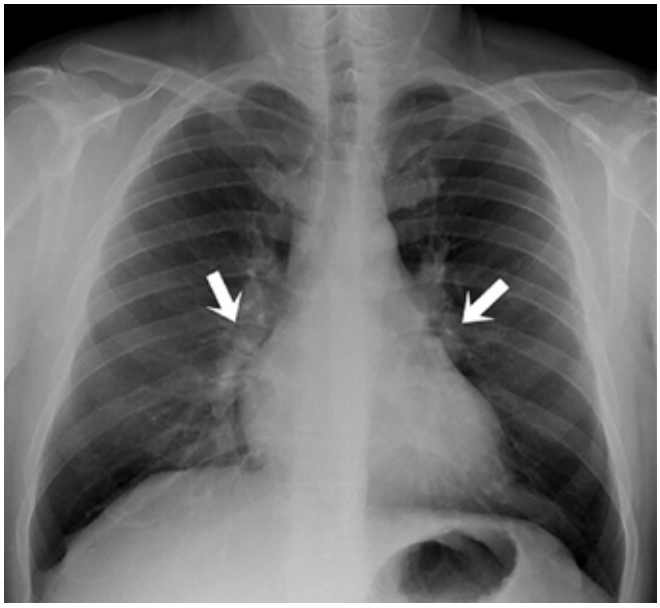
Just a painful swelling of the ankles?

A 41 year old man presented with a three week history of acute painful swelling of both ankles accompanied by local erythema and heat, morning stiffness of short duration, and fever. During the fourth week, a tender nodular erythematous eruption developed in both legs, suggestive of erythema nodosum. The man had no personal, family, or epidemiological medical history. He reported no previous infections, trauma, or new medications. On physical examination, his temperature was 38.2°C. There was erythema, oedema, pain, and heat in both ankles, with a normal range of movement in the ankle joints, and lesions compatible with erythema nodosum on the anterior aspect of both legs.

Laboratory results showed

- Leucocytosis of 14.5×10^9/L (reference range 4-10) with neutrophilia
- Erythrocyte sedimentation rate of 56 mm in first hour (1-20)
- C reactive protein 9.53 mg/dl (‹0.5)
- Normal results on tests of renal, liver, and thyroid function
- Normal levels of calcium (serum and urinary); 1,25-dihydroxyvitamin-D, and angiotensin converting enzyme
- Negative blood and urine cultures, viral serology (Epstein-Barr, cytomegalovirus, HIV, hepatitis, parvovirus-B19), interferon-gamma-release-assay, and negative autoantibody test results

Chest radiography showed mild bilateral hilar lymphadenopathy (figure). A chest computed tomography scan and 18F-FDG-PET/computed tomography scan confirmed symmetrical bilateral hilar lymphadenopathy, with no other findings. Ankle ultrasound showed bilateral peri-articular soft tissue oedema, without synovitis or tenosynovitis.



Bronchoalveolar lavage fluid contained 45% lymphocytes, 11.1 CD4/CD8 cell ratio, and negative bacteriological examination. Bronchial aspiration showed polymorphonuclear cells. Pulmonary function tests were normal.

1 What is the most likely diagnosis?

2 Are further tests needed to confirm the diagnosis?

3 How is this condition managed and what is the prognosis?

Submitted by Gisela Eugénio, João Tavares, Mary Marques, Cátia Duarte, and J A P da Silva

Patient consent obtained.

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If you would like to write a Case Review for Endgames, please see our author guidelines at http://bit.ly/29HCBAL and submit online at http://bit.ly/29yyGSx

answers

CASE REVIEW

Just a painful swelling of the ankles?

1 The combination of arthralgia (with ankle involvement), erythema nodosum, and bilateral hilar lymphadenopathy make Löfgren syndrome, an acute form of sarcoidosis, the most likely diagnosis.

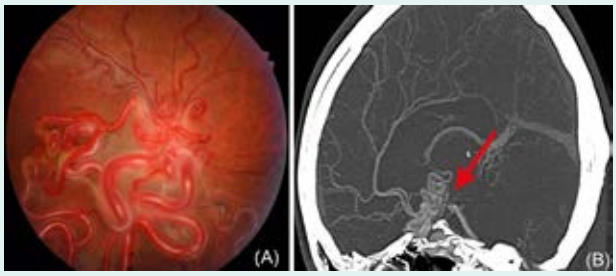
2 The diagnosis of Löfgren syndrome relies on clinical history, examination findings, and exclusion of other diagnoses. A hilar lymph node biopsy is often needed to verify a diagnosis of sarcoidosis; however, the presence of typical features of Löfgren syndrome (bilateral hilar lymphadenopathy, erythema nodosum, and arthritis/arthralgia) confers a high diagnostic specificity, and hence invasive diagnostic tests are usually not needed.

3 The main treatment for musculoskeletal symptoms and erythema nodosum (both cardinal manifestations of Löfgren syndrome) is non-steroidal anti-inflammatory drugs (NSAIDs) and bed rest. In refractory cases, a short period of oral corticosteroids might be given. Löfgren syndrome generally has a good prognosis, with ›80% of patients achieving complete remission in 3-24 months.

Wyburn-Mason syndrome

A 22 year old woman reported a superior visual field defect in her right eye. The defect had remained unchanged for years and she had no history of trauma or headache. Her best corrected visual acuity was 10/20 (right) and 20/20 (left). Funduscopy of the right eye showed a retinal arteriovenous malformation with large calibre convoluted, tortuous retinal vessels extending from the disc (figure A). Magnetic resonance imaging of the brain revealed an arteriovenous malformation in the right suprasellar region, near the optic chiasm (figure B, arrow). Given the patient's risk profile, conservative management was preferred.

Wyburn-Mason syndrome is a rare disease in which vascular dysgenesis affects the retina and brain. It can also be associated with vascular abnormalities of the maxilla or mandible, which can cause substantial bleeding during dental procedures. Patients with large retinal arteriovenous malformations should therefore undergo brain imaging.



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Patient consent obtained.
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Use a ruler

The ABCDE system—a mnemonic for asymmetry, border, colour, diameter, and evolution—is a well established way to evaluate suspicious skin lesions. A survey from North America finds that, when it comes to assessing diameter, it's much better to use a ruler to measure the lesion than to hazard a guess (*JAMA Dermatol*). Although dermatologists were more accurate than medical students or non-specialists in estimating lesion size by eye, they managed to judge only half the lesions to within 1 mm of their actual size.

Ventricular drains

Although the insertion of an external ventricular drain can be a lifesaving procedure in cases of intracranial haemorrhage and traumatic brain injury, it carries a risk of introducing infection into the central nervous system. A prospective multicentre survey in the UK and Ireland estimates this risk at around 10% overall (*J Neurol Neurosurg Psychiatry*). The risk of infection was greater if the catheter remained in place for more than eight days or if repeated sampling from the catheter was carried out. However, the type of catheter and whether it had been impregnated with antibiotics or silver made no difference.

Congenital abnormalities and autism spectrum disorders

Hypospadias and cryptorchidism are congenital abnormalities of the male reproductive tract linked to prenatal androgen deficiency. There is also a suggestion that abnormal gestational exposure to androgens might be part of the aetiology of autism spectrum disorder. If so, one would expect there to be an association between the two conditions, and that's what a study of nearly a quarter of a million live male singleton births in Israel reports (*Am J Epidemiol*). Even so, the association was weak, with odds ratios of less than 2 for autism spectrum disorder among boys with these urogenital tract anomalies, which makes it hard to conclude anything definite.

The effect of testosterone on cardiovascular biomarkers

On the subject of androgens, trials have shown that testosterone replacement in older men with low testosterone levels has a modest benefit on sexual function and mood. But is there a price to be paid in terms of increased cardiovascular risk? The trials were too small to provide an answer in terms of clinical events, but an investigation of biochemical markers suggests not (*J Clin Endocrinol Metabol*). After a year of testosterone treatment there were small reductions in cholesterol and insulin but no changes in other indicators of glucose metabolism or in markers of inflammation or fibrinolysis.

Cholera in Haiti

A microbiologist writing in the *London Review of Books* points out that the current scandal over aid workers in Haiti is neither the first nor the worst. When the United Nations flew in Nepalese troops after the 2010 Port-au-Prince earthquake they brought cholera with them. The disease spread rapidly and by June 2014 there had been 700 000 cases and more than 8000 deaths. Genome sequencing of cholera bacteria showed beyond doubt that the soldiers had been the source, but the UN denied responsibility until December 2016, when Ban Ki-moon made a public apology.

Cite this as: *BMJ* 2018;360:k986