MANAGEMENT OF PATIENTS WITH REPAIRED CONGENITAL HEART DISEASE

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The population of children and adults who have undergone repair or palliation of congenital heart disease (CHD) is on the rise[1]. The advances in echocardiography, anesthesia, intensive care, and perhaps more importantly early and more definitive surgical intervention and improved surgical techniques, have increased the survival of babies born with complex cardiac anomalies into adulthood[2].

In the year 2000, there were about 785,000 adult patients with congenital heart disease living in the U.S, the majority of them having undergone reparative cardiac surgery in infancy or childhood[3]. This population is likely to increase tremendously over the next few years as estimates predict and an annual addition of the 10,000 to 30,000 patients/year with doubling of survival rate in patients with complex physiology[1].

As life expectancy of adults with CHD has dramatically improved, noncardiac medical and surgical issues typical of any adult population have become more prominent[1]. Over the next few years, more and more patients with repaired/palliated CHD are likely to be scheduled for non-cardiac surgery. The anesthesiologist caring for these patients faces the challenge of dealing with the complications that may arise intra- or postoperatively.

McGowan emphasizes the importance of addressing the outcome and problems associated with specific CHD lesions as well as their repair in order to formulate rational perioperative management strategies. According to him, "the anesthesiologist caring for patients with

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repaired/palliated CHD undergoing noncardiac surgery needs to have a complete understanding of the physiology of the lesion, the type and natural history of repair, and the likely interactions of all of these with the planned procedure."[1]

What is the outcome of repaired CHD?

Stark defines corrective surgery for CHD as that which achieves and maintains a normal cardiac function, results in normal life expectancy and requires no subsequent surgical or medical therapy[4]. Based on this, very few types of repairs are truly curative. These would include PDA ligation, early repair of isolated ASD or VSD[1,5]. All other forms of repair have the potential for residua and sequelae that can become progressive as patients grow older[1].

Overview of common types of CHD

Congenital heart disease occurs in 6 to 8 out of every 1000 births[5]. Grossly, common defects discussed in this review include VSD, PDA, ASD, TOF, TGA, coarctation of the aorta and "single ventricle" defects (e.g. tricuspid atresia, hypoplastic left heart syndrome etc.).

Three physiological factors are important in determining the type of defect and the timing of surgical intervention for CHD[6]. Defects are classified according to the amount of oxygenated blood delivered to the systemic circulation (cyanotic vs. acyanotic), the amount of pulmonary blood flow (increased, normal or decreased), and the presence or absence of obstructive lesions[6].

As such, acyanotic CHD can be further classified into two categories:

1) acyanotic disease with increased pulmonary blood flow (PBF) (namely left-to-right shunts such as ASD, VSD, PDA).

2) acyanotic CHD disease with normal blood flow (mainly obstructive lesions such coarctation of the aorta).

On the other hand, cyanotic CHD is also divided into defects with increased pulmonary blood flow (e.g. TGA, truncus arteriosus) vs. defects with decreased pulmonary blood flow (TOF, tricuspid atresia).

The significance of such a classification is to emphasize the hemodynamic abnormalities associated with each type of lesions and to understand the rationale of surgical repair[6].

Acyanotic CHD with increased PBF (left-to-right shunt)

Shunt lesions whether intracardiac (ASD, VSD) or extracardiac (PDA) operate by the same principle. Blood flow through the shunt depends on the size of the shunt and the relative resistances on either side of the shunt[7]. Grossly, if defect is large enough not to impede blood flow (i.e. non-restrictive defect), then the main determinant of the degree of shunting across this defect is the resistance on both sides of the shunt[8]. If the defect opening is small (i.e. restrictive defect), then the degree of shunting will depend more on the size of the defect and less on the resistance across the defect[8].The drop in PVR occurring shortly after birth creates the driving force for the left-to-right shunting of blood across the defect.

1 - Ventricular Septal Defect (VSD)

Whether located in muscular or membranous portion of the septum, the hemodynamics associated with VSD depend mostly on the size of the defect. Small defects account for about 85% of all VSDs[9]. The extent of shunting is limited by the size of the defect therefore the blood flow across the defect is minimal and decreases with time. Most of these defects close spontaneously (90% close by the age of 6 years)[6] with those in the muscular portion of the interventricular septum closing sooner than those in the membranous part[6]. Children with small defects are usually followed up with echocardiography until the defect closes.

Larger non-restrictive VSDs become more problematic as there is more blood shunted across the defect. The physiology of the shunting results in volume overload of the RV, RA and the pulmonary circulation that is transmitted to the left side of the heart as well[8]. Initially, the increased blood volume returning to the LV increases the stroke volume (Frank-Starling mechanism), however with time the volume overload can

lead to LV dilatation, systolic dysfunction and symptoms of left-sided heart failure as early as 2-3 months of age (failure to thrive, tachypnea, lower respiratory tract infections)[3,6].

The volume overload of the pulmonary vasculature leads to increased PAP and pulmonary arteriolar hypertrophy with time with PHTN setting in as early as the age of 2^3. The initial medial hypertrophy of the pulmonary arterioles, intimal proliferation and fibrosis, and occlusion of capillaries and small arterioles are potentially reversible. As the disease progresses, the more advanced morphologic changes (necrotizing arteritis and plexiform lesions) are irreversible; resulting in obliteration of the pulmonary vascular bed, leading to increased pulmonary vascular resistance[10]. The flow across the VSD decreases as PVR increases; once the PVR exceeds the SVR, reversal of flow occurs (Eisenmenger syndrome)[3,5]. When the shunt reverses (that is, when right-to-left shunting occurs), cyanosis and erythrocytosis develop.

Eventually, most patients with the Eisenmenger syndrome have one or more of the following:

1) symptoms of a low cardiac output (such as dyspnea on exertion, fatigue, or syncope).

2) subtle neurologic abnormalities (such as headache, dizziness, or visual disturbances) due to erythrocytosis and hyperviscosity.

3) symptoms of congestive heart failure[11].

In addition, arrhythmias (atrial fibrillation/flutter, ventricular tachycardia) and hemoptysis (secondary to bleeding bronchial vessels or pulmonary infarction) are common[3,11]. Both arrhythmias and massive hemoptysis account for most of the sudden death in patients with Eisenmenger syndrome[11]. Cerebrovascular accidents frequently occur mainly because of hyperviscosity but they can also be the result of paradoxic emboli[11]. Renal dysfunction is also a common problem; more than one third of adult patients with cyanotic congenital heart disease have evidence of glomerulopathy (proteinuria; elevated serum creatinine concentration; or abnormal urinalysis results with hematuria, sterile pyuria, or casts)[12]. Renal dysfunction is mainly the result of diminished

renal blood flow secondary to erythrocytosis and as such the incidence of renal abnormalities increases with the degree and duration of cyanosis and accompanying erythrocytosis[11].

Surgical repair of VSD

Reparative surgery for VSD is usually done electively between 3-9 months of age[3]. Surgery is performed before 2 because of increased risk of irreversible PHTN after this age[3]. Surgical repair of isolated VSD is done using CPB to close the defect by direct sutures or patching [3]. The closure is usually performed through the right atrium or pulmonary valve to avoid a ventriculotomy[6]. Transcatheter closure of VSD is less commonly used[3].

Postoperative sequelae

Early repair: Most patients who undergo repair do well[8]. For patients with good left ventricular function prior to surgery, life expectancy after surgical correction is close to normal[3]. Problems that may arise after early repair may be due to any residual defect (especially with multiple VSDs), aortic regurgitation after repair of perimembranous VSD, LVOT or RVOT obstruction secondary to patching, heart block requiring temporary or long term pacemaker support[5].

Late repair: Patients who had late repair of VSD deserve special consideration as they are at increased risk of LV dysfunction and decreased EF long after repair[13]. Residual hemodynamic problems occur when repair is delayed to late childhood because by then the change in PVR has become irreversible. After closure of VSD, the RV is forced to pump against an increased PVR. It must suddenly change from pumping against a volume load to support a pressure load[8]. Similarly, the LV must pump a smaller volume load against SVR without having the advantage of pumping any extra volume through the VSD and into the lower pressure right-sided circulation[8]. The LV function may be further compromised if the RV is significantly dilated with the interventricular septum shifting into the LV[8]. The altered LV geometry further impairs LV ejection and thus decreases CO. It is therefore important for any patient with known late repair to be evaluated before any major procedure to assess LV

function and EF.

Patients with Eisenmenger syndrome: These patients are particularly vulnerable to the alterations in hemodynamics associated with surgery or anesthesia. Any minor decrease in SVR can increase right-to-left shunting and can therefore precipitate cardiovascular collapse[3].

Because noncardiac surgery in patients with the Eisenmenger syndrome is associated with a high perioperative mortality rate it should be avoided, if possible[14]. When surgery is necessary, a number of peri, intra and postoperative considerations should be followed. Prolonged fasting should be avoided perioperatively. The use of antibiotic prophylaxis of endocarditis is recommended depending on the type of surgery[3].

The choice of general or epidural-spinal anesthesia is controversial. The latter technique causes sympathetic blockade resulting in systemic arterial vasodilation and hypotension. On the other hand, many of the agents used for induction and maintenance of general anesthesia depress myocardial function and reduce systemic vascular resistance.

Intraoperative monitoring of BP is critical. Systemic arterial hypotension should be treated aggressively (with methoxamine or phenylephrine for example) or intravenous volume replacement if the patient is hypovolemic[5]. Blood loss should be minimized and excessive bleeding should be promptly treated with blood products. In addition all IV lines used intraoperatively should be equipped with air filters to prevent the occurrence of paradoxical air embolism[3].

Patients with Eisenmenger syndrome are at increased risk for post-operative arrhythmias and DVT. It is preferable to closely observe postoperative patients in an intensive care unit setting. Early ambulation should be encouraged to prevent thromboembolism and subcutaneous heparin should be considered when prolonged immobilization is anticipated.

2 - Atrial Septal Defect (ASD)

The physiology of ASD is that of left-to-right shunting with volume

overload of the RA and RV. The extent of shunting is not as significant as compared to shunting across VSD mainly because the atrial system is a lower pressure system. In ASD, signs of congestive heart failure are late manifestations therefore patients may go undetected until later in childhood or even adulthood.

Surgical repair for ASD is electively done between 2-5 years of age as a prophylaxis for late pulmonary HTN, paradoxic emboli and arrhythmias[8].

Device closure of secundum ASD can be performed percutaneously under fluoroscopy and TEE or with intracardiac echo guidance[15]. Other types of ASDs have to be closed by patching or surgical suturing using CPB (via a median sternotomy or right anterior thoracotomy)[6].

Postoperative sequelae

In one of the earliest long-term follow-up studies on patients with repaired ASD, Murphy et al. found that patients with ASDs repaired at ages less than 25 had similar survival rates compared to age-matched control subjects[16].

However patients who underwent repair when they were older than 25 years old had decreased long-term survival compared to control subjects[16]. Decreased survival is probably related to the long-standing deleterious effects of volume overload on the right-sided cardiac chambers and the pulmonary vasculature including development of PHTN, atrial enlargement predisposing patients to atrial arrhythmias (atrial flutter and atrial fibrillation) and stroke secondary to atrial thrombosis[2].

Acyanotic Congenital Heart Disease with Normal PBF

Coarctation of the aorta

The physiology of coarctation of the aorta consists of a pressure overload on the LV (secondary to increased afterload) leading to compensatory LV hypertrophy to sustain CO. Another type of

compensatory mechanism is the formation of collateral blood vessels in the intercostals arteries to supply blood to the descending aorta therefore bypassing the coarct site[5]. The presence of such collaterals should be kept in mind when performing procedures such as epidural anesthesia in patients with a history of coarctation.

Another important consideration is that this type of CHD is associated with other defects such as bicuspid aortic valve and cerebral aneurysms in respectively 50% and 10% of patients[2].

Surgical repair

Repair for coarctation of the aorta is usually performed electively at 2 to3 years of age[6]. Timing of surgery is controversial because of the high recurrence rate in infants (up to 25%)[6]. After the age of 3, the risk of recurrence becomes theoretically less than 2%[6]. Repair is done by either subclavian flap arterioplasty or coarctation segment resection with end-to-end anastomosis with prophylaxis needed for prevention of endarteritis[3].

Postoperative sequelae

Long-term follow-up of patients after surgical correction reveals an increased incidence of premature cardiovascular disease and death[3].

Patients with repaired coarctation can demonstrate recoarctation, aneurysms at the coarctation site, residual hypertension, stroke secondary to either cerebral aneurysm or bacterial endocarditis[5].

The prevalence of recoarctation reported in the literature varies from 7 to 60% depending on the definition of recoarctation, the length of follow-up and the age at surgery[3]. For example, recoarctation was found to be more common with end-to-end anastomosis repair as compared to subclavian flap angioplasty[5].

Recoarctation is usually addressed with balloon dilatation if the obstruction is relatively localized. In the presence of a long-segment narrowing, surgical intervention may be necessary[3].

Aneurysm formation is also one of the recognized sequelae of coarct repair with a reported incidence of 2 to 27%[3]. Aneurysms are particularly common after Dacron patch aortoplasty and usually occur in the segment

of the aorta opposite to the patch. Parikh et al. demonstrated aneurysmal formation in 5% of patients after Dacron patch repair but not after subclavian flap angioplasty or end-to-end anastomosis[17]. It is very important that these higher risk patients get followed up after repair with Doppler ultrasound or MRI for aneurysmal formation or recoarctation[3].

Another possible complication of reparative surgery for coarctation is systemic hypertension in the absence of residual coarctation. The pathophysiologic mechanism underlying residual hypertension is not very well understood but is thought to be due to residual abnormal vascular reactivity in the arterial system[2]. Some studies suggested increased risk of residual hypertension when reparative surgery is done at an older age[18], but more recent studies suggest that residual hemodynamic abnormalities are common even after early repair[19].

Patients with normal BP at rest can demonstrate systolic hypertension in response to physical exercise[2]. The significance of exercise induced systolic hypertension is still unknown but is thought to contribute to the increased LV mass frequently seen in patients after surgery. In long-term follow-up studies, LV hypertrophy is seen in 25-33% of repaired coarct patients[1]. It is by itself a predictor of future cardiovascular complications[1].

Other problems that might occur postoperatively include endarteritis at the coarct site with embolic manifestation usually restricted to the lower extremities[3]. Any co-existent bicuspid aortic valve may become stenotic or regurgitant as patients grow older so periodic assessment of possible aortic valve disease is warranted even after successful reparative surgery[3].

Cyanotic Congenital Heart Disease

1 - Tetralogy of Fallot

Tetralogy of Fallot (TOF) is one of the most common cyanotic lesions in infancy[2]. The functional problems in TOF are the presence of VSD, RV outflow tract obstruction (at valvular, infundibular or

pulmonary arterial level) and right-to-left shunting resulting in decreased PBF[5]. The extent of right-to-left shunting depends mainly on the magnitude of the RV outflow tract obstruction as well as the SVR[5].

Surgical repair

Treatment of TOF has shifted from palliation with subsequent repair to early total repair[6]. Symptomatic children are now repaired at any age, and elective surgical repair in asymptomatic infants is usually done during the first 6 months of life[3]. Currently, earlier corrective surgery has allowed more than 85% of children with TOF to survive to adulthood[20].

Repair of TOF consists of right ventriculotomy, patch closure of VSD, resection of infundibular stenosis and patch augmentation of RVOT[8]. If the pulmonary valve is too small, then the RV incision is extended across the pulmonary valve annulus and onto the pulmonary artery[8]. Because of the common finding of a small annulus (especially in younger infants), repair often involves the placement of a transannular patch. Although the patch is effective in relieving obstruction, it distorts the pulmonary valve apparatus and pulmonary regurgitation inevitably occurs with time[2,8].

Thus, earlier repair has minimized the deleterious consequences of long-standing cyanosis but this is often at the expense of transannular patch enlargement of the RVOT, which may predispose patients for later complications due to pulmonary regurgitation[3].

Anatomical variants in coronary artery anatomy alter the surgical approach in patients with TOF. When an anomalous coronary artery crosses the RVOT making transection not feasible, an extracardiac conduit can be placed between the RV and PA bypassing the RVOT obstruction[3].

Postoperative sequelae

Most of the complications relate to abnormal RV loading and to problems associated with RVOT reconstruction[1]. Progressive RV dysfunction, arrhythmias and sudden cardiac death are major problems after repair[1].

Progressive RV systolic dysfunction may be the result of many interplaying factors but in most cases it is the result of long-term volume overload secondary to pulmonary regurgitation (especially after transannular patch repair)[2]. Patients with RV dysfunction may be completely asymptomatic or can manifest with decreased exercise tolerance or even signs of right-sided heart failure[1].

Since RV dysfunction predisposes patients to increased cardiac morbidity and mortality (by increasing the risk of arrhythmias and sudden cardiac death), cardiac imaging namely echocardiography and MRI should be used before any major procedure to quantify the degree of RV dysfunction[3]. Cardiac MRI has become particularly useful because it allows to assess RV function as well as pulmonary regurgitation[21] and to image potential sites for RVOT obstruction[1]. The value of MRI over echocardiography in the evaluation of the RV is becoming increasingly appreciated in complex CHD lesions (such as TOF, TGA) where the CO is greatly dependent on RV function[21].

Atrial and ventricular arrhythmias are common sequelae after TOF repair. Atrial arrhythmias may be present in up to one-third of patients after repair and are a major source of morbidity[22]. Ventricular arrhythmias may occur early or years after repair[23]. Factors such as chronic pulmonary regurgitation, RV dysfunction, RV scarring at the surgical incision site can lead to the development of arrhythmias. The incidence of ventricular arrhythmias (RBBB, complete heart block, PVBs) and non-sustained ventricular tachycardia on ambulatory Holter or exercise testing is considerable and increases with age[1]. These types of arrhythmias do not predispose patients to develop sustained ventricular tachycardia. Sustained ventricular tachycardia are less prevalent than other types of arrhythmias but they are believed to be responsible for incidence of SCD in patients after repair[2].

There is little information in the medical literature concerning perioperative considerations for patients with repaired TOF. Because of the complexity of problems arising after repair, it is impossible to design an algorithm that fits all patients[1].

However in light of the physiology of the repair and its sequelae, some issues are important to address before any major procedure.

First, the degree of RV dysfunction needs to be assessed with consideration given to interventional catheterization for lesions amenable to improvement (e.g. RVOT or PA obstruction, residual VSD, ventricular arrhythmias)[1].

Second, the potential for positive-pressure ventilation to compromise CO (especially in patients with significant pulmonary regurgitation and RV dysfunction) should be recognized[1]. By increasing intrathoracic pressure, positive-pressure ventilation increases RV afterload and therefore decreases RV filling leading to underfilling of the LV as well and decreased CO[1].

Third, any increase in PVR especially in the setting of pulmonary regurgitation and RV dilation can compromise RV filling[1].

For patients with repaired TOF undergoing surgery, it is very important to 1) maintain adequate contractility and filling volume of RV and 2) avoid fluctuations in HR, BP as the combination of tachycardia, hypotension, acidosis is particularly detrimental in these patients[1].

2 - Transposition of the great arteries (TGA)

This defect, characterized by atrial-ventricular concordance with ventricular-arterial discordance, is one of the most common cyanotic lesions encountered in the newborn period[2,6]. Because of the existence of two parallel circulations instead of the normal in series circulation, survival depends on intracardiac or extracardiac mixing of the two circulations[6].

Surgical repair of TGA

Reparative surgery is usually done either in the neonatal period or during the first 6 months of life[5]. Palliation with balloon atrial septostomy can be used to stabilize hemodynamically compromised infants before corrective surgery can be performed[3]. Two reparative procedures exist: the atrial switch operation (Mustard or Senning) developed in the 1950's and 1960's replaced nowadays by the arterial switch repair adopted in the

1980's[24]. However in the current adult population with repaired TGA, the atrial switch is still the most encountered type of procedure[3].

A) Atrial switch repair:

The atrial switch repair is an example of physiologic repair whereby the parallel circulation is transformed into a circulation in series[1]. Blood is redirected at the atrial level using either baffles made of Dacron or pericardium (Mustard procedure) or atrial flaps (Senning procedure)[3]. As a result, systemic venous return is diverted through the mitral valve to the LV and out to the PA whereas the pulmonary venous return is diverted through the tricuspid valve into the RV and then into the aorta[1,3].

By virtue of this repair, the RV and tricuspid valve are left in series with the aorta and therefore the RV functions as the systemic ventricle supporting the systemic circulation[1,3].

Functional outcome of atrial switch repair

Patients with atrial switch repair have shortened life expectancy compared to the normal population (70-80% survival rate at 20 to30 years' follow-up in one series of patients with Mustard repair[25]).

The long-term prognosis for cardiac function is not good[1]. The RV has a limited capacity to function as the systemic ventricle, therefore progressive deterioration of the RV function occurs with time often with concomitant tricuspid regurgitation[26] (echocardiographic evidence of moderate to severe systemic RV dysfunction in up to 40% of patients on long-term follow-up[3]). Progressive RV dysfunction along with tricuspid regurgitation lead to the development of signs and symptoms of right-sided heart failure[1]. Even asymptomatic patients with Mustard repair were found to have decreased exercise tolerance (less increase in HR and blunted increase in SV with exercise as compared to control subjects)[5]. The importance of exercise challenge probably lies in demonstrating abnormal ongoing RV physiology in otherwise asymptomatic patients.

Arrhythmias also complicate the course of atrial switch repair for TGA. Sinus rhythm is found in only 35% of patients 10 years after surgery[5]. Arrhythmias can be the result of the reparative surgery itself

(septectomy/septostomy of the atrial septum with excision of one or more internodal pathways)[5] or can result from the progressive RV dysfunction resulting in atrial enlargement[1,5]. Significant arrhythmias include atrial flutter (occuring in 20% of patients by 20 years of age[3]) and sinus node dysfunction mainly sick sinus node syndrome warranting occasionally pacemaker insertion[1]. Sudden cardiac death occurs in 10% of patients 8 to 10 years after atrial repair of TGA, possible because of arrhythmias and hemodynamic complications (mainly RV dysfunction)[27].

Other problems associated with atrial repair include baffle obstruction with obstruction of the SVC return being the more common than either IVC obstruction or pulmonary venous obstruction[8]. Pulmonary venous return obstruction can lead to pulmonary edema acutely or pulmonary hypertension chronically[8].

B) Arterial switch repair:

In contrast to the atrial switch, the arterial switch procedure is an anatomic reparative surgery whereby the arterial trunks (aorta and PA) are transected and reanastomosed with the contralateral root and the coronary arteries are excised and reimplanted into the proximal neoaorta[1,3]. The major advantages of this type of repair are the restoration of the LV as the systemic pump and the long-term maintenance of sinus rhythm mainly because there is no manipulation of the atrial septum[3].

Functional outcome of arterial switch repair

Long-term follow-up on patients with arterial switch repair is lacking because most of these patients have not yet reached adulthood[3]. The currently available studies have so far shown good LV function and normal exercise capacity in patients with this type of repair[28,29].

Recognized sequelae of arterial switch repair include supravalvular pulmonary or aortic stenosis at the anastomotic sites (which can be addressed by balloon angioplasty), neoaortic regurgitation as well as coronary occlusion[3]. So far, a 25-30% incidence of neoaortic regurgitation, usually trivial to mild, has been reported[1]. The development of severe regurgitation has been rare[1]. In addition, the significance of coronary ostial lesions found in 3-5% of patients on coronary

angiography is still unclear[1]. Myocardial ischemia secondary to stenosis at the aortocoronary anastomosis has been identified by angiography or wall motion abnormalities on echocardiography[5]. However no myocardial infarct has been reported yet[1]. It remains to be determined by future studies whether neoaortic regurgitation or coronary ostial lesions will predispose patients to significant morbidity or mortality on long-term follow-up.

"Single Ventricle" CHD: the Fontan Physiology

The Fontan procedure is a palliative surgical procedure that directs systemic venous return to the pulmonary arteries without passing through a subpulmonary ventricle[3]. It is usually performed in patients with a "functional single" ventricle such as patients with tricuspid atresia or hypoplastic left heart syndrome[3].

The initial Fontan procedure consisted of an anatomosis between the right atrial appendage and the pulmonary arteries. This type of repair was associated with right atrial dilatation with secondary arrhythmias and thrombosis. Nowadays, several modifications of this procedure exist. These include the modified Fontan procedure, fenestrated Fontan and cavopulmonary anastomosis (a.k.a. "partial Fontan procedures")[8]

1) Modified Fontan

When the total systemic return (of both SVC and IVC) is directed to the pulmonary arteries, the procedure is called modified Fontan procedure[8]. In the modified Fontan, IVC blood flow is directed to the pulmonary arteries via either a lateral tunnel or an extracardiac conduit[30]. The SVC itself is directly connected to the right and left pulmonary arteries. This surgical arrangement allows all the systemic venous return to passively enter the pulmonary circulation[1,8]. With such a circuit, cardiac output becomes preload limited. Blood returning to the single ventricle depends on maintaining a pressure gradient between the systemic venous return, the pulmonary vasculature and the single ventricle[1]. Patients with a modified Fontan circuit have higher than

normal RA pressure and systemic venous pressure both having an adverse effect on CO[8].

Both the lateral tunnel circuit and the extracardiac conduit have been used to achieve a modified Fontan circuit. The lateral tunnel circuit was introduced in the mid 80's and consists of connecting the IVC to the PA using a prosthetic baffle and a portion of the lateral atrial wall[30]. The main advantage of this circuit is that it has the potential to grow with the child therefore it can be created in children as young as 1 year[30]. Another important improvement compared to the classic Fontan is that this connection allows minimal amount of atrial tissue exposed to high pressure therefore decreasing the risk of future arrhythmias[8,30].

The extracardiac conduit was introduced in the 1990's and consists of a tube graft connecting the IVC to the PA. This circuit allows minimal suturing of the atrium and leaves the atrium at low pressure minimizing the risk of arrhythmias. The disadvantage of this circuit is that it does not have the potential to grow so it can only be offered in older patients[30].

2) Fenestrated Fontan

The fenestrated Fontan consists of the same design as the modified Fontan, however a punch hole is placed in the tube graft connecting the IVC and SVC. The punch hole is supposed to create a right-to-left shunt allowing approximately 20% of the venous return to cross from the RA to the LA thereby increasing CO[8].

3) "Partial" Fontan (cavopulmonary anastomosis)

In the "partial Fontan" technique, the SVC alone is anastomosed to the pulmonary arteries while the blood flow from the IVC enters the physiologic LA and mixes with pulmonary venous blood (thus creating a physiologic left-to-right shunt)[8]. The classic Glenn shunt is a cavopulmonary anastomosis connecting the SVC to the right PA[8]. The left PA is left separated from the right PA and from the SVC, therefore systemic venous return is directly only to the right lung[3,8]. A more preferred form of cavopulmonary anastomosis is the bidirectional Glenn shunt whereby the SVC is connected to both right and left PA's[3,8].

In both fenestrated Fontan and classic/bidirectional Glenn shunt, cardiac output is increased (secondary to left-to-right shunting) at the expense decreasing systemic oxygen saturation. By allowing part of the systemic venous return to bypass the pulmonary circulation both of these circuits result in lower levels of venous congestion. The advantages of the bidirectional Glenn shunt over the fenestrated Fontan consist in achieving lower IVC pressure (because the entire IVC blood return is dumped into the LA) with a larger increase in CO. This is usually at the expense of a greater degree of arterial desaturation and cyanosis.

In any type of "single ventricle" defect, performing a Fontan circulation at birth is not feasible[30]. This is because the pulmonary vascular resistance is still increased for several weeks after birth. Furthermore caval veins and pulmonary arteries are usually too small for cavopulmonary anastomosis[30]. Therefore, a staged approach is usually undertaken allowing the body to adapt progressively to the altered hemodynamic loads. Initially in the neonatal period, management is mainly focused on providing both adequate CO and well balanced yet limited blood flow to the pulmonary vasculature (via PA banding, BT shunt etc.). Once the pulmonary vasculature is reasonable developed (usually after age of 3 months), the cavopulmonary connection (mainly bidirectional Glenn shunt) is introduced[30]. Because of the deleterious effects of long-term arterial desaturation and secondary cyanosis, the cavopulmonary anastomosis is converted at around 1 year of age to a modified Fontan circuit. A fenestrated Fontan can be performed as an intermediate stage before achieving a modified Fontan circuit[30].

Physiology of Fontan circulation

In all of its forms, the Fontan circulation operates by passive flow of the systemic venous return to the pulmonary vasculature and then to the single ventricle. As such, PBF and CO are the result of the pressure differential existing between the "upstream" component of the circuit (consisting of the caval veins and the PA) and the "downstream" component (the pulmonary veins/atrium/single ventricle system)[1]. The systemic venous pressure of an ideal Fontan circulation is approximately

10-15 mmHg and the pulmonary venous atrial (functional left atrium) pressure is approximately 5-10 mmHg; this allows a transpulmonary gradient driving pressure of 5-8 mmHg[1,8]. Anything that affects this gradient might compromise the single ventricle filling and subsequently the CO[1].

Adequate flow in the upstream component of the circuit depends on 1) an unobstructed venous return from IVC and SVC, 2) adequate systemic venous return (i.e. preload), 3) patent anastomotic connections between the caval veins and pulmonary arteries, 4) low intrathoracic pressure[1].

At the level of the lungs and pulmonary circulation, Fontan physiology requires low PAP (<15-20 mmHg), low PVR, unobstructed pulmonary arterial/pulmonary venous flow and normal lung parenchyma[1,8]. At the level of the single ventricle, CO is maintained by adequate ventricular filling, normal AV valve, adequate diastolic and systolic function and normal sinus rhythm[1]. Small alterations in ventricular function (particularly diastolic function), circuit efficiency (e.g. increased PVR secondary to pneumonia) or the onset of arrhythmias can lead to decreased CO and symptomatic deterioration[3].

Functional outcome of the Fontan circulation

Arrhythmias: Arrhythmias (atrial flutter/fibirillation, sick sinus syndrome, sinus bradycardia) and heart block occur in more than 20% of patients by 10 years after surgery[1]. Arrhythmias are more common in patients with older versions of the Fontan procedure whereby part of the atrial wall is incorporated into the circuit leading to progressive atrial dilatation and hypertrophy[3,30]. Another possible cause could be the damage to the sinus node (or its arterial supply) associated with extensive atrial manipulation in older versions of the Fontan procedure. Recent data show a lower incidence of arrhythmias in patients with cavopulmonary anastomosis or modified Fontan procedure; however this may be partly due to the shorter length of follow-up.

Atrial flutter/fibrillation carries a significant morbidity and require medical therapy, ablation or at times conversion of the old Fontan circuit

to an extracardiac cavopulmonary anastomosis[1,3,30]. Sinus node dysfunction and complete heart block may require pacemaker insertion[3].

Thromboembolic events: The incidence of thromboembolic complications in the Fontan circuit varies from 6 to 25%[3]. Thrombus formation can be the result of atrial dilatation, supraventricular arrhythmias, systemic venous dilatation with blood stasis, the presence of prosthetic material in the circuit etc.[3,30]. Patients with a Fontan physiology suffer from increased systemic venous pressure with secondary hepatic congestion. As such, they develop coagulation factor abnormalities seen in other types of chronic hepatic congestion namely protein C, protein S and antithrombin III deficiency[30]. Other coagulation factor abnormalities have also been reported less consistently in the medical literature. The result is a "hypercoagulable" state that contributes to the increased incidence of thromboembolic events in Fontan patients (stroke, pulmonary emboli etc.) A particularly detrimental complication would be the development of multiple pulmonary microemboli leading to the progressive onset of obstructive pulmonary vascular disease with subsequent increase in PVR[30]. There are no guidelines concerning how to use anticoagulation in Fontan patients. Patients with a history of arrhythmias, previous thromboembolic event can particularly benefit from anticoagulation[3].

Protein-losing enteropathy: Protein-losing enteropathy (PLE), characterized by loss of serum proteins into the gut, occurs in 4 to 13% of patients after a Fontan procedure[3]. PLE is thought to be the result of chronic state of venous congestion leading to the development of lymphatic telangectasia with leakage of albumin, immunoglobulins, lymphocytes, chylomicrons into the gut[3,30]. The clinical manifestations of PLE include generalized edema, pleural effusion, immunodeficiency, chronic diarrhea etc. The dagnosis of PLE is confirmed by finding an elevated alpha$_1$- antitrypsin stool clearance[3]. The diagnosis of PLE is a poor prognostic factor in Fontan patients[30]. The 5-year survival rate after diagnosis is approximately 46 to 59%[31,32]. Conservative therapy for PLE (high-protein, low-fat diet, albumin infusions, diuretics, afterload reducing agents) has a high failure rate[3,30]. The majority of patients will

require modification of the Fontan circulation (e.g. creation of an atrial fenestration to decrease venous congestion); some patients may even require cardiac transplanatation[30].

Peri- intra- and postoperative considerations in Fontan patients

For patients with a Fontan circulation undergoing noncardiac surgery, certain perioperative considerations should be kept in mind. First, assessment of the functional level of the cardiovascular system cannot be overemphasized. Echocardiography should be used to assess both ventricular function and the patency of circulation[1]. Some patients may even require a more invasive approach (catheterization, radiofrequency ablation)[1].

The use of invasive monitoring depends on the individual patient and the type of surgery to be performed. Central venous cannulation offers the advantage of monitoring venous filling and PAP however it carries an increased risk of venous return impairment and the possibility for thromboembolic complications.

Intraoperatively, the main target would be to focus on maintaining adequate CO. Patients with a Fontan circulation have decreased compensatory mechanisms and even a slight compromise in CO can be disastrous. Effort should be made to ensure adequate preload, good ventricular filling and contractility while avoiding an increase in afterload[1]. Because of the passive nature of blood flow from the systemic veins to the pulmonary circulation, any increase in PVR can compromise ventricular filling and CO.

Therefore, it is very important to avoid any increase in PVR that might be precipitated by hypoxia, hypercarbia, acidosis, excessive mean airway pressure, compression of the lung by pleural effusion etc[8]. Positive-pressure ventilation at relatively high lung volume can increase PVR. Positive-pressure ventilation can compromise the pulmonary blood flow by increasing PA pressure mainly by transmission of increased intrathoracic pressure but also by compression of pulmonary vasculature secondary to alveolar distention[1].

A postoperative complication of surgery, usually occurring in older

Fontan patients, is end-organ dysfunction (namely liver and kidneys)[1]. Organ perfusion, already compromised because of limited CO, can further deteriorate after surgery (possibly secondary to anesthesics or intraoperaive blood loss)[1]. The risk of thromboembolic complication after surgery can be decreased by using subcutaneous heparin or low molecular weight heparin along with adequate hydration and early ambulation[1].

Conclusion

The increase in the population of adults with repaired congenital heart disease over the past years has put forth new issues pertaining to the problems and complications ongoing after reparative surgery. This review, although by no means comprehensive, deals with some of the common ongoing problems in these patients that need to be addressed before any type of major elective surgery.

Abbreviations and acronyms

PDA: Patent ductus arteriosus

ASD: Atrial septal defect

VSD: Ventricular septal defect

TOF: Teralogy of Fallot

TGA: Transposition of the great arteries

PVR: Pulmonary vascular resistance

SVR: Systemic vascular resistance

RA: Right atrium/atrial

RV: Right ventricle/ventricular

LA: Left atrium/atrial

LV: Left ventricle/ventricular

AV: Atrioventricular

PBF: Pulmonary blood flow

HTN: Hypertension

PHTN: Pulmonary hypertension

PA: Pulmonary artery/arterial

PV: Pulmonary vein/venous

PAP: Pulmonary artery pressure

CBP: Cardiopulmonary bypass

LVOT/RVOT: Left/right ventricular outflow tract

BP: Blood pressure

TEE: Transesophageal echocardiography

SVC: Superior vena cava

IVC: Inferior vena cava

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