Congenital Heart Disease in Adults

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"In science generally, to solve one set of problems may be to create or discover a whole new set and of no science ... is this more true than that of medicine." (1)

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Introduction:

Congenital heart disease is defined as "the presence at birth of a gross structural abnormality of the heart, great arteries or great veins that is actually or potentially of functional significance (2). While some congenital heart defects were recognized to exist by the late 1800's, there was only limited clinical interest in these defects. In William Osler's classic textbook *The Principles and Practice of Medicine*, published in 1892, he devoted only 5 pages in his 1100 page textbook to these "Congenital affectations of the heart (3)." In his words, "these have only a limited clinical interest, as in a large proportion of cases, the anomaly is not compatible with life, and in others nothing can be done to remedy the defect or even relieve the symptoms."

By the 1930's, there was a growing interest in congenital heart disease and pioneering work in cataloguing the various types of defects was performed by Maude Abbott who published her *Atlas of Congenital Cardiac Disease* in 1936 (4). Early attempts at cardiac shunt procedures were made in the 1940's, for the first time offering some treatment for a few patients (5) (6). By the 1950's, the development of the heart-lung bypass machine made the era of cardiac surgery possible which has profoundly altered the survival patterns of patients with congenital heart disease. From these origins, pediatric cardiology has became a thriving subspecialty.

The Magnitude of the Problem:

The incidence of congenital heart disease has been reported to be anywhere from 0.4-0.9% of live births (7) (8) (2). This does not include bicuspid aortic valves which occur in 1-2% of live births (9). Assuming an average prevalence of 0.8%, there are approximately 32,000 new cases of congenital heart disease per year in the United States (10) and 1.5 million new cases world-wide (11). The incidence of congenital heart disease does not appear to be changing substantially over time. What has changed substantially over the last twenty to thirty years is the survival data for these patients. A wide variety of surgical techniques including palliative, partially corrective, and corrective operations have been designed which have dramatically affected the survival as well as the quality of life for patients with congenital heart disease. Approximately 20,000 cardiac surgical procedures are performed for congenital heart disease per year in the United States (12). It is largely because of cardiac surgery that it is now estimated more than 85% of children with congenital heart disease will survive to adulthood. The population of adults with congenital heart disease is estimated to be over 500,000 and is increasing at a rate of approximately 5% per year (12). It has been conservatively estimated that there will be close to 1 million adults with congenital heart disease in the United States by the year 2000. Thus, there is a substantial population of adults with congenital heart disease in the United States who require care.

Traditionally, congenital heart disease has been in the domain of pediatricians and pediatric cardiologists. Internists and "adult" cardiologists, in contrast, traditionally have received little training in the diagnosis and management of patients with congenital heart disease as these types of patients were thought to be rarely encountered in the practice in adult internal medicine. However, largely because of the success of cardiac surgery as well

as other improvements in both the medical management and catheter interventions, the vast majority of patients with congenital heart disease can be treated and most will survive to an adult age. Thus, there is an increasing population of patients with congenital heart disease who will be seen by internal medicine physicians.

Pediatric cardiologists have often been reluctant to relinquish the care of their patients with congenital heart disease to their adult medicine colleagues, often due to concerns that so-called "adult physicians" would not have the knowledge adequate to provide for the care of these patients, especially some of the more complicated defects (13) (14). However, there is a small but growing group of adult cardiologists who are interested in maintaining a collaborative effort with pediatric cardiologists in taking care of this patient population. Thus, the subspecialty of adult congenital heart disease is a relatively new but growing area within cardiology (15) (16) (17) (18).

There are many excellent textbooks and reviews which describe the diagnosis of congenital heart disease, including the physical exam, ECG and chest-xray findings of various defects (19) (20) (21). Echocardiography plays an essential role in the diagnosis and follow-up of patients with congenital heart disease (22). Rather than focusing on the diagnostic evaluation, this review will focus on the long-term management of patients with congenital heart disease and on issues specifically related to the care of <u>adult</u> patients with congenital heart disease.

Types of congenital heart disease seen in adults:

Common examples of congenital defects that may be missed in childhood and thus diagnosed for the first time in adult life include atrial septal defects, pulmonic stenosis, bicuspid aortic valve and coarctation of the aorta (23). In some patients with unoperated congenital heart disease, the defect is relatively mild and surgical or other intervention has not been required. Alternatively, patients may have a significant structural abnormality that is amenable to surgical intervention. This category of patients often includes those patients in whom the diagnosis of congenital heart disease was not made in childhood and they are now presenting for the first time as adults. Finally, there are those patients in whom a significant defect or structural abnormality is present but who are not amenable to operation. This category includes patients in whom the cardiac defects are severe and there are no reasonable options for palliative or corrective surgery. At other times, patients will be encountered in whom operative intervention would have been reasonable in childhood but there is now severe pulmonary vascular disease which has rendered the patient inoperable (i.e. so-called Eisenmenger syndrome).

When most internists think about congenital heart disease in adults, they primarily consider the occasional patient in whom a new diagnosis of congenital heart disease is made. In reality, the majority of adults with congenital heart disease will have had some type of surgical intervention in the past. In centers with clinics devoted to congenital heart disease in adults, only 15-20% of patients have not had previous surgical intervention. The remaining 80-85% will have had at least one surgical intervention in the past (24) (16) (19). Thus, the majority of adults with congenital heart disease requiring long term care will have had some type of operative intervention in the past.

Patients who have had some sort of operative intervention for their congenital heart disease fall into two major categories. In one group, survival to adulthood was expected despite the presence of their congenital defect but their quality of life or longevity has been enhanced due to their operative intervention. In the second group of patients, their survival to adulthood is related solely or chiefly due to their surgical intervention. That is, these are patients who would not have survived their childhood without operative intervention.

Specific Cardiac Defects Seen In Adults, Survival Patterns, And Recommendations For Longterm Follow-Up:

Bicuspid aortic valve

Certain types of congenital defects are commonly associated with survival to adulthood. The most common of these is bicuspid aortic valve which is reported to occur in 1-2% of the general population (9). Bicuspid aortic valve is usually an isolated congenital defect but there are associated defects in approximately 20% of patients, the most common being coarctation of the aorta. Patients with bicuspid aortic valves may have a normal lifespan without ever developing evidence of significant valvular disease. In many cases, however, the bicuspid aortic valve becomes progressively stenotic, resulting in symptoms of aortic stenosis (25). In these cases, patients usually present in their 4th or 5th decade with symptoms of aortic stenosis. Alternatively, the bicuspid aortic valve may be regurgitant, leading to progressively worsening aortic regurgitation and ultimately to congestive heart failure (26). Patients with bicuspid aortic valves are at significantly increased risk of developing infective endocarditis, often leading to significant worsening of their aortic insufficiency (27). Patients with bicuspid aortic valves need to be instructed about the importance of prophylaxis against subacute bacterial endocarditis. In addition, while not commonly appreciated, patients with bicuspid aortic valves have structural abnormalities of their thoracic aorta (i.e. cystic medial necrosis) and are at risk for aortic dissection (28) (29) (30).

Endocarditis risk: High.

Recommendations for follow-up: Patients with bicuspid aortic valves that are functionally normal or minimally regurgitant require periodic follow-up by a cardiologist. Patient with significant stenosis or regurgitation require more intensive follow-up. Subspecialty care is usually not required.

Coarctation of the aorta

In approximately 20% of patients with coarctation of the aorta, the diagnosis is not made until adolescence or adulthood(31). It is the most common congenital cardiac defect seen in Turner's syndrome. The most common form of coarctation of the aorta seen in adults is a discrete narrowing of the aorta at the level of the ligamentum arteriosus (just distal to the origin of the left subclavian artery). Patients are hypertensive in the upper extremities but have substantially lower blood pressures and decreased pulses in their lower extremities. In addition to hypertension, patients are also at risk for developing left ventricular failure. Fifty to 85% of patients have an associated bicuspid aortic valve. Patients are also at risk for accelerated coronary artery disease. Aneurysms of the circle of Willis may also be present.

Once the diagnosis of coarctation of the aorta has been made and the presence of significant obstruction (>50mmHg) has been confirmed, correction of the coarctation is warranted. The average age at death for unoperated coarctation of the aorta is 30 and more than 75% of patients will be dead by the age of 50 if left unrepaired (32) (33). Surgical treatment options include resection of the coarcted segment with a patch angioplasty or resection with placement of an interposition tube graft (34). In general, the operative risk is somewhat higher for adults than for children and should be performed by surgeons with experience in congenital heart surgery (35) (36). Overall, operative mortality is generally low but patients are at risk for spinal cord ischemia (occurs in 1-2%) (37) (38). Balloon aortoplasty and stenting is also an option but is still considered experimental (39) (40) (41) (42). Head-to-head comparison of balloon versus surgical techniques for primary treatment of coarction have not been performed and neither technique has been proven superior at this point.

Despite correction of the coarctation, either by surgical or balloon catheter means, coarctation may recur and thus the patient must be monitored for evidence of recurrent obstruction. Aneurysm formation at the site of either aortoplasty or surgical repair is also recognized as a complication but its clinical significance is not well understood. The majority of patients who are repaired in late adolescence or adulthood will remain hypertensive after correction of their coarctation although their hypertension is usually much easier to control after relief of the coarctation.

Endocarditis risk: Unoperated coarctation = moderate risk. Post-op with low or absent gradient = low risk.

Recommendations for follow-up: All patients require follow-up by a cardiologist, preferably a subspecialist. Patients are at risk for re-coarctation, residual hypertension, aortic dissection, aortic aneurysm formation, and complications associated with an associated bicuspid aortic valve.

Atrial septal defects

Atrial septal defects are a common form of congenital heart disease and are one of the most common diagnoses among adults with congenital heart disease. While atrial septal defects are usually diagnosed and treated in childhood, the defect may be missed as the patients are usually asymptomatic and the physical exam findings can be relatively subtle(23) (43). Ostium secundum defects (in the central portion of the atrial septum) are the most common, occurring in 75% of cases. The presence of a left to right shunt results in a volume overload of the right atrium, right ventricle and pulmonary arteries which places patients at risk for developing right heart failure and atrial arrhythmias. While survival to adulthood is the rule, life expectancy is decreased. Survival beyond the age of 40 to 50 occurs in less than 50% of patients with large shunts and the majority of surviving patients over the age of 50 will be symptomatic with exertional dyspnea, fatigue and/or palpitations (23) (44) (45).

Some patients may develop pulmonary vascular obstructive disease resulting in pulmonary hypertension. In some cases, reversal of shunting across the defect occurs as pulmonary vascular resistance equals or exceeds that of the systemic vascular resistance, i.e. Eisenmenger syndrome. The development of Eisenmenger syndrome is unusual in atrial septal defects, occurring in less than 10% of patients(46). It is not clear whether the pulmonary vascular disease occurs as a result of the increased pulmonary blood flow from the intracardiac shunt or whether this in fact represents a primary pulmonary vascular disease (e.g. primary pulmonary hypertension) with an incidental atrial septal defect (19).

Because of the long-term morbidity and mortality associated with unrepaired atrial septal defects, surgical closure of the defect is recommended when the diagnosis is made unless there is evidence of severe pulmonary vascular disease. In general, earlier closure results in a better outcome. Repair in childhood is almost always associated with return of cardiac chamber sizes to normal (47). Repair in adulthood, however, is associated with persistent right atrial and right ventricular dilation in as many as 80% of patients (48) (49). Patients who have evidence of right ventricular failure preoperatively will have persistent post-operative failure as well. Data from the early surgical experience (1956 to 1960) with atrial septal defect closure at the Mayo Clinic demonstrated that closure of the defect in childhood or adolescence results in normal long term survival (50). Closure of the defect after the age of 25 also appeared to result in good survival, although decreased compared to an age-matched control population. Closure of atrial septal defects after the age of 40 was associated both with increased morbidity and mortality but survival was improved as compared with historical controls. Because of the higher risk of surgery in older patients, some authors have suggested that surgery may not be beneficial in older patients (51) (52). A more recent surgical series (surgery between 1971 and 1991) involved only adult patients (mean age of 44 years, 35% over the age of 50) and demonstrated a low operative mortality (1.3%) and good 10-year survival (92%). Finally, a study by Kostantinides, et al

addressed this issue with a retrospective comparison of surgical or medical therapy in 179 patients age 40 or older with ostium secundum atrial septal defects (53). Eighty-four patients were treated surgically and 95 were treated with medical therapy. The mean age was somewhat older in the medical therapy group (57 years versus 54) but other baseline variables were similar. A clear benefit was demonstrated for the surgical group with a 5-year survival of 98% versus 93% and 10-year survival of 95% versus 84%. In the surgical group, 32% of patients had improvement of their New York Heart Association class versus only 3% in the medical group. Conversely, worsening NYHA class was seen in 11% of surgical patients versus 34% of medically managed patients. There was no difference in the risk of arrhythmias between the two groups.

Thus, patients with atrial septal defects should undergo closure of the defect at the time of diagnosis unless the patient is elderly (predicted life-expectancy less than 10 years). A variety of devices are being developed to allow percutaneous closure of atrial septal defects and some devices will likely be commercially available in the near future. However, there are no studies comparing device closure to surgical closure and these devices should be considered experimental at present (54).

Endocarditis risk: Ostium secundum atrial septal defect, unoperated or post-op = low risk. With associated mitral valve pathology (e.g. cleft valve or mitral valve prolapse) = moderate risk.

Recommendations for follow-up: Patients with uncomplicated ostium secundum atrial septal defects repaired in childhood are unlikely to have long-term complications and do not require special follow-up. However, patients repaired as adults should have long-term follow-up by a cardiologist. In addition, patients with elevated pulmonary pressures at the time of surgery, patients with more complicated defects (e.g. ostium primum defects), and patients with atrial arrhythmias or ventricular dysfunction pre-operatively require long-term follow up.

Pulmonic stenosis

Pulmonic stenosis is another congenital defect in which survival to adulthood is expected. The degree of stenosis does not necessarily correlate well with symptoms (19). Patients with mild to moderate pulmonic stenosis are usually asymptomatic and often are not detected until adulthood. Survival appears to be normal for patients with mild to moderate obstruction (55). The degree of stenosis may progress over time so patients with moderate or severe stenosis require serial evaluation. For patients with symptomatic moderate or severe pulmonic stenosis, pulmonary valvuloplasty is now the treatment of choice (56) (57). With balloon valvuloplasty, the stenosis can be successfully relieved in nearly all cases, thus avoiding the need for operative intervention. Patients may develop some pulmonic insufficiency after the procedure although this is uncommon and is usually clinically not important. Restenosis requiring reintervention is exceedingly rare.

Endocarditis risk: Mild stenosis = low risk. Moderate to severe stenosis = moderate risk.

Recommendations for follow-up: Patients with mild stenosis (peak gradient <50mmHg) do not require specific cardiac follow-up. Patients with moderate to severe stenosis require follow-up and may require intervention.

Ventricular septal defect

Other shunt lesions such as ventricular septal defects and patent ductus arteriosus are usually detected in childhood because of the prominent murmurs associated with these defects. Ventricular septal defects are one of the most common forms of congenital heart disease and are commonly diagnosed in childhood. Spontaneous closure of ventricular septal defects is fairly common in childhood and can occur rarely in adults (19). In general, once a moderate to large ventricular septal defect is detected, surgical closure in early childhood is recommended (58). If these large defects are not repaired, patients are

at risk for developing progressive pulmonary vascular disease over time with reversal of their shunting (i.e. Eisenmenger syndrome). Patients with a large ventricular septal who are seen in adulthood nearly always have evidence of Eisenmenger physiology. Occasionally, an adult patient may present with a ventricular septal defect with a moderate-sized shunt. A careful evaluation is needed to assess for the presence and severity of pulmonary hypertension in these patients. If pulmonary vascular resistance is not prohibitively elevated (i.e. less than 2/3 systemic vascular resistance), the defect should be closed surgically. Patients with evidence of Eisenmenger physiology should be managed medically (see below). Adult patients who have a small ventricular septal defect have normal survival but are at lifelong risk of bacterial endocarditis, usually involving the tricuspid valve. All patients with ventricular septal defects, including those with surgically closed ventricular defects as well as patients with small, unoperated defects are at risk for ventricular arrhythmias (59) Approximately 5% of patients with ventricular septal defects develop aortic regurgitation

Endocarditis risk: Unoperated VSD = moderate risk. S/p spontaneous closure of VSD = low risk. Post-op without residual VSD = low risk. Post-op with residual VSD = moderate risk.

Recommendations for follow-up: Patients with a history of a VSD in childhood who have no audible murmur should be evaluated for spontaneous closure versus Eisenmenger syndrome. Patients with spontaneous closure do not require special follow-up. Patients with small defects do not require cardiology follow-up. The following problems require cardiology follow-up: residual VSD (patch leaks), pulmonary hypertension at the time of surgery (may progress post-op), concomitant aortic valve disease or other associated cardiac lesions, late repair of moderate or large defects, and patients with atrial or ventricular arrhythmias.

Patent ductus arteriosus

Like ventricular septal defects, the diagnosis of patent ductus arteriosus is usually made in childhood but occasionally may present in adulthood. All patients with a patent ductus arteriosus are at risk for endocarditis, regardless of the size of the shunt. An exception to this rule is the occasional patient with a "silent" ductus seen on echocardiography but no clinical evidence of a shunt. In this case, no intervention is required(60). Patients with a small patent ductus arteriosus usually have normal survival but closure of the ductus is still recommended to decrease the risk of endocarditis. Patients with larger left to right shunts usually begin to have symptoms of dyspnea due to the left ventricular volume overload or palpitations from atrial arrhythmias as they reach adult age. These patients should be referred for closure of the ductus to improve survival(61). Closure of a patent ductus can be done either via surgical techniques or catheter-based techniques. A variety of PDA closure devices exist and can be deployed at the time of initial catheterization in experienced centers (62) (63). Patients with a large shunt are at risk for developing severe pulmonary vascular disease with shunt reversal (e.g. Eisenmenger syndrome). Once severe pulmonary vascular disease has developed, closure of the ductus is contraindicated.

Endocarditis risk: "Silent" PDA = low risk. All other PDAs = moderate risk. Post-operative patients = low risk. S/p device closure = continue antibiotic prophylaxis for 6-months post-procedure once complete closure has been documented.

Recommendations for follow-up: Patients with small defects or patients s/p surgical closure do not require specific cardiac follow-up, although recanalization of a closed ductus arteriosus can occasionally occur. Patients with moderate to large shunts repaired in adulthood and any patient who has undergone device closure need continued cardiac follow-up. Patients with clinically "silent" PDA do not require follow-up.

Forms of cyanotic congenital heart disease seen in adults:

Among the cyanotic forms of congenital heart disease, survival to adulthood is less common but has been well reported. The most common forms of cyanotic congenital heart disease seen in an adult patient population are the Eisenmenger syndrome (due to an unrepaired shunt) and tetralogy of Fallot.

Tetralogy of Fallot

The components of tetralogy of Fallot include a large ventricular septal defect, an aorta which overrides both the right and left ventricle, stenosis of the right ventricular outflow tract (which may be below, at or above the pulmonic valve level), and right ventricular hypertrophy. This combination of defects results in cyanosis due to inadequate pulmonary blood flow and right to left shunting across the ventricular septal defect. The vast majority of patients present with cyanosis at birth. Survival in patients with unoperated tetralogy of Fallot decreases progressively with increasing age. Only 66% of patients with tetralogy of Fallot will be alive at one year, 40% at three years, 11% by the age of twenty, 6% by the age of 30, and 3% by the age of forty (64). However, survival into the sixties has been reported in rare cases.

Initial attempts at surgical treatment of tetralogy of Fallot were palliative operations to increase pulmonary blood flow and thus decrease the level of cyanosis. Operations such as the Waterston shunt (anastomosis between the ascending aorta and the right pulmonary artery), the Potts shunt (anastomosis between the descending thoracic aorta and the left pulmonary artery), and the Blalock-Taussig shunt (anastomosis between the subclavian artery and the ipsilateral pulmonary artery) provided substantial relief of cyanosis and improved survival and quality of life for these children. However, some of these shunts (especially the Waterston and Potts shunts) were associated with significant long-term complications, most importantly the development of severe pulmonary vascular obstructive disease and pulmonary hypertension due to the increased pulmonary blood flow. Currently, patients with tetralogy of Fallot are treated with primary cardiac repair in childhood (58).

Patients with unrepaired tetralogy of Fallot may present for the first time in adulthood, although this is uncommon. When patients present as unoperated adults with tetralogy of Fallot, surgical correction is usually recommended provided that there is no significant comorbidity. Surgical correction includes closure of the ventricular septal defect and relief of right ventricular outflow tract obstruction. Other associated defects may need to be addressed as well (e.g. atrial septal defect, aortic regurgitation). It is imperative that prior to surgery a detailed evaluation of the cardiac anatomy needs to be undertaken in order to optimally plan surgery. In experienced centers, the operative mortality in adults undergoing repair of tetralogy of Fallot is similar to that in a pediatric population (i.e. in the range of 2-4%) (65) (66) (67) (68) (69). In order to achieve optimal results, this type of surgery should be performed by a cardiac surgeon with experience and expertise in congenital heart surgery in both pediatric and adult populations (35) (36).

More commonly, adults with tetralogy of Fallot will have been diagnosed in childhood and patients will have had some type of surgical intervention. In these post-operative patients, it is imperative to define the specifics of their surgical intervention (i.e. palliative shunt versus corrective surgery) in order to provide appropriate long-term follow-up. Most patients will have had intracardiac repair of their defect but patients may be encountered who have only had a palliative procedure. Patients who have had a long-standing Potts or Waterston shunt (initially created to increase pulmonary blood flow and thereby decrease cyanosis) will invariably have severe, inoperable pulmonary vascular disease (70). Patients who have received a Blalock-Taussig shunt may have continued symptomatic improvement for decades but are still considered to benefit from intracardiac repair as adults (65) (71). Most patients will have undergone intracardiac repair of their

defect which includes closing the ventricular septal defect and relieving right ventricular outflow tract obstruction. Long-term survival data for tetralogy of Fallot patients who have undergone repair indicates that their survival, while good, is not normal (72) (73). Increased age at the time of repair, preoperative erythrocytosis, use of an right ventricular outflow patch, and a previous Waterston or Potts shunt were all noted to adversely affect long-term outcome.

While surgical repair of tetralogy of Fallot clearly improves both quality of life and longevity, there are significant long-term sequelae of this operation (74). While the surgery is usually quite successful in relieving right ventricular outflow tract obstruction, this is often at the expense of causing pulmonic regurgitation. Pulmonary regurgitation is usually well tolerated for years but, over time, results in a significant volume load on the right ventricle leading to significant right ventricular dysfunction. Pulmonic valve replacement may be required(75). In some patients, right ventricular outflow tract obstruction may not be adequately relieved or may recur, resulting in a pressure overload on the right ventricle. Residual ventricular septal defects may be present. They are usually small and hemodynamically insignificant although they do place the patient at increased risk for endocarditis. Aneurysms of the right ventricular outflow tract may occur, although they are usually clinically insignificant. Patients who have undergone repair of tetralogy of Fallot in childhood may have late post-operative left ventricular dysfunction which is thought to be due to the volume overload from the right to left shunting that was present preoperatively. (72) (69) (76).

Most significantly, patients with repaired tetralogy of Fallot are at risk for arrhythmias. Atrial arrhythmias are common and patients may have atrial fibrillation or, more commonly, a reentrant rhythm originating from the surgical atriotomy site (clinically appears as atrial flutter) (77) (78). Patients are also at risk for developing ventricular tachycardia, especially if a surgical incision was made in the right ventricular outflow tract (79). Both atrial and ventricular arrhythmias may become manifest years after the original surgery and can be worsened or exacerbated by residual hemodynamic defects such as significant pulmonary regurgitation or residual right ventricular outflow tract obstruction. Finally, patients who have had successfully repaired tetralogy of Fallot are at risk for late sudden death, with rates reported to range from 0.5% to 7% (80) (72) (66) (65). It is presumed that sudden death late after cardiac repair in patients with repaired tetralogy of Fallot is due to ventricular arrhythmias although this has never been clearly demonstrated(19).

Endocarditis risk: Unoperated = moderate risk. Post-operative = low risk, unless there is a residual VSD, aortic regurgitation, presence of a prosthetic conduit.

Recommendations for follow-up: All patients with tetralogy of Fallot, whether unoperated or post-operative, should have long-term follow-up with a cardiologist dedicated to the management of patients with congenital heart disease.

Eisenmenger syndrome

The other common cause of cyanosis in adults with congenital heart disease is the Eisenmenger syndrome. The Eisenmenger syndrome is defined as pulmonary vascular disease and cyanosis resulting from any intracardiac or systemic-to-pulmonary circulation connection (46). In this syndrome, a large defect is present at birth which allows a significant left to right shunt. The increased volume (and often increased pressure) in the pulmonary vascular bed from the shunt leads, over time, to progressive pulmonary vascular obstructive disease and elevation of pulmonary pressures and pulmonary vascular resistance (81). As pulmonary vascular resistance increases, the magnitude of the left to right shunting decreases. At the point where pulmonary vascular resistance equals or exceeds systemic vascular resistance, right to left shunting occurs. Symptoms are common by the time patients reach adulthood with progressive fatigue, dyspnea and cyanosis being

the most common complaints (46). Patients may present with: 1) a history of congestive heart failure in infancy which improved (as pulmonary vascular resistance increased), followed by the gradual onset of cyanosis and dyspnea, 2) a history of cyanosis in childhood, often having had some type of surgical intervention, or 3) with no history of cardiac disease in childhood (82).

Pathologically, the disease appears identical to primary pulmonary hypertension but the long term prognosis is actually quite different (83). Survival data for patients with Eisenmenger syndrome is actually encouraging. Approximately 80% of patients will be alive at 10 years after the diagnosis has been made, 77% at 15 years, and 42% at 25 years (84) (85). Thus, while these patients clearly do not have a normal lifespan, they can do surprisingly well for many years. The most common causes of death in patients with Eisenmenger syndrome include sudden death, congestive heart failure, cardiac or non-cardiac surgery, hemoptysis, brain abscess, stroke, and pregnancy (46) (86) (85). On reviewing this list, it is clear that at least some deaths in patients with Eisenmenger syndrome are potentially avoidable. Thus, the careful management of these patients involves avoidance of potentially dangerous situations such as pregnancy, general anesthesia, and other situations which might upset the delicate balance between pulmonary and systemic vascular resistance in these patients.

Patients with Eisenmenger syndrome have varying degrees of cyanosis and are at risk for multiple complications associated with chronic cyanosis. An understanding of these complications is critical in the appropriate management of these patients (19). One of the most dramatic sequelae of chronic cyanosis is the development of erythrocytosis. Chronic hypoxemia stimulates red blood cell production, resulting in an increase in total red blood cell mass. This, in turn, results in an increase in whole blood viscosity which may result in symptoms of hyperviscosity (headache, fatigue, dizziness, visual disturbances, and parenthesis). In the majority of cyanotic patients, the degree of erythrocytosis is appropriate and does not require intervention. Repeated phlebotomies performed in an attempt to decrease red blood cell mass will result in iron deficiency and, since iron deficient red cells are actually less deformable than iron-replete red cells, may result in worsening of hyperviscosity symptoms (87).

Patients with chronic cyanosis also have abnormal hemostasis (19) (88) (89). Patients have a hemorrhagic diathesis which is attributable, at least in part, to an acquired abnormality in one of the large multimers of von Willebrand factor. Cyanotic patients may bleed from relatively minor injuries and can have significant hemorrhagic complications from surgery. Paradoxically, these patients are also at risk for thrombus formation, especially in the pulmonary vascular bed. Thus, patients may present with hemoptysis or pulmonary hemorrhage due to thrombosis of the pulmonary arteries (resulting in pulmonary infarction) or due to rupture of abnormal vessels in the pulmonary vascular tree. Hemoptysis may be relatively minor or may be massive and life-threatening.

Because of the increased red blood cell turnover, cyanotic patients are at risk of developing pigment gallstones (calcium bilirubinate) and are at risk for cholecystitis (19). Patients with Eisenmenger syndrome are at risk for paradoxical emboli as venous thrombi can easily traverse the cardiac defect and can result in devastating complications such as stroke or, less commonly, peripheral arterial embolization. Also, because of the chronic cyanosis patients, frequently develop hypertrophic osteoarthropathy resulting in arthralgias and bone pain. Chronically cyanotic adults frequently have hyperuricemia which is thought to reflect abnormal renal clearance of uric acid due to clinically occult renal disease (90). Renal insufficiency may not be suspected as the serum creatinine level may not reflect the severity of underlying renal dysfunction (91). While hyperuricemia is common, clinical episodes of gout are infrequent. Finally, patients with chronic cyanosis are at risk of brain abscess because of their right to left shunting (19).

Treatment strategies for managing patients with Eisenmenger's syndrome are largely preventative measures (19) (46). Dehydration should be avoided as it can worsen symptoms of hyperviscosity and can compromise already marginal renal function. Certain

categories of medications should be scrupulously avoided in these patients. Any medication which causes systemic vasodilatation will decrease systemic vascular resistance while pulmonary vascular resistance remains fixed. In this setting, right to left shunting will increase and cardiac output may fall precipitously. For this reason, vasodilators are contraindicated and anesthetic agents need to be carefully chosen. Anticoagulants and antiplatelet agents should be avoided since these patients have a hemorrhagic. Non-steroidal antiinflammatory drugs or other drugs with potential nephrotoxicity are also to be avoided. It is important to maintain sinus rhythm if possible in these patients as they may decompensate with loss of their atrial contribution to filling. Hormonal therapy may be detrimental as well. Estrogens are contraindicated due to the increased risk of thrombosis. Progesterone-only agents associated with fluid retention may also be contraindicated. Pregnancy should be scrupulously avoided because of the high maternal and fetal mortality. Permanent sterilization is the most safe and effective means of preventing pregnancy in this patient population.

Finally, it is important to optimize the level of hemoglobin. It should be recognized that erythrocytosis is an appropriate and normal response in these patients and is not a risk factor for cerebral thrombosis, as was previously thought (91). In fact, a history of phlebotomy and microcytosis are strongly associated with an increased risk of cerebrovascular events (91). Phlebotomy is only indicated in patients with erythrocytosis who have an unstable syndrome manifested by an increasing hematocrit with moderate to severe symptoms of hyperviscosity. Preoperative phlebotomy prior to either surgical intervention or invasive procedures can be performed to improve the hemostatic parameters in a patient but should be done with great care. Patients undergoing phlebotomy should be in an iron-replete state and should receive isovolumic volume replacement during phlebotomy (19).

Patients with Eisenmenger syndrome by definition are not candidates for primary repair of their cardiac defect because of their pulmonary vascular disease. That is, if repair of their intracardiac defect is undertaken, it is done so only with a very high operative morbidity and mortality with no survival benefit (92). Transplantation has been used as a treatment option for this patient population. Because of the pulmonary vascular disease, patients require lung transplantation. If the cardiac defect is amenable to surgical repair, then lung transplantation can be combined with repair of the cardiac defect. For those patients in whom the cardiac defect is complicated or not amenable to simple repair, combined heart-lung transplantation is performed. The results of transplantation in Eisenmenger patients have been disappointing. While the operative mortality for a standard cardiac transplant is in the range of 1-2%, the operative mortality for transplant surgery in a patient with Eisenmenger syndrome is substantially higher (as high as 20-30% in some small series) (93) (94) (95). Likewise, the long-term survival has also been disappointing. Patients who undergo a single or bilateral lung transplant have a 1-year survival of 70-80% but less than 50% are alive at 4 years post-transplant (96) (97). Survival for combined heart-lung transplant is 60-80% at 1 year and at 10 years less than 30% of patients remain alive (96) (97) (98). Since the average survival for patients with Eisenmenger syndrome without any intervention is at least 10-15 years from the time that the diagnosis is made, it is difficult to be enthusiastic about transplant surgery for patients with Eisenmenger syndrome in the hopes of improving their survival. For this reason, transplantation is reserved for patients who are exhibiting symptoms that suggest that they are high risk for an adverse outcome (e.g. syncope, hemopytsis, and right heart failure). Late referral (when the patient is severely decompensated) results in even higher operative mortality. Thus, the criteria for who should be referred for transplant and the appropriate timing of referral for transplant in the Eisenmenger patient population has not been determined.

Endocarditis risk: Overall risk is low but results can be devastating. Prophylaxis is recommended.

Recommendations for follow-up: All patients with Eisenmenger syndrome or severe pulmonary vascular disease in a postoperative patient should have long-term follow-up with a cardiologist dedicated to the management of patients with congenital heart disease

Post-operative patients with complex congenital defects.

As mentioned previously, the majority of patients seen in a clinic for adults with congenital heart disease will have had some type of operative intervention in the past. Surgery for congenital heart disease can be considered to be curative, reparative, or palliative(19). With curative cardiac surgery, the cardiac defect is completely repaired and there are no post-operative residua, sequelae or complications. Only a few truly curative procedures exist, including ligation and division of a patent ductus arteriosus, or closure of a ventricular septal defect or atrial septal defect with no evidence of pulmonary vascular disease. For more complicated defects, corrective or reparative surgery may be performed which may include repair of defects, relief of obstruction, and otherwise attempting to restore normal circulation. Prosthetic materials such as patches, valves, and conduits may be utilized. Reparative surgery may result in normal circulation but is not curative since there are obligatory post-op residua and sequelae and these patients require long-term cardiology follow-up. Surgery for tetralogy of Fallot falls into this classification. Finally, some procedures are considered palliative. In palliative procedures, no attempt is made to repair or reconstruct the basic structural defect. Rather, the goal is to improve functional status of the patient, most commonly by increasing pulmonary blood flow. Table I is a brief listing of cardiac surgical procedures performed in patients with congenital heart disease with a brief description of the surgery and its intent (palliative versus corrective). A full discussion of the various types of post-operative patients with congenital heart disease is beyond the scope of this review. General recommendations follow below.
Endocarditis risk: Patients at highest risk include those with prosthetic valves, prosthetic valved conduits, and certain palliative shunts (Potts shunt and Blalock-Taussig shunt). Unrepaired cyanotic defects, non-valved conduits, left-sided regurgitant lesions, valve stenosis, and high-velocity intracardiac shunts are considered to be moderate risk(19). Nearly all post-operative patients with complex congenital heart disease should receive endocarditis prophylaxis.
Recommendations for follow-up: All patients with a history of cardiac surgery for complex congenital heart disease should have long-term follow-up with a cardiologist dedicated to the management of patients with congenital heart disease

General issues in the management of congenital heart disease in adults:

Pregnancy and contraception

There are a number of important issues that relate specifically to the long-term management of adults with congenital heart disease. One of the most important issues in adolescents and adults with congenital heart disease is the advisability and risks of pregnancy as well as the need for and the appropriate type of contraception. For certain types of congenital defects, pregnancy is absolutely contraindicated while in other conditions carries with it at least a mild to moderate risk. For other types of congenital defects, pregnancy may be extremely well tolerated and would not be contraindicated. In some patients with complicated anatomy, especially after complex cardiac repairs, the risks of pregnancy are actually unknown. For this reason, pregnancy in patients with congenital heart disease should be planned and the risks of pregnancy to the mother need to be addressed prior to conception (99) (100) (101).

Pregnancy in patients with Eisenmenger syndrome or severe pulmonary hypertension of any cause is contraindicated because of the high maternal and fetal mortality (102) (103) (104) (105). In addition, pregnancy is absolutely contraindicated for

patients with evidence of decompensated heart failure and/or severe obstructive lesions (106). Patients with Marfan syndrome with aortic root dilatation are at risk for aortic dissection and rupture during pregnancy and pregnancy is contraindicated (107). A similar risk of aortic dissection and rupture exists for patients with unrepaired coarctation of the aorta (108). In addition, the fetus is at risk for growth retardation due to inadequate uterine blood flow. Other cardiac conditions seen in patients with congenital heart disease represent a moderate or high risk state in which pregnancy is not absolutely contraindicated. This includes patients with prosthetic valves (especially those requiring chronic anticoagulation) and patients with repaired coarctation of the aorta or patients with Marfan syndrome without evidence of aortic dilatation (as these patients are still at risk for aortic dissection or rupture during pregnancy). Patients with moderate obstructive lesions or impaired ventricular function are also at increased risk but may successfully tolerate a pregnancy (107). Patients with chronic cyanosis are at increased risk for pregnancy although maternal morbidity and mortality are highly dependent on other features of their cardiac defect (e.g. presence of obstructive lesions, ventricular dysfunction, and pulmonary hypertension) (109). Fetal mortality in cyanotic patients is highly dependent on the level of maternal cyanosis with severely cyanotic patients having a much higher incidence of fetal wastage than those patients with milder degrees of cyanosis.

In this expanding population of patients, there are a growing number of young women who have undergone some type of surgical repair or palliation in whom the risks of pregnancy are not well known (110). In some complex forms of congenital heart disease, creative surgery has resulted in an "unusual" circulation that may function well in the normal, non-pregnant state. However, the effects of volume loading as well as other effects of pregnancy on these altered circulations is not well known. An example of this is the Mustard operation which was formerly the operation of choice performed for d-transposition of the great arteries. In patients with d-TGA, the aorta arises from the right ventricle and the pulmonary artery from the left ventricle. Thus, desaturated blood returns to the body and fully oxygenated blood returns to the lungs. Some form of intracardiac shunting is required for neonatal survival. In the past, the operation of choice for this defect was the redirection of venous blood flow to the heart through an atrial switch operation known as the Mustard procedure. After this operation, systemic venous return is baffled across the mitral valve into the left ventricle and thus out into the pulmonary circulation. Pulmonary venous return is then baffled across the tricuspid valve into the right ventricle. In patients with a "Mustard circulation," the right ventricle functions as the systemic ventricle. This atrial switch operation is now rarely performed as it has been replaced by the arterial switch procedure in which the aorta and pulmonary artery are transected above the semilunar valves and the aorta is then connected to the left ventricle and the pulmonary artery to the right ventricle. The coronary arteries are relocated to the neoaorta to restore normal coronary circulation. While this operation has proven to be superior to the atrial switch operation, there is a substantial population of patients now in or approaching adult age and child-bearing years who have undergone the atrial switch operation in childhood (111). There is limited data available that indicates that young women who are s/p Mustard operation are able to tolerate pregnancy fairly well although they do have an increased miscarriage rate (112).

An even more complicated situation are patients who have had a Fontan operation. The Fontan procedure was developed for patients with a functional single ventricle. The basic premise of the Fontan operation is that the systemic venous return is connected directly to the pulmonary artery, resulting in non-pulsatile flow to the pulmonary vascular bed. The single ventricle (whether it be a morphologic right or left ventricle) then functions as the systemic ventricle and the pulmonary and systemic circulations are separated. This is clearly a palliative operation as it does not restore normal circulation but, in ideal candidates, this operation has been quite successful in improving survival and quality of life (113). The implications of pregnancy in patients who have undergone a Fontan procedure are not well known. Some successful pregnancies as well as a few significant

complications have been reported in a small number of patients (114). There does appear to be an increased risk of miscarriage in these patients.

Thus, a patient who has a history of congenital heart disease (especially with a history of some type of operative repair in childhood) needs to have a thorough evaluation prior to contemplating pregnancy. This evaluation should include investigation of what the primary anatomy was and the exact details of any operative repair. In addition, evaluation for post-operative residual defects or long-term sequelae needs to be carefully performed. In those patients in whom pregnancy is contraindicated or not desired, permanent sterilization is recommended as the safest and most effect means of contraception (99). The use of standard-dose estrogen oral contraceptives is generally though to be contraindicated, especially in cyanotic patients or patients with significant pulmonary vascular disease, due to the increased risk of thrombosis. Low-dose estrogen contraceptives may be an acceptable alternative. Progestational agents such as the minipill or Norplant have been used in patients with congenital heart disease, although their is relatively little data. Depoprovera is often associated with significant fluid retention which makes it undesirable for use in patients with heart disease. Intrauterine devices (IUDs) are contraindicated due to the risk of infection and bleeding. Barrier methods are unreliable and are not recommended (115) (19) (99).

In addition, patients contemplating pregnancy need to have genetic counseling to be aware of the risk of congenital heart disease in their offspring. Most forms of congenital heart disease are multifactorial in origin, demonstrating non-Mendelian inheritance and only an estimate of risk to the fetus can be made. In patients with congenital heart disease who have children, the risk of congenital heart disease in their offspring is substantially higher than for the general population. For mothers with congenital heart disease, the risk of their child being born with congenital heart disease is in the range of approximately 7% (2.5-18%). When a male with congenital heart disease fathers a child, the risk of congenital heart disease in the offspring is lower, in the range of 2% (1.5-3%). The greater risk associated with the affected parent being the mother is thought to reflect cytoplasmic inheritance (inheritance of all mitochondria and all mitochondrial DNA from the mother) (116). Other factors influencing the risk of congenital heart disease in the offspring include the severity of the cardiac defect in the parent and the number of other affected individuals in the family. For the 10% of patients with single gene disorders (e.g. Noonan syndrome, Holt-Oram syndrome, CATCH-22, William's syndrome, and Marfan syndrome) or chromosomal defects (e.g. Trisomy 21, 13, or 18, Turner syndrome) a more precise estimate of fetal risk can be made (117)(118).

Noncardiac surgery in patients with congenital heart disease

Patients with congenital heart disease may be at increased risk during non-cardiac surgery and require careful preoperative evaluation and intraoperative management. The preoperative evaluation should include acquiring information regarding the underlying cardiac defect, any reparative or palliative procedures performed, and a recent assessment for residual defects or long-term sequelae. The critical data to be defined include the presence, magnitude and direction of shunting, the presence and magnitude of pulmonary hypertension, assessment of ventricular function, assessment for valve stenosis or regurgitation, and assessment of the function of any prosthetic material (conduits, valves, etc.) (119) (19). From here, common sense takes over as there is relatively little data regarding the appropriate management of patients with congenital heart disease undergoing non-cardiac surgery. Clinical risk indices which have been defined for cardiac patients undergoing non-cardiac surgery have been derived from patients with acquired forms of heart disease, namely ischemic heart disease and congestive heart failure. There is little data on surgical outcomes of various anesthetic approaches in patients with congenital heart disease. Anesthesia medications must be carefully chosen and administered (120) (121). In patients with intracardiac shunts, the effects of anesthetic medications on the magnitude of

intracardiac shunting need to be carefully evaluated. In patients with pulmonary hypertension and intracardiac shunting, any agent which decreases systemic vascular resistance will increase the magnitude of right to left shunting. Conversely, agents which increase systemic vascular resistance will decrease the magnitude of right to left shunting. Maneuvers which decrease pulmonary vascular resistance will increase left to right shunting while maneuvers that increase pulmonary vascular resistance will increase right to left shunting. Patients with intracardiac shunts are at risk for paradoxical emboli and careful management of intravenous lines is mandated. In general, preoperative consultation with a cardiologist knowledgeable about congenital heart disease is recommended. Cardiac anesthesia is preferable when available.

Insurance in adults with congenital heart disease.

Nearly all patients in the United States have some type of access to health care in childhood, whether through federal or states programs or private insurance through their parents. However, as these patients reach adulthood, their previous health insurance coverage is lost and they often have difficulty obtaining personal health insurance (122) (123). Congenital heart disease is the ultimate "preexisting condition." While it is clear from a medical point of view that all types of congenital heart disease are not the same, from the viewpoint of insurance carriers the multiple types of defects are often lumped together. In fact, a history of cardiac surgery is usually a greater obstacle for obtaining health or life insurance than is the severity of the cardiac disease itself. Thus, patients with severe cardiac disease who are inoperable but have had poor follow-up for their condition may be able to obtain health insurance whereas a patient who has had successful cardiac surgery and has a reasonably good outlook from a cardiac standpoint may be denied insurance. Similar to health insurance, life insurance is often not available for patients with many types congenital heart disease. These are significant issues for young adults.

Lifestyle issues in adults with congenital heart disease.

Congenital heart disease is a chronic illness that has lasting effects on both the patient and his/her family (124) (125). For many of these patients, their cardiac condition dominated their childhood. When patients with congenital heart disease are looked at as a group, there is no evidence of intellectual impairment. It is unclear whether or not chronic cyanosis results in cognitive impairment but it is generally thought that the mean IQ for cyanotic patients falls well within the range of normal (126). The academic performance of patients with congenital heart disease is typical of patients with interrupted schooling due to other causes (127). Patients with congenital heart disease may have difficulty finding employment as employers may be reluctant to hire employees with known heart disease, despite the existence of legislation which should protect such individuals (i.e. Americans with Disabilities Act, National Rehabilitation Act) (128). There is some evidence that patients with congenital heart disease may be at increased risk of psychiatric illnesses such as depression and panic disorders, although this is not clearly proven (129) (19). As patients who have had to deal with a chronic illness, patients with congenital heart disease develop a variety of coping mechanisms to deal with their disease. Common coping mechanisms seen especially in adolescents and young adults include denial of their disease or failure to acknowledge the seriousness of their disease, rebellion (which may be manifested as noncompliance or refusal of medical care), or passivity where the patient refuses to take an active role in their health care (19).

Patient knowledge of their disease:

Somewhat surprisingly, patients with congenital heart disease often have only a limited understanding of their disease process and its complications (125) (130). For

patients diagnosed and treated in childhood, the focus on disease education may have been directed towards the parents and the patient may have been too young to comprehend or may have been excluded from discussion of their condition (either deliberately or inadvertently). When patients remain stable for substantial periods of time, they may forget details of their condition and may minimize the importance of their disease. Many post-operative patients incorrectly view themselves as "cured" and find the details of their cardiac history unimportant. One recent study demonstrated that in a group of 50 adult patients attending a clinic devoted to adult congenital heart disease, only 54% knew their diagnosis (131). When given a heart diagram, only 26% could mark their defect correctly. Perhaps even more disturbing, only 16% understood the term "endocarditis" and only 22% recognized the term "antibiotic prophylaxis. In a similar series from the Mayo Clinic, 50% of patients could define endocarditis but only 43% had an adequate understanding of hygiene measures and antibiotic prophylaxis (132). Patients usually have a similar lack of knowledge regarding the advisability of pregnancy, the genetic risk in their offspring, risks of non-cardiac surgery, limitations on their physical activity, and their risk for acquired heart disease such as coronary artery disease. Thus, education is a vital component of the management of adults with congenital heart disease.

Summary.

As this overview demonstrates, the long-term care of adults with congenital heart disease can be complex. These patients have been traditionally cared for by pediatric cardiologists who usually develop long-term relationships with the patient and their family members. Pediatric cardiologists are at times reluctant to relinquish care of these patients to their colleagues in adult cardiology. This reluctance arises partly from fear that adult cardiologists will not understand the disease process and its management. Traditional training in adult cardiology does not include much exposure to congenital heart disease. Thus, adult cardiologists who are interested in treating patients with congenital heart disease require additional training (133). In addition, it is important to maintain a collaborative relationship with our colleagues in pediatric cardiology. It has been proposed that congenital heart disease in adults be identified as a new subspecialty within cardiology and that specialized centers be organized and identified where adults with congenital heart disease can receive adequate treatment (15) (134) (16) (17) (18) (135). Important components to be included in these centers include both pediatric and adult cardiologists (including echocardiographers, electrophysiologists, and interventionalists), cardiac surgery (with ability to provide both pediatric and adult surgery), the availability of a transplant program, cardiac anesthesia, and an obstetric program with an interest in high risk pregnancy.

Most adult patients with congenital heart disease would benefit from being seen by a specialist in congenital heart disease. However, not all patients with congenital heart disease require referral to a specialist. Any adult with a history of cyanosis or who is currently cyanotic should be referred to a cardiologist with interest in congenital heart disease. Any patient with complex congenital heart disease who is unoperated should be referred to a specialist. Most post-operative patients should also be seen at a specialized center due to the long-term sequelae of most cardiac operations. Exceptions to this rule include patients with uncomplicated cardiac shunts such as atrial septal defects, ventricular septal defects, and patent ductus arteriosus who were repaired in childhood and have no evidence of pulmonary hypertension. Likewise, patients with small, hemodynamically insignificant atrial septal defects or ventricular septal defects do not require follow-up in a specialty clinic.

In summary, there is an increasing population of adults with congenital heart disease who will be encountered in the practice of internal medicine. While patients with previously undiagnosed congenital heart disease will still be encountered, the vast majority of adults with congenital heart disease will have had some sort of operative intervention in

childhood. These patients have special needs which can best be provided for by physicians with a special interest in congenital heart disease. George Elliott commented that "In science generally, to solve one set of problems may be to create or discover a whole new set and of no science ... is this more true than that of medicine (1)." This is particularly true in the field of adult congenital heart disease where amazing advances in the management of these patients, particularly through advances in cardiac surgery, have resulted in cures for some and substantial improvement in lifestyle and lifespan for others for what were many times in the past fatal diseases. As surgical techniques change, this field will continue to change with the long-term complications of different surgical techniques resulting in an ever-changing spectrum of patients and problems to be encountered.

Table I.

Procedure	Description	Intent	Result
Blalock-Taussig	Subclavian artery to pulmonary artery anastomosis	PAL	Increases pulmonary blood flow
Central shunt	Conduit or anastomosis between aorta and pulmonary artery	PAL	Increases pulnonary blood flow
Damus-Kaye-Stansel	Pulmonary artery end-to-side anastamosis to aorta, valved conduit between RV and main pulmonary artery	COR	Increases blood flow to aorta and pulmonary artery when there is aortic stenosis and two ventricles, reestablishes RV to PA continuity
Fontan	Anastomosis or conduit between right atrium and pulmonary artery	PAL	Increases pulmonary blood flow in cases of univentricular heart or tricuspid atresia
Glenn	SVC to pulmonary artery anastomosis	PAL	Increases pulmonary blood flow
Arterial switch, Jatene	Transection of aort and pulmonary artery with reimplantation onto the proper ventricles, coronary arteries reimplanted	COR	Creates normal relationship between the ventricles and great arteries in transposition
Hemi-Fontan	SVC to pulmonary artery anastomosis with baffle placed in right atrium so that IVC blood flow goes across ASD to left heart	PAL	Increases pulmonary blood flow and sets stage for eventual complete Fontan
Konno	Replacement of aortic valve with aortic valve annular enlargement	COR	Alleviates subaortic obstruction and replaces abnormal aortic valve
Mustard	Atrial switch with intraatrial baffle made of pericardium	COR	Reestablishes proper flow sequence to pulmonary artery and aorta in D-transposition of great arteries

Procedure	Description	Intent	Result
Norwood (first stage)	Pulmonary artery anastomosis to aorta, conduit from aorta to main pulmonary artery	PAL	Increases flow to aorta for subaortic obstruction with single ventricle
Potts	Descending aorta to pulmonary artery shunt	PAL	Increases pulmonary blood flow
PA band	Constrictive band around main pulmonary artery	PAL	Decreases pulmonary blood flow
Rashkind	Atrial septostomy with catheter balloon	PAL	Increases mixing of blood for transposition of the great arteries or tricuspid atresia
Rastelli	Valved conduit from RV to PA, establish continuity between LV and aorta with closure of VSD	COR	Increases pulmonary flow, may reestablish proper sequence of flow to aorta and pulmonary artery
Ross	Pulmonary autograft to aorta, pulmonary homograft	COR	Correction for aortic stenosis. Avoids mechanical and bioprosthetic valve
Senning	Atrial switch with intraatrial baffle made of atrial wall flaps	COR	Reestablishes proper flow sequence to pulmonary artery and aorta in transposition of the great arteries
Waterston	Ascending aorta to right pulmonary artery anastomosis	PAL	Increases pulmonary blood flow

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