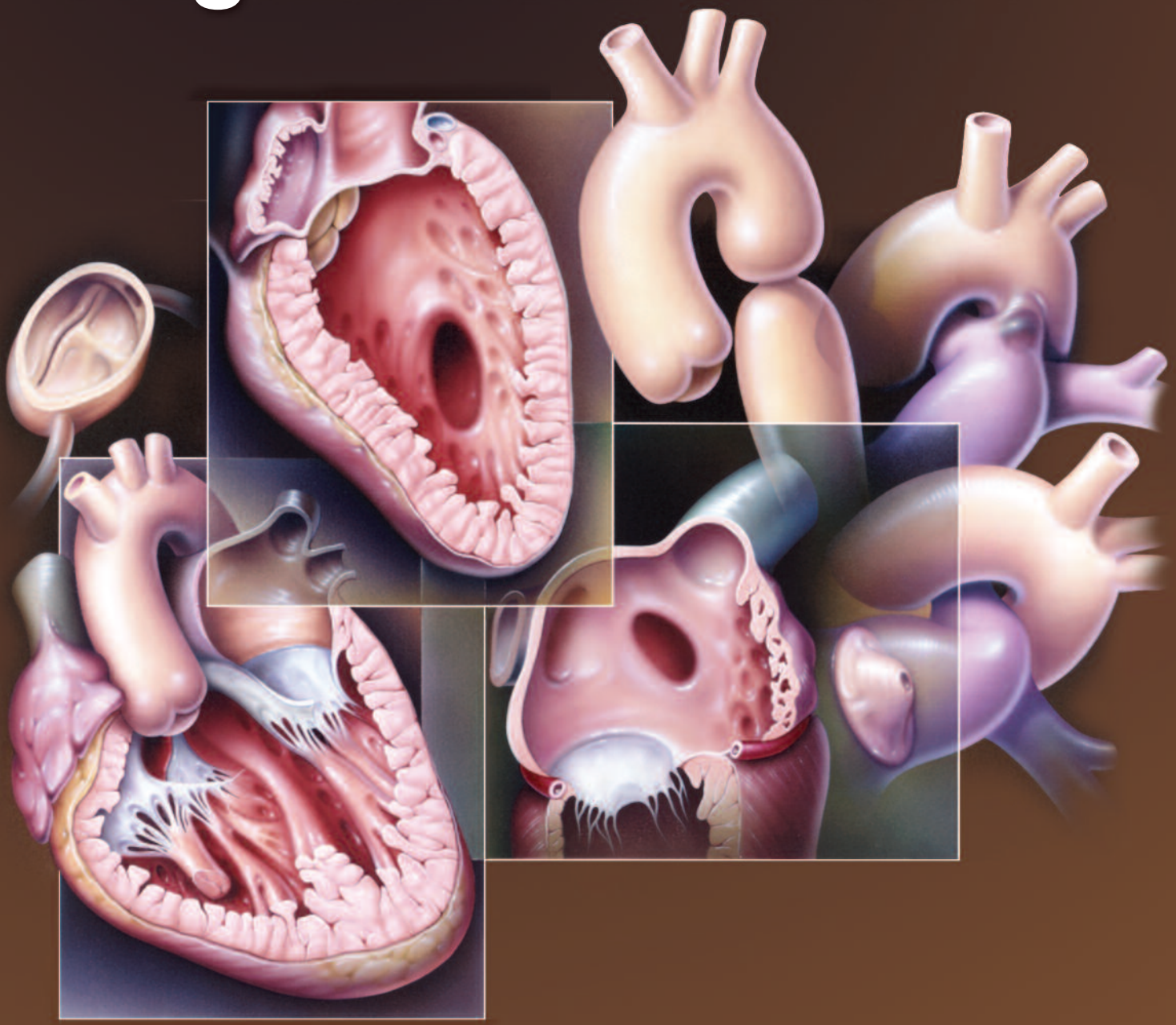
FEATURE

Care of adults with congenital heart disease



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With improved survival and longevity, GPs will have increasing exposure in the day-to-day medical care of adults with congenital heart disease. Many of these patients will face complications such as arrhythmias, heart failure and re-operation or intervention.

Key points

- **There are now more adults than children with congenital heart disease (CHD).**
- **Adults with CHD frequently have residual defects after childhood surgery.**
- **Common problems faced by adults with CHD include arrhythmias, heart failure, re-operation or catheter interventions, family planning and loss to cardiac follow up.**
- **Uncommonly, a new diagnosis of CHD is made in adulthood.**
- **Most GPs are likely to care for adults with CHD and play a crucial role in the detection of new complications, day-to-day medical care, including immunisations, and prevention of loss of the patient to regular cardiac follow up.**

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Congenital heart disease (CHD) occurs in 0.8% of neonates, making it the most common birth defect. With improvements in surgical and medical care, 95% of children with CHD now survive, so that there are currently more adults than children with CHD. GPs are now likely to care for patients with CHD. Common problems that GPs may face when initially assessing this patient population include new-onset arrhythmias, heart failure, worsening exercise capacity and medical management in pregnancy. GPs can also play a role in advising patients about vocational and lifestyle choices and prevention of loss to medical follow up. Due to complexities in the management of this patient group, most adults with complex CHD are followed at a specialised adult congenital heart disease (ACHD) centre. Intermittent review of adults with less complex CHD at these centres is also recommended.

The following cases are presented to illustrate important management issues in adult patients with CHD and are based on real-life scenarios. They represent patients with a small selection of the various diagnoses of CHD that may be seen in general practice.

Case 1. Follow up of adult patients with previously diagnosed CHD

Case scenario

A 42-year-old man was sent to hospital by his GP with palpitations and breathlessness. After a rate-controlling agent was given, electrocardiography showed atrial flutter with 3:1 AV conduction and right bundle branch block (Figure 1). He had undergone surgery as a child for tetralogy of Fallot, but because he felt well he had received no regular medical follow up since he was a teenager. Echocardiography showed severe pulmonary regurgitation with resultant severe right ventricular dilatation as is commonly seen after repair of tetralogy of Fallot (Figure 2). After being cardioverted, subsequent cardiac MRI demonstrated that the right ventricle was three times the normal size with reduced systolic function (Figure 3).

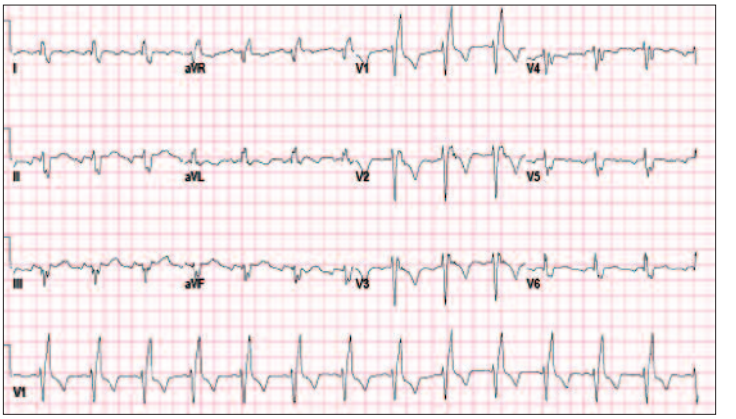
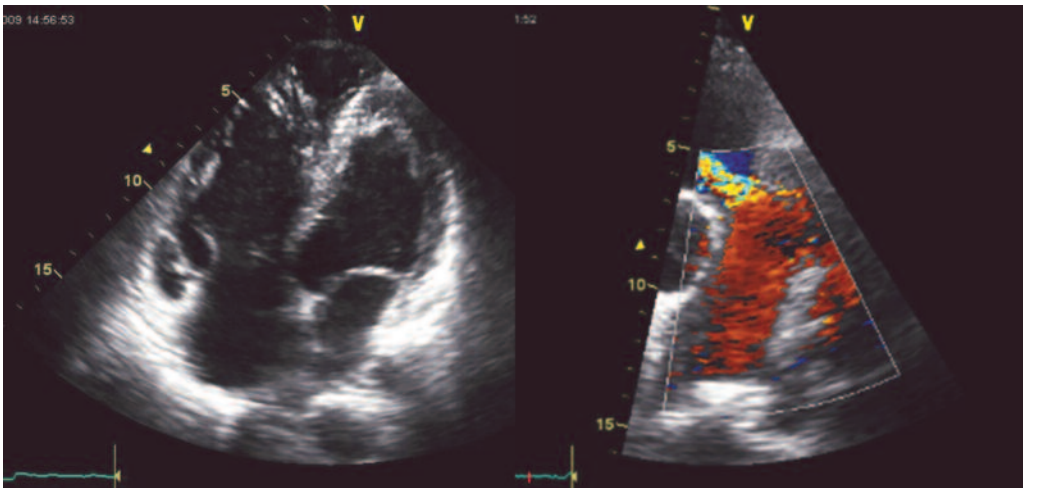
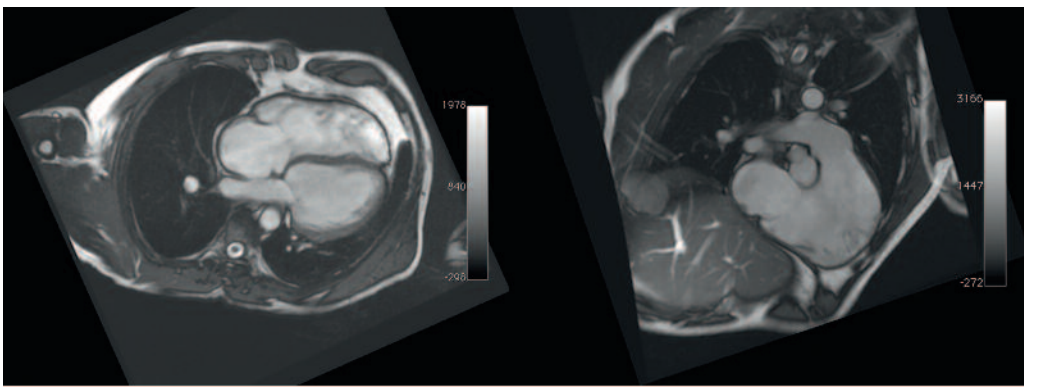


Figure 1. ECG showing atrial flutter with 3:1 AV conduction with right bundle branch block. Flutter waves are best appreciated in leads V4, II and aVL.



Figures 2a and b. Four-chamber echocardiogram view showing a very dilated right ventricle (a, left). Short axis view showing severe pulmonary regurgitation seen as red flow arising from branch pulmonary arteries (b, right). There is often laminar flow (without turbulence) back from the pulmonary arteries and the severity of pulmonary regurgitation can be missed.



Figures 3a and b. Cardiac MRI showing dilated right ventricle in four-chamber view (a, left). Aneurysmal right ventricular outflow tract (b, right).

Recommendations for endocarditis prophylaxis in adults with CHD

Endocarditis prophylaxis is recommended in adults with congenital heart disease (CHD) and:

- previous endocarditis
- unrepaired or palliated cyanotic heart disease
- a prosthetic heart valve
- prosthetic material within six months of repair or intervention
- prosthetic material adjacent to residual defect following repair.

Standard endocarditis prophylaxis (taken one hour before the procedure, alternative agents when intravenous formulation is required): amoxycillin 2 g. In patients who are allergic to penicillin: clindamycin 600 mg or cephalexin 2 g.

The patient underwent a pulmonary valve replacement and right atrial arrhythmia surgery. Six months later, his exercise capacity was the best it had been in years and he attended regular medical reviews. The right ventricular size reduced, but remained moderately dilated. He understood that the current valve would eventually become dysfunctional and need replacement but this would probably be performed with a stented valve in the cardiac catheterisation laboratory. Following valve replacement, endocarditis prophylaxis was mandatory at times of risk (see the box on this page listing patients with CHD who require endocarditis prophylaxis).

Commentary

Most GPs are likely to have some patients who underwent cardiac surgery or were diagnosed with CHD during childhood. Most patients with CHD have residual defects and are thus rarely cured. To improve symptoms and to prevent long-term complications, including sudden death and arrhythmias, many patients require repeat operations (sometimes numerous) and catheter interventions. The most common type of cardiac surgery in adults with CHD is the pulmonary valve replacement. Timing of pulmonary valve replacement in patients with previous repair for tetralogy of Fallot is determined by right ventricular size and function, and development of symptoms. In the patient in the current case study, right ventricular size did not return to normal size because surgery was performed too late. Arrhythmias are common in adults with CHD. As with other structural cardiac defects, atrial and ventricular arrhythmias can be a reflection of the underlying structural problem that needs to be addressed. Similar to the patient who presented in this case, adults with CHD often gradually adjust to their functional limitations, so that they claim to be asymptomatic.

Patients who require specialised follow up at ACHD centres

Specialised follow up at adult congenital heart disease (ACHD) centres is required in patients:

- with unrepaired or palliated cyanotic heart disease
- with conduit connecting heart to pulmonary artery
- with transposition of the great arteries
- who underwent the fontan procedure
- with repaired or previous tetralogy of Fallot
- with coarctation of the aorta
- with complex or large atrial or ventricular septal defects.

Intermittent follow up of patients with less complex congenital heart disease is also recommended at specialised ACHD centres.

Cardiac MRI is an important imaging modality for adults with CHD because it allows more accurate anatomical assessment of structures that can be difficult to image by echocardiography (e.g. right ventricle, pulmonary arteries) and can provide functional assessments that are not available with other noninvasive modalities (e.g. degree of systemic to pulmonary artery shunting, differential blood flow to each lung). Cardiac MRI is highly specialised and demanding because of the need for cardiac and respiratory gating, complex algorithms/sequences and often complex anatomy. Thus there are usually only one to two sites in each state or territory where cardiac MRI for CHD is routinely available.

Many adults are lost to medical and specialised CHD follow up. GPs can play a crucial role in directing these patients to appropriate ACHD care centres. All patients with cyanotic and complex CHD would normally be followed at a specialised ACHD centre (see the box on this page showing examples of patients who need follow up at ACHD centres). We would also recommend at least one review at specialised ACHD centres of other patients who had surgery in childhood.

GPs are also the first line of medical care for adults with CHD when they experience febrile illnesses. The first symptom of endocarditis is often a prolonged fever without a clear focus. In this situation, blood cultures are recommended before commencing antibiotics. Additionally, adults with complex CHD or those with significant residual valvular or ventricular dysfunction should routinely receive the influenza vaccine.

Case 2. Follow up and transition from paediatric to adult care and lifestyle choices

Case scenario

An 18-year-old man was in the process of being transitioned to an ACHD centre by his paediatric cardiologist and presented for his first review in the adult's clinic. He had a history of neonatal repair of coarctation of the aorta with a goretex patch. He was an elite athlete at school and was keen on joining the police force but was not sure whether he would be accepted, given his history of cardiac surgery. He was also treated with bronchodilators for asthma. His blood pressure was 150/70 mmHg, otherwise his clinical examination was normal. Echocardiography showed mild concentric left ventricular hypertrophy and mild flow acceleration across the prior repair site. Cardiac MRI showed only mild narrowing at the site of previous repair (Figure 4) consistent with the echocardiographic findings. Ambulatory blood pressure monitoring showed an average blood pressure of 140/70 mmHg. Exercise stress testing was performed to a high level with a significant hypertensive response to a peak of 240/60 mmHg. He was subsequently commenced on amlodipine 5 mg with good blood pressure control.

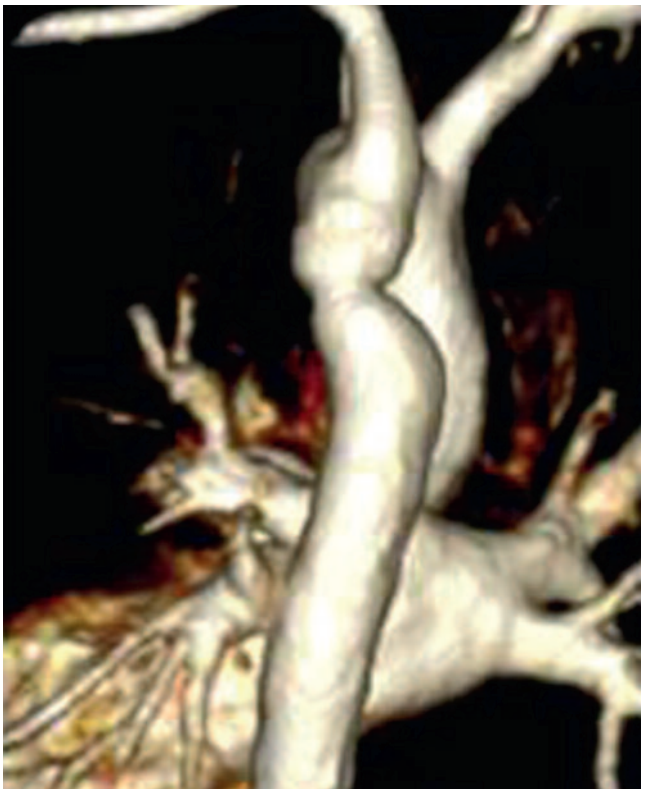
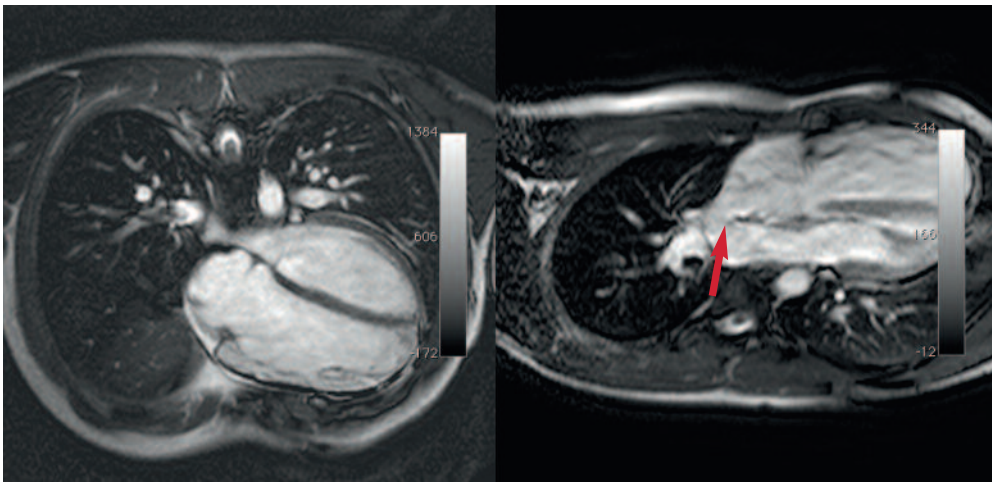


Figure 4. Cardiac MRI showing mild narrowing at previous coarctation repair site.

Commentary

Patients with childhood coarctation repair continue to require monitoring for management of hypertension and associated complications, including premature coronary artery disease, left ventricular hypertrophy and diastolic dysfunction. Monitoring of repair site complications, including re-coarctation and aneurysms, is also required. Most patients with coarctation have a bicuspid aortic valve and about 10% have cerebral aneurysms that may also contribute to morbidity. Despite successful anatomical repair of coarctation, most patients will ultimately end up being hypertensive due to multiple factors including increased arterial stiffness,



Figures 5a and b. Four-chamber cardiac MRI showing dilated right heart chambers (a, left). Sinus venous defect demonstrated with superior vena cava seen to drain into both right and left atria (b, right; see arrow).

endothelial dysfunction and re-programming of sympathetic and renin-angiotensin responses. MRI or CT imaging of the aorta is recommended to ensure early management of patients with possible postoperative complications.

Transitioning to adult services is a critical time due to the potential for loss to follow up. GPs can play an important role in ensuring young adults with CHD are suitably transitioned to adult care. Patients with prior surgical repair of congenital heart conditions may require specialist clearance or be precluded from participating in certain activities or vocations. Additionally, lifestyle choices (e.g. jobs or exercise training with heavy lifting) are discouraged in some patient groups (e.g. patients with aortic dilatation). Counselling against recreational drug use and risk-taking behaviour, which can have an especially deleterious effect to the health of this young group of patients, is important. Many adults with CHD have applications for life insurance declined or the price of premiums is prohibitive.

Case 3. Pregnancy in adults with CHD

Case scenario

A 22-year-old woman with a history of complete transposition of the great arteries (aorta arising from the right ventricle, pulmonary artery arising from the left ventricle) with a neonatal arterial switch (operation where aorta/coronary arteries and pulmonary trunk are transected and connected back to anatomically correct position) presented to her GP as she was planning to become pregnant. The patient underwent a cardiac evaluation before conception. Cardiac examination demonstrated a short systolic ejection murmur. Echocardiography demonstrated normal chamber dimensions with normal biventricular function. There was mild aortic regurgitation and mild flow acceleration across the right ventricular outflow tract and into the branch pulmonary arteries. Right ventricular systolic pressure was 40 mmHg. Cardiac MRI two years prior had also demonstrated mild narrowing of the branch pulmonary arteries as is sometimes seen after this type of surgery. She was advised that she was at low risk of maternal cardiac complications.

In the subsequent pregnancy, an obstetric ultrasound and fetal echocardiogram did not find a cardiac abnormality in the unborn fetus. She was reviewed in the multidisciplinary high-risk obstetric clinic where a delivery plan involving an early epidural and vaginal delivery were recommended to reduce maternal cardiac complications. The patient went into spontaneous labour at 40 weeks' gestation. The delivery plan was adhered to and a healthy child was born after an uncomplicated vaginal delivery. The mother was discharged after three days without any cardiac complications.

Commentary

Many women with CHD are at low risk of cardiac complications during pregnancy. Conditions in which pregnancy is contraindicated include: pulmonary hypertension; severe aortic stenosis or left ventricular outflow tract obstruction; conditions where there is moderate-to-severe left ventricular dysfunction (or systemic right ventricular dysfunction, for example, in some patients with transposition of the great arteries); and Marfan syndrome where there is a dilated aortic root. For other women with CHD, the risk of cardiac complications in pregnancy is intermediate and a careful assessment before pregnancy can help with determining safety of pregnancy and whether any intervention is required prior to pregnancy. Other issues that need to be considered before pregnancy in women with CHD include: the risk of neonatal complications (increased in certain types of CHD such as cyanotic heart disease or woman with single ventricles); the risk of transmission of CHD to the offspring (e.g. less than 5% in most forms of CHD); the safety of cardiac drugs in pregnancy; the delivery plan; and lastly whether pregnancy can have detrimental long-term effects on heart and valve function (currently a contentious issue).

In most women with significant residual cardiac disease, a vaginal delivery with an early epidural is considered safest to reduce cardiac complications, but the mode of delivery is often dictated by obstetric factors.

Case 4. Newly diagnosed CHD in adults

Case scenario

A 24-year-old man was found to have a systolic ejection murmur at the time of a diving medical. A further examination by his GP demonstrated a widely split second heart sound. He subsequently underwent an echocardiogram that showed a dilated right atrium and right ventricle without a demonstrable atrial septal defect on standard views. He proceeded to a transoesophageal echocardiogram that demonstrated a sinus venous defect with anomalous drainage of the right upper pulmonary vein to the superior vena. A cardiac MRI further delineated the anatomy and measured the right ventricle to be approximately twice the normal size and the pulmonary to systemic blood flow ratio was 2.5 (Figure 5). This type of atrial septal defect was not suitable for closure with a catheter-delivered device so he underwent cardiac surgery.

Commentary

The diagnosis of CHD is uncommon in adulthood but requires an elevated index of suspicion. Most patients seen at ACHD centres were diagnosed in infancy. Echocardiography is diagnostic in most types of CHD. Echocardiography is indicated if there is the suspicion of structural heart disease, including a long systolic murmur or any diastolic murmur, particularly in the presence of symptoms such as breathlessness, chest pain or palpitations. Other indications can include the finding of dilated heart chambers, increased pulmonary blood flow or heart failure on chest x-ray. In young adult patients with hypertension where there is the suspicion for coarctation of the aorta, echocardiography can be considered after a clinical examination has been performed, looking for weak foot pulses or reduced blood pressure in the lower extremities.

Conclusion

The number of adults with CHD will continue to grow. Many of these patients will face complications such as arrhythmias, heart failure and re-operation or intervention. GPs will have increasing exposure in the day-to-day medical care of adults with CHD as these patients survive longer. GPs can play an important role in preventing these patients being lost to cardiac follow up. **CT**

Further reading

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COMPETING INTERESTS: None.