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# History and evolution of the treatment of adult congenital heart disease

Surgical developments and other advances mean that more congenital heart disease patients are reaching adulthood and requiring the support of a team that includes cardiologists, nurses, psychologists, and social workers with knowledge of adult CHD.

**ABSTRACT: Cardiology experts around the world, including many Canadians, have contributed to dramatic surgical, interventional, and diagnostic advances since the 1930s. These developments began when Dr Helen Taussig established the pediatric cardiology clinic at Johns Hopkins Hospital in Baltimore in 1930 and Dr Maude Abbott of Montreal published the *Atlas of Congenital Heart Disease* in 1936. The first surgical procedure was ligation of a patent ductus arteriosus performed by Dr Robert Gross at the Children's Hospital in Boston in 1938. Intracardiac repair first became possible with the development of cardiopulmonary bypass technology in the 1950s, followed in the 1970s by the development of deep hypothermia with circulatory arrest, which made lengthier surgeries possible. Interventional techniques went hand in hand with surgical advances. Balloon dilatation of the pulmonary valve was first described in the 1950s and became widely used after static balloon dilatation was introduced in 1982. Balloon atrial septostomy was developed in 1966 to promote mixing at the atrial level and dramatically improved the outcome for newborns with complete transposition of the great arteries. Beginning with innovative use of X-ray imaging, diagnostic techniques supported both surgical and nonsurgical interventions. Right heart catheterization became available in the late 1940s and left heart catheterization was developed in the 1950s. The advent of two-dimensional echocardiography in the 1970s permitted a major step forward in the treatment of congenital heart disease (CHD), as did the establishment of standardized nomenclature. Canadian doctor Wilfred Bigelow determined how to use total body hypothermia for open heart surgery in 1953, and the first open heart procedure in Canada was performed by Dr John Callaghan in Edmonton in 1954. In British Columbia, Dr Ross Robertson performed a Blalock-Taussig shunt, closed a patent ductus arteriosus, and repaired a coarctation of the aorta at Vancouver General Hospital in 1947. In the late 1950s Dr Harold Rice built the first cardiopulmonary bypass machine used at St. Paul's Hospital. Because of the many advances made since the 1930s, children born with CHD today are much more likely to grow to adulthood, but they are also likely to require multiple operations for scarring and narrowing of arteries or veins and insertion or replacement of conduits and valves. Patients with moderate to severe disease are rarely cured and face a lifetime of repeat surgical and interventional procedures. Each year, BC Children's Hospital registers approximately 500 newly diagnosed CHD patients and moves 300 previously diagnosed patients from pediatric to adult care. Approximately 150 patients per year will require follow-up in an adult CHD clinic. A review of advances in the treatment of CHD reveals dramatic progress beginning in the 1930s and continuing to the present. Cardiology experts around the world, including many Canadians, have contributed to a variety of surgical, interventional, and diagnostic developments.**

*This article has been peer reviewed.*

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## Surgical developments

The organized study of congenital heart disease (CDH) began with the establishment of Dr Helen Taussig's pediatric cardiology clinic at Johns Hopkins Hospital in Baltimore in 1930[1] and the publication of Dr Maude Abbott's incredible atlas describing 1000 CHD cases in 1936.[2] The first surgical procedure was ligation of a patent ductus arteriosus (PDA) performed by Dr Robert Gross at the Children's Hospital in Boston in 1938.[3] Dr Taussig had observed that some children became progressively more cyanotic with spontaneous closure of the ductus arteriosus and proposed using an arterial to pulmonary artery shunt. She convinced Dr Alfred Blalock of the merit of this idea and eventually Blalock collaborated with his technician, Vivien Thomas, to construct a shunt from the right subclavian artery to the right pulmonary artery in a cyanotic child. A report on the procedure was published in 1945.[4] Also in 1945, Drs Crafoord and Nylin of Stockholm performed surgery on a patient with coarctation of the aorta.[5] In 1948, Sir Russell Brock, working in Guy's Hospital in London, England, published a report describing three cases of pulmonary stenosis that were repaired with pulmonary valvotomy.[6] In 1950, Drs Blalock and Hanlon performed atrial septectomy using a surgical clamp devised by Vivien Thomas.[7] With the development of cardiopulmonary bypass technology, intracardiac repair became possible. The first procedure done with the use of a heart-lung machine was for closure of an atrial septal defect and was performed by Dr Gibbon in Philadelphia in 1953.[8] Later that year, Dr Lillehei, working in Minneapolis, performed open heart surgery using cross-circulation between the child and a parent. This procedure was found to have a high mortality rate, which led Lillehei and his colleagues to develop a pump oxygenator.[9] However, even with this innovation the preservation of blood flow to the brain was not always optimal and surgeons had to work quickly until the development of deep hypothermia with circulatory arrest in the early 1970s made lengthier surgeries possible.

## Interventional and diagnostic techniques

Interventional techniques went hand in hand with surgical advances. Although balloon dilatation of the pulmonary valve was described in 1953 by Rubio-Alvarez and colleagues,[10] the procedure did not become widely used until Kan and colleagues[11] introduced static balloon dilatation in 1982. Balloon atrial septostomy, developed in 1966 by Drs Rashkind and Miller,[12] promoted mixing at the atrial level and dramatically improved the outcome for newborns with complete transposition of the great arteries. There was an explosion of catheter-based therapies in the 1980s, including balloon dilatation for repair of coarctation of the aorta and stenotic valves, shunts, and conduits. The development of stents vastly improved long-term results. Various devices became available to address fistulae, patent ductus arteriosus, and atrial septal defects. A major advance was the development of prosthetic pulmonary valves by Dr Bonhoeffer[13] and prosthetic aortic valves by Drs Cribier[14] and Webb[15] in the 2000s.

Diagnostic techniques, beginning with Dr Taussig's innovative use of X-ray imaging, supported both surgical and nonsurgical interventions. Right heart catheterization became available in the late 1940s and left heart catheterization was developed in the 1950s. M-mode echocardiograms, first available in the 1960s, were helpful, but it was the advent of two-dimensional echocardiography in the 1970s that permitted a major step forward. Important advances in pathology included the establishment of standardized nomenclature by Richard and Stella Van Praagh working in Toronto, Chicago, and then Boston, and by Robert Anderson, working in London, England.

**Canadians have been at the forefront of improvements for patients with congenital heart disease.**

## Canadian contributions

Canadians have been at the forefront of improvements for patients with congenital heart disease, beginning with Dr Maude Abbott of Montreal, who wrote the *Atlas of Congenital Heart Disease* already mentioned. Dr Wilfred Bigelow[16] of the Toronto

General Hospital determined how to use total body hypothermia for open heart surgery in 1953. The first open heart procedures in Canada were for closure of an atrial septal defect and a ventricular septal defect and were performed by Dr John Callaghan in Edmonton in 1954. Dr William Mustard at the Hospital for Sick Children in Toronto significantly advanced the care of patients with complete transposition of the great arteries with his atrial switch operation (Mustard procedure) in 1963.[17]

In British Columbia, Dr Ross Robertson performed a Blalock-Taussig shunt, closed a patent ductus arteriosus, and repaired a coarctation of the aorta at Vancouver General Hospital in 1947. Dr Jack Stenstrom started performing PDA ligations and Blalock-Taussig shunts in Victoria in 1949. In 1957, Dr Peter Allen, with the assistance of Drs Phil Ashmore, Bill Trapp, and Ross Robertson, performed the first open heart procedure at Vancouver General Hospital, closing an atrial septal defect in a 9-year-old boy.[18] In the late 1950s, Dr Harold Rice built the first cardiopulmonary bypass machine used at St. Paul's Hospital.[19] He had a very personal reason for wanting to do this: his wife, Dorothy, was born with an atrial septal defect, and in 1953 she was the fourth patient in the world and the first patient at the Mayo Clinic to have open heart surgery under hypothermic cardiac arrest. An anomalous pulmonary vein discovered at surgery could not be repaired until the advent of cardiopulmonary bypass, and she had a second procedure in 1958 at the Mayo Clinic when she was in her late forties. Drs Bob Gourlay, Ted Musgrove, and Gerry Coursley closed an atrial septal defect in a 12-year-old girl using Dr Rice's machine at St. Paul's Hospital in 1960. Cardiac catheterization was first performed at Vancouver General Hospital by Drs Morris Young and Dennis Vince, starting in the mid-1950s. Dr Doris Kavanagh performed the first cardiac catheterization at St. Paul's Hospital in 1959. The need for this procedure was great. After her first successful study, Dr Kavanagh was asked by Dr Young if she could catheterize some of his patients and he sent her a list of 400 patients who had been waiting for as long as 4 years.

**In the early days of cardiac surgery, intracardiac repairs could only be performed on older children.**

## Further developments

In the early days of cardiac surgery, intracardiac repairs could only be performed on older children. Congenital heart lesions frequently resulted in too much or too little blood flow to the lungs. Infants and young children with reduced blood flow to the lungs were palliated with arterial shunts, either Blalock-Taussig (subclavian artery to pulmonary artery), Potts (descending aorta to pulmonary artery), or Waterston (ascending aorta to pulmonary artery), and those with excess blood flow to the lungs were palliated with pulmonary artery banding. The flow through these arterial shunts was difficult to control and pulmonary hypertension was a significant risk. Dr Glenn felt that venous shunts would be superior, anastomosing the superior vena cava to the pulmonary artery in 1959.[20] Many patients had repeat operations with ligation of arterial shunts and replacement with right and/or left Glenn shunts. When the child patient reached an adequate size, usually around age 4, intracardiac repair was performed, the shunts were ligated, or the pulmonary band was removed. Dr Fontan developed total right heart bypass for patients with single-ventricle physiology in 1971[21] and subsequent modifications to improve hemodynamics were developed by him and Dr de Leval.[22] In the early 1980s, Dr Aldo Castenada perfected neonatal repairs at the Boston Children's Hospital.[23]

## Adult congenital heart disease care in BC

As in the past, many children born with congenital heart disease today will require multiple operations as they grow to adulthood for various reasons, including scarring and narrowing of arteries or veins and insertion or replacement of conduits and valves. Patients with moderate to severe disease are rarely cured and face a lifetime of repeat surgical and interventional procedures.

Based on a Canada-wide incidence rate of 12 to 14 cases per 1000 live births,[24] 500 to 600 infants with CHD are born per year in British Columbia. Data suggest that as of 2010 over 24 000 individuals with CHD born in BC had survived to adulthood. Some of these adults have simple defects and have little need for medical care. However, over 12 000 adults have moderate to severe defects and will require lifelong care by an array of health professionals with expertise in the field of CHD.

BC Children's Hospital currently registers approximately 500 newly diagnosed patients with CHD every year and moves 300 patients from pediatric to adult care each year. Approximately 50% of these patients, or 150 per year, will have moderate to severe CHD and require follow-up in an adult CHD clinic. These patients need ongoing evaluation to determine whether they require further intervention or medical management. About 110 of these patients (60%) can be expected to require specialized continuing care for optimal quality of life. A smaller but significant number of individuals present later in childhood or early adulthood with congenital defects that have gone undetected due to the sometimes insidious nature of CHD progression, and like patients with known CHD, these newly diagnosed patients may need advice regarding pregnancy risks and cardiac surgery options.

The range of abnormalities, the complexities of postoperative anatomy, and the challenges of multisystem involvement mean a full understanding of CHD is now well beyond the education and experience of the typical cardiologist caring for adult patients. To care for these patients, practitioners require knowledge and training in congenital heart disease, adult cardiology, and general medicine, and the support and expertise of a multidisciplinary team (nurses, psychologists, social workers) who have knowledge of CHD.

A recent study from Quebec has shown that these complex patients have higher rates of hospitalization, more visits to emergency rooms, greater use of outpatient cardiologist care, and more days in critical care.[25] The Canadian Cardiovascular Society,[26] American College of Cardiology,[27] and European Society of Cardiology[28] have all recognized the urgent need for trained medical staff, allied health personnel, and specialized clinics to deliver appropriate care to this rapidly growing population of adults with CHD.

## Summary

Many advances have followed the first successful ligation of a patent ductus arteriosus in 1938. Intracardiac repair became possible with the development of cardiopulmonary bypass technology in the 1950s, while lengthier surgeries became possible after the development of deep hypothermia with circulatory arrest in the 1970s. Interventional techniques have accompanied surgical advances, and a variety of imaging innovations have supported the development of both surgical and nonsurgical interventions. The many advances made since the 1930s mean that children born with CHD today are much more likely to grow to adulthood. However, they are also likely to require multiple operations for scarring and narrowing of arteries or veins and insertion or replacement of conduits and valves, and to require the support and expertise of a multidisciplinary team with knowledge of CHD.

**BC Children's Hospital currently registers approximately 500 newly diagnosed patients with CHD every year and moves 300 patients from pediatric to adult care each year.**

**Competing interests**

None declared.

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