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Surgical and interventional management of adult congenital heart disease

A growing population of patients with adult CHD is being helped today by rapidly developing surgical techniques and catheter-based technologies.

ABSTRACT: Survival of pediatric patients after surgery for congenital heart disease has consistently improved over the past 2 decades. As a consequence, more young adults are presenting with both historical and contemporary repairs, and with both anticipated and completely unanticipated complications. Surgery for adult congenital heart disease continues to evolve in Canada, and even the recent rapid progress in percutaneous and hybrid approaches to congenital heart disease has had no impact on the volume or the complexity of surgical cases. Technological advances now permit the treatment of both relatively simple and very complex anatomical and pathophysiological problems using percutaneous techniques. Managing adult congenital heart disease patients now means choosing from the surgical and interventional options available and determining which cases will truly benefit from novel therapies and which will require only what has always been done. We will continue to need well-integrated congenital heart disease programs that permit collaboration between adult and pediatric medical and surgical subspecialists.

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Over the past 2 decades the survival of pediatric patients after surgery for congenital heart disease (CHD) has consistently improved, with in-hospital mortality rates now routinely below 3%. With this advance more children are now surviving and presenting as young adults with both historical and more contemporary repairs and therefore with both anticipated and completely unanticipated complications. This means we must look at the different levels of complexity of adult CHD and the array of surgical and interventional options, and determine which cases will truly benefit from novel therapies and which will require only what has always been done.

Surgical management

Modern surgical management of congenital heart disease began on 26 August 1938 with the first documented successful ligation of a patent ductus arteriosus (PDA) in a 7-year-old patient by Dr Robert Gross in Boston.[1] The operation was scheduled while surgeon-in-chief Dr William Ladd was on vacation for fear that Ladd would not allow the groundbreaking surgery to go ahead. Later this led to an irreparable rift between the two surgeons, despite confirming that native defects of the heart and great vessels were amenable to surgical repair.

As Dr Gross stated in his case report, at that time 50% of the children affected by PDA could be expected to die in infancy and the remainder would suffer the consequences of heart failure and, potentially, Eisenmenger syndrome, which would lead to an early death in the third or fourth decade of life. PDA ligation is now performed at the bedside in the neonatal intensive care unit in 30 minutes or less.

Since this beginning, cardiac surgery for congenital defects has made great strides, but at every step there has been tension between the possible benefits of innovation and the risk of causing more harm than existing therapies or the natural history of the disease. This tension was very apparent in the 1980s when therapy for transpo-

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sition of the great arteries underwent a revolution as atrial-level repair was replaced by an arterial-level operation developed principally by Dr Jatene of the University of Sao Paulo. For the first time, the coronary arteries of children were dissected and transposed from the pulmonary root to the neoaorta, a procedure that would have been considered impossible in 1975. After repeated efforts, Jatene and his team developed a programmatic approach to the management of these children; even so, Jatene's review of his first 116 patients revealed an early mortality rate of almost 21%, with only 33% of surviving patients having normal function and a good anatomic repair.[2] Over this same period, centres experienced with atrial-level repairs achieved early mortality rates of 2% to 3%. However, 25 years later it appears that the long-term benefits of a systemic left ventricle clearly outweigh the risks of a more complex neonatal operation as these children have grown into adults with improved survival and better exercise tolerance.[3] Thus the challenge for management of congenital heart disease remains: How do you decide which procedure is safest when one of the procedures is novel and the disease process is unknown?

The different forms of adult congenital heart disease requiring management (**Table**) can be grouped as follows:

- Category 1. Previously undiagnosed pediatric disease that presents in adulthood.
- Category 2. Anticipated residual disease following a pediatric surgical procedure or palliated pathology.
- Category 3. Unanticipated deterioration following a pediatric surgical procedure or palliated pathology.

Understandably, most new lesions will initially present as category 3 disease. Once several patients with similar anatomy are identified, what was initially thought of as unanticipated becomes anticipated and a new algorithm for care is developed.

Category 1 disease

The most common category 1 condition leading to surgical referral 15 years ago was ostium secundum atrial septal defect (ASD). Now with the refinement and expansion of catheter-based technologies surgeons see fewer isolated atrial septal defects. Interestingly, the increased scrutiny of patients with atrial septal defects for possible catheter-based closure has identified a large number of patients with associated anomalous pulmonary venous return who cannot undergo catheter-based closure and are therefore referred for more complex surgical repair with venous baffling of the anomalous veins. So despite the large number of ASD cases being managed in the catheterization laboratory, the identification of underdiagnosed patients means the number of surgical cases has actually increased.

Asymptomatic ventricular septal defects are surprisingly common as an incidental echocardiographic finding but few are found to have a significant shunt. Clefts and parachute-morphology mitral valves may present in adulthood with a gradual progression of regurgitation or stenosis and can be occasionally misinterpreted as a more routine anatomy amenable to conventional repair. This can lead to the rare intraoperative consultation for the congenital heart disease surgeon in a colleague's operating room, and heightens the importance of having a well-integrated CHD program with collaboration between adult and pediatric medical and surgical subspecialists.

Category 2 disease

The most common category 2 condition leading to surgical referral remains pulmonary insufficiency associated with tetralogy of Fallot (TOF). Up until the 1980s, the vast majority of surgical repairs for TOF

Table. Forms of adult congenital heart disease requiring management.

	Complexity	Examples	Outcome
Category 1 disease Previously undiagnosed pediatric disease presenting in adulthood	Low	Atrial septal defect, ventricular septal defect, double aortic arch, cleft mitral valve	Good
Category 2 disease Anticipated complications from pediatric repair or palliated pathology	Moderate	Pulmonary insufficiency following repair for tetralolgy of Fallot, mitral regurgitation following repair for atrioventricular septal defect, failure of Fontan repair, systemic right ventricle failure following atrial switch procedure	Varies
Category 3 disease Unanticipated complications from pediatric repair or palliated pathology	High	Neoaortic valve failure following arterial switch, systemic ventricular dysfunction following repair for ALCAPA (anomalous left coronary artery from pulmonary artery)	Unknown

involved a vertical incision crossing the pulmonary valve and rendering it incompetent. Over time the volume load on the right ventricle creates dilatation and eventually a decrease in ventricular function that can lead to symptoms. A great deal of research has been performed to determine the right ventricular volumes above which an adverse surgical outcome can be expected and whether earlier restoration of pulmonary competence will lead to a more rapid return to normal ventricular dimensions. Other considerations include timing of valve implantation, valve type, and the possibility of future percutaneous valve implantation.

Category 3 disease

The most complex and challenging cases for the surgeon involve category 3 disease, as patients in this category generally have survived the most severe forms of pediatric CHD. They often have diminished systemic ventricular function, either due to a right ventricle in the subaortic position or as a consequence of multiple episodes of cardiopulmonary bypass. There is presently little evidence to guide timing or determine the type of repair that is best for these patients.

For patients who have received a classic right atrial to pulmonary anastomosis as the final stage of their single-ventricle palliation, their atrium eventually dilates and becomes an inefficient capacitance chamber for systemic venous drainage, which leads to decreased forward flow into the pulmonary circulation and abdominal congestion. These patients are considered to have a failed or failing Fontan circulation,[4] and as a result are candidates for the more contemporary Fontan repairs that have been developed. Much remains poorly understood about this patient population. Similarly, when patients with a subaortic right ventricle begin to develop ventricular dysfunction, their generally young age and tolerance of cyanosis can mean they tolerate pulmonary edema and diminished cardiac output much better than older ischemic cardiac patients. As single-ventricle anatomy and multiple sternotomies make the prospect of implanting a ventricular assist device daunting, CHD patients with minimal symptoms but severe dysfunction are often overlooked for this therapy and therefore have great difficulty qualifying for cardiac transplantation.[5] Establishment of a separate congenital heart disease transplant list has been discussed in several countries, but does not yet exist in North America.

Surgery for adult congenital heart disease continues to evolve in Canada. Even the recent rapid progress in percutaneous and hybrid approaches to complex heart disease has had no impact on the volume or the complexity of surgical cases seen in our centre. Instead, with an ever-growing population of pediatric CHD patients who will require reintervention in future, we expect to be providing more options for patient management and working to minimize the complexity of procedures that will be needed over time. As the pediatric CHD population continues to age, more late postoperative complications will be recognized and the number of patients with category 3 disease will increase. And as we saw with the work of Dr Gross and Dr Jatene, surgeons will continue to weigh possible benefits against the risk of a new intervention, and in time new surgical treatments will evolve.

Interventional management

Technological advances now allow us to treat both relatively simple and very complex anatomical and pathophysiological problems using percutaneous techniques. Interventions for adult CHD have for some time employed devices and techniques that are referred to as "structural heart disease interventions."[6] These interventions require the operator to have expertise in congenital heart disease and the unique surgeries and connections that many of these patients have had in the past. Broadly speaking, interventions fall into two main categories: those for closing abnormal connections and those for relieving obstructions.

Closing abnormal connections

The primary goal in closing an abnormal connection is to obtain or improve separation of the oxygenated arterial circulation from the deoxygenated venous circulation. Considerations include the effect on symptom status, the vasculature, cardiac chambers, and/or physiology. Closure of connections can also be performed to prevent paradoxical embolization, depending on the clinical scenario and the lesion in question. Small connections that have no significant hemodynamic effect or present little risk for paradoxical embolization are often followed clinically.

Although there are several devices available to close abnormal vascular connections, the most commonly used devices worldwide are occluders manufactured by St. Jude Medical in St. Paul, Minnesota. These devices are made from a deformable nickel and titanium alloy that allows passage through a catheter and rely on a screw-release mechanism that allows the device to be controlled until it is deliberately released (**Figure 1**). Different series are designed specifically for closure of atrial septal defects, patent foramen ovale, patent ductus arteriosus, and vascular connections.

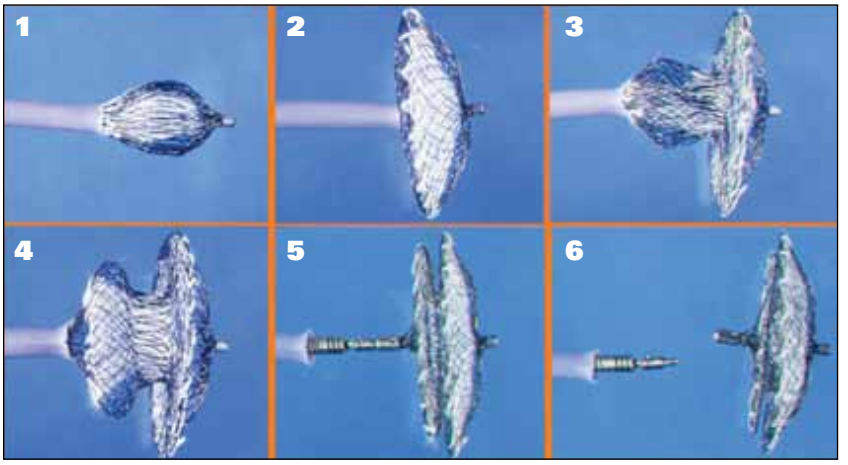

Figure 1. Steps in the deployment of the Amplatzer septal occluder.

Step 1: Delivery sheath is withdrawn to expose left atrial retention disc. **Step 2:** Left atrial disc is fully deployed to prepare for pulling entire mechanism back to atrial septum. **Step 3:** Waist of device (sized to fit defect) is deployed. **Step 4:** Delivery sheath is pulled back to right atrium to begin deployment of right atrial retention disc. **Step 5:** Device is fully deployed, with left and right retention discs positioned on either side of atrial septum, and placement and security of device are checked. **Step 6:** Delivery cable is unscrewed and removed.

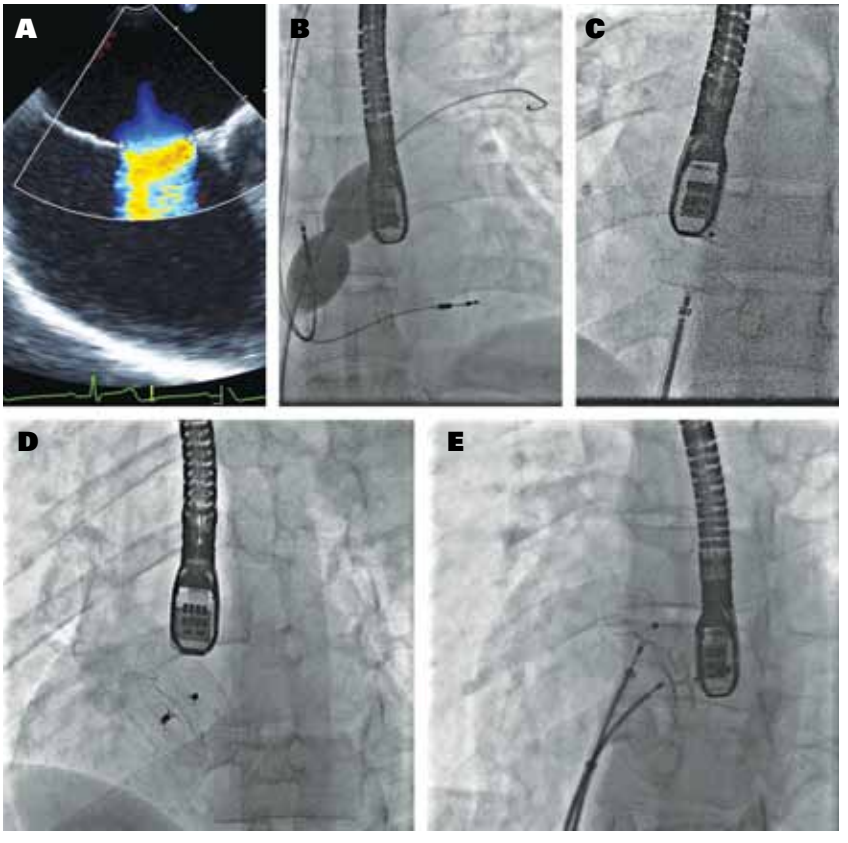

Figure 2. Atrial septal defects

Atrial septal defects. There are various atrial septal defects that can be repaired with closure devices, with the most common being the ostium secundum atrial septal defect. The direction and magnitude of the shunt in an ASD patient is determined by the size of the defect and the relative compliance of the right and left ventricle. The shunt occurs predominantly from the left atrium to the right atrium and can lead to right atrial and right ventricular dilation, arrhythmias, and right-sided heart failure. Although it is rare to develop pulmonary hypertension, if this does occur closure should be undertaken only after careful consideration.

The commonly used Amplatzer atrial septal defect occluders come in sizes ranging from 4 to 40 mm, with retention discs that are 8 to16 mm larger than the waist. All patients are prescreened with a transesophageal echocardiogram (TEE) to confirm there is an adequate rim of atrial septal tissue to safely anchor the closure device and ensure there are no other associated congenital heart defects that would preclude percutaneous closure. After the size of the defect is established using transesophageal or intracardiac echocardiography, the device is positioned and released once the position and stability are confirmed. Although large defects and multiple defects are more challenging

Figure 2. A: Image from a transesophageal echocardiogram (TEE) combined with color doppler ultrasound reveals the size of an atrial septal defect and shows blood flow through the defect from the left atrium (top) to the right atrium. **B:** Sizing balloon is inflated in a defect so that the balloon waist can be measured and the appropriate device size selected. **C:** A 14-mm septal occluder in position with the left atrial disc deployed. **D:** A 34-mm septal occluder in position. **E:** Two devices positioned but not yet released—a septal occluder (higher to left) and a cribriform patent foramen ovale occluder with a narrow waist to close multiple defects.

to correct than small single defects, they can often be closed successfully (Figure 2).[7]

Patent ductus arteriosus. In the developing fetus, the ductus arteriosus between the pulmonary artery and the descending aorta is the connection that enables passage of oxygenated blood from the mother's placenta directly to the systemic circulation. When this fails to close shortly after birth, blood shunts from the aorta to the pulmonary artery. Indications for closure include a significant shunt that affects the left heart chambers or pulmonary pressures. More controversially, closure may also be considered as a way to reduce the long-term risk of endarteritis in the event of systemic bacteremia.

Patent ductus arteriosus comes in a variety of configurations. When small, such defects may be closed with coils, while larger defects may be closed with one of many Amplatzer devices (Figure 3).

Fistulas. Fistulas are connections between vascular structures and can be arteriovenous or venovenous. Fistulas can also occur between a cardiac chamber and vein or artery. Indications for fistula repair include the establishment of hemodynamic significance, deoxygenation, and endocarditis. Closure of a fistula commonly involves a vascular plug (Figure 4).

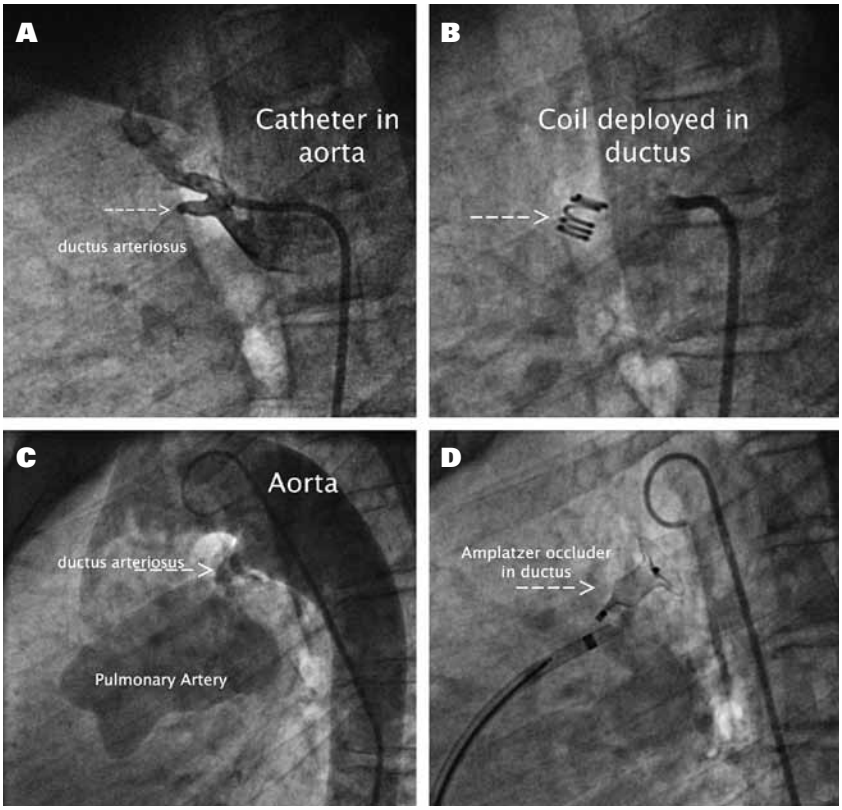

Figure 3. Patent ductus arteriosus

Figure 3. A: Patent ductus shown from contrast injection from the aorta. The small defect results in minimal evident blood flow into the pulmonary artery in this image. **B:** The defect shown in image A has been closed using Cook "Flipper" coils (Cook Medical, Bloomington, IN). **C:** A larger patent ductus is shown with flow evident from the aorta into the pulmonary artery. **D:** Defect shown in image C successfully closed with an Amplatzer duct occluder (St. Jude Medical, St. Paul, MN).

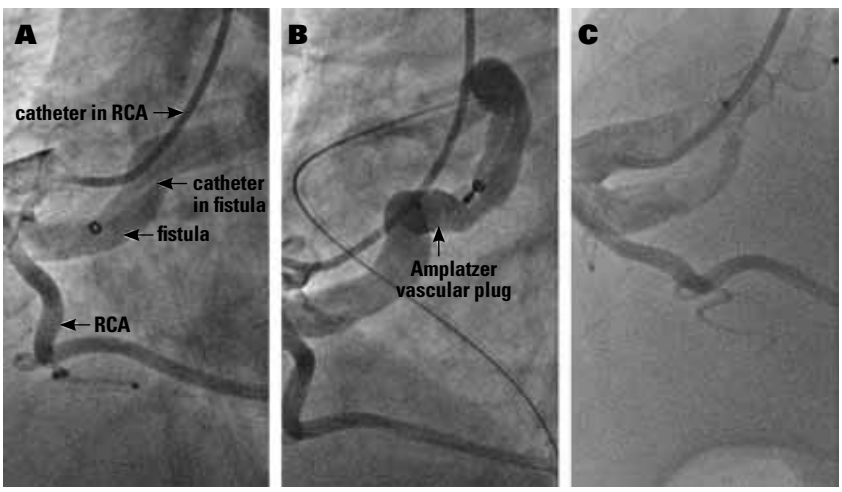

Figure 4. Atrial septal defects

Figure 4. A: Large fistula (centre) from the right coronary artery to the right atrium in a 45-year-old man who had previously experienced an episode of bacterial endocarditis and right atrial vegetation that was successfully treated with antibiotics. **B:** Delivery cable for 12-mm Amplatzer vascular plug can be seen traversing the right atrium from the lower right corner of the image toward the upper left and entering the fistula, where the cable will be used to position and deploy the plug. **C:** Successful occlusion of fistula after deployment of the plug.

Relieving obstructions

Relieving obstructions in native vascular or postsurgical connections involves basic interventional cardiac and vascular procedures. Typically, the structure to be treated is crossed with a guide wire and then prepared with a balloon. Although the balloon may be a definitive treatment, contemporary procedures usually employ stents to improve the immediate and long-term result. A variety of wires are available along with balloon and stent products that range from several millimetres to 30 mm in diameter. Stents are typically made from stainless steel and alloys of platinum and iridium or cobalt and chromium, and may be used as bare metal scaffolds or with a covering of polyethylene to reduce the risk of vascular rupture (**Figure 5**).

Venous obstructions. There are no clear indications for treating obstructions involving the venous circulation. However, even a small pressure gradient can be important in inhibiting venous return. Also, since the pressure is typically assessed and measured at rest, a low pressure gradient can be expected to worsen on exercise when greater venous return is required to increase cardiac output. An example of a repair for an extreme case of obstructed venous return is shown in **Figure 6**. The patient originally had complex congenital heart disease and was palliated with a number of surgical procedures over many years before undergoing heart transplantation. Given the significantly abnormal anatomy, the transplant surgeon needed to construct unique connections to provide venous return from the patient to the donor heart. In the postoperative period, one of these connections from the head and neck did not remain patent and the connection from the inferior vena cava was restrictive because it was necessary to use a portion of a pre-existing conduit. These anomalies were successfully treated by percutaneous stent placement in a two-stage procedure. Treatment also included working directly with a device manufacturer to create a custom-made covered stent, a requirement in cases where the anatomy is very complex and there is no product readily available.[8,9]

Figure 5. Stents commonly used for treating congenital heart disease.

From left: expanded and unexpanded Cheatham-Platinum (CP) stents. **From right:** expanded and unexpanded, covered versions of CP stents (NuMed, Hopkinton, NY).

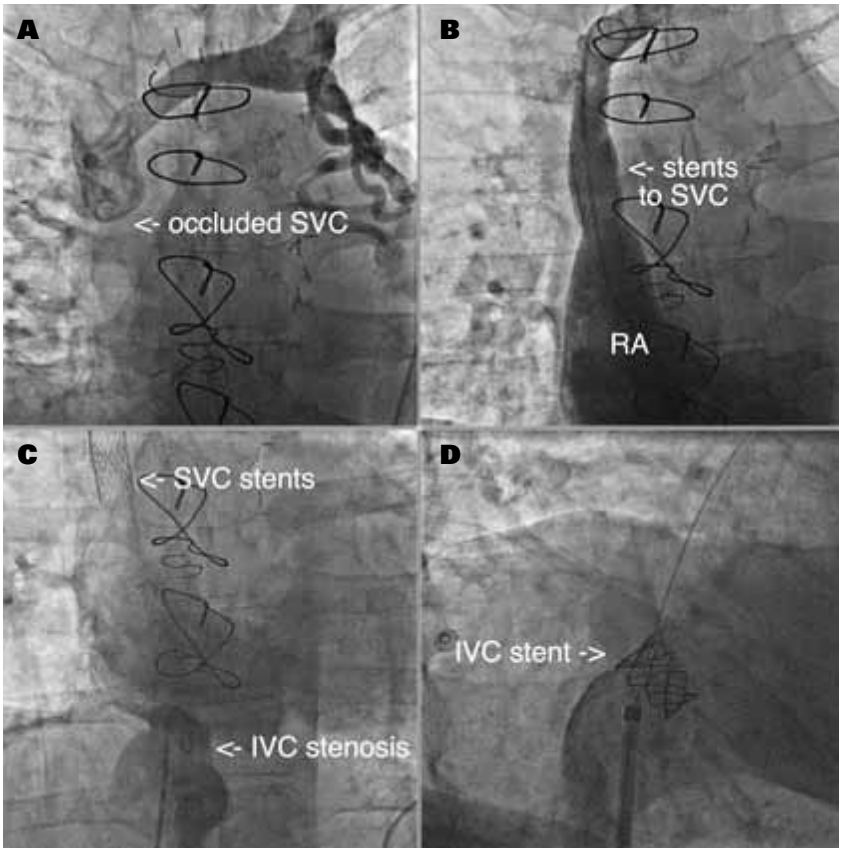

Figure 6. Venous obstructions

Figure 6. A: Obstructed connection from the superior vena cava to the right atrium in a heart transplant patient. **B:** Obstruction seen in image A treated with three percutaneous stainless steel Genesis stents (Cordis Corp, Fremont, CA). **C:** Heart of same transplant patient showing stenosis at the inferior vena cava connection to the right atrium as a result of a remnant of synthetic conduit left from a previous surgical procedure. **D:** Stenosis seen in image C treated with placement of a custom-made 16-by-22-mm Cheatham-Platinum stent (NuMed, Hopkinton, NY).

Aortic and valvular obstructions. Indications are usually clearer for treating aortic and valvular obstructions; these issues can often be resolved with interventional procedures as shown in **Figure 7**.

Aortic coarctation is an example of an obstruction that can be successfully managed without complex surgery using a contemporary approach that employs covered stents. In general, a peak-to-peak systolic pressure gradient of less than 20 mm Hg is an indication for intervention, although other factors, such as the presence of systemic hypertension, left ventricular hypertrophy, or extensive collaterals, will also influence the management decision.

Although the more common example of percutaneous aortic valve implantation has been well described,[10] percutaneous implantation was originally used for valves in the pulmonary position for patients with significant right ventricular outflow tract obstruction or severe pulmonic regurgitation.[11] The first valve available was approved in Canada in 2007. The Melody valve (Medtronic, Minneapolis, MN) is constructed from bovine jugular vein and is indicated for relief of obstruction and/or regurgitation in right ventricular to pulmonary artery conduits ranging from 14 to 22 mm. The number of adult patients who can benefit from implantation of this valve has been limited despite improved techniques, largely because of the small valve size. Recently, the SAPIEN XT valve from Edwards Lifesciences has become the valve of choice because of the availability of larger sizes more suitable for use in adults.

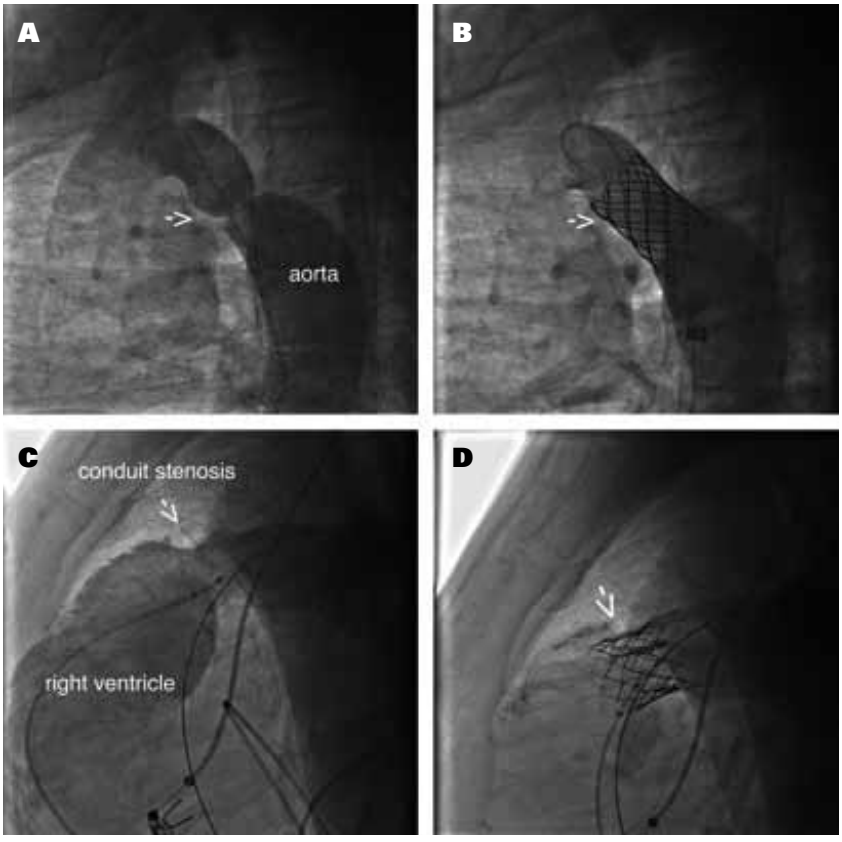

Figure 7. Aortic and valvular obstructions.

Figure 7. A: Focal coarctation of the aorta. **B:** Stenosis seen in image A and problem of associated pressure gradient are resolved with placement of covered stent (NuMed Inc, Hopkington, NY). **C:** Stenosis in conduit from right ventricle to pulmonary artery. **D:** Stenosis seen in image C is resolved with placement of a large stent (Johnson and Johnson, New Brunswick, NJ), which also helps address significant pressure gradient and pulmonary valve incompetence by providing an anchoring site for a SAPIEN transcatheter heart valve (Edwards Lifesciences, Irvine, CA).

Aortic coarctation is an example of an obstruction that can be successfully managed without complex surgery using a contemporary approach that employs covered stents.

Summary

Since the first successful ligation of a patent ductus arteriosus by Dr Robert Gross, cardiac surgery for congenital

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defects has made great strides. Today management for adult CHD addresses problems in patients with previously undiagnosed pediatric disease that presents in adulthood, anticipated residual disease following a pediatric surgical procedure, and unanticipated deterioration following a pediatric surgical procedure. Technological advances also now allow us to treat both relatively simple and very complex problems using percutaneous techniques. Broadly speaking, interventions fall into two main categories: those for closing abnormal connections and those for relieving obstructions.

With a growing population of patients with adult congenital heart disease and the developing technologies available to us, we will continue to need well-integrated CHD programs that permit collaboration between adult and pediatric medical and surgical subspecialists. We will also need to consider which is the most appropriate surgical or interventional option for each patient.

Competing interests
None declared.

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