Review

Clinical update on adults with congenital heart disease

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The number of patients with congenital cardiac disease reaching adulthood is increasing steadily. Many adults with such disease face both medical and surgical difficulties. Most clinicians know very little about basic cardiac defects, their natural history, complications after surgery, and adequate management of these patients. We aim to provide an overview of the most frequently encountered cardiac lesions and long-term complications and to outline an up-to-date approach to their management. We present a series of hypothetical cases and discuss their management.

Introduction

We use a hypothetical case format presentation to emphasise clinical relevance and to be of interest to the practising clinician. Hopefully, this review will raise awareness of adult congenital heart disease and stimulate further interest in this specialty.

Panel 1: **Atrial septal defect: case 1**

A 43-year-old businessman presents to the accident and emergency department having had palpitations for 12 h. He admits to having increasing shortness of breath over the past two years, and having to rest after climbing three flights of stairs. On examination, his pulse was irregular at a rate of 110 beats per minute, and his jugular venous pulsation (JVP) 6 cm over the sternal angle. He also had a mild right ventricular impulse, a fixed split of S2 (second heart sound) and an ejection murmur 2/6 best heard at the second left intercostal space. Chest radiography showed a dilated right ventricle and increased pulmonary blood flow. The electrocardiogram showed atrial fibrillation. Transthoracic echocardiography confirmed the presence of a secundum atrial septal defect with moderate right ventricular dilation.

Atrial septal defects are some of the most common congenital cardiac malformations in adults, representing up to 40% of acyanotic shunt lesions in patients older than 40 years. They arise from either excessive resorption of the septum primum or from deficient growth of the septum secundum, and are occasionally associated with anomalous pulmonary venous connection (about 10%). The degree of left-to-right atrial shunting depends on the size of the defect and the diastolic filling properties of the two ventricles. A substantial shunt (Qp/Qs >1·5/1·0) will probably cause symptoms over time, and the movement of such patients will become progressively more restricted with age. Effort dyspnoea is seen in about 30% of patients by their third decade whereas supraventricular arrhythmias (atrial fibrillation or flutter) and right heart failure develop in about 10% of patients by age 40 years.[1]

Lancet 2003; **362:** 1305–13

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Treatment

Haemodynamically unimportant atrial septal defects (Qp/Qs <1·5) do not need closure, except for the prevention of paradoxical emboli in patients who have had cryptogenic stroke.[2] In the absence of clinically significant pulmonary hypertension, closure is recommended for severe defects (Qp/Qs >1·5, or those associated with right ventricular volume overload).[3–8] In patients without symptoms, indications for closure are somewhat controversial. In those younger than 40 years, severe defects should probably be closed.[4] The appropriate treatment for patients who are older than 40 years is in some dispute, although results of one randomised clinical trial which showed an overall survival benefit among surgical patients would suggest that closure is advisable.[9]

Surgical closure of these defects can be done by primary suture closure or by an autologous pericardial or synthetic patch. Surgical mortality in adults with no pulmonary hypertension should be less than 1%, with a low morbidity related mainly to the development of perioperative arrhythmias (atrial flutter or fibrillation, or junctional rhythm).[3,4,6,10] The use of devices to close defects percutaneously under fluoroscopy and trans-oesophageal echocardiographic guidance[11] is gaining popularity (figure 1). Indications for closure by devices are the same as for surgical closure but selection criteria are stricter than for surgery. Devices are available only for patients with one secundum atrial septal defect and with an adequate septal margin for proper device support. Anomalous pulmonary venous connection precludes the use of this technique. This procedure is safe and effective when done by a skilled cardiologist, and major complications (eg, device embolisation, atrial perforation)

Search strategy

We searched the McGill library health sciences, Cochrane DSR (1990–2003), and MEDLINE (1990–2003) in all languages. We used the terms atrial septal defects, ventricular septal defects, patent ductus arteriosus, bicuspid aortic valve and aortopathy, coarctation of the aorta, transposition of the great vessels or D transposition, tetralogy of Fallot, and Fontan in combination with adult congenital heart disease. We mainly selected publications from the past 13 years, but also included frequently referenced landmark and highly regarded older publications. The Canadian Consensus on Adult Congenital Heart Disease and book chapters were also included because they provided a more extensive overview of subjects that were beyond the scope of this seminar.

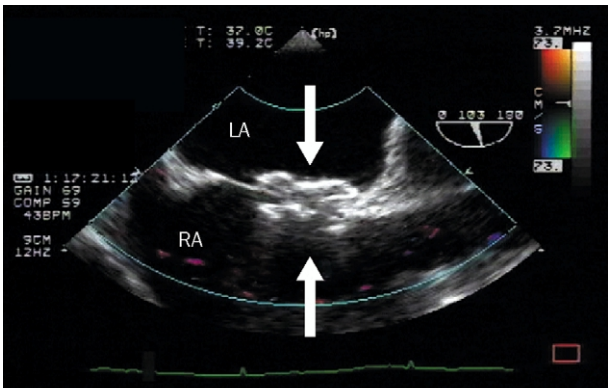

Figure 1: **Transoesophageal echocardiographical image of a secundum atrial septal defect after closure with Amplatzer device**

LA=left atrium. RA=right atrium. Arrows point at both sides of the Amplatzer device after insertion.

arise in less than 1% of patients. Complete closure is achieved in 80% or more of patients. Long-term follow-up data, however, are not available.[12,13] Notwithstanding, closure by device can be appealing to a patient wishing to avoid the consequences of surgery (general anaesthesia, cardiopulmonary bypass, pain, scar, and time for convalescence), or to a patient believed to be at high surgical risk.

Of note, surgical closure of atrial septal defects does not prevent the development of atrial fibrillation, atrial flutter, or stroke[4,5] especially when patients are operated on after age 40 years.[3] The use of a Cox-Maze procedure at the time of repair in patients after age 40, with a history of atrial flutter or fibrillation, is being studied,[14,15] and should be considered in this patient (panel 1). Closure of this patient's defect is necessary in view of its haemodynamic importance (right ventricular dilation) and the patient's symptoms. Surgical closure with a concomitant atrial Maze procedure should be considered since the patient has established atrial fibrillation. However, the long-term outcome of the Maze procedure in such patients remains unknown. Closure of the defect with a device, with medical management of the arrhythmia, is another option.

Panel 2: **Patent ductus arteriosus: case 2**

A 72-year-old woman is referred by her family doctor for atypical chest pain. Risk factors for coronary artery disease are few. Physical examination revealed a grade 1/6 continuous murmur in the left subclavian area. Transthoracic echo confirmed a small colour flow jet area seen between the aorta and the pulmonary artery in the suprasternal view, suggestive of a small patent ductus arteriosus. Left-heart chambers are of normal size and function.

The incidence of isolated persistent patency of the ductus arteriosus is estimated at one in 2000 to one in 5000 births. The ductus arteriosus derives from the left sixth primitive aortic arch and connects the proximal left pulmonary artery to the descending aorta, distal to the left subclavian artery. Occasionally, the ductus fails to close at birth and presents as a potential clinical problem.

Physiological consequences of a patent ductus arteriosus depend on the degree of left-to-right shunting, which is determined by both the size of the duct and the difference between systemic and pulmonary vascular resistances.[16] A small ductus accompanied by a small shunt does not cause significant haemodynamic disruption but might predispose to endarteritis, especially if accompanied by an audible murmur.[17] A moderate sized duct and shunt put a volume load on the left atrium and ventricle that results in dilation and often dysfunction of the left ventricle, and development of atrial fibrillation, or both. A large duct results initially in left-ventricular volume overload (which disappears after pulmonary hypertension develops), with a progressive rise in pulmonary artery pressure. This rise in pressure leads to high pulmonary vascular resistance and eventually, irreversible pulmonary vascular changes and systemic pulmonary pressures.

Treatment

Closure of a clinically detectable patent ductus arteriosus, in the absence of irreversible pulmonary hypertension, is usually recommended, although controversial, to avoid infective endarteritis.[18,19] The risk of endarteritis in a patient with a small silent patent ductus arteriosus is regarded as negligible, and closure of such small ducts is therefore not recommended.[17] Over the past 20 years, the effectiveness and safety of transcatheter device closure for ducts smaller than 8 mm has been established.[20,21] Complete ductal closure is achieved in more than 85% of patients, 1 year after device placement, with a mortality rate of less than 1%. Patients who undergo transcatheter closure avoid general anaesthesia, thoracotomy, postoperative pain, and long convalescence, which makes this procedure very appealing. In centres with appropriate resources and experience, transcatheter device occlusion should be the method of choice for ductal closure.

Surgical closure, by ductal ligation or division, or both, has been done for over 50 years with a slightly better closure rate, but greater morbidity and mortality, than that for closure with device.[22,23] Surgical closure of a duct should be reserved for patients with ducts larger than 8 mm in diameter or done in centres with no cardiologists specialising in such interventional techniques. Patients who undergo duct closure by device or surgery should be examined periodically and have an echocardiogram to assess possible recanalisation. The risk of late endarteritis from a clinically silent residual shunt, after device implantation or surgical closure, is unknown,[24] and appropriate clinical management of such patients is unclear. Some would recommend closure by device in this patient (panel 2) to eliminate the risk of endarteritis. In our view however, there is no evidence for that approach.[25] Others might choose a non-interventional approach with the use of prophylactic treatment for subacute bacterial endocarditis when needed.

Panel 3: **Bicuspid aortic valve: case 3**

A 25-year-old woman (Gravida one, pregnancy none, abortion none) is referred at 18 weeks' pregnancy for a heart murmur. The patient is asymptomatic. Her blood pressure is 90/60 mm Hg. On examination she had an ejection click best heard at the apex, a grade 3/6 systolic ejection murmur best heard at the second right intercostal space, with radiation to the carotids with a soft diastolic murmur grade 2/6 heard clearest along the left sternal border. A transthoracic echocardiogram confirmed the presence of a bicuspid aortic valve with moderate aortic stenosis (aortic valve area 1·0 cm^2), and mild-to-moderate aortic regurgitation. Cardiac MRI showed a dilated ascending aorta measuring 50 mm and no evidence of coarctation (figure 2).

Bicuspid aortic valve has a male preponderance of 4 to 1. This lesion accounts for about half the cases of surgically important isolated aortic stenosis in adults. A bicuspid aortic valve consists of two cusps, often of unequal size, the larger usually containing a false raphe. This lesion generally arises in isolation but is associated with other abnormalities in 20% of patients, the most common of which is coarctation of the aorta and patent ductus arteriosus.

At least half the patients with a bicuspid aortic valve have no complications, although there is always the risk of endocarditis. Mild aortic stenosis or regurgitation from bicuspid aortic valve generally progresses as the patient ages, but the rate is variable.[26] Enlargement of the aortic root resulting from cystic medial changes in patients with bicuspid aortic valve has a high prevalence,[27,28] and occurs irrespective of altered haemodynamics or age.[29] Aortic dissection is rare.

Treatment

Bicuspid aortic valves need intervention for stenosis when symptoms (exertional dyspnoea, angina, pre-syncope, or syncope) are present and should be considered before pregnancy when the stenosis is severe. Intervention for asymptomatic severe aortic stenosis to allow safe pregnancy is controversial, but severe aortic stenosis has proved to be a risk factor for cardiac decompensation during pregnancy.[30] Bicuspid aortic stenosis can be treated with balloon valvuloplasty if the patient is younger than 30 years and if the valve is not calcified.[31] Other treatment options include open aortic valvotomy, or valve replacement with a mechanical valve, a biological valve, or a pulmonary autograft (Ross procedure).[32]

Prophylactic surgery for proximal aortic dilation (>55 mm),[33] in the context of bicuspid aortopathy, seems better than waiting for the aorta to dissect or rupture, although there is no agreement about the diameter at which referral for surgery is appropriate. With the added haemodynamic load of pregnancy (50% increase in cardiac output) and the high oestrogen concentrations that tend to lessen the strength of the aortic wall, any patient with a dilated ascending aorta should be closely monitored during their pregnancy.

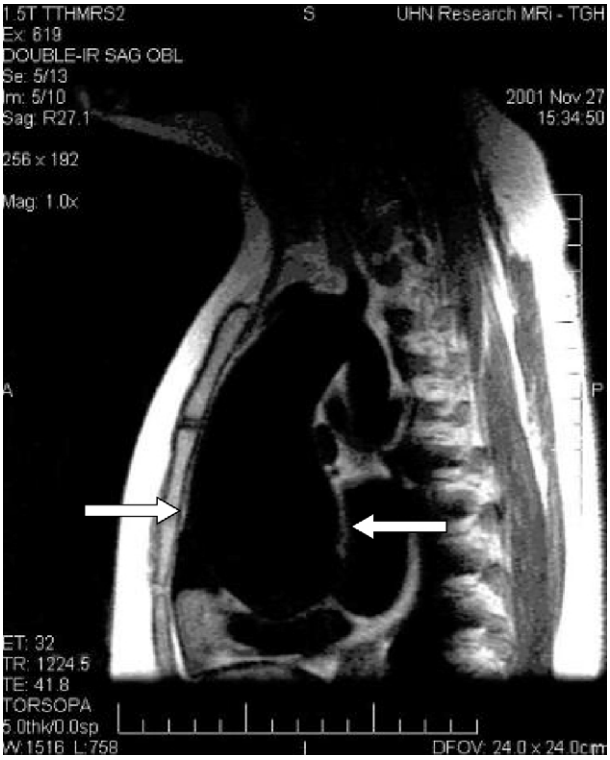

Figure 2: **MRI of a dilated ascending aorta in a patient with a bicuspid aortic valve**
Arrows indicate the sides of the dilated ascending aorta.

This patient (panel 3, figure 2) would be best treated with close medical follow-up and echocardiography every 3 months or so, to look for signs of progressive aortic root dilation. Treatment with β blockers (if blood pressure is normal) and bedrest, with or without early delivery, should be considered when rapid progression of the ascending aortopathy is seen or the ascending aorta reaches more than 55 mm. Vaginal delivery at term with a rapid (forceps-assisted) second stage labour would be acceptable if her ascending aortopathy remained stable during pregnancy. Prophylactic replacement of the ascending aortic root with a bioprosthetic aortic valve should be considered before other pregnancies in this patient if her ascending aorta is 55 mm or more or if she has severe aortic stenosis, or both.[30,33]

Panel 4: **Coarctation of aorta: case 4**

A 44-year-old man with a history of coarctation repair in his childhood is referred for persistent mild systolic hypertension. The patient is otherwise asymptomatic. The blood pressure is 145/80 mm Hg on the right arm, 120/80 mm Hg on the left arm, 120/80 mm Hg on the right leg with a mild radiofemoral delay. MRI confirms the presence of a mild discrete narrowing at the site of previous coarctation repair and cardiac catheter shows a withdrawal gradient of 20 mm Hg between the proximal and distal descending aorta.

Coarctation of the aorta is seen most frequently in men with a ratio of almost 3 to 1. It usually resides in the region of the ligamentum arteriosum. It might be discrete or associated with hypoplasia of the aortic arch and isthmus. Related abnormalities include bicuspid aortic valve in 50–85% of cases. Despite initial successful correction of coarctation, the risk of late systemic hypertension (due to residual or recurrent coarctation, or merely to abnormal aortic wall compliance) in these patients is fairly high. If inadequately controlled, hypertension leads to an increased risk of premature death from late heart failure. Other causes of premature death might be coronary artery disease with or without aortic rupture or dissection.[34,35] Careful long-term follow-up and aggressive management of patients with hypertension is therefore recommended.[33] Local complications such as re-coarctation and aneurysm formation at the site of previous transcatheter or surgical correction should be identified through periodic chest radiography, echocardiogram, MRI, or spiral CT examinations.

Treatment

All patients with serious coarctation or recoarctation having proximal hypertension, a withdrawal gradient greater than 20 mm Hg at angiography or an echo peak gradient of more than 20 mm Hg in the absence of extensive collaterals or less than 20 mm Hg in their presence, warrant treatment to eliminate the gradient and reduce the risk of long-term complications.[33] Balloon dilatation with stent insertion in patients with native coarctation and recoarctation can be done with good immediate and medium-term results in adolescents and adults[36,37] (figure 3A, 3B, and 3C) and should

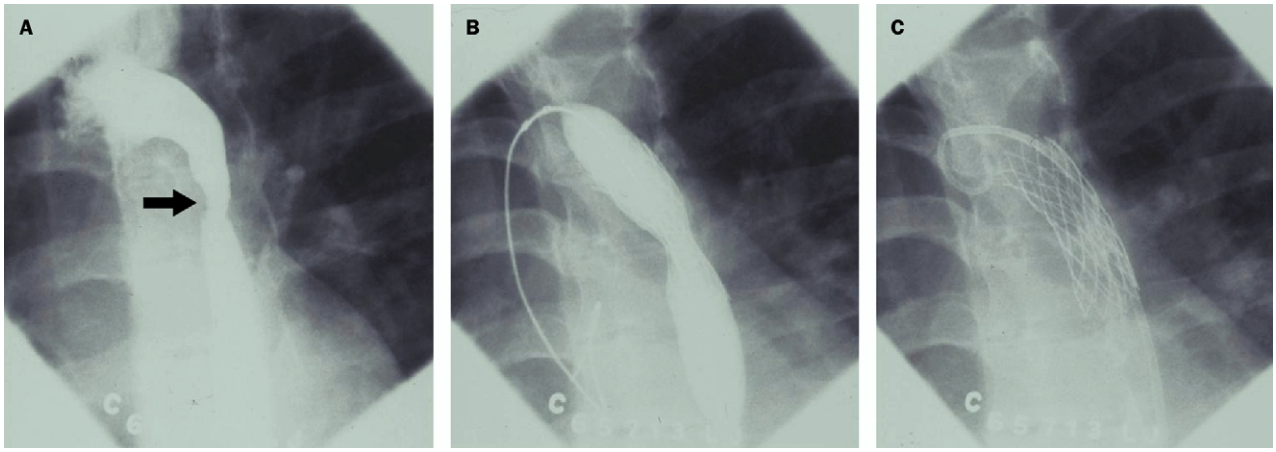

Figure 3: **Angiograms of mild re-coarctation of the aorta (A); balloon dilatation and stenting of the re-coarctation segment of the descending aorta (B); and stented descending aorta (C)**
Arrow in (A) indicates re-coarctation segment.

probably be the procedure of choice when the anatomy is suitable and expert skills are available. If the anatomy is not suitable (ie, long tunnel-like stenosis) surgery might be needed. After surgical repair of isolated aortic coarctation, the obstruction is usually relieved with minimum mortality (<2%). However, mortality is increased for reoperation (5–15%).

This patient (panel 4) has mild re-coarctation of the aorta (peak gradient >20 mm Hg) at the site of previous surgical repair with superimposed mild systolic hypertension. Balloon dilatation and stenting of the re-coarctation segment (if the anatomy is suitable) should be done to reduce proximal systolic blood pressure. The procedure should only be done, however, by experienced cardiologists. Surgical revision or antihypertensive medication, or both, might also be appropriate.

Panel 5: **Tetralogy of Fallot: case 5**

A 46-year-old woman is brought to the emergency room by ambulance after a witnessed collapse. Sustained ventricular tachycardia was ended (figure 4) and the patient successfully revived. The patient has a history of repaired tetralogy of Fallot in childhood. She was lost to follow-up. Physical examination showed a right ventricular impulse and a low-pitched diastolic murmur grade 3/6 best heard along the left sternal border. Transthoracic echo showed a dilated right ventricle and severe pulmonary regurgitation. Cardiac MRI confirmed a severely dilated right ventricle (right ventricular end-diastolic volume 350 mL—normal range up to 108 mL/m^2) (figure 5), severe pulmonary regurgitation, and fairly normal right ventricular systolic function. Electrophysiological studies confirmed the presence of easily inducible ventricular tachycardia from the right ventricular outflow tract, and angiography shows normal pulmonary and coronary arteries.

Tetralogy of Fallot is the most common form of cyanotic congenital heart disease after 1 year of age, with a frequency of almost 10% of all congenital heart disease. The defect is caused by anterocephalad deviation of the outlet septum resulting in four features: (1) a non-restrictive ventricular septal defect; (2) an overriding aorta (<50% override); (3) obstruction of the right ventricular outflow tract which may be infundibular, valvar, or (usually) both, with or without supravalvar or branch pulmonary artery stenosis; and (4) consequent right ventricular hypertrophy.

Reparative surgery is usually done early in infancy, by closure of the ventricular septal defect with a Dacron patch and relief of the right ventricular outflow tract obstruction. Obstruction relief might include resection of infundibular muscle and insertion of a right ventricular outflow tract or transannular patch—a patch across the pulmonary valve annulus that disrupts the integrity of the pulmonary valve and causes important pulmonary regurgitation. Significant pulmonary regurgitation is almost always encountered when the transannular patch repair technique is used. Pulmonary regurgitation is usually well-tolerated indefinitely, if mild to moderate. Severe chronic pulmonary regurgitation might be well tolerated for 20 years or more, but could then lead to symptomatic right ventricular dysfunction,[38] and increase the susceptibility to ventricular tachycardia and sudden cardiac death.

Residual obstruction of the right ventricular outflow tract can arise in the infundibulum, the pulmonary valve, and the main pulmonary trunk or branches of the left and right pulmonary arteries, or both branches. Right ventricular dilation in this setting is usually due to longstanding free pulmonary regurgitation,[38] but might result from operative injury. Substantial tricuspid regurgitation might occur because of right ventricular dilation, which then leads to further dilation of the right ventricle.

Atrial tachyarrhythmia arises in about a third of adults and contributes to late morbidity and even mortality.[39,40] It usually takes the form of atrial flutter or intra-atrial re-entrant tachycardia. Often indicative of haemodynamic

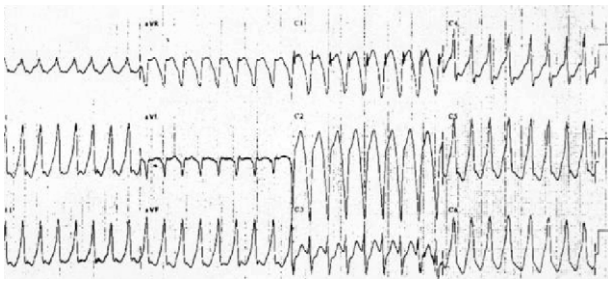

Figure 4: **Electrocardiogram showing ventricular tachycardia in a patient after repair of tetralogy of Fallot**

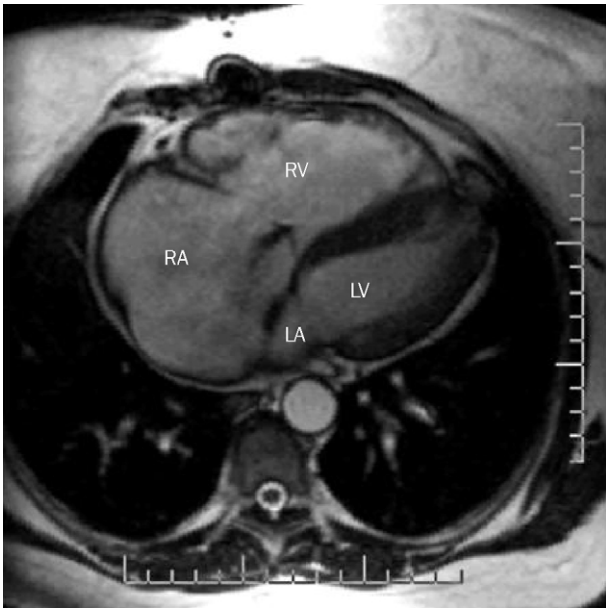

Figure 5: **MRI showing right right ventricular dilation in a patient after repair of tetralogy of Fallot**

LA=left atrium. LV=left ventricle. RA=right atrium. RV=right ventricle.

trouble (significant right ventricular dilation and dysfunction, increased tricuspid regurgitation), the substrate is most likely a surgical scar in the atria and the trigger, atrial dilation.

Sustained monomorphic ventricular tachycardia is rare, compared with other types,[41] and is highly associated with pronounced right ventricular dilation. The QRS duration from the standard surface ECG has been shown to correlate well with right ventricle size in these patients.[42,43] A maximum QRS duration of 180 ms or more is a highly sensitive marker for sustained ventricular tachycardia and sudden cardiac death in adults with previous tetralogy repair,[42] although its positive predictive value is low. Dilation of the right ventricle is thought to trigger ventricular tachycardia, whereas surgical scars (near the right ventricular outflow tract or ventricular septal defect patch) form the arrhythmia substrate. The reported frequency of sudden death, presumably due to arrhythmia, in late follow-up series is 0·5–6% over 30 years, accounting for about a third to a half of late deaths.[44,45] Older age at repair, severe left ventricular dysfunction,[46] postoperative right ventricular hypertension, transannular patching (causing free pulmonary regurgitation), and an accelerated rate of QRS prolongation are all predictors of sudden death in these patients.[41,44,45]

Treatment of complications

Replacement of the pulmonary valve (with either a homograft or porcine bioprosthesis) might be necessary for severe pulmonary regurgitation leading to right ventricular dilation, sustained arrhythmias or symptoms, or both. Such replacement might also be needed for a grossly calcified pulmonary valve. It has a low operative risk[47,48] and leads to symptomatic improvement.[48,49] Timely pulmonary valve replacement, before irreversible severe right ventricular dilation and systolic right ventricular dysfunction begins, is a major clinical goal.[49] Concomitant tricuspid valve annuloplasty might also be necessary when at least moderate tricuspid regurgitation is present.

Patients presenting with sustained atrial flutter, atrial fibrillation, or ventricular tachycardia, should undergo a thorough assessment of their haemodynamics and should have residual haemodynamic lesions repaired—eg, significant right ventricular dilation from pulmonary regurgitation with resulting tricuspid regurgitation that needs pulmonary valve replacement and tricuspid valve annuloplasty.[50,51] Radiofrequency ablation, after mapping for atrial re-entry tachycardia, now yields better results than before for classic atrial flutter or incisional atrial re-entrant tachycardia, or both, and should be done either percutaneously (if there is no need for concomitant surgery) or intraoperatively at the time of surgical repair of underlying haemodynamic lesions.[50] For atrial fibrillation, a biatrial maze procedure should also be considered, and ideally done at reoperation. Likewise, transcatheter (when surgery is unnecessary) or concomitant intraoperative ablative procedures of the ventricular tachycardial pathway should be done when appropriate.[50] Antiarrhythmic medications and the new generation of atrial antitachycardia pacemakers (for supraventricular tachycardia) can be used as adjunct treatments. Patients who were resuscitated after sudden cardiac death and are in need of surgery without a haemodynamic substrate should probably receive an automated implantable cardioverter defibrillator (AICD).

This patient (panel 5, figures 4 and 5) should probably have pulmonary valve replacement since the right ventricle is severely dilated, with repair of the ventricular septal defect patch as well as cryoablation of the ventricular tachycardia focus at the time of surgery. Pulmonary valve replacement will lead to a smaller right ventricle and cryoablation of the ventricular tachycardia focus would eliminate the arrhythmia substrate and probably prevent a further episode of sudden cardiac death.[50]

Panel 6: **Transposition of the great arteries: case 6**

A 32-year-old man who had the Mustard procedure for D-transposition of the great arteries presents with increasing exertional dyspnoea and fatigue. Cardiac examination revealed a right ventricular impulse, a loud single S2 (second heart sound) and a holosystolic murmur best heard at the left sternal border. Transthoracic echo confirmed the presence of severe systemic tricuspid regurgitation and severe systemic right ventricular systolic dysfunction (ejection fraction 15%).

In patients with complete transposition of the great arteries, the connections between the atria and ventricles are concordant (normal), and the connections between ventricles and great arteries are discordant. Thus, the pulmonary and systemic circulations are connected in parallel rather than the normal in-series connection. In one circuit, systemic venous blood passes to the right atrium, the right ventricle, and then to the aorta. In the other, pulmonary venous blood passes through the left atrium and ventricle to the pulmonary artery. This situation is fatal unless the two circuits mix. About half of patients with transposition of the great arteries have additional abnormalities, most often a ventricular septal defect.

The most common previously done surgical procedure seen in adults is the atrial switch operation. Patients will have had either a Mustard or a Senning procedure. Blood is redirected at atrial level with a baffle made of Dacron or pericardium (Mustard operation) or with atrial flaps (Senning operation), to achieve physiological correction. Systemic venous return is diverted through the mitral valve into the subpulmonary morphological left ventricle

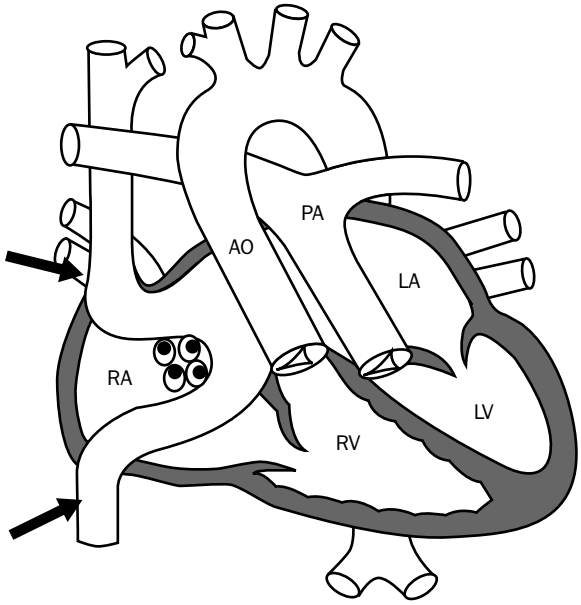

Figure 6: **Diagram showing atrial baffle in a patient with D-transposition of the great arteries**
RA=right atrium. RV=right ventricle. LA=left atrium. LV=left ventricle. PA=pulmonary artery, AO=aorta. Arrows indicate systemic venous baffle, and black dots pulmonary venous baffle.

and the pulmonary venous return is rerouted via the tricuspid valve into the subaortic morphological right ventricle. This repair, however, leaves the morphological right ventricle to support the systemic circulation (figure 6). After atrial baffle surgery, most patients reaching adulthood will be in New York Heart Association class I–II.[52–55] During 25 years of follow-up, about half these patients will have moderate systemic dysfunction of the right ventricle[52–55] with only a few presenting with symptoms of congestive heart failure. Severe systemic tricuspid regurgitation is present in about a third,[53,54] which exacerbates right ventricular dysfunction. Atrial flutter arises in 20% of patients by age 20[56,57] and progressive sinus node dysfunction is seen in half the patients by that time.[53,55,57] These rhythm disturbances are thought to be a result of atrial and sinus node damage at the time of atrial baffle surgery. Baffle leak or obstruction can also occur.

The atrial switch operation was gradually replaced by the arterial switch operation (Jatene) in the 1980s, but few of these patients have yet become adults. Blood is redirected at the great artery level by switching the aorta and pulmonary arteries such that the morphological left ventricle becomes the subaortic ventricle and the morphological right ventricle becomes the subpulmonary ventricle. Reliable data for the clinical outcome in adults after the arterial switch procedure should be available over the next decade. Clinical arrhythmia promises to be less of a problem in this group of patients, but concerns about the development of supra neopulmonary artery stenosis, ostial coronary artery disease, and progressive neoaortic valve regurgitation warrant serial follow-up.[58]

Treatment

The benefits of angiotensin-converting enzyme inhibitors in patients with systemic right ventricular dysfunction after an atrial switch have not been established,[59,60] and are being investigated in a double-blind multicentre randomised trial of ramipril. Occasionally, patients with a failed Mustard or Senning operation might need heart transplantation. Alternatively, a conversion procedure to an arterial switch, after re-training of the left ventricle with a pulmonary artery band, could be considered, but few data for the outcome of such a procedure are available in adults.[61–64]

The degree of right ventricular dilation and systolic dysfunction should be confirmed by MRI or multiple gated acquisition examinations. An angiotensin-converting enzyme inhibitor with or without other heart failure therapies could be tried in this patient (panel 6). If no clinical improvement is noted, however, a switch conversion procedure (pulmonary artery banding with left ventricular training followed by pulmonary artery debanding, and arterial switch with take down of the atrial baffle) should be used. Tricuspid valve replacement in this patient would probably lead to worsening of right ventricular function and is not recommended. Cardiac transplantation could also be considered at an appropriate time.

Panel 7: **Fontan procedure: case 7**

A 26-year-old college student with a diagnosis of tricuspid atresia palliated by the Fontan procedure goes to her local emergency room because of 6 h of continuous rapid palpitations. The patient is haemodynamically stable, and a cardiac monitor reveals atrial fibrillation with a ventricular response of ten beats per minute.

The Fontan procedure is the palliative, non-curative, surgical treatment for patients with univentricular hearts. The principle is diversion of the systemic venous return directly to the pulmonary arteries without the need for a subpulmonary ventricle. Many modifications of this procedure have been described, eg—direct atrio-pulmonary connection, total cavopulmonary connection, and extracardiac conduit. Progressive deterioration of functional status with time is the rule,[65–67] with survival at 10 years after the procedure reported to be 60–71%.[66–68] The most common complications after a Fontan procedure include atrial flutter or fibrillation, right atrial thrombus formation, obstruction of the Fontan circuit, and ventricular dysfunction.

Atrial flutter or fibrillation are common (15–20% at 5-year follow-up),[69–72] and increases with duration of follow-up.[69,71] They are associated with serious morbidity (especially the development of atrial thrombi within hours),[73] and can lead to profound haemodynamic deterioration. Such patients need prompt and expert medical attention. The combination of atrial incisions and multiple suture lines at the time of Fontan surgery with increased right atrial pressure and size probably accounts for the high frequency of atrial arrhythmias in these patients. Obstruction of the Fontan connection should be ruled out in all patients presenting with new onset atrial arrhythmias.

The reported frequency of thromboembolic complications in the Fontan circuit varies from 6% to 33%, dependent on the diagnostic method used and the length of follow-up.[74–76] Right atrial thrombus formation relates to the presence of atrial flutter or fibrillation,[73] right atrial dilation, right atrial smoke (spontaneous echo contrast), and the presence of artificial material used to construct the Fontan circuit.[74]

Part obstruction of the Fontan connection leads to exercise intolerance, atrial tachyarrhythmias, and right-sided heart failure. Sudden total obstruction presents as

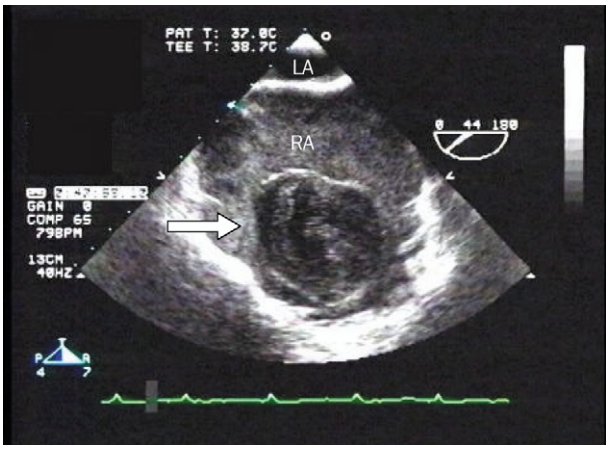

Figure 7: **Transoesophageal echocardiogram showing a right atrial clot**
LA=left atrium. RA=right atrium. Arrow indicates right atrial clot.

sudden death. Protein-losing enteropathy is seen in about 2–3% of patients after the Fontan procedure.[66,77] Patients present with generalised oedema, ascites, pleural effusion, or chronic diarrhoea. The diagnosis is confirmed by low serum albumin and protein and high α1-antitrypsin stool clearance. The prognosis is poor, with a 5-year survival of 46–59%.[72]

Treatment of complications

In patients with atrial fibrillation or flutter, prompt anticoagulation and transoesophageal echo assessment to rule out atrial thrombi before cardioversion is recommended (figure 7). Long-term management with antiarrhythmic therapy is successful in less than 50%. Transcatheter atrial ablation can be done in specialised centres, with a 50% success rate.[78] For recalcitrant cases, surgical revision with antiarrhythmic surgery is recommended.[79]

For established thrombus, thrombolytic therapy or surgical removal of the clot and conversion of the Fontan circuit have been described. Long-term anticoagulation is recommended for patients with known thrombi.[80] Some centres anticoagulate all Fontan circuits for the rest of the patient's life. For patients with Fontan obstruction, surgical revision of the Fontan connection is usually needed.[81] Alternatively, balloon angioplasty with or without stenting can be used when appropriate. Patients with protein-losing enteropathy might be candidates for creation of a fenestration in the atrial septum or revision of the Fontan. Alternatively, subcutaneous heparin, octreotide treatment, and prednisone treatment have also been tried with variable success.[77] No particular treatment seems more successful than any other.

In this patient (panel 7), who is haemodynamically stable, ventricular rate should be controlled with digoxin or other agents, and prompt anticoagulation with heparin should be started. The patient should be transferred to a highly specialised hospital for transoesophageal echo to rule out right atrial clot before cardioversion. Fontan obstruction as the cause of atrial fibrillation (conduit obstruction with secondary right atrial stretch) should be ruled out either at the time of transoesophageal echo or by cardiac catheterisation. Long-term coumadin is recommended for all patients with Fontan obstruction who have a history of atrial fibrillation. Maintenance of sinus rhythm after cardioversion is probably best achieved with amiodarone, although the success rate is low at 50%. Antitachycardia pacing, catheter ablation or surgical revision, or both, with concomitant biatrial maze procedure are used as second-line treatment in patients with Fontan obstruction and chronic or paroxysmal atrial fibrillation that is unresponsive to medical management. Other causes of atrial fibrillation such as ethanol binge or hyperthyroidism should also be ruled out.

Conflict of interest statement
None declared.

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Uses of error

Sources of error: from procedures to end-of-life decisions

Trevor Duke

As a registrar common sources of error were procedures, as a consultant they may be end-of-life decisions. Years ago I used a 14 F central line stillette to insert an intercostal catheter. Almost immediately the child became as pale as the bed sheet on which he was lying. An intercostal vessel had been perforated and blood was filling the pleural cavity. This boy survived, but never again would I use anything but blunt dissection to place an intercostal catheter. Later, when the Seldinger wire of a subclavian line needed unexpected force to be removed, I learnt that unless all lumens of the catheter draw back venous blood, the transducer shows a venous pressure wave, and a radiograph shows the line to be in an appropriate position, the catheter might be in the wrong place. I learnt, by this error, that in doing any procedure I must go though a checklist to be certain it has been done correctly, rather than default to the hope that all is well.

Two more recent management errors are vivid: both involve hasty decisions at the end of life. I thought a moribund 13-month-old girl with meningococcal septicaemia and unrecordable blood pressure might be saved by extracorporeal membrane oxygenation. The decision had to be quick, but by the time cannulation was done and artificial circulation established, removing the sterile drapes showed a barely recognisable purple and oedematous body for the grieving family to bid farewell. Heroics are not always what parents want. They sometimes recognise futility before the doctors. Yet even futility can be ambiguous. One courageous family forced me to help them salvage something valuable out of unspeakable tragedy. A 3-year-old girl had had a fatal head injury. At admission to the intensive care unit she had no brain-stem function, severe hypotension, and a grossly distended abdomen. We quickly assessed the injuries as non-survivable and the child as not being a viable organ donor. I discussed this with her parents who were devastated, but listened calmly. They asked about organ donation and requested that we keep her alive until other family members arrived to say good-bye. We agreed to provide ventilatory support for her as long as it would take for relatives to come, but to stop all other treatment and monitoring, since prolonged ischaemia meant her organs were not suitable for donation. That night the unit was very busy and in the next ten hours I stopped by her room only briefly to observe the family's vigil and inquire if the time had come to withdraw support completely. The mother asked me to reconsider the issue of organ donation. Only then did I realise that although accepting the inevitability of her daughter's death, she held on to the hope that it might somehow become more than a meaningless tragedy. An echocardiograph showed a very slowly beating, but well contracting heart. I ventilated the child more, put in a central line and started infusions of dopamine and adrenaline. The heart function improved, the girl became an organ donor, and her family said a final farewell.

Rapid decisions are necessary in intensive care, but even in the face of apparent futility, haste in closing a life that has been nurtured and loved for years is not what families wish for. In running audit meetings I have learnt that honestly declaring ones own errors is necessary to empower junior staff to learn from their mistakes . . . and from mine.

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