

Adults With Congenital Heart Disease

An Expanding Population

Continued progress in diagnosing and managing infants and children with congenital heart disease will likely result in a better prognosis and improved survival rate for adults.



By Brian W. McCrindle, MD, MPH, FRCPC

Case

Mr. CR, 29 years old, presents with complaints of decreasing exercise tolerance and occasional palpitations for the past three months. He has never been very active, but currently admits to having difficulty climbing two flights of stairs. He has palpitations when at rest, lasting between five and 10 seconds, and occurring about three to four times per day. There are no associated symptoms, except for one episode in the last two weeks, which lasted two minutes and was associated with lightheadedness.

He says when he was an infant he had “a shunt,” and at age four he had “a hole in the heart closed, and a patch to open the artery.” He last saw a cardiologist at the pediatric hospital before he finished high school. He was supposed to see a cardiologist in the general hospital, but never kept the appointment. His last medical contact was in a walk-in clinic three years ago when he had an episode of bronchitis. He currently smokes. He is taking sertraline for depression, and says he needs to take antibiotics when he goes to the dentist.

He had difficulty finishing high school, and now works as a receptionist for a local hotel. He lives with his parents, and is not currently in a relationship.

The case continues on page 35.

In this article:

1. What's new in managing congenital heart disease in adults?
2. What risk factors are specific to adult patients?
3. What are the treatment options?

Case Cont'd

On clinical examination, Mr. CR has the following:

Height 170 cm, weight stable at 75 kg.

Left arm blood pressure 126/85 mmHg sitting, pulse rate 72 bpm with extra beats.

A right thoracotomy and a median sternotomy scar.

Prominent right ventricular (RV) impulse, no thrills.

Normal S1, single S2, no extra sounds.

Grade II/VI harsh ejection systolic murmur heard best at the mid-left sternal border (LSB) radiating to the base, with a grade II/VI low-pitched diastolic murmur.

Chest X-ray shows moderate cardiomegaly with clear lung fields.

ECG shows normal sinus rhythm with monomorphic premature ventricular contractions (PVCs), a widened QRS interval measuring 195 milliseconds, left axis deviation and complete right bundle-branch block (RBBB).

What steps should be taken?

See Case Discussion on page 39.

Advances in the management of patients with congenital heart disease have been some of the seminal medical achievements in the past century. The vast majority of newborns with congenital heart disease will now survive to adulthood, including those with severe and previously lethal lesions. This has resulted in a growing population of adult survivors. In Canada, the projected number of adult survivors will grow from 94,000 in 1996 to 124,000 by the end of 2006.¹ It is estimated that the number of adults with congenital heart disease has now exceeded the number of

children with congenital heart disease. The current status of these adults will often reflect the evolving expertise in diagnosis and management over decades. Some patients have had procedures that are no longer used, or may suffer from complications from unrepaired or palliated congenital heart disease. Many of the complications seen in adults with congenital heart disease have contributed to changes in management of newborns and children with the disease. For example, cases of late ventricular failure, arrhythmias and sudden death in adult patients with transposition of the great arteries who have had atrial baffle procedures (*i.e.*, the Mustard procedure) have led to performing the arterial switch procedure in newborns.

The health-care system's capacity to address the specific care issues and medical morbidity for this expanding population was the subject of a recent Bethesda Conference of the American College of Cardiology.¹⁻⁶ It is recognised that the complexity of congenital heart disease and the impact of its past management is well beyond the training and experience of the average adult cardiologist.

While pediatric cardiologists are experts at managing congenital heart disease, they are disadvantaged in dealing with the special care issues of adult patients. This had led to the development of a specialty within adult cardiology, working in partnership with many pediatric cardiologists. In the U.S., many adult patients fall through the cracks, due to difficulties with medical insurance and a lack of developed medical expertise. In Canada, this effort is particularly well-advanced and characterised by specialised centres grouped into the Canadian Adult Congenital Heart (CACH) Network. This network provided the first recommendations (which have been recently updated) for care for adults with congenital heart disease.⁷⁻⁹ These recommendations provide lesion-specific, best evidence-based guidelines for monitoring and management. The recommen-

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Table 1

Disorders That Should Be Seen At Congenital Heart Disease Centres

- Aorto-left ventricular fistula
- Atrioventricular septal defects
- Coarctation of the aorta
- Complete transposition of the great arteries
- Congenitally corrected transposition of the great arteries
- Coronary artery anomalies (except incidental findings)
- Criss-cross heart
- Cyanotic congenital heart disease (all)
- Double outlet ventricle
- Ebstein anomaly
- Eisenmenger syndrome
- Fontan procedure
- Heterotaxy syndromes
- Infundibular right ventricular outflow obstruction of significance
- Mitral atresia
- One ventricle (also called double inlet, double outlet, common, single, primitive)
- Partial anomalous pulmonary venous connection
- Patent ductus arteriosus (not closed)
- Pulmonary atresia (all forms)
- Pulmonary hypertension complicating congenital heart disease
- Pulmonic valve regurgitation (moderate or greater)
- Pulmonic valve stenosis (moderate to severe)
- Pulmonary vascular obstructive disease
- Sinus of Valsalva fistula/aneurysm
- Subvalvar or supra-valvar aortic stenosis
- Tetralogy of Fallot
- Total anomalous pulmonary venous connection
- Tricuspid atresia
- Truncus arteriosus
- Valved conduits
- Ventricular septal defect with
 - absent valves
 - aortic regurgitation
 - aortic coarctation
 - mitral disease
 - RV outflow tract obstruction
 - straddling tricuspid and/or mitral valve
 - subaortic stenosis

Modified from the CCS Consensus Conference 2001: Recommendations for the Management of Adults With Congenital Heart Disease.

dations identify the types of lesions that require the expertise of specialised care (Table 1) and those that may be adequately managed by less specialised providers in the community (Table 2).

Adults with congenital heart disease are at risk for a number of complications and may have specific health-care issues. Families usually develop strong relationships with their pediatric cardiologist, and the patient's lack of symptoms along with the notion they have been "cured" may contribute to a failure to make the appropriate transition to adult care. A health-care team can facilitate a successful transition with a graduated, comprehen-

sive, individualised and coordinated program to educate the patient about specific ongoing care needs. Such a program also addresses the patient's prognosis and provides support. Each of the issues listed in Table 3 should be discussed with the patient.

Pediatric providers should also ensure that medical information is transferred appropriately at the time of transition. Some adults have had associated congenital and acquired disease associated with developmental, cognitive, and physical disabilities. In addition, health anxiety and psychiatric illness may be

present, and these may impact the patient's capacity to make informed decisions about his/her medical condition. Counselling may be necessary for issues, such as sexuality and the capacity for work and economic independence. Many adults with congenital heart disease, including those with repaired or minor lesions, may have difficulty obtaining health, life, and disability insurance. The vast majority of adults with congenital heart disease require no exercise limitations, and some individuals may benefit from a rehabilitation program.

Some adults may have associated non-cardiac anomalies or acquired disease that requires surgical intervention. Adequate preoperative evaluation is necessary. Adults with moderate or severe congenital heart disease may need specialised anesthesia and surgical care at the time of non-cardiac surgery. Managing anticoagulation and other medications and endocarditis prophylaxis require advance planning. These patients are also at increased risk of post-operative complications.

The risks associated with pregnancy increase with the severity of the underlying congenital heart disease and the clinical status of the patient. The presence of pulmonary hypertension is a risk factor for maternal morbidity and mortality, as are poor functional class, arrhythmias, cyanosis, heart failure, important systemic outflow obstruction, and a history of cerebral ischemia. Maternal cyanosis is associated with fetal and neonatal complications.

The patient should be made aware of specific risks associated with pregnancy, and counselled on the appropriate use of contraception. Pregnancy in patients at intermediate or high risk should be planned and closely monitored by a multidisciplinary team with expertise in adults with congenital heart disease. Genetic

counselling should be offered, as there is an increased risk of congenital heart disease in offspring. Fetal echocardiography and genetic testing should be encouraged. Medications should also be reviewed for their potential teratogenic effects.

Table 2

Lesions That Can Be Treated In The Community

Valves

Isolated aortic valve disease

Isolated mitral valve disease (except parachute mitral valve and similar anomalies)

Mild pulmonic valve stenosis

Isolated tricuspid valve disease (except Ebstein anomaly)

Shunts

Secundum atrial septal defect (closed, no residual shunt, no arrhythmia, no pulmonary hypertension)

Ductus arteriosus after complete closure with no residual shunt

Ventricular septal defect (small and isolated, or repaired with no residual shunt)

Repaired partial anomalous pulmonary venous connection

Modified from the CCS Consensus Conference 2001: Recommendations for the Management of Adults With Congenital Heart Disease.

Table 3

Specific Care Issues

Transition to adult care

Understanding of medical condition

Capacity for informed consent

Cognitive and physical disabilities

Psychosocial issues, psychiatric illness

Ability to obtain insurance

Exercise recommendations and rehabilitation

Care needs at the time of non-cardiac surgery

Importance of dental care, endocarditis prophylaxis

Contraception, pregnancy, recurrence risk in offspring, genetic counseling

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Table 4

Medical Morbidity

Cyanosis—erythrocytosis, low platelet counts, iron deficiency, hyperuricemia, pulmonary hypertension and therapy

Residual disease, mechanical problems

Ventricular failure

Pulmonary hypertension

Heart block, tachyarrhythmias

Sudden death

Reoperations, reinterventions, cardiac transplantation

Medications, adverse effects

In addition to having specific health-care issues, adults with congenital heart disease are often at risk for certain medical complications depending on their particular lesion and its past management. Some adults continue to have unrepaired or palliated lesions that cause central cyanosis. Some patients may be cyanosed due to the superimposition of pulmonary vascular obstructive disease and pulmonary hypertension. Oxygen desaturation can induce erythrocytosis, which increases blood viscosity and may eventually cause symptoms. This may also be associated with low platelet counts, iron deficiency, and hyperuricemia. Many patients with acquired pulmonary hypertension or Eisenmenger syndrome can survive for decades with a relatively good quality of life.

Adult patients who have had lesions repaired in a past era may suffer from residual disease, such as leaks at the sites of septal defect closure or residual or recurrent valvular obstruction or regurgitation. Some mechanical problems, such as obstruction in baffle pathways, may result

following the Mustard or Fontan procedure. Many patients face treatment with transcatheter interventions, reoperations or cardiac transplantation. Some patients, particularly those with longstanding ventricular volume or pressure overload, may develop ventricular dysfunction and failure. The right ventricle seems to be particularly vulnerable. This is especially so when it is subjected to chronic volume overload of severe pulmonary regurgitation after repair of tetralogy of Fallot, or when required to serve as the systemic pumping chamber (*i.e.*, with transposition of the great arteries after atrial baffling or in congenitally corrected transposition of the great arteries).

Atrial tachyarrhythmias may manifest in patients with atrial volume or pressure overload (*i.e.*, after the Fontan procedure or after extensive atrial surgery, such as with atrial baffle procedures). Heart block can be associated with

particular lesions (*i.e.*, congenitally corrected transposition of the great arteries) or acquired at the time of surgery (*i.e.*, at ventricular septal defect closure). Such patients have pacemakers that require monitoring and maintenance. Ventricular tachyarrhythmias may manifest in patients who have surgical scarring from ventricu-

lotomy or ventricular failure due to volume or pressure overload. Arrhythmias are associated with an important risk of sudden death, and require complete evaluation and appropriate therapy.

Complications appear to be specific to the underlying congenital heart disease and its management. Patients who have had the Fontan procedure for a single ventricle anatomy are also at risk for thrombosis and thromboembolic events, as well as for protein-losing enteropathy.



**For a good move
see page IBC**

Case Discussion

Several aspects of Mr. CR's case are typical of about half of adults who had moderate or severe congenital heart disease as children. He remains limited in terms of work and social achievements, suffers from depression and is dependent on his parents. Mr. CR's knowledge of his congenital heart disease is vague, and he has failed to make the appropriate transition to adult care. Mr. CR originally had palliation, followed by repair of tetralogy of Fallot — one of the conditions for which annual followup within a specialised adult congenital cardiac program is recommended.

Upon referral to a specialised clinic, an echocardiogram was performed which showed normal LV function with a dilated and poorly functioning RV, an aneurysm of the RV outflow tract, free pulmonary regurgitation and normal branch pulmonary arteries.


Often during repair, which may include transannular patching of the RV outflow tract, the pulmonary valve is disrupted, resulting in severe pulmonary regurgitation. This is well-tolerated for many years, but many adult patients develop RV failure and may benefit from pulmonary valve implantation. While all patients have complete right bundle branch block, a QRS duration of 180 milliseconds or more is known to be associated with sustained ventricular tachycardia and sudden death. RV hypertrophy, fibrosis, dilation, surgical scarring, and aneurysms are predisposing factors.

A cardiac catheterization was performed to assess the patient's hemodynamics and to perform electrophysiologic testing to further characterise his arrhythmia. He underwent pulmonary valve implantation with resection of the RV outflow aneurysm and placement of an automatic implantable defibrillator. He had an unremarkable post-operative course with some improvement in his exercise tolerance. However, the RV remains dilated with moderately reduced function.

Because of the above complications, some patients require medical therapy. They should be monitored and counselled regarding the potential adverse effects of medications.

The majority of current adult survivors have mild or adequately repaired congenital heart disease that can be managed in the community by health-care providers without specialised exper-

tise. These patients usually have an excellent prognosis with a minimal risk of complications (Table 4). Nonetheless, such patients still require education and counselling. They may have problems with transition to adult care, psychosocial issues related to misperceptions and insurance problems. Patients should also be advised of the increased risk that congenital heart disease may be hereditary and may affect their offspring.

Continued progress in diagnosing and managing infants and children with congenital heart disease will likely result in a better prognosis and an improved survival rate for adults with congenital heart disease in the future. 

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