education

ART OF MEDICINE

The "lonely predicament"

I am sometimes asked to see children in families where a death is anticipated. I help them understand more about the illness and provide a safe space in which they can express their feelings about what is happening. But surprisingly often, my young patients make it quite clear that this is not something they want to do.

In the literature I found a paper that struck a chord. It described the helplessness, apprehension, and loneliness of children of a dying parent—hence the phrase "lonely predicament" in its title.[1] The children studied chose to talk mainly about their lives in general rather than their parent's illness and were subsequently found to be doing better than a comparison group, who had not had the chance to talk to someone.

So I continue to find myself engaged in lively discussions with children in this situation about their passions: a steady focus on their lives and dreams while the world around them feels so uncertain.

C S Lewis described the death of his mother poignantly: "It was sea and islands now; the great continent had sunk like Atlantis." This paper suggests that we may all have a useful role in a child's world at such a time: that of helping them hold on to who they are in spite of what is happening. We may not be able to replace "the great continent," but we can sometimes be a port in a storm.

Gillian Colville, consultant clinical psychologist, St George's University Hospitals NHS Foundation Trust, London, UK gcolvill@sgul.ac.uk

1 Rosenheim E, Reicher R. Children in anticipatory grief: the lonely predicament. *J Clin Child Psychol* 1986;15:115-9.

Cite this as: *BMJ* 2015;350:h3346

We welcome contributions to this column via our online editorial office: https://mc.manuscriptcentral.com/bmj

FAST FACT—INTRINSIC RENAL PATHOLOGY

Most cases of acute kidney injury are pre-renal, so a high index of suspicion for intrinsic renal disease is required. In particular, the presence of haematuria or proteinuria on urine dipstick test should always be a prompt.

Other suggestive features include:

- Signs and symptoms of vasculitis
- Significant proteinuria based on laboratory measurement
- Recent change in medications such as proton pump inhibitors, penicillin based antibiotics, and NSAIDs

BMJ Learning

For more information visit BMJ Learning
http://bit.ly/1XA3wBd

PRACTICE UPDATES

The management of nausea and vomiting of pregnancy and hyperemesis gravidum

Nausea and vomiting of pregnancy (NVP) should be diagnosed only if the onset occurs in the first trimester and other causes have been ruled out first, says a new Green-top Guideline from the Royal College of Obstetricians and Gynaecology. Severity can be scored with the pregnancy-unique quantification of emesis (PUQE) score. Diagnose hyperemesis gravidum in cases of protracted NVP with >5% loss of prepregnancy body weight, dehydration, and electrolyte imbalance. Manage mild NVP with oral antiemetics such as cyclizine. Consider inpatient management if patients have continued nausea and vomiting and are unable to keep down oral antiemetics, or if NVP are associated with ketonuria or weight loss (>5% of body weight) despite oral antiemetic. Admit patients with a confirmed or suspected comorbidity such as urinary tract infection and inability to take oral antibiotics.

http://bit.ly/29YdbeD

Alair bronchial thermoplasty system for adults with severe, difficult to control asthma

Alair bronchial thermoplasty uses radiofrequency energy delivered during bronchoscopy to reduce the amount of smooth muscle in the airway walls to improve symptoms in adults with severe, difficult to control asthma. A NICE Medtech innovation briefing (MIB) concludes that the procedure is associated with some patient benefits such as improved quality of life and morning peak expiratory flow, with mixed evidence over other outcomes such as hospital visits.

http://bit.ly/29WSxdS

CPD/CME

You can gain CPD points from your reading by recording what you have read in your appraisal folder. You should try to link your reading back to a learning need and also consider how you plan to improve your practice as a result of your learning.
http://learning.bmj.com

We print a statement on financial interests and patient partnership with each education article because they are important to us. We have resolved to reduce the involvement of authors with financial interests that *The BMJ* judge as relevant. We encourage and make clear how patients have been involved and shaped our content. More details can be found on thebmj.com.

Congenital heart disease in adults

CPD/CME
1 CREDIT

Bejal Pandya, Shay Cullen, Fiona Walker

Grown-up Congenital Heart Disease (GUCH) Clinic, Barts Heart Centre, St Bartholomew's Hospital, London, UK

Correspondence to: F Walker fiona.walker@bartshealth.nhs.uk

This is an edited version, full version on thebmj.com

Heart disease is the most common birth defect, affecting nine in 1000 babies born in the UK. The spectrum of the underlying lesions ranges from a simple septal defect to more complex structural abnormalities.

Most of those born with cardiac defects lead active lives and survive well into late adulthood. These adults with congenital heart disease have often had previous surgical corrections or percutaneous catheter treatments and are left with residual structural defects, valve lesions, or ventricular dysfunction.

NHS England has recently issued a review on the care of patients with congenital heart disease.[1] Specialist centres have been evaluated against agreed standards and designated or commissioned accordingly, in order to provide varying levels of care across a geographical network. This review provides practical advice for non-specialists to help understand the longer term problems associated with congenital heart disease and how these are managed.

What are the usual clinical findings in adults with congenital heart disease?

Findings on clinical examination may differ between adults with congenital heart disease. Table 1 lists the common variants.

The electrocardiogram usually shows sinus rhythm, with a normal PR interval. However, axis deviation is common after surgical corrections due to conduction abnormalities and rotation of the heart after manipulation.

Box 1 lists the symptoms that should prompt discussion with a specialist.

WHAT YOU NEED TO KNOW

- Adults with congenital heart disease are generally reviewed annually by a specialist congenital cardiologist
- Before general anaesthesia, ensure patients are reviewed by a specialist congenital cardiologist
- Offer women preconception counselling by a specialist
- Progesterone only contraception is safe for all women with congenital heart disease
- Adults with congenital heart disease are highly susceptible to infective endocarditis

Box 1 | Red flag symptoms in patients with congenital heart disease

- Progressive or paroxysmal breathlessness—consider new onset of arrhythmia
- Palpitations—consider new arrhythmia or deteriorating valve or ventricular function
- Fevers, malaise, weight loss—consider infective endocarditis until proved otherwise
- Rapidly increasing weight or peripheral oedema—consider arrhythmias or heart failure

What imaging is most helpful for routine assessment during follow-up?

Most adults with congenital heart disease are asymptomatic, therefore imaging and functional tests are used to assess the progression of disease and monitor residual lesions (eg, ventricular function or valve regurgitation). Several advanced imaging techniques are widely used (box 2), which can evaluate repaired or newly presenting congenital heart disease.[1]

What types of lesions are diagnosed?

Many congenital heart disease lesions will be diagnosed and repaired in childhood. Some, however, are

Table 1 | Typical, abnormal, and atypical findings in adults with congenital heart disease

	Findings		
Measurements	Normal	Abnormal	Atypical but normal
Pulse	Regular	Irregular	Absent in arms after Blalock-Taussig shunt (see infographic) or left subclavian artery flap repair of coarctation of the aorta
Blood pressure (measured in all four limbs)	Normotensive for age	Hypertensive for age or >20 mm Hg difference between arms and legs then consider coartaction of the aorta or re-coarctation of the aorta	Absent in left arm; left subclavian flap repair of coarctation of the aorta
Peripheral oxygen saturation	>95%	<90%	Fontan circulation with fenestration; aortopulmonary collaterals; cyanotic heart disease (finger clubbing)
Auscultation (use anatomical landmarks to describe location of murmurs)	Heart sounds: S1 (mitral and tricuspid valve closure) and S2 (aortic and pulmonary valve closure)	S3 (ventricular filling sound), S4 (increased resistance to ventricular filling)	Split S2: right bundle branch block, atrial septal defect, pulmonary stenosis Ejection click: bicuspid aortic valve Systolic clicks: Ebstein's anomaly of the tricuspid valve

diagnosed for the first time in adults, especially atrial septal defects and coarctation of the aorta.

Atrial septal defect (ASD)

An atrial septal defect is a hole in the atrial septum, which allows blood to shunt from the left to right atrium. This causes right heart volume overload and dilatation. Secundum atrial septal defect is one of the commonest types, with other forms including sinus venosus atrial septal defect and primum atrial septal defect (fig 1). Closure of an atrial septal defect is indicated if there is right heart dilatation and the oxygen saturation and pulmonary vascular resistance is normal. Symptoms are subtle and include breathlessness, fatigability, and exercise intolerance. Atrial septal defects can be closed surgically (all forms) or percutaneously with a device (secundum atrial septal defect only, fig 2). Percutaneous and surgical closures are low risk interventions, if there are no major comorbidities such as coronary disease or renal impairment. There is no upper age limit for closure of an atrial septal defect. After closure there is a risk of atrial fibrillation, stroke, and heart failure, as with late or adult diagnosis there is often severe enlargement of the right atrium and right ventricle, which may persist.[5]

Ventricular septal defect (VSD)

Ventricular septal defects allow shunting of blood from the left to right ventricle. When diagnosed incidentally in a healthy asymptomatic adult, these defects are often small or restrictive and of no haemodynamic relevance. The main risk to health is an increased susceptibility to infective endocarditis, most likely from an oral source. It is therefore good practice to remind patients to maintain good dental hygiene.[6] If a large ventricular septal defect is present in adults, it will cause volume loading and dilatation of the left ventricle. If a large defect is present from childhood, this will cause pulmonary hypertension and Eisenmenger's syndrome (see box 3).

Patent ductus arteriosus (PDA)

The ductus arteriosus is a connection between the pulmonary artery and aorta, which is present in the fetal circulation. If it remains patent, it can cause dilatation of the left ventricle. A continuous murmur may be heard. Symptoms are generally absent, although some report palpitations and awareness of a rapid heart rate. Closure is indicated if left ventricular dilatation is present. Most can be closed percutaneously with a device.

Coarctation of the aorta (CoA)

Coarctation of the aorta is a narrowing of the aorta, distal to the origin of the left subclavian artery. Consider this condition if systemic hypertension is detected in a young adult (<40 years). The femoral pulses may be absent or weak. Relief of coarctation of the aorta is indicated if there is systemic hypertension or a more than 50% luminal narrowing, or both, and can usually be achieved by percutaneous stenting (fig 3). Symptoms are generally absent, although leg claudication may occur with severe coarctation.

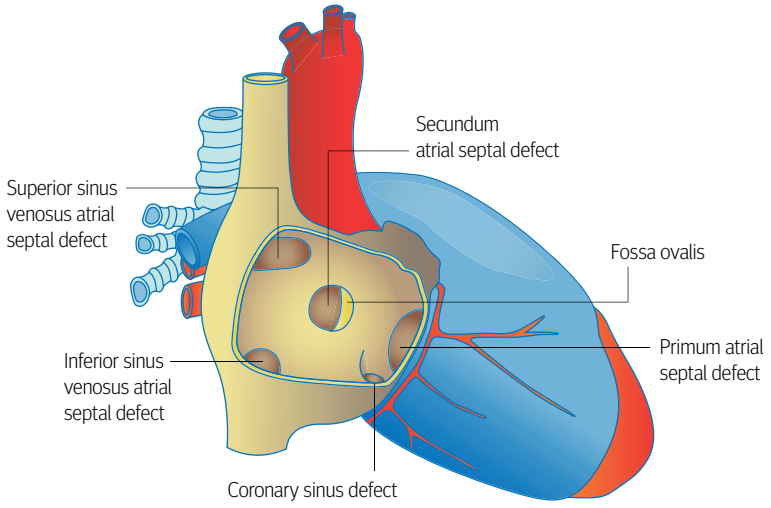


Fig 1 | Various types of atrial septal defects

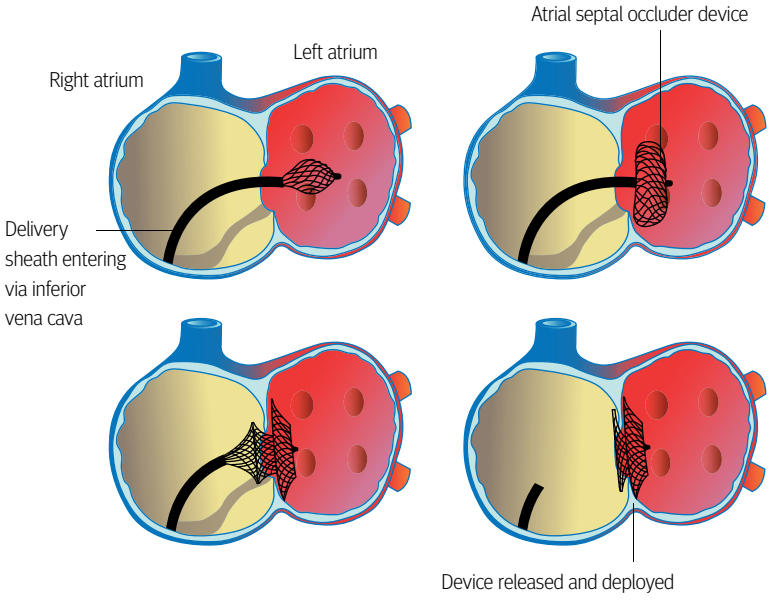


Fig 2 | Closure of secundum atrial septal defect using percutaneous device

Box 2 | Advanced imaging techniques for congenital heart disease

- **Echocardiography**—transthoracic echocardiography (TTE) is the mainstay for routine assessment and surveillance of adults with congenital heart disease. TTE can make a qualitative assessment of ventricular function, and Doppler echocardiography assesses the severity of haemodynamic or valve lesions. Tricuspid regurgitation jet can be used to estimate right heart pressures. TTE is done at each clinic review. With TTE, ultrasound has to penetrate between ribs and through the chest wall, therefore fat and heavy set chest musculature is a barrier to good image quality. If TTE images are poor, transoesophageal echocardiography (TOE) is an alternative
- **Cardiovascular magnetic resonance imaging (CMRI)**—CMRI provides 3D and 4D (real time blood flow) imaging. It is accurate for defining morphology and quantifying ventricular function.[2 3] If CMRI is contraindicated owing to the presence of metallic implants or implantable electronic devices, then computed tomography (CT) is an alternative for providing 3D images
- **CT**—current CT scanners, electron beam CT (EBCT), or multislice spiral CT have a fast image acquisition time, which reduces scan time and therefore radiation dose. CT only provides qualitative (observational) rather than quantitative (volumes, ejection fractions %) data. CT uses ionising radiation, and must be used judiciously as patients potentially require several scans over their lifetime[4]

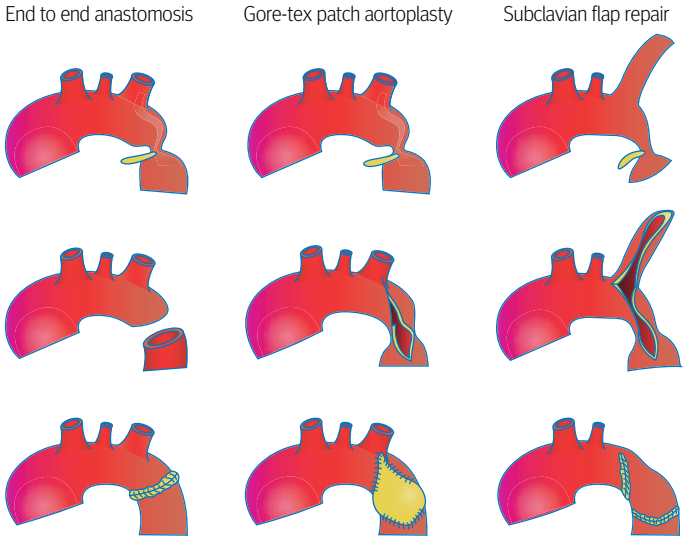


Fig 3 | Various types of repair of coarctation of the aorta

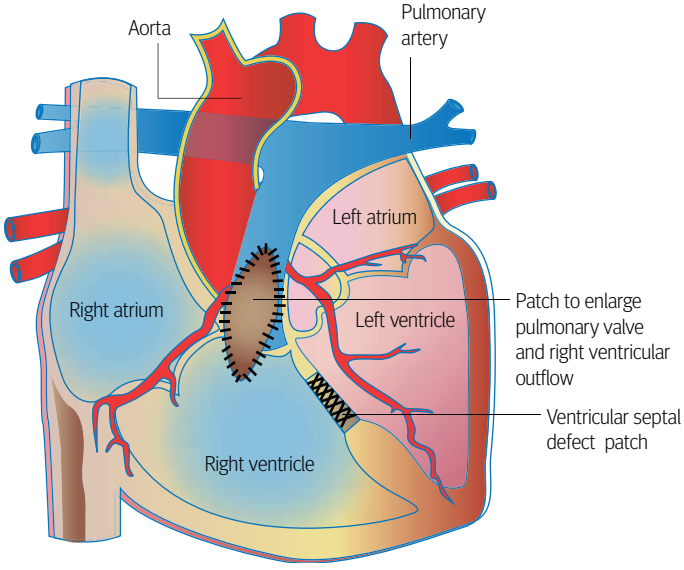


Fig 4 | Repair of tetralogy of Fallot with transannular patch. The ventricular septal defect is closed with a patch and the subpulmonary stenosis is relieved with a patch across the subpulmonary area and pulmonary artery

Box 3 | Mechanisms underlying cyanotic congenital heart disease

- Eisenmenger's syndrome—an uncorrected left to right intracardiac shunt (eg, ventricular septal defect) causes a high pulmonary vascular resistance, shunt reversal, and desaturation
- Palliative surgeries—these create systemic (aorta) to pulmonary arterial shunts
- Complex congenital heart disease lesions, whereby a biventricular repair is not possible, leading to a "mixed circulation" of saturated and desaturated blood and single ventricle physiology

HOW PATIENTS WERE INVOLVED IN CREATING THIS ARTICLE

This article was submitted before we asked authors to involve patients and report any contributions.

Pulmonary stenosis (PS)

Congenital pulmonary stenosis might be subvalvular, at valve level, or supravalvular. In adults it is often diagnosed when an incidental ejection murmur is heard and echocardiography detects outflow tract obstruction. The condition is usually asymptomatic, but if there is severe obstruction then breathlessness, syncope, or presyncope might occur. Consider treatment in patients with symptoms or severe pulmonary stenosis (peak gradient >70 mm Hg), or both. Valvular stenosis can be relieved by percutaneous balloon dilatation, whereas subvalvar and supravalvar stenosis require surgical correction.

Which repaired lesions are seen and how are they managed?

A wide spectrum of repaired lesions is seen in adults, ranging from simple repairs such as patch closure of an atrial septal defect, to the creation of highly complex circulations such as the Fontan palliation (see the infographic for scars associated with previous surgery). The most commonly encountered repaired lesions are:

Atrial and ventricular septal defects (ASD)

Repairs in infancy are generally curative. Patients do not require long term follow-up unless there is residual left ventricular dilatation (in case of ventricular septal defect) or right ventricular dilatation (atrial septal defect).

Atrioventricular septal defect (AVSD)

Late complications in patients with repaired atrioventricular septal defects include progressive left atrioventricular valve regurgitation, subaortic stenosis, and atrioventricular block. At specialist follow-up, patients undergo echocardiography to assess the atrioventricular valves, and routine electrocardiography for atrioventricular conduction. Approximately 15% of patients require repeat surgery.[7]

Tetralogy of Fallot (TOF)

Surgical repair of tetralogy of Fallot (fig 4) in infancy or childhood incorporates patch closure of ventricular septal defect, resection of the infundibular septum, and transannular patch augmentation of the right ventricular outflow tract. All of those who undergo transannular patch augmentation have residual severe pulmonary regurgitation, and pulmonary valve implantation is needed during follow-up.[8-11] Late complications include ventricular (4%) and atrial arrhythmias (3%), heart failure, and sudden death (2%); therefore all patients need specialist lifelong follow-up.

Coarctation of the aorta (CoA)

Adults with previous surgical repair of coarctation of the aorta might have had a variety of surgical procedures (fig 3). Re-coarctation can occur in all types of repair, whereas aneurysm formation is more common in those who undergo repair by a Gore-tex patch aortoplasty. Consider re-coarctation in those with new hypertension or worsening hypertension.

Transposition of the great arteries

In transposition of the great arteries, the aorta arises from the right ventricles and the pulmonary artery from the left ventricles. There are two types of surgical repair. Before 1980, an atrial switch repair was performed (the Mustard or Senning operations, fig 5). This involved complex intracardiac suturing to create tunnels (baffles), which redirect systemic and pulmonary venous return. The right ventricle remains subaortic, supporting systemic cardiac output. Late complications include right ventricular dysfunction, heart failure, and arrhythmias. In the 1980s, the arterial switch became the repair of choice (fig 6), restoring normal anatomy with a subaortic left ventricle. The coronaries are reimplanted, so late complications include ostial coronary stenosis and neovalvular regurgitation.

Is antibiotic prophylaxis required against infective endocarditis?

Data from randomised controlled trials are lacking on the use of antibiotic prophylaxis in congenital heart disease, and guidelines vary between the UK, Europe, and North America. Concerns have been raised about the UK National Institute for Health and Care Excellence guidelines,[12] which have more or less abandoned antibiotic prophylaxis. Patients with congenital heart disease are a highly susceptible group.[12] Many specialists still recommend prophylaxis for specific patients; our own service recommends prophylaxis with mechanical valves and for those with a history of endocarditis. Good oral hygiene should be reiterated for all patients.[13 14] A high index of suspicion and low threshold for taking blood cultures in this patient population is also advised because treating infective endocarditis after oral antibiotics leads to a much more protracted and potentially worse outcome.

How are patients with cyanotic congenital heart disease managed?

Cyanosis is defined as a deoxygenated haemoglobin concentration of more than 50 g/L, and peripheral oxygen saturations are often less than 85%. Patients have central and peripheral cyanosis with finger clubbing and are susceptible to cerebral abscesses,

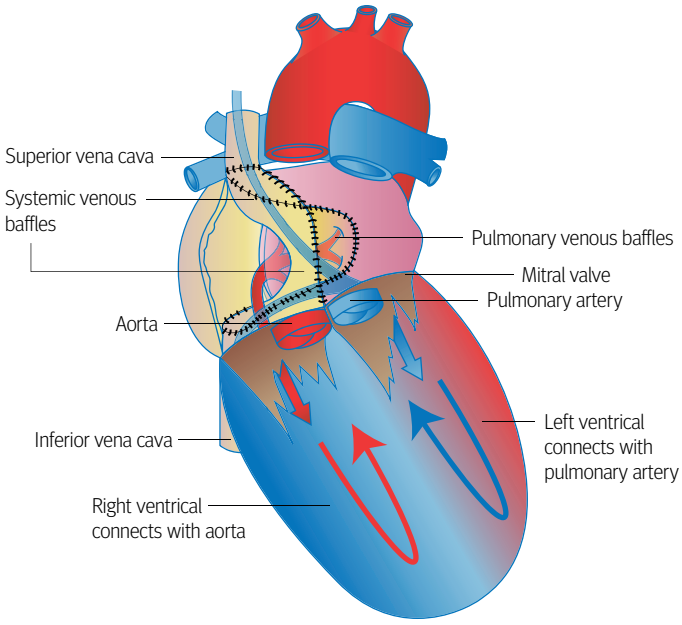


Fig 5 | Atrial switch (Mustard) repair of transposition of the great arteries. Blood flow is diverted through surgically created tunnels (baffles) in order to correct blood flow so that desaturated blood flows into the pulmonary artery and oxygenated blood flows physiologically into the aorta

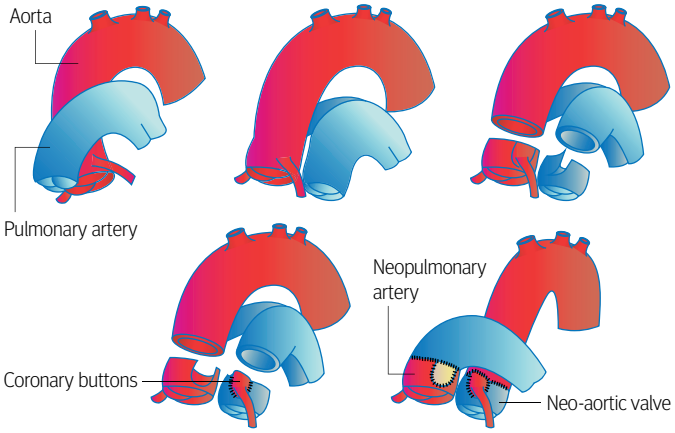


Fig 6 | Arterial switch repair of transposition of the great arteries. This provides an anatomical correction whereby the great vessels are transposed and the native pulmonary valve becomes the valve for systemic outflow and the aortic valve becomes the valve for outflow to the pulmonary artery. This procedure also requires re-implantation of the coronary arteries

Table 2 | Pregnancy risk in various heart disorders

WHO 1 (low risk)	WHO 2 (small increased risk)	WHO 2 or 3 (small increased risk mortality or moderate morbidity)	WHO 3 (significantly increased risk maternal mortality or morbidity)	WHO 4 (extremely high risk of maternal mortality or morbidity)
Repaired atrial septal defect	Unoperated atrial septal defect, unoperated ventricular septal defect	Mild left ventricular impairment	Mechanical valves	Pulmonary hypertension
Repaired ventricular septal defect	Repaired tetralogy of Fallot	Hypertrophic cardiomyopathy	Systemic right ventricle	Severe left ventricular impairment ejection fraction <30%
Ectopy	Arrhythmias	Repaired coarctation of the aorta	Fontan	Previous peripartum cardiomyopathy with residual left ventricular impairment
Mild pulmonary stenosis		Marfan syndrome aorta <45 mm	Cyanotic	Severe symptomatic aortic stenosis
Repaired patent ductus arteriosus			Complex congenital heart disease	Severe mitral stenosis
Repaired anomalous pulmonary veins			Aortopathy in Marfan syndrome 40-45 mm	Aorta >45 mm in Marfan syndrome
Mitral valve prolapse			Aortopathy in bicuspid aortic valve 40-45 mm	Aorta >50 mm in bicuspid aortic valve

WHO=World Health Organization.

arrhythmias, and heart failure. Different mechanisms underlie cyanotic congenital heart disease (box 3).

Clinically, there is erythrocytosis and increased haemoglobin levels (often >200 g/L). There is a prothrombotic (due to polycythaemia) and bleeding tendency (dysfunctional low platelet levels). Patients are susceptible to venous thromboembolism as well as epistaxis and increased bleeding after surgical procedures. Venesection is not indicated unless symptoms of hyperviscosity such as headache, myalgia, or blurred vision are intolerable and the haematocrit is more than 65%.[15] Iron deficiency can occur with venesection, leading to microcytosis, which is associated with adverse outcome. Oral or intravenous iron supplementation may be needed to maintain normal red cell indices. General anaesthesia and pregnancy are high risks for this patient group.

How does congenital heart disease affect women's health?

Contraception advice

Congenital heart disease lesions with residual chamber dilatation are associated with atrial or ventricular dysrrhythmias, or both. This increases the risk of thromboembolism and means that for many women, contraceptives with oestrogen are best avoided. Progesterone only contraceptives are in the main safe and preferred, especially for those with more complex congenital heart disease—eg, Fontan and people with cyanosis.[16]

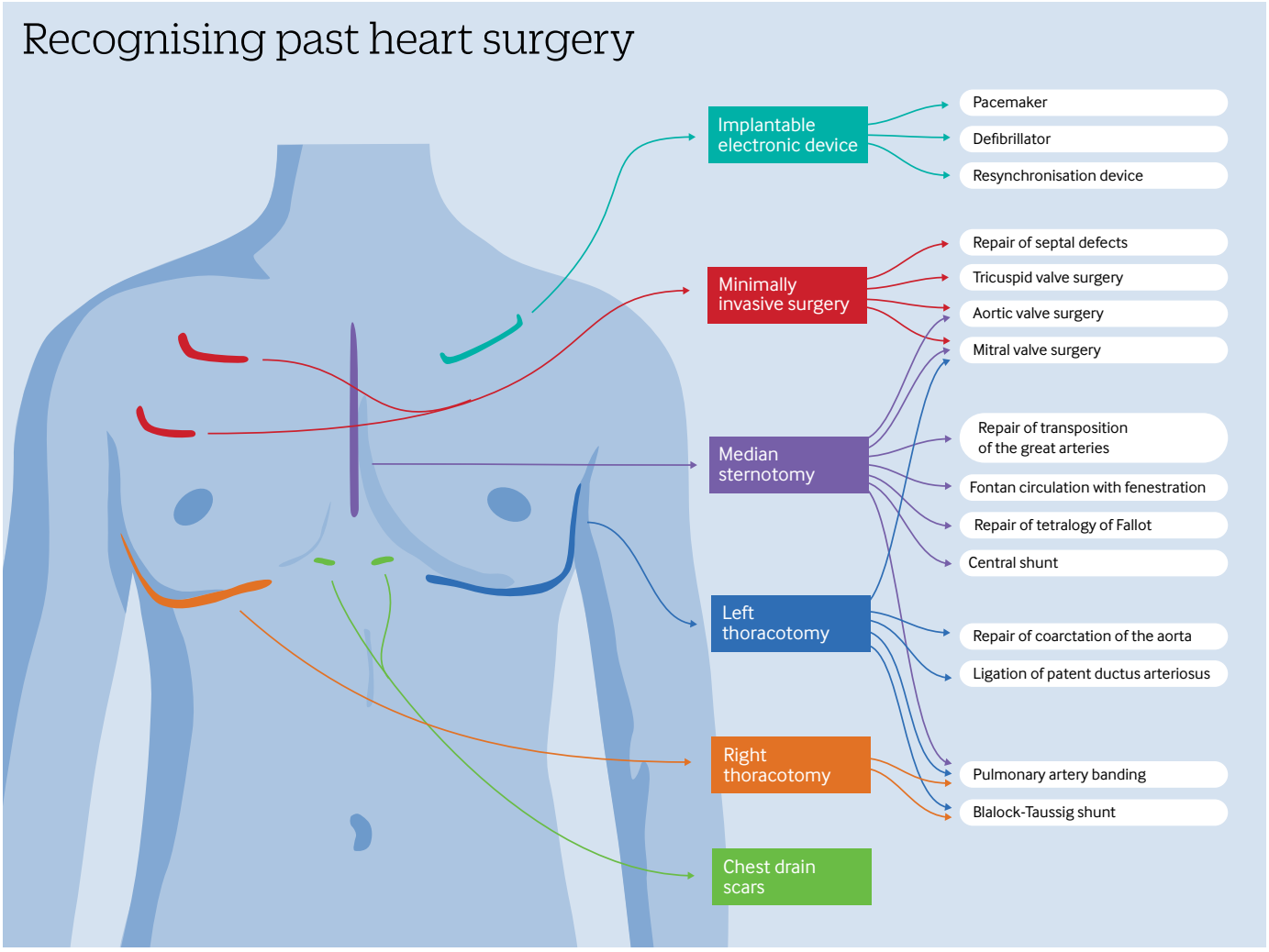
Advice in pregnancy

Most women with congenital heart disease have their babies at term and can have normal vaginal deliveries. However, there is a 4-5% increased risk of congenital heart disease in babies born to mothers with the condition. Women with congenital heart disease who are considering pregnancy should receive preconception counselling by a specialist congenital cardiologist. The European Society of Cardiology has published guidelines for managing pregnancy in women with heart disease and the risk in various cardiac disorders (table 2).[17]

Competing interests: None declared.

Cite this as: *BMJ* 2016;354:i3905

Find this at: http://dx.doi.org/10.1136/bmj.i3905



What is the best way to manage neurogenic bowel dysfunction?

Doreen McClurg,[1] Christine Norton[2]

A single episode of faecal incontinence can precipitate a fear of repetition and may lead to reduced social activity and isolation.[1] Bowel dysfunction, faecal incontinence, and constipation have a prevalence of around 70% in people with central neurological disease such as Parkinson's disease, stroke, multiple sclerosis, or spinal cord injury.[2]

Tools are available aid to conversation about bowel dysfunction (fig 1). Constipation may lead to difficult evacuation, abdominal pain and bloating, haemorrhoids, anal fissures, rectal bleeding, prolapse, and autonomic dysreflexia. Hospital admission for impaction occurs more than twice as frequently as in healthy people.[3-5] Management to ameliorate either incontinence or constipation risks precipitating the other. The condition is time consuming and arduous and causes anxiety to the patient and care givers.[6]

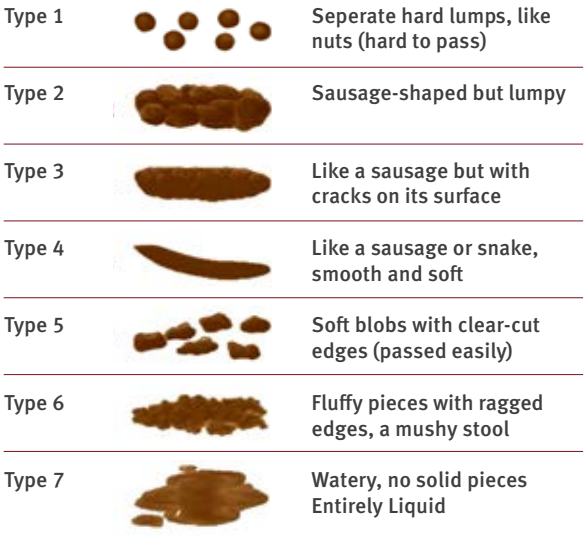


Fig 1 | Bristol Stool Chart

What is the evidence of the uncertainty?

Various approaches have been tried for faecal incontinence and constipation in these patients, but limited research is available to develop recommendations for care. An updated Cochrane review in 2014 identified 20 randomised controlled trials (902 participants) comparing different management strategies (see table on thebmj.com). Five studies that reported on the use of cisapride and tegaserod were excluded from this update owing to adverse cardiovascular effects. Limited evidence from individual trials suggests improvement in bowel function with a bulk forming laxative (psyllium), an isosmotic macrogol laxative, abdominal massage, transanal irrigation, and sacral nerve stimulation. Some suppositories were effective in aiding bowel movements, particularly with morning use. One study found digital evacuation of stools to be more effective than oral or rectal drugs. Overall, the review concluded that the evidence was of very poor quality as most studies are small and not reliably reported.[2]

Using the same search criteria, we updated the Cochrane searches and identified eight additional randomised controlled trials (367 participants) on the role of physical interventions such as daily standing and abdominal massage, acupuncture, electrical stimulation, and drugs. However, these additional studies are small and at high risk of bias (see table on thebmj.com).

Is ongoing research likely to provide relevant evidence?

We searched clinicaltrials.gov for ongoing studies and found one trial, which is our own Health Technology Assessment funded trial comparing abdominal massage plus advice with advice alone in people with multiple sclerosis (200 participants), which will report

WHAT YOU NEED TO KNOW

- Constipation and faecal incontinence are common in patients with central neurological disease and may prove difficult to manage
- Limited evidence and clinical expertise suggest that diet modification, oral laxatives, rectal stimulants, digital stimulation, manual evacuation of faeces, and abdominal massage are options that may be tried
- Quality data to support these approaches are lacking, and trials are needed in mixed groups of patients to explore the efficacy of common approaches alone and in combination

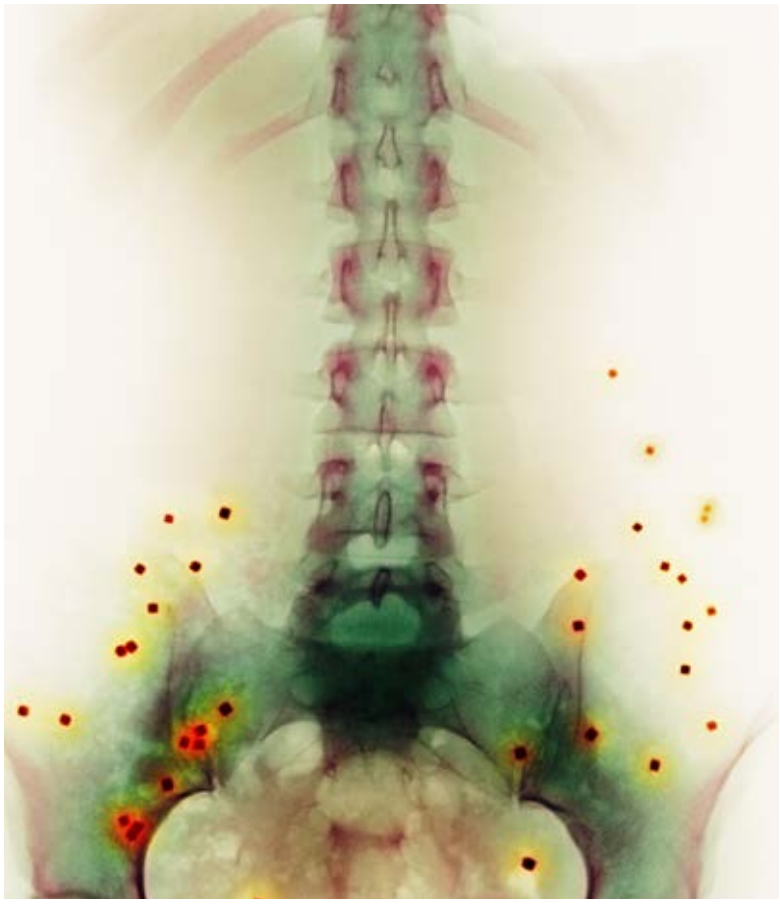
EDUCATION INTO PRACTICE

Do you routinely ask patients with neurological conditions whether they have bowel symptoms?

[1]Nursing Midwifery and Allied Health Professions Research Unit, Glasgow Caledonian University, Glasgow G4 0BA, UK
[2]King's College London School of Medical Education, Florence Nightingale School of Nursing and Midwifery, London, UK
Correspondence to: D McClurg Doreen.mcclurg@gcu.ac.uk

This is one of a series of occasional articles that highlight areas of practice where management lacks convincing supporting evidence. The series advisers are Sera Tort, clinical editor, and David Tovey, editor in chief, the Cochrane Library. To suggest a topic for this series, please email us at uncertainties@bmj.com

This is an edited version, full version on thebmj.com



CNRI/SPL

Fig 2 | Constipation markers. Coloured x ray of silicone markers (dots, highlighted yellow) that are being used to diagnose the severity of a patient's constipation. These markers are still in the digestive system six days after being swallowed. Normally, food passes through the digestive system in a day or two. The image also shows the bones (green) of the pelvis (across bottom) and the spine (down centre), as well as the lowest ribs (top)

HOW PATIENTS WERE INVOLVED IN CREATION OF THIS ARTICLE

We discussed a draft of this paper with five patients with neurogenic bowel dysfunction. They stressed the need for information on self management strategies such as diet and use of laxatives and on drugs that could make them constipated, as often they might receive misleading information (such as all fibre is good).
Establishing a routine, if possible, was important for both patients and their carers. They wanted to know how to decide which adjunctive treatment may be tried when symptoms did not respond. In patients with faecal incontinence, containment of faecal matter was a major challenge.
With their comments, we reduced the use of medical terminology in the article and highlighted the impact of bowel dysfunction on quality of life of these patients

RECOMMENDATIONS FOR FUTURE RESEARCH

As most commonly used approaches are poorly researched, a need exists for adequately powered randomised controlled trials as below:

- Population—People with various neurological conditions
- Interventions—Individual and combined approaches for constipation and faecal incontinence
- Outcomes—Standardised outcome measures such as the Neurogenic Bowel Dysfunction Score,[18] which records frequency of defecation, time spent trying to defecate, frequency of faecal or flatal incontinence, interventions such as drugs or digital stimulation, and perianal skin problems; acceptability of interventions to patients
- Additionally, research on innovative faecal containment devices is needed

in 2017.[15] A qualitative study is ongoing in patients with multiple sclerosis to evaluate the effect of bowel dysfunction on quality of life and their experience in accessing services.

What should we do in the light of the uncertainty?

Cisapride and tegaserod should not be prescribed. Guidance on faecal incontinence from the National Institute for Health and Care Excellence (NICE) suggests that clinicians should proactively ask about symptoms as patients may not volunteer the information.[16] Explain to patients that, although evidence is poor, clinicians have some experience with a variety of options and it may be a case of trying the simpler ones such as diet modification to start with.

On the basis of guidance from the Multidisciplinary Association of Spinal Cord Injured Professionals,[17] we recommend a stepwise approach to establish a regular pattern that is comfortable for the patient, using any number of the possible interventions alone or in combination:

- Establish an appropriate diet—for example, avoid insufficient or excessive fibre, attempt three meals a day, and aim for liquid intake of at least 1.5 L per day.
- Aim to establish a bowel habit by a routine of attempting defecation when peristalsis is likely to be maximal (after meals).
- Make sure the patient can access the toilet or is supported in doing so as needed.
- Review drugs that could exacerbate either constipation (for example, analgesics) or faecal incontinence (for example, laxatives or antacids).
- Consider oral laxatives or rectal suppositories alone or in combination.
- A continence nurse specialist could teach abdominal massage, manual evacuation, digital stimulation, or transanal irrigation to the patient or carer to aid self management.

Consider referral to a specialist service if symptoms remain unsolved. A combination of anorectal physiology tests, gut transit studies (fig 2), and endoanal ultrasonography will assist in determining whether further management, such as surgery, is appropriate.

Competing interests: None declared.
Cite this as: *BMJ* 2016;354:i3931
Find this at: http://dx.doi.org/10.1136/bmj.i3931

CASE REVIEW

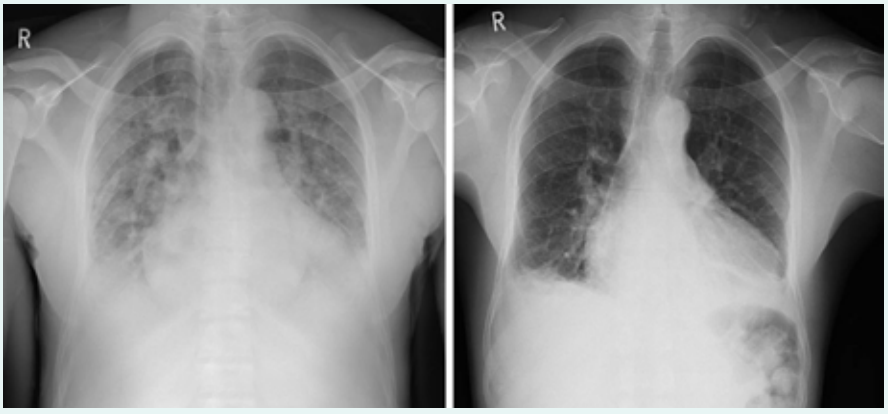
Now you see it, now you don't

A 40 year old black British man with known sickle cell anaemia presented to the emergency department with a nine hour history of left-sided pleuritic chest pain. The patient also had pain in both arms and legs. Three weeks earlier, he had had a productive cough treated with oral antibiotics in the community. A chest radiograph at that time showed no consolidation, and his symptoms had since resolved.

On admission, he reported excruciating pain, his oxygen saturations were 81% on air, and his respiratory rate was 30 breaths/min. Clinical examination was unremarkable except for reduced air entry throughout the lung fields, and he was apyrexial. His haemoglobin level was 60 g/L (baseline 69 g/L) and white cell count was 23×10^9/L with neutrophilia. His ECG showed no ischaemic changes. His admission chest radiograph can be seen in the figure (left).

He was treated with high flow oxygen, intravenous fluids and both simple and opioid analgesia. Four hours later he was more settled, his respiratory rate had decreased, and widespread crepitations—worse on the right—had developed. The patient was admitted to the haematology ward and received automated red cell exchange transfusion, oxygen therapy, intravenous fluids, analgesia, incentive spirometry, and antibiotics.

The figure (above) shows his second chest radiograph, performed 36 hours later.

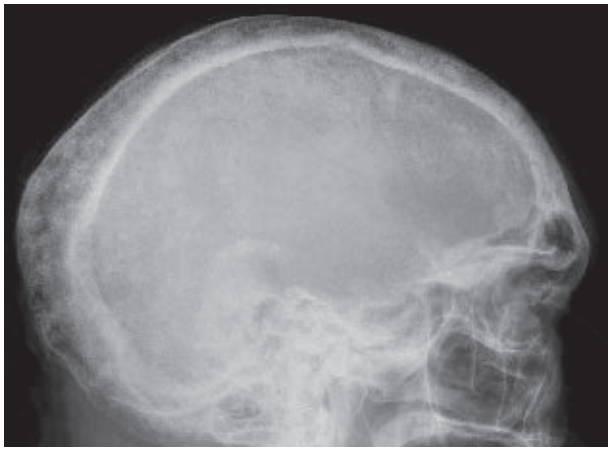


Patient's chest radiographs: at presentation (left), and 36 hours later (right)

1 What are the diagnosis and differential diagnoses?
2 What improvements does the second radiograph show, and why have they occurred?
3 What is the long term management of a patient such as this in the community and in the specialist haematology setting?

Submitted by Lauren Berg (lauren.berg09@imperial.ac.uk), Anietie Ekong, Susan Rowe, and Dimitris A Tsitsikas
Patient consent obtained.
Cite this as: *BMJ* 2016;354:i3714

If you would like to write a Case Review for inclusion in the Endgames section please see our updated author guidelines at **http://bit.ly/29HCBAL** and submit via our online editorial office at **http://bit.ly/29yyGSx**



SPOT DIAGNOSIS

Plain radiography of the skull after investigation for raised calcium

A 63 year old man was being investigated for raised serum calcium and alkaline phosphatase. Plain radiography of the head was performed after a bone scan showed increased uptake in the patient's skull. What diagnosis does the radiograph suggest?

Submitted by David C Howlett (david.howlett@nhs.net) and Joseph Dalby Sinnott
Patient consent obtained.
Cite this as: *BMJ* 2016;353:i2759

answers

CASE REVIEW **Now you see it, now you don't**

1 The diagnosis is acute chest syndrome. Differential diagnoses include pulmonary embolism and vaso-occlusive crisis of the ribs.
2 The removal of sickled erythrocytes during automated red cell exchange transfusion (ARCET) has led to resolution of the peribronchial cuffing and ground glass opacification, as well as reduction in azygous vein size and pleural effusion size. There is residual bibasal atelectasis.
3 Community based preventive measures include antibiotic prophylaxis, immunisations, and daily folic acid supplementation. Any single life threatening episode of acute chest syndrome (ACS) or recurrent episodes of ACS are indications for long term therapy with oral hydroxyurea or regular blood transfusions in the specialist setting.

SPOT DIAGNOSIS **Plain radiography of the skull after investigation for raised calcium**

The radiographic findings are consistent with a diagnosis of Paget's disease of the bone.

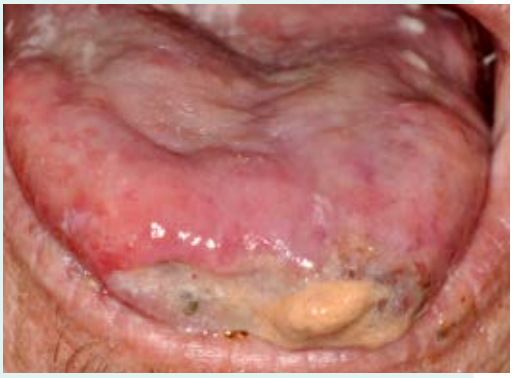
Tongue tip ischaemic necrosis after head and neck radiotherapy

A 60 year old smoker with a history of radiotherapy for soft palate squamous cell carcinoma had had a painful tongue tip for three weeks (fig 1). Clinically there was progressive destruction of the tongue. Imaging and tissue biopsy confirmed obliterative endarteritis with complete occlusion of his left lingual artery and near occlusion of the right. Smoking cessation and administration of dipyridamole and pentoxifylline resulted in complete healing. Both agents reduce inflammation and inhibit platelet aggregation. Pentoxifylline also facilitates oxygenation by enhancing red blood cell deformability. Clinicians should be aware of this rare and late complication of radiotherapy, and the contribution of chronic smoking. Treatment should be initiated by specialists. Autoimmune or infectious diseases and cancer can mimic this condition.

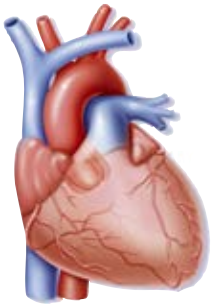
John C Devine, Deborah Boyd, Natasha Fullerton, Stergios Doumas (steliosdoumas@hotmail.com), Southern General Hospital, Glasgow G51 4TF, UK

Patient consent obtained.

Cite this as: *BMJ* 2016;353:i2370



Heart success



The opposite of heart failure is heart success—meaning the recovery of function in a previously impaired heart. Patients whose left systolic ejection fraction improved with treatment comprised 16.3% of a series of 2166 participants of median age 65 from heart failure clinics at Emory University, Georgia, USA (*JAMA Cardiol* doi:10.1001/jamacardio.2016.1325). At three years, the age and sex adjusted mortality in this cohort was 4.8% in patients with heart failure with recovered ejection fraction compared with 16.3% in patients with heart failure with reduced ejection fraction and 13.2% in patients with heart failure with preserved ejection fraction.

Multiple sclerosis and birth date in the UK

Multiple sclerosis rates vary across the UK, as do live birth rates by season. But after adjusting for the temporal and regional variations in the live births of the UK population, investigators found that there is a statistically significant season of birth effect in patients with multiple sclerosis, with an increased risk of disease in the peak month (April) compared with the trough month (November) (odds ratio 1.24, 95% confidence interval 1.10 to 1.41) (*JAMA Neurol* doi:10.1001/jamaneurol.2016.1463).

Maternal flu vaccination and baby flu

Advice about influenza vaccine for pregnant women on NHS Choices states that this also protects babies "for the first few months of their lives." But in a double blinded, placebo controlled randomised trial of trivalent flu vaccine conducted in South Africa in 2012-13, the protection to infants was only substantial in the first eight weeks of life, as confirmed by polymerase chain reaction tests for influenza (*JAMA Pediatr* doi:10.1001/jamapediatrics.2016.0921). From eight weeks on there was a steep decline in antibodies.

Midlife fitness and stroke

The time to be fit is in middle life. The message comes through most recently from an analysis of data from 19 815 adult US citizens in the Cooper Center Longitudinal Study, which measured cardiorespiratory fitness using treadmill tests (*Stroke* doi:10.1161/STROKEAHA.115.011532). Tracking of Medicare data showed that 808 participants had later been admitted with stroke. After adjustment for hypertension, diabetes mellitus, and atrial fibrillation, high midlife cardiorespiratory fitness still emerged as a major protective factor against subsequent stroke (hazard ratio 0.63, 95% confidence interval 0.51 to 0.79; fourth and fifth fifths versus first fifth).

Rheumatoid survival improvement

Good control of rheumatoid arthritis has been shown to normalise life expectancy in individuals. A study based on the UK Health Improvement Network shows how this has translated into a population effect by comparing patients with a first diagnosis between 1999 and 2006 with those having a diagnosis between 2007 and 2014 (*Ann Rheum Dis* doi:10.1136/annrheumdis-2015-209058). The hazard ratio for death compared with the general population of Britain has declined from 1.56 to 1.29.

Questions to ask about surgery

Older patients and their families often have difficulty in deciding about surgery, yet fully informed consent is especially important when the risks may be high. A qualitative study of shared decision making in this context came up with three domains of questions that need to be addressed: "Should I have surgery?" "What should I expect if everything goes well?" and "What happens if things go wrong?" (*JAMA Surg* doi:10.1001/jamasurg.2016.1308). The final list included 11 questions within these domains.

Delirium Day

Public delirium was an annual feature in many societies through history. On one or more days of the year, people could abandon themselves to worship Bacchus or become Lords of Misrule or carnival revellers. But Delirium Day, 30 September 2015 in Italy was not like that. It was the appointed date of the first nationwide point prevalence study to assess delirium in adults aged 65 or more admitted to acute and rehabilitation hospital wards (*BMC Med* doi:10.1186/s12916-016-0649-8). An acute confusional state occurred in more than one out of five patients in these settings: nothing to do with delirious happiness.

Cite this as: *BMJ* 2016;354:i4076