The Adult with Congenital Heart Disease: You've come a long way blue baby!

Lisa W. Forbess, M.D.

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[University of Texas Southwestern Medical Center at Dallas]

Lisa W. Forbess, M.D.

Associate Professor of Internal Medicine

Division of Cardiology

UT Southwestern Medical Center Dallas

My clinical and research interests are in valvular and congenital heart disease and echocardiography. Care for the adult patient with congenital heart disease is a challenging subspecialty of cardiology that requires a team of highly dedicated cardiologists and cardiac surgeons in regional centers of excellence. I work closely with both adult and pediatric interventional cardiologists, imaging experts, electrophysiologists, heart failure and transplant cardiologists, and cardiac surgeons at UTSW and Children's Medical Center Dallas. The high risk obstetric service, cardiac anesthesia, specialists in pulmonary hypertension, and geneticists are also vital to the care of these most challenging patients here at UTSW.

Introduction

Adult patients with congenital heart disease are an expanding population and now outnumber pediatric patients with congenital heart disease. Remarkable improvement in the survival of patients with congenital heart has occurred over the past 70 years due to advances in cardiac surgery and cardiology. It is now estimated that approximately 85% of patients survive into adult life. [1] It is estimated that there are over 1,000,000 adult patients with congenital heart disease in the United States today. In fact, this number is likely an underestimate given the number of defects that are completely repaired in childhood, who no longer follow up as cardiac patients. There are many challenges in the care of adult congenital patients, not exclusive to the care of their cardiac issues. There are numerous social, financial, and psychological issues and comordibities which make these patients even more complex. A team of individuals with experience and knowledge of adult congenital heart disease is essential in providing state of the art care. Regional centers of excellence are best suited to care for these most challenging patients.

Patent Ductus Arteriosus

Understanding of the fetal circulation dates back to the second century when Galen described the fetal connections between the aorta and pulmonary artery and the left and right atria.[2] A patent ductus arteriosus (PDA) is the persistent opening of the connection between the aorta and the pulmonary artery beyond its expected time of closure during the first few days of life.(Figure 1) PDAs are quite common, comprising approximately 8% of all congenital cardiac defects.

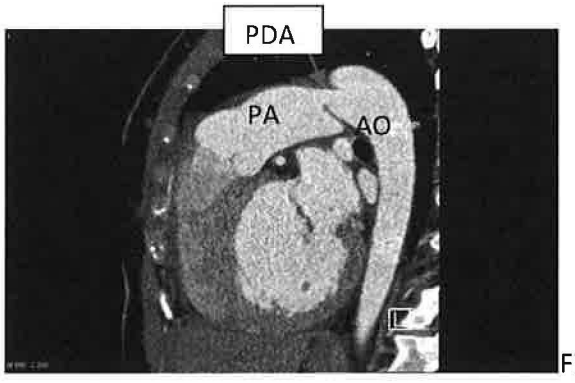


Figure 1 Patent Ductus Arteriosus.

From http://www.radrounds.com/photo/patent-ductus-arteriosus-pda PA=pulmonary artery, AO=aorta, PDA=patent ductus arteriosus

Symptoms are related to the size of the ductus. A large non-restrictive ductus with a left-to-right shunt can cause cardiac failure and pulmonary hypertension, while a small restrictive PDA is associated with an

increased risk of infective endarteritis but not a risk of cardiac failure. Because of the risk of heart failure or endarteritis, it is recommended that PDAs that persist after the age of 2 years be closed with surgical ligation or transcatheter techniques. Historical records show that as far back as 1888, the surgeon John Munro performed ligation of a patent ductus in an infant cadaver in Boston.[2] Dr. Robert Gross performed the first successful surgical closure of a patent ductus in an asymptomatic seven year old girl at Children's Hospital Boston on August 26, 1938.[3] This procedure was the first successful congenital heart operation. Surgical techniques for PDA ligation evolved over the years with excellent results. In 1976, Portsmann and Wierny reported the first percutaneous closure of a patent ductus using an Ivalon (foam) plug.[4] The disadvantage of this first device was the large delivery system, and it could only be used in large children and adults. Technology advanced with Rashkind and colleagues reporting of the first successful deployment of a percutaneously delivered double umbrella disk device in an infant weighing 3.5kg in 1979.(Figure 2) [5]

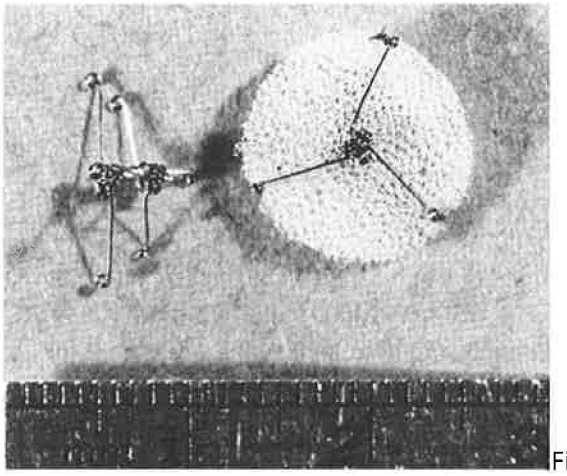


Figure 2. Rashkind double umbrella PDA occluder. On the left, device skeleton in profile. On the right, device with foam disks attached.[5]

Today, most PDAs are closed in the catetherization lab using embolization coils or the Amplatzer Duct Occluder (AGA Medical, Golden Valley, MN) with very high rates of success and very low morbidity and mortality.(Figures 3,4) Infants weighing more than 5kg can generally have percutaneous closure. Extremely large defects continue to require surgical intervention. The major risks of the percutaneous method include incomplete closure of the ductus, hemolysis, device embolization, stenosis of the left pulmonary artery, and endocarditis. With the latest technology, these complications are rare. The total mortality reported to the National Institute for Clinical Excellence in 2007 was 1 out of 2317 or 0.04%.[6] The newest technology permits closure of PDAs up to 12mm in diameter with residual shunting seen in less than 5% of cases at one year follow up. Persistence of the patent duct is often seen in premature infants. Indomethacin can be used but surgery is frequently required in these tiny infants and can now be performed safely in the neonatal intensive care unit through a small muscle sparing left lateral thoracotomy. Surgical closure can also be performed with video assisted thoracoscopic surgery, which was introduced in 1991 by Laborde and colleagues.[6]

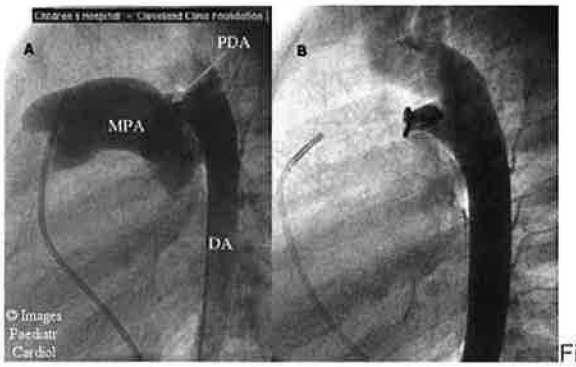


Figure 3. A. Aortogram demonstrating the patent duct. MPA=main pulmonary artery, DA=descending thoracic aorta, and PDA=patent ductus arteriosus B. Aortogram after placement of **coils** in PDA, no residual shunting seen. From www.med.yale.edu/.../coil_placement.jpg

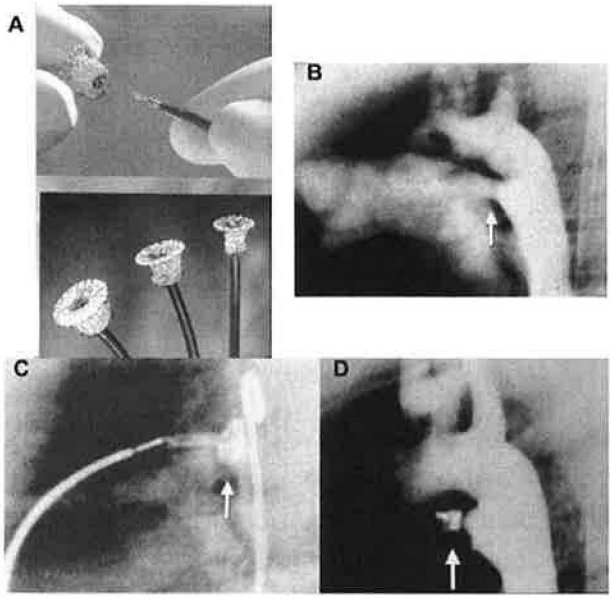


Fig. 3 - Example of arterial duct occlusion using the Amplatzer occlusion system. A) Shows the device and its release cable. B) Aortogram showing an extensive arterial duct (arrow). C) Amplatzer prosthesis being manipulated by the release cable to adjust its position (arrow). D) Aortogram showing total occlusion of the duct after release of the device (arrow).

Figure 4. PDA closure with the Amplatzer Duct Occluder

From http://www.scielo.br/img/revistas/rbccv/v18n4/4a09f3.gif

Surgery for Cyanotic Heart Disease

In the early 1900s, Maude Abbott did pioneering work in the field of congenital cardiac pathology, laying the ground work for the great clinical work done by Dr. Helen Taussig. In the 1930s, Dr. Taussig began to diagnose congenital heart disease in living patients at Johns Hopkins University. Using simple techniques including a physical examination, a chest roentgenogram, and an electrocardiogram, she was able to identify specific cardiac defects. Dr. Taussig believed that her cyanotic patients who suffered from too little pulmonary blood flow would benefit from surgical creation of a ductus arteriosus. She hoped that Dr. Robert Gross, who had performed the first successful congenital heart operation by closing a patent duct, would be able to create an artificial ductus in a patient. Legend has it that Dr.

Gross told her he was not interested in creating ductuses, only in closing them. She then approached Dr. Alfred Blalock who had been attempting to create an animal model of pulmonary hypertension. In 1944, Dr. Blalock, with the help of his lab assistant Vivien Thomas, operated on a one month old cyanotic child and performed the first subclavian artery to pulmonary artery shunt operation, known as the Blalock-Taussig shunt.(Figure 4).[7] The child had a stormy postoperative course but did survive.

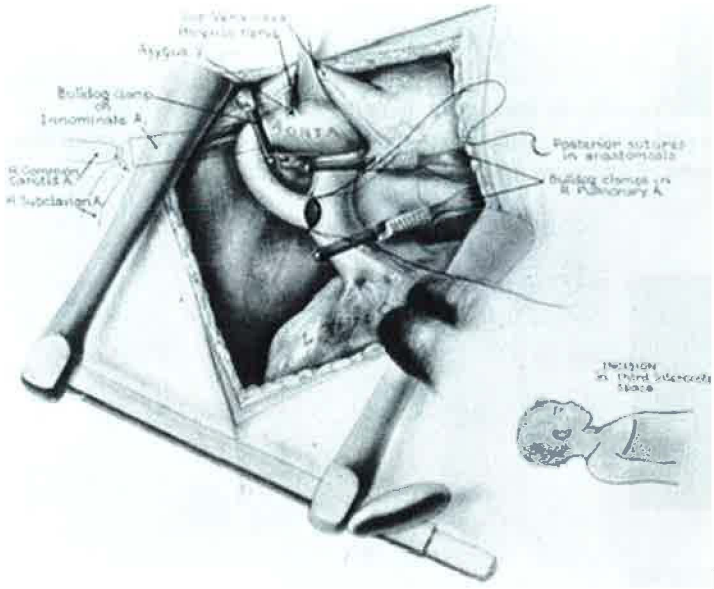


Figure 4. Classic Blalock-Taussig shunt. [7]

The subclavian artery turned out to be a very good size vessel to use to improve cyanosis. The subclavian artery was able to augment pulmonary blood flow and decrease the degree of cyanosis without causing excessive pulmonary blood flow or causing pulmonary hypertension. Modified versions of the Blalock-Taussig (BT) shunt are done today and use a Gore-tex tube graft (usually 3-5mm in diameter) to shunt blood from the innominate or subclavian artery to one of the branch pulmonary arteries.(Figure 5)

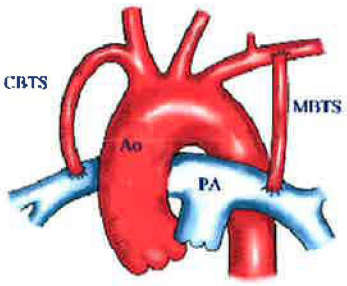


Figure 5. CBTS=classic Blalock-Taussig shunt MBTS=modified Blalock-Taussig shunt. From http://www.crkirk.com/chdguide/surgery/images/bts.gif

Other shunt operations were introduced; however the modified BT shunt is the one most commonly used today. In 1946, Dr. Willis Potts of Chicago performed a shunt operation by creating a direct connection from the descending thoracic aorta to the left pulmonary artery.[8](Figure 6a)

In 1962, Dr. David Waterston of London, created another type of systemic to pulmonary artery shunt by making a connection between the descending thoracic aorta and the left pulmonary artery.[9](Figure 6b) Both the Potts and Waterston shunts proved unsatisfactory because they frequently caused excessive pulmonary blood flow and led to severe pulmonary hypertension. In 1966, Dr. Denton Cooley in Houston devised a central shunt from the ascending aorta to the pulmonary artery which is also used on occasion today.(Figure 6c)

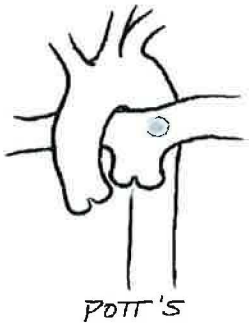


Fig 6a http://www.ctsnet.org/graphic/Pall5.jpg

Fig 6b http://www.ctsnet.org/graphic/Pall4.jpg

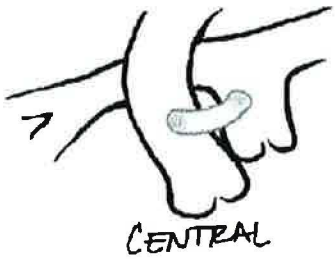


Fig 6c http://www.ctsnet.org/graphic/Pall3.jpg

Another type of shunt operation was devised by William Glenn of Yale University to increase pulmonary blood flow in patients with a variety of congenital lesions. In 1958, Glenn performed a superior vena cava-to-right pulmonary artery anastomosis, called a Glenn shunt. (Figure 7) [10]

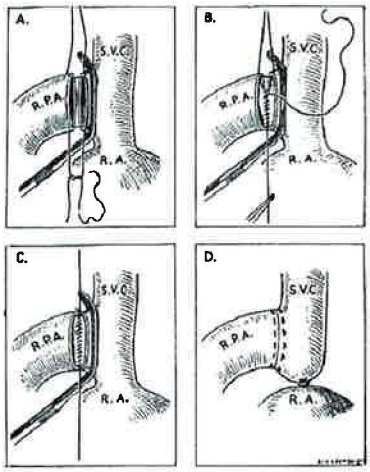


Figure 7. Classic Glenn shunt. R.P.A=right pulmonary artery, S.V.C.=superior vena cava. Illustration shows anastomosis of the SVC to the distal right pulmonary artery. [8]

Today, a modified form of this procedure, called a bidirectional Glenn, is commonly used as one of the operations used in performing a complete right heart bypass in patients with single ventricle physiology. The bidirectional Glenn allows for the superior vena cava blood to flow into both pulmonary arteries, maintaining the confluence of the pulmonary arteries. (Figure 8)

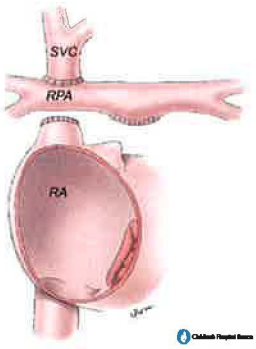


Figure 8 Bidirectional Glenn shunt

http://www.childrenshospital.org/cfapps/mml/viewBLOB.cfm?MEDIA_ID=1836

Coarctation of the Aorta

Coarctation of the aorta is a narrowing or constricted segment of the thoracic aorta near the ligamentum arteriosum which may involve a discrete narrowing (or short segment) of the aorta (figure 9), a long segment, or be may be more complex and involve hypoplasia of the ascending aorta and aortic arch (figure 10).

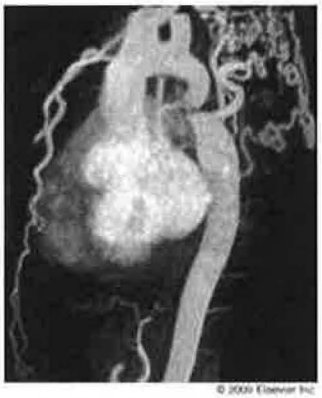


Figure 9 Sagittal oblique gadolinium enhanced maximum intensity projection MR image. There is a focal narrowing of the descending aorta in the region of the ligamentum arteriosum (juxtaductal) with post-stenotic aortic dilatation. There are multiple collateral vessels into the descending aorta via internal mammary and intercostal arteries. From http://download.imaging.consult.com/ic/images/S1933033207745421/gr2-midi.jpg

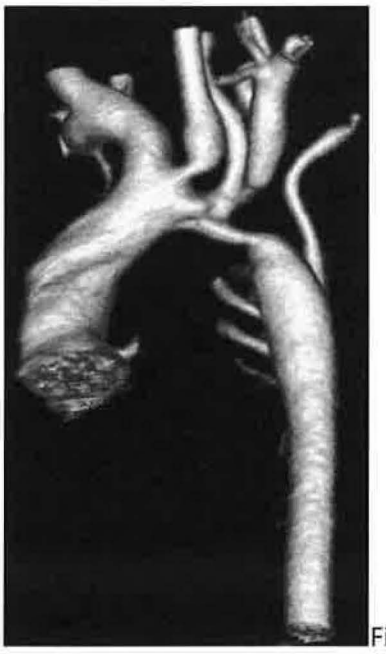


Figure 10 64 slice CT angio showing complex coarctation with arch hypoplasia and severe proximal stenosis of the left carotid artery.

Coarctation of the aorta is one of the more common congenital anomalies, comprising 5-8% of congenital defects. Coarctation is commonly associated with other congenital lesion including bicuspid aortic valve, subaortic stenosis, mitral valve anomalies such as parachute mitral valve, ventricular septal defect, and cerebral artery aneurysms. In 1944, Clarence Crafoord of Stockholm, Sweden carried out the first successful repair of coarctation in an 11 year old boy. [11] The operation took six hours, and the aorta was cross clamped for 2 hours. The narrowed segment was excised and the aorta was reapproximated. Over the years, various surgical techniques have been developed to treat coarctation of the aorta, including a subclavian flap repair (which sacrifices the left subclavian artery to repair the coarctation, [12](fig 11a), Dacron patch augmentation of the coarct with patch aortoplasty (figure 11b), and extensive resection of the coarctation with end to end anastomosis of the aorta (figure 11c). Surgery is often successful but recurrent coarctation is not uncommon, with recurrent coarctation is defined by catheterization gradient >20mmHg.

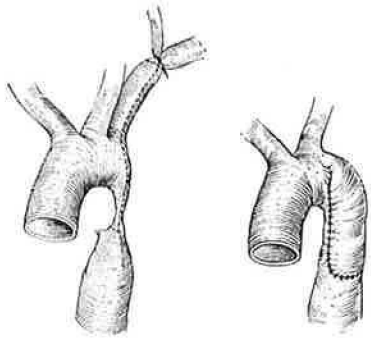


Figure 11a. Subclavian flap repair for coarctation of the aorta.[12]

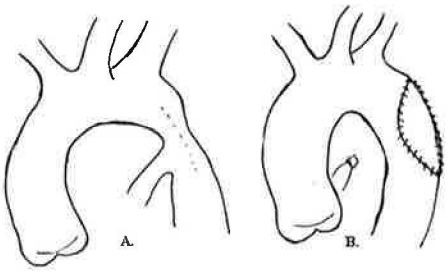


 Figure 11b. Patch aortoplasty repair for coarctation. From http://www.health.gov.mt/impaedcard/issue/issue19/Omejei/Fig08.jpg

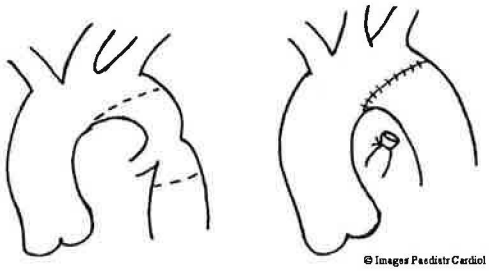


Figure 11c. Extended resection of the coarctation to the under surface of the aorta with end to end anastomosis. From http://www.health.gov.mt/impaedcard/issue/issue19/Omejei/Fig08.jpg

Patching of the coarcted segment with a Dacron patch has been associated with aneurysm formation at the site of repair. Lifelong monitoring of the patient is recommended due to the risk of recoarctation, aneurysm formation, dissection, and the development of hypertension. Hypertension, particularly exercise induced hypertension, may develop years after repair and may be seen in the presence or absence of recurrent coarctation. An associated intrinsic abnormality of the wall of the aorta predisposes patients with coarctation to dissection or rupture. Catheterization techniques are commonly used today in the treatment of coarctation. Singer first reported use of balloon dilation of coarctation in 1982, and Lock et al reported the first case from Children's Hospital Boston in 1983 with nine balloon dilations in eight patients ranging from 7 days of life to 22 yrs of age and mixed results. [13] Treatment for primary unoperated coarctation remains controversial today. Balloon angioplasty with or without stenting is often performed today in both older children and adults for both treatment of both primary coarctation and recurrent coarctation. In 1991, Perry and Locke at Children's Hospital Boston reported successful stenting of coarctation .[14] Surgical repair is necessary for optimal treatment of neonatal, infant, and early childhood coarctation. For many adult patients found to have coarctation, primary stenting is an excellent option unless the coarctation is calcified, tortuous, or involves a very long segment of the thoracic aorta. In adults who require surgery, the aorta is not as elastic as in a child so extended resection with end to end anastomosis may not be possible. In that case, resection of the coarctation often requires placement of an interposition graft. For extremely tortuous and calcified coarctations, a bypass graft can be placed from the ascending to descending thoracic aorta, without need for resection of the coarcted segment.(Figure 12)

Fig. 2 - Postoperative chest computed tomography with three-dimensional reconstruction, showing patent ascending aorta to descending aorta extra-anatomic bypass (§), the previous obstructed graft (*) and involution of all aneurysms

Figure 12. Extra-anatomic ascending to descending aorta bypass http://www.scielo.br/img/revistas/rbccv/v23n4/en_a20fig02.jpg

Recurrent coarctation is most often successfully managed in the catheterization laboratory with balloon dilation with or without stenting and avoids the need for surgery. The morbidity for operation of primary or recurrent coarctation in adults can be considerable and includes bleeding, pleural effusion, lung contusion, recurrent laryngeal nerve palsy, phrenic nerve injury, hypertension, and paraplegia secondary to spinal cord ischemia (more common in patients with poor collateral circulation)

It is not uncommon for coarctation to present in adult life as a cause of secondary hypertension or in association with other congenital heart lesions, such as bicuspid aortic valve disease. It is essential to fully examine pulses, looking for a delay between the radial and femoral pulses or diminished or absent pedal pulses. The murmur of coarctation is typically a systolic murmur heard posteriorly between the scapulae. Documentation of blood pressure readings in all four extremities is helpfu. A complete transthroacic echocardiogram should make the diagnosis of coarctation; however the descending thoracic aorta is not well visualized by transthoracic echocardiography in adults. In addition, a gradient in the descending thoracic aorta may not be present if most of the flow to the lower body is via collaterals.

Closed Techniques in Cardiac Surgery

Shortly after Blalock and Taussig's landmark contribution to the field of cardiac surgery, some thought that patients with pulmonic stenosis might benefit from a direct surgical intervention on the pulmonary valve. T. Homes Sellors in Lodon, performed the first successful pulmonary valvotomy in December 1946 on a patient with tetralogy of Fallot and bilateral pulmonary tuberculosis, felt too sick to undergo a Blalock-Taussig shunt.[8] Sellors passed a long tenotomy knife directly through the right ventricle and

incised the stenotic pulmonary valve. In 1950, Russell Brock treated a patient with pulmonary valve stenosis with a similar technique. He directly accessed the right ventricle (without use of cardiopulmonary bypass) and used a valvulotome to incise the pulmonary valve. The Brock procedure became the standard surgical approach for pulmonic stenosis until the development of open heart surgery. (Figure 13)[8] Today, pulmonary valvotomy is almost exclusively performed in the catheterization laboratory with excellent short and long term results unless the valve is dysplastic.

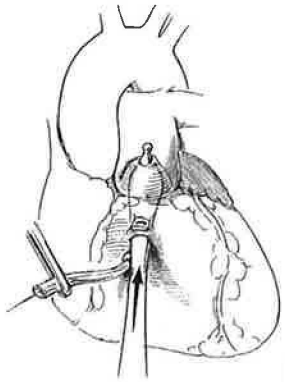


Figure 13 Brock procedure shown with insertion of a valvlotome through the right ventricle to incise the stenotic pulmonary valve.[8]

Other closed heart operations performed in the closed heart era were critical in the treatment of congenital heart disease. In 1948, Blalock and Hanlon reported a complex technique for excising the atrial septum in patients who needed to have mixing of systemic and pulmonary venous blood in order to survive (as in transposition of the great vessels).[8] The Blalock-Hanlon procedure was used until the percutaneous balloon atrial septostomy (BAS) procedure became commonplace. Balloon atrial septostomy was performed in the catheterization laboratory by Rashkind and Miller in 1966.[15]

In 1952, Dr. Gross and colleagues were the first to report successful first closure of an atrial septal defect using the atrial well technique. A rubber funnel was sutured into the atrial wall, and the ASD was closed by feel through the blood that filled the funnel (meaning there was no direct visualization of the defect).(Figure 13) [8]

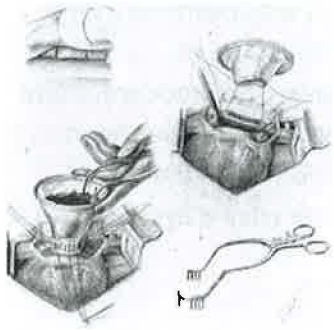


Figure 13 Gross' atrial well technique for closure of an ASD[8]

In 1952, Muller and Dammann attempted to limit the amount of shunting in a patient with a ventricular septal defect (VSD) by placing a band on the pulmonary artery.(Figure 14)

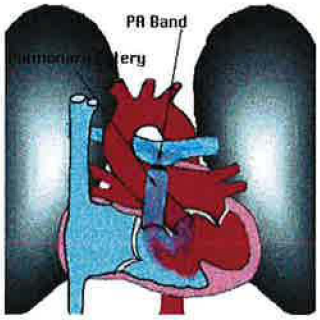


Figure 14 Pulmonary artery band procedure for patient with a large VSD and heart failure shown. http://www.rnceus.com/vsd/band.gif

The pulmonary artery band procedure became an important way to treat heart failure in VSD patients. Even after open heart techniques became possible, PA banding was still performed. In the early days of open heart surgery, mortality rates for repairs in infants exceeded 75% and took many years to decrease. Today, surgical repair of large VSDs can be performed safely in infancy; however, pulmonary artery banding continues to be used in certain cases. Device closure of muscular ventricular septal defects can be performed in the catheterization laboratory provided there is an adequate rim of muscular septum to anchor the device and the device does not impinge on tricuspid valve function. (Figure 15) Device closure is not possible for other types of VSDs (perimembranous, inlet, or supracristal ventricular septal defects).

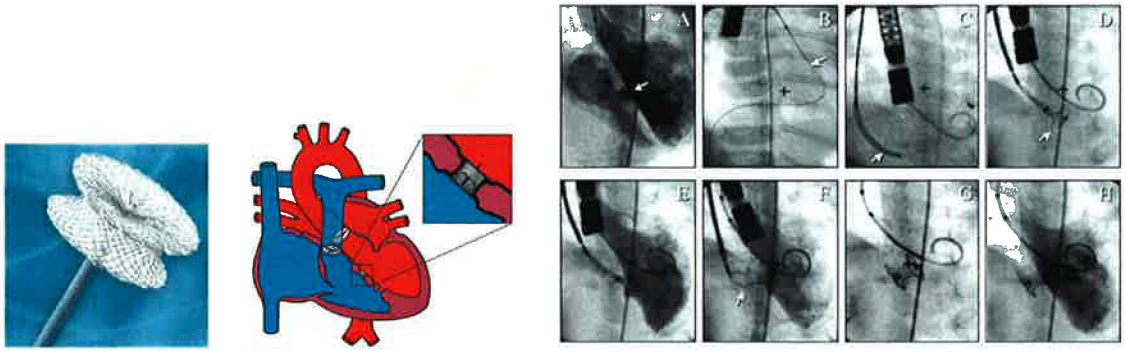


Figure 15. left, Amplatzer Muscular VSD Occluder; center, heart diagram showing final positioning of the device; right Fluoroscopic and angiographic steps in the closure of a muscular VSD. **A**. LV angiogram demonstrates the presence of a single mid-muscular VSD; **B**. The wire is across the VSD from the left ventricle; **C.** The delivery sheath is in the left ventricle from the right internal jugular and the device is being advanced; **D**. The left sided disc is advanced into the left ventricle. **E**. LV angiogram to confirm the position of the left ventricular disc; **F**. The right disc is deployed in the right ventricle; **G**. The device is released from the delivery cable; **H**. LV angiogram shows complete closure of the muscular VSD with the Amplatzer muscular VSD device.

Left: from http://www.nature.com/nrcardio/journal/v2/n11/thumbs/ncpcardio0351-f3.jpgCenter: from http://www.amplatzer.com/portals/aga/images/uploaded_images/vsd/fig3_placement.gif
right: from http://www.fac.org.ar/ccvc/llave/c007/fig03.jpg

Open Heart Surgery

In the mid 1900s, it became clear to surgeons that a bloodless field would be required to repair intracardiac defects. The first successful open heart operation was performed by John Lewis at the University of Minnesota in 1951 with closure of an ASD using hypothermia and systemic venous inflow occlusion.[16] Pioneering work at Jefferson Medical College in Philadelphia by Dr. John Gibbon and his lab assistant and wife Mary Hopkinson led to the development of a heart lung machine.(Figure 16)

Figure 16 John Gibbon and a prototype heart lung machine, 1953http://www.nature.com/nm/journal/v9/n10/images/nm937-F1.jpg

Between 1950 and 1955, 5 centers were actively developing heart lung machines. Companies like General Motors and IBM were instrumental in support of this research. However, early surgeries done with cardiopulmonary bypass had exceedingly high rates of mortality. Attempts at open heart surgery were made on the sickest patients, and major complications including death were frequent. Clotting of the circuits and introduction of air bubbles into the circuit led to systemic embolization, and destruction of blood components led to major and often fatal postoperative bleeding. The pumps required large priming volumes, failsafe roller pumps , and a means of oxygenating the blood safely and effectively. After devoting almost three decades on the development of a heart lung machine, Dr Gibbon performed the first successful open heart procedure on May 6, 1953, using cardiopulmonary bypass with a screen oxygenator to close an atrial septal defect in an 18 year old girl with right heart failure.[16] 5 other patients in his series died, and he abandoned the procedure.[16] Simultaneously, other centers including the Mayo Clinic, the University of Minnesota, University of Toronto, and Wayne State were actively engaged in the development of a heart-lung machine.

In the 1950s, the technique of cross circulation was used at the University of Minnesota in which a human donor served as the heart lung machine. (Figure 17)[17]

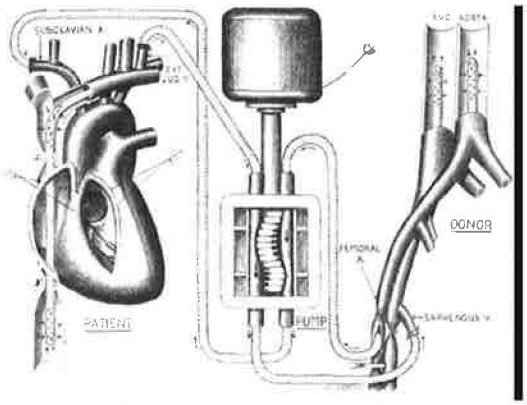


Figure 17 Diagram of cross circulation circuit used by Lillehei. [17]

Between March 1954 and July 1955, C. Walton Lillehei at the University of Minnesota used cross circulation to repair a variety of intracardiac defects on 45 infants and children, ranging from 4 months to 14 years of age.[18] 28 (62%) survived surgery and were discharged. Of those, 8 early survivors died, 20 were reported alive in 2009.[17] These results were astonishing. However, cross circulation was limited since it could only provide adequate oxygenation and flow rates for infants and small children and risked 200% mortality, ie death of the donor and the patient with congenital heart disease . Thanks to an influx of funding for medical research in the in the three decades after World War II, great advances in production of the heart lung machine, surgical instruments and surgical techniques were made. In May 1955, Lillehei and others switched to use of the DeWall bubble oxygenator.[16] By the mid 1960s, many companies were making affordable heart lung machines and use of cardiopulmonary bypass became widespread. (As a side noted, Rene Favoloro performed the first coronary artery bypass

in 1967 at the Cleveland Clinic.[19]) With improvements in surgical technique and cardiopulmonary bypass, neonatal heart surgery became a real possibility in the 1970s. In 1969 in Auckland, New Zealand, Sir Brian Barratt-Boyes pioneered the technique of profound hypothermia and circulatory arrest to repair complex defects in neonates. Barrat-Boyes in New Zealand and Aldo Castaneda in Boston focused their efforts on the correction of complex congenital lesions in infants and newborns. Today, complex congenital heart repairs can be done safely and successfully in the first few days of life. For example, some babies with tetralogy of Fallot and inadequate pulmonary blood flow can undergo surgical repair in the first several days of life, avoiding the need for a palliative systemic to pulmonary artery shunt procedures.

Pacemaker

Lillehei and colleagues noted that early surgical repair of intracardiac defects was complicated by transient or persistent complete heart block in approximately 10% of cases, constituting an important source of major morbidity and mortality for patients undergoing open heart surgery for repair of congenital heart defects. In December 1958, Lillehei and colleagues reported the first use of a percutaneously inserted myocardial electrode at the University of Minnesota.[20] Pacemaker technology has evolved with implantation of permanent transvenous leads and subcutaneous or subpectoral generator implantation. However, surgical implantation of leads and generator is still necessary for some of the more complex single ventricle patients, in order to avoid placing leads in the systemic ventricle .

Single Ventricle Physiology

Single ventricle patients have mixing of the pulmonary and systemic circulation and currently undergo a series of operations to separate the two circulations known as the Fontan procedure. A variety of congenital defects have this mixed type of circulation and may have a single right ventricle, left ventricle, or hypoplasia of one of the ventricles to such a degree that it is not capable of adequate pump function for either the systemic of pulmonary circulation. Infants with this type of circulation may be critically ill at the time of birth due to severe cyanosis (lack of adequate pulmonary blood flow) or from heart failure due to excessive pulmonary blood flow. It is also possible for an infant to be well compensated with adequate balance of the two circulations without the need for urgent intervention. Before it was discovered that prostaglandin E1 could maintain patency of arterial duct, many cyanotic infants would die within the first few days or weeks of life without surgical intervention to increase or decrease the degree of pulmonary blood flow. In 1977, Barratt-Boyes reported that a continuous infusion of prostaglandin E1 could maintain patency of arterial duct in a newborn infant and maintain a

type of fetal circulation until a surgical or catheter based intervention could be performed. Today, many infants can be stabilized on prostaglandins and then transported safely to a specialized congenital heart center. For infants with single ventricle physiology who require an increase in pulmonary blood flow in the newborn period, a modified Blalock-Taussig shunt can be performed. For infants that are stable over the first few months of life, the first operation in the Fontan pathway is the bidirectional Glenn shunt which diverts flow from the superior vena cava to the branch pulmonary arteries. [10] This procedure cannot be performed in the newborn period due to the high pulmonary vascular resistance seen in the neonatal period. For complete separation of the pulmonary and systemic circulation, a second or third operation is required to divert the inferior vena cava blood directly to the pulmonary circulation. This complete right heart bypass procedure was first reported by Fontan and Baudet in 1971 for palliation of tricuspid valve atresia, where there is an atretic tricuspid valve and a diminutive (inadequate) right ventricle. [21] The original Fontan procedure included the placement of one valve at the junction of the inferior vena cava and the right atrium and a second valve between the roof the right atrium and the left pulmonary artery.(Figure 18a) It turned out that right atrial function contributed little to this type of circulation, and it was noted that the right atrium became severely dilated. Modifications of the initial procedure were made to optimize the hydrodynamic flow from the vena cavae to the pulmonary arteries. The modern Fontan is typically a total cavopulmonary connection (TCPC) [22] using a Goretex tube (or lateral tunnel) to divert IVC blood directly to the pulmonary arteries (figure 18b) or an extracardiac conduit Fontan which uses a tube graft outside the heart to connect the vena cavae to the pulmonary arteries. (Figure 18c) For optimal hemodynamics this type of circulation requires having good systolic and diastolic ventricular function, little atrioventricular valve regurgitation, and low pulmonary vascular resistance. In the Fontan, a single ventricle pumps blood through the entire body, and the pulmonary circulation fills passively.

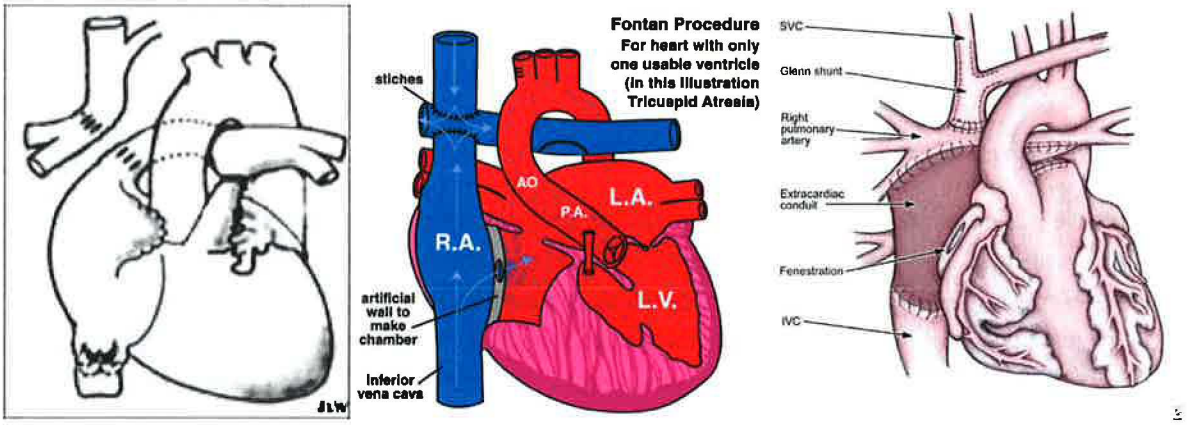


Fig 18a, Original Fontan[21] 18b, Total Cavopulmonary Connection 18c, Extracardiac Fontan

Patients undergoing the early types of Fontan procedure experienced significant postoperative morbidity and were often hospitalized for a number of weeks with persistent pleural effusions and ascites due to the increased atrial pressure. Postoperative morbidity has decreased in recent years with

fenestration of the Fontan (shown figure 18b), where a hole is punched in the conduit which allows for right-to-left shunting and minimizes the accumulation of pleural and ascetic fluid. After adaptation to the new circulation, the fenestration can be closed percutaneously in the catheterization laboratory in many cases. Current mortatlity rates have decreased to less than 5% with better patient selection, staging of repair, and modifications of surgical technique. Cardiopulmonary exercise capacity is subnormal, approximately 70% of predicted primarily because they do not have the ability to augment cardiac output with this type of circulation. Unfortunately, we have many adult Fontan patients who have undergone the older types of Fontan surgery and are now failing Fontans. Poor ventricular function, atrioventricular valve regurgitation, and chronic elevations of systemic venous pressure lead to a host of complications including, hepatic congestion and cardiac cirrhosis, atrial arrhythmias, and a poorly understood debilitating entity called Protein Losing Enteropathy(PLE). PLE is thought to be related to elevated portal venous pressure, and unfortunately treatment options are limited. Stenting of the Fontan baffle may be helpful. Systemic corticosteroids have been used with some success. Cardiac transplantation is an option, albeit at higher risk than noncongenital heart transplant patients. However, results with modern Fontan surgery are mixed and relate to the patient's underlying anatomy. Arrhythmia and heart failure continue to be a major sources of morbidity and mortality for these patients.

Transposition of the Great Vessels

Transposition of the Great Arteries (TGA) is a congenital defect where the aorta and the pulmonary artery arise from the wrong ventricle resulting in deoxygenated to blood to circulate in the body and oxygenated blood to circulate in the lungs.(Figure 19)

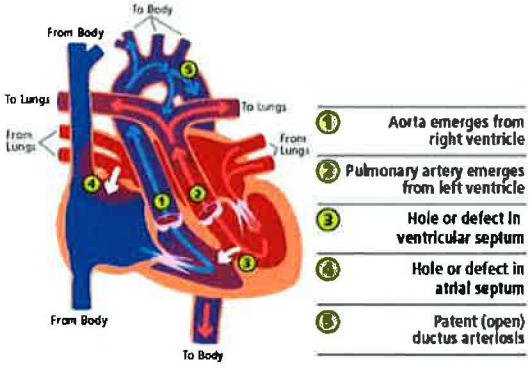


Figure 19, Diagram of Transposition of the Great Arteries http://www.eurekalert.org/multimedia/pub/web/6222_web.jpg

In the absence of an interatrial or interventricular communication to allow for mixing of the oxygenated and unoxygenated blood, survival is not possible. Additional congenital cardiac lesions are common and include ventricular septal defect (in up to 45% of patients), left ventricular outflow tract obstruction (in up to 25% of cases), coarctation of the aorta (in up to 5%), and coronary artery anomalies.[23] In 1950 (in the closed heart surgery era, prior to the development of the heart lung machine), Blalock and

Hanlon reported the first atrial septectomy procedure to allow mixing of the pulmonary and systemic circulations in patient with TGA.[24] The first attempts to switch the aorta and the pulmonary artery to completely correct the defect failed. Coronary artery transfer was required and proved to be too difficult with techniques available in the 1950s. An atrial repair was proposed as an alternative. In an atrial repair or atrial switch operation, the systemic venous flow (or vena cava flow) is baffled over to the left ventricle, and the pulmonary venous flow is baffled over to the right ventricle.(Figure 20) Ike Senning performed the first successful atrial switch operation in 1958 in London using an ingenious technique of using flaps of atrial tissue to create the baffle.[25] In 1964, William Mustard at Toronto Hospital for Sick Children performed a second type of atrial repair using pericardial tissue to create the baffle.[26] The atrial switch operation was typically performed at 6-12 months of age after a Blalock-Hanlon procedure.

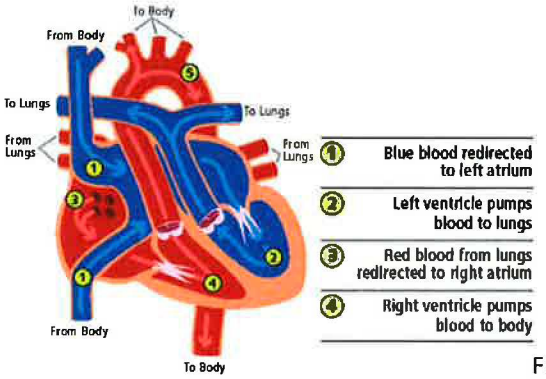


Figure 20 Diagram of the atrial switch operation
http://faculty.washington.edu/alexbert/MEDEX/Spring/PedCardiac/tga-mustard.gif

Although the atrial switch operation could be done with low mortality, the long term viability of the systemic right ventricle was uncertain and complications were reported. Baffle leaks and baffle obstruction are not uncommon. In addition, over time sinus node dysfunction, atrial flutter, and diminution of right ventricular function, and heart failure are common. Unfortunately, sudden cardiac death is a common cause of late death following the atrial switch operation, possibly triggered by supraventricular arrhythmia.[27] For ture anatomic correction of the defect, switching the great vessels and moving the coronary arteries would be required. The first attempts at switching the great vessels in the mid 1950s had failed, as transfer of the coronary arteries proved to be too difficult at that time. The first successful arterial switch operation was performed in 1975 by Dr. Jatene in Sao Paolo, Brazil. Jatene operated on a 40 day old child with transposition of the great arteries and a ventricular septal defect.(Figure 21) [28] In 1982, he reported his series of 116 operations on patients ranging from 1 month to 7 yrs of age where 58 (20.7%) died immediately postoperatively, and there were 5 late deaths.

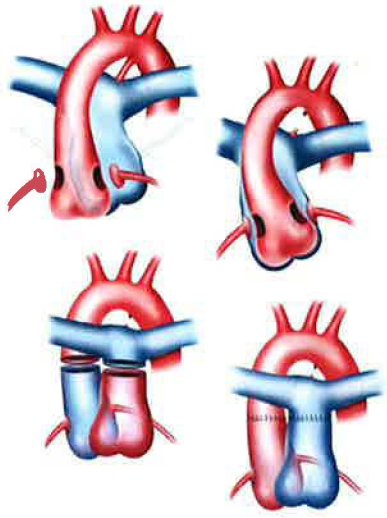
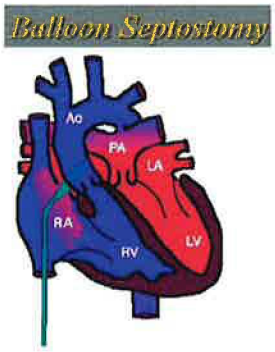


Figure 21. Diagram of the Arterial Switch Operation or Jatene Repair. Top left, harvest of the coronary button; top right, ligation and division of the arterial duct; bottom left, transection of the aorta and pulmonary artery with transfer of the coronary arteries to the neoaorta and bringing the pulmonary artery anterior to make the new pulmonary trunk.

http://radiology.rsna.org/content/247/3/617/F13.large.jpg

Since the Mustard and Senning atrial switch operations were so successful for patients with transposition and intact ventricular septum (with mortality rates less than 5%), the more difficult Jatene arterial switch operation (ASO) was not routinely used until the late 1980s. With improvements in surgical technique and the experience of Barratt-Boyes in New Zealand Castaneda in Boston, surgical mortality rates for the arterial switch operation decreased dramatically. In experience centers today, the mortality rate for repair of simple transposition is <1%.

Today, prostaglandins are initiated at the time of diagnosis and then a balloon atrial septostomy (figure 22) is performed in the catheterization laboratory rather than a Blalock-Hanlon surgical procedure.



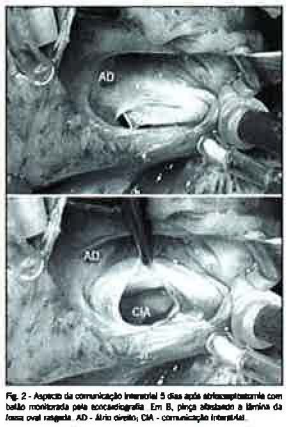


Figure 22 Diagram of Balloon Septostomy

Inspection of balloon septostomy 5 days after procedure

http://home.cc.umanitoba.ca/~soninr/Heart%20Diagrams/TGA/TGA1.JPG http://www.scielo.br/img/revistas/abc/v84n3/a10fig02.gif

With adequate mixing and resuscitation, an infant can be stabilized and have surgical correction of the defect within the first few days to weeks of life. Two major long term concerns following the arterial switch have been the fate of the pulmonary valve in the aortic (high pressure) position and the status of the coronaries after transfer. Serial surveillance of the neoarterial trunks and coronaries is necessary for these patients. Most of the patients who have undergone the Jatene repair remain in the care of pediatric cardiologists but are expected to transition to the care of the adult cardiologists in the coming years.

Atrial Septal Defects

As previously mentioned, atrial septal defects (ASDs) were initially closed in the era of closed heart surgery using the atrial well technique or systemic inflow occlusion and hypothermia. Today, many ASDs can be closed in the cardiac catheterization laboratory. The most common ASD is the secundum type which involves a defect in the midatrial septum where there is a rim of atrial septal tissue which can be used to anchor a closure device.(Figure 23) The less common types of ASDs including the sinus venosus, primum, and coronary sinus defects require surgical closure since these defects do not have an adequate rim of septal tissue for seating the device and placement of a device would impinge upon a valve or venous return to the heart flow.(Figure 23)

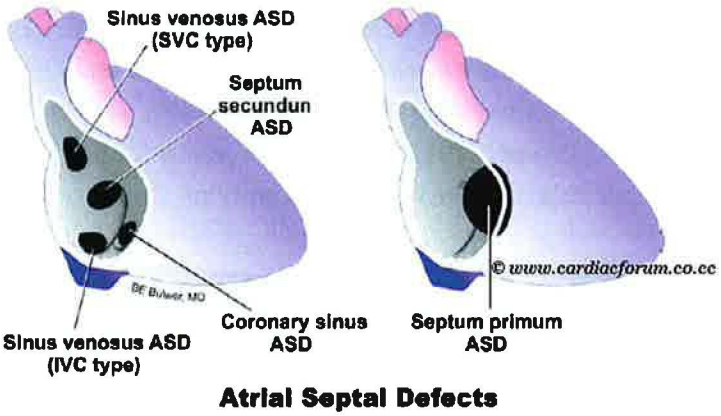


Figure 23

http://www.echoblog.co.cc/wp-content/uploads/2008/10/asd1.jpg

The first percutaneous ASD closure was performed by by King and Mills in 1976.[29] Several modifications of the first device have been made. Today, the most commonly used device is the Amplatzer Atrial Septal Occluder (AGA Medical, Golden Valley, MN).(Figure 24) It is a self-centering device with 2 expandable round discs made of nitinol (an alloy of titanium and nickel) wire and polyester mesh. Device size ranges from 4-38mm which is the diameter of the waist. The device is delivered prograde across the defect with full heparinization and monitoring overnight in the hospital. Patients are treated with aspirin for a minimum of 6 months, and Clopidogrel may be added for 1-6 months to decrease risk of thromboembolic complications, at the discretion of the physician. There are rare reports of thrombosis of these devices, most often successfully treated medically.[30] Thrombosis has frequently been reported in patients with atrial fibrillation or those without full evaluation for hypercoaguable state prior to device implantation.[30] There are case reports of erosion of the aortic wall causing pericardial effusions, aortic rupture, and sudden death.[31] Erosion of the mitral and significant mitral regurgitation has also been reported.(Joseph Forbess personal communication) Follow up is recommended at 1, 6, and 12 months and then at yearly intervals. High intensity activity may be restricted for 3 months.

Figure 24, Amplatzer Setpal Occluder (AGA Medical, Golden Valley, Mn) http://www.childrenshospital.org/cfapps/mml/viewBLOB.cfm?MEDIA_ID=301

No randomized trials comparing surgical and percutaneous ASD closure have been conducted, but nonrandomized data is available. A multicenter trial with a mixed pediatric and adult population using the Amplatzer ASD Occluder reported a complete closure rate in 97.5% at 6 month follow up.[32] 80% of adult defects were able to be closed in the catheterization laboratory with lower rates seen in children.[32] Device migration was reported in in 0.11% or 37/35,000 Amplatzer implantations.[32] Higher complication rates are seen with larger defects and those with insufficient rim.

A recent review of reported complications of Amplatzer Septal Occluder by MAUDE database review and STS review documents 223 adverse events, including 17 deaths and 152 surgical rescues.[33] Based on an estimated 18,333 implants of the ASO device between July 2002 and June 30 2007, a perforation rate of 0.28% clustered in the first 6 months but reported up to 3 years. Erosion of the left atrial appendage or or the aorta were most common. The mortality rate was 0.093 % for device closure vs 0.13% for surgical closure ($p=0.649$).[33] An important finding was the significant mortality for surgical management of device complication reported at 2.6%.[33] When complications do occur, they can be serious and often require urgent surgical intervention with a significant mortality rate. Continued post marketing surveillance is essential as the field moves forward with this interventional technique.

Summary

Due to the great advances in cardiology, cardiac surgery, and critical care, patients with congenital cardiac disease are surviving in large numbers. We find ourselves caring for a rapidly expanding population of patients who are entering into adulthood. These patients provide a host of challenges for the health care system. We have a number of these patients many of whom have been lost to follow up for a number of years for a variety of reasons. Many lack insight into their disease and frankly did not know they needed to continue to be under the care of a specialist in congenital heart disease. Many have lost health insurance and do not have the means to access the health care system. Unfortunately, many of these patients present when they are critically ill or pregnant. In addition, many are under the care of physicians who do not fully understand their congenital heart disease and receive substandard care. Fortunately, we are also seeing a number of patients who have been thoughtfully and carefully transitioned from care in pediatric cardiology to an adult congenital specialist. Many are faced with residual defects, complex arrhythmias, implantable defibrillators and pacemakers, pulmonary hypertension, heart failure, and high risk pregnancies, not to mention acquired illnesses including obesity, diabetes, and hypertension. It is not uncommon for us to make a new diagnosis of congenital heart disease in adults ranging from a simple bicuspid aortic valve to large, hemodynamically significant complex defects. Unfortunately, we have a number of unrepaired patients with Eisenmenger syndrome who are chronically cyanotic with good medium term survival but poor quality of life and few options except for avoidance of phlebotomy and use of the endothelin receptor antagonists, Bosentan and Ambrisentan. Some of our patients are end stage and are reasonable candidates for orthotopic heart

transplantation, albeit at higher risk than noncongenital patients. We just beginning to see patients who have benefited from repairs in infancy and who we expect will have many fewer complications than our older adult congenital patients. In addition to the complex medical issues involved in caring for the adult congenital patient, the issues of insurability, disability, pregnancy, contraception, and psychosocial issues require a team of specialists who are not only knowledgeable but highly dedicated to providing the highest quality care for these patients.

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