Congenital heart disease: Complexities and considerations

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During normal fetal circulation the fetus receives oxygen directly from the mother through the placenta. Blood enters the fetus's right atrium, and about two-thirds of it then flows through the foramen ovale into the left atrium, bypassing the lungs. Blood from the left atrium passes into the left ventricle and on to the aorta. The remainder of blood from the right atrium flows through the pulmonary artery and is shunted away from the lungs to the aorta through the ductus arteriosus. Blood from the aorta then enters the umbilical arteries and flows into the placenta and is oxygenated. The oxygenated blood flows back through the umbilical cord to the liver, is bypassed through the ductus venosus into the inferior vena cava, and arrives in the right atrium of the heart. At birth the umbilical cord is clamped and the baby no longer receives oxygen from the mother. With the first breaths the lungs begin to expand and the ductus arteriosus and the foramen ovale both close.

The heart is completely formed by 8 weeks of gestation, but if problems occur during the crucial steps of cardiac development congenital heart disease (CHD) can result. The incidence of CHD Canada-wide is estimated to be 12 to 14 cases per 1000 live births[1] and includes a spectrum of abnormalities, from minor narrowing of a blood vessel to malformation of one or more cardiac valves and chambers.

Since the introduction of cardiac surgery and the first successful ligation of a patent ductus arteriosus in the 1930s, the care of patients with CHD has been characterized by dramatic progress and widespread optimism. Historically, most patients with CHD died in infancy or childhood. In the past 4 decades, however, advances in medical care, surgery, and interventional cardiology have changed this poor prognosis, and today over 90% of patients with CHD reach adulthood.

Analysis of administrative data from Quebec indicates that in the year 2000 there was a prevalence rate of 4.09 cases per 1000 adults for all CHD and 0.38 cases per 1000 adults for severe lesions,[2] and that an equal number of adults and children were affected by CHD. Since 2000 the number of adults with CHD has grown to exceed the number of children with CHD. The prevalence of severe CHD in adults increased by 85% from 1985 to 2000 and by 55% from 2000 to 2010, consistent with the concept that the greatest survival benefit has occurred in those with more severe forms of CHD.[2] In 2010, adults represented 66% of the entire CHD population.

One result of the successful treatment of children with CHD is an increasing number of medically complex adult CHD patients who live with uncertainty regarding complications and prognosis. Few of these patients

have been cured and most have been left with residual defects or abnormalities that will lead to problems later in life (e.g., valvular dysfunction, stenosis in previously implanted conduits, ventricular dysfunction, arrhythmias, heart failure, pulmonary hypertension, thromboembolism, complications associated with pregnancy). Issues unique to adults with CHD include long-term and multisystemic effects of single-ventricle physiology, cyanosis, systemically positioned right ventricles, complex intracardiac baffles, and complex arrhythmias. Acquired heart diseases such as hypertension and coronary artery disease may have a significant negative impact on patients with delicately balanced circulation. Even simple procedures may pose additional risk in some patients (e.g., insertion of a jugular line in a patient with intracardiac baffles can result in puncture of the baffle and death). The majority of these patients will require lifelong cardiac-focused medical care and repeat cardiac surgery or interventional procedures. As well, a number of these patients will be developmentally delayed, have learning difficulties, and/or psychological, social, and financial difficulties that present barriers to proactive health management. Many of these patients will require genetic counseling, reproductive counseling, advice about insurability, and guidance regarding appropriate leisure activities, employment restrictions, and career choices.

The majority of adult CHD patients should be seen at least once by an adult CHD specialist to determine the most appropriate care, and certainly patients with moderate to severe disease merit ongoing follow-up by an expert multidisciplinary team dedicated to the care of adults with CHD. There is strong evidence to show that referral to and management by such a team decreases morbidity and mortality in this complex patient population.[3]

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At some time in their medical careers, most physicians in BC will encounter patients who have CHD. Physicians are often at a loss when dealing with these medically complex patients and can benefit from knowing more about common issues and available resources.

In this theme issue we attempt to bridge some knowledge gaps with four articles by adult congenital heart disease experts from the Pacific Adult Congenital Heart Disease program based at St. Paul's Hospital in Vancouver. The first article, by Dr Marla Kiess, provides some historical context and reviews the current state of care for adults with CHD in BC. The second article, by Drs Andrew Campbell and Ronald Carere, describes current surgical and noninvasive interventions for managing adult patients with CHD.

In the third article, Dr Jasmine Grewal and colleagues consider specific medical issues in adult CHD patients related to pregnancy, pulmonary hypertension, and arrhythmias. Finally, in the fourth article, Ms Karen LeComte and colleagues discuss the need to ensure a successful transition and transfer when a patient moves from pediatric to adult care.

The goal of these articles is not to outline specific management principles, but rather to highlight some complexities and considerations faced by these patients so that physicians can make appropriate referrals and provide better care for adults with congenital heart disease.

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