Perioperative Issues in Patients with Congenital Heart Disease

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At the conclusion of this lecture, participants will be able to 1) identify key pathophysiologic issues in patients who have undergone common reparative procedures for congenital heart disease and 2) devise appropriate perioperative management plans based on this knowledge of long-term outcome issues in this population.

Scope of the Problem

The population of children and adults who have undergone successful palliation of various forms of congenital heart disease (CHD) continues to increase and live longer. Reasons are multiple, including expanded indications, early, more definitive surgery and other interventional techniques, and improved surgical, interventional, catheterization, electrophysiological, and intensive care management. The numbers are not entirely clear and based upon certain assumptions, but current estimates suggest that there are at least 1.2 million adults (>16 yr) living with various forms of CHD (ACHD) in the United States; of these, perhaps as many as 80,000–100,000 have complex forms (defined as lesions *other than* repaired secundum or sinus venosus atrial septal defect [ASD] without residuae [including pulmonary hypertension or dysrhythmias], isolated patent foramen ovale [PFO] or small ASD, mitral valve prolapse, mild pulmonic stenosis, ligated or occluded patent ductus arteriosus [PDA], congenital aortic valve disease [isolated, mild gradient, without aortic root dilation, without prior intervention], isolated restrictive or repaired ventricular septal defect [VSD]) (1–7).

Current birth, incidence, and survival rates predict an annual increase of 10,000 to 30,000 patients/yr in the CHD population, including a doubling in the survival of patients with complex physiology. As life expectancy has improved and the risk of cardiac operation(s) has decreased, noncardiac medical and surgical issues typical of adult/aging populations have become more prominent (e.g., pregnancy, scoliosis and joint surgery). For example, data from at large ACHD centers (Mayo Clinic, Toronto, and London) show that as many as 50% of these patients with complex physiology are 40 yr of age or older and that overall ~60%–70% can be categorized as medium or high risk (significant risk for premature death, reoperation, ventricular dysfunction, severe dysrhythmias, and other complications).

These data have led to broad-reaching and somewhat controversial recommendations about who and where should provide care for ACHD patients (1–7). In summary, these recommendations include: 1) complex ACHD patients should be followed in, and have all of their care coordinated by, *regional/supraregional centers* that have multi-disciplinary ACHD programs and staff; such centers would be expected to serve ~5–10 million in population; 2) every adult cardiology/cardiac surgery program and medical cardiologist should affiliate with a specialized, dedicated ACHD program; 3) every pediatric cardiology program should identify a specialized, dedicated ACHD program for the transition of their patients; 4) patients with moderate or severe ACHD should be transferred to a dedicated ACHD program for urgent or acute care, most cardiac catheterization and electrophysiology procedures, and both cardiac and noncardiac surgery. Facilities and expertise required at the regional/supraregional level include the following: 1) several/group ACHD specialists; 2) pediatric cardiologists; 3) medical cardiologists and internists; 4) congenital cardiac surgeon(s) [each with >100 cases/yr]; 5) cardiac anesthesiologists*; 6) echocardiography, including transesophageal echocardiography (TEE) and intraoperative TEE*; 7) diagnostic and non-coronary interventional catheterization*; 8) electrophysiology* including pacemakers, implantable cardiac defibrillators (ICDs), ventricular tachycardia/ventricular fibrillation (VT/VF) management, electrophysiology (EP) surgery, complex ablations for atrial dysrhythmias and ventricular tachycardia (VT) 9) exercise testing; 10) cardiac, lung, heart-lung transplantation; 11) nuclear medicine, cardiac computed tomography (CT)*, cardiac magnetic resonance imaging (MRI)*; 12) cardiac pathology; and 13) high-risk obstetrics, genetics, rehab services, social services, database (*listed modalities marked with an asterisk must be supervised and performed by physicians with specific skills and knowledge in CHD). Of particular note, the guidelines for anesthesia include the presence of a dedicated cardiac

anesthesia team with expertise in the management of CHD and ACHD that performs consultative services, interacts at all levels with other members of the ACHD team, and anesthetizes patients with CHD.

In summary, the face of the problem has changed with the last 5–10 yr (8–36). This change has been driven by early, more definitive (less palliative) surgery, reduced mortality, increasing survival into adulthood with the possibility of improved functional outcomes, and hence a shift of focus to morbidity, functional status, quality of life issues, and resource utilization. The remainder of this talk will attempt to summarize key pathophysiologic issues that arise in patients with various forms of congenital heart disease.

Who Is Actually Repaired?

Very few types of congenital heart disease are truly "fixed," if one defines "fixed" as having a high likelihood of minimal or no significant residual problems or sequelae. Lesions in this category may include PDA ligation, uncomplicated repair of secundum ASD (within the first decade), and uncomplicated repair of isolated VSD (within the first couple of years of life). Virtually all other forms carry substantial risk of residual and potentially progressive structural, contractile, hemodynamic, electrophysiologic, and/or end-organ abnormalities. As will hopefully be evident from the ensuing discussion, knowing the outcome and problems with specific lesions and repairs is the basis for formulating rational perioperative management strategies.

General Pathophysiological Themes

Overall, the chronic presence and/or potential to develop or exacerbate *low systemic cardiac output* is probably the single most important consideration in approaching the care of these patients; this is especially true in the perioperative setting. The ways for this to occur are multiple and, to some extent, lesion-specific. There are a number of related themes associated with various forms of congenital disease that are likely to carry increased perioperative risk. Some prominent ones include:

1) *progressive contractile dysfunction*—risk factors include chronic pressure and/or volume loads, chronic arrhythmias (which can also be the result of chronic abnormal loading, e.g., tetralogy of Fallot [TOF]), repeated cardiac operations, cyanosis (?), various congenital and acquired cardiomyopathies, etc. Examples include TOF (right ventricular [RV] pressure and/or volume loading after "complete" repair), truncus arteriosus (truncal valve regurgitation, recurrent RV pressure loading due to conduit stenosis and/or distal pulmonary artery [PA] stenoses), atrioventricular (AV) canal repair (AV valve insufficiency), and aortic valve abnormalities. Progressive dysfunction of the RV when it is functioning as the systemic ventricle (e.g., hypoplastic left heart syndrome, Mustard or Senning correction of transposition) should also be expected over time.

2) *the pressure- and/or volume-loaded ventricle*—even when contractile function is preserved, abnormal hemodynamic loading is likely to complicate management and substantially increase perioperative risk. The major abnormality associated with pressure or volume loading is increased demand resulting from increased wall tension; inadequate coronary blood flow, abnormal control of coronary vasomotor tone, and numerous abnormalities associated with the molecular program associated with hypertrophy are also involved. Overall, these changes make the abnormally loaded ventricle much more sensitive to altered supply-demand relationships and hence less tolerant to common perioperative events such as tachycardia, hypotension, and anemia (and especially combinations thereof).

3) *pulmonary hypertension*—increased PA pressures can occur on a mechanical basis (e.g., PA stenoses in TOF or at the site of a Blalock-Taussig shunt insertion) or as a result of idiopathic or acquired pulmonary vascular disease (the latter most often a result of large left-to-right shunts or other lesions associated with left atrial hypertension, such as severe cardiomyopathies and mitral stenosis lesions). All types chronically pressure load the RV (as above). Types associated with pulmonary vascular disease can be reactive and hence potentially exacerbated by stresses such as intubation, surgical stress and stimulation, pain, hypoxemia, and acidosis. It is thus important to know the site(s), mechanism(s), degree, reactivity, and response to pulmonary vasodilators in these patients.

4) *end-organ dysfunction*—this is a poorly defined area at present, but is becoming increasingly important as patients survive longer with various palliated forms of CHD. It appears that chronic cyanosis, low systemic cardiac output, and/or high venous pressures (e.g., Fontan physiology) may contribute over time to the development of liver and renal insufficiency (potentially central nervous system dysfunction as well). This chronic underlying injury (which may not be revealed by routine clinical laboratory tests such as blood urea nitrogen/creatinine ratio or liver function tests) may predispose some patients to severe acute perioperative dysfunction in response to relatively minor changes in organ perfusion and oxygen delivery resulting from surgical manipulation, volume shifts, or drugs (e.g., volatile anesthetics, nonsteroidal antiinflammatory drugs).

In summary, most lesions are palliated rather than completely repaired. Palliated lesions frequently continue to have some aspect of abnormal hemodynamics

and circulation, along with other consequences such as CHF, hypoxemia, cyanosis, polycythemia, and pulmonary vascular disease. Lesions that may be classified as "successfully palliated" or "corrected" can still have significant residual problems that can either be static or, more likely, lead to progressive dysfunction over time; these include ventricular dysfunction, residual shunts, residual obstructive and/or regurgitant lesions, arrhythmias, and pulmonary hypertension.

Preoperative Optimization/Effective Use of the Consultant Cardiologist

I personally believe that "clearance" from a pediatric or adult cardiologist is not a useful concept. The anesthesiologist caring for these patients for noncardiac surgery needs to have a comprehensive understanding of the pathophysiology of the lesion, the type and natural history of its repair, and the likely interactions of all of these with the planned procedure. He/she needs to be able to use this knowledge to assess existing information, order and interpret new tests, and initiate and coordinate operative and perioperative planning. Few cardiologists have sufficient understanding of what is actually involved surgically or anesthetically (e.g., extent of trauma or blood loss during spinal fusion, CO_2 insufflation and its potential consequences during laparoscopic surgery, ventilation issues during airway surgery); they need to be gently educated in these issues to provide the best assessment and advice.

The cardiologist can be used to better define pathophysiologic issues specific to the particular lesion and in that particular patient, but again, one needs to have a pretty good idea of what questions to ask for this to be most effective, and one may also need to define the surgical and perioperative risks to justify additional investigations. The cardiologist can and should be used to 1) help assess the likelihood, severity, and complications of issues such as CHF, pulmonary overcirculation, valvular regurgitation and obstruction, contractile dysfunction, potential dysrhythmias, in addition to the overall functional status of the patient; 2) recommend and help evaluate tests to better clarify these issues; 3) recommend medical interventions to better optimize patient condition (e.g., increased diuresis or other CHF therapies, dobutamine or milrinone "tune-up" [dilated cardiomyopathy], anti-arrhythmic therapies, response to pulmonary vasodilators [e.g., inhaled nitric oxide], etc.).

We have found the diagnostic and interventional catheterization laboratory to be particularly useful for improving patient status before major procedures. Some examples of recent interventions that were undertaken based on preoperative consultation and planning by the cardiac anesthesiologist (i.e., driven by anesthesiology consultation rather than initial cardiology input) include the following: 1) RV outflow tract (RVOT) dilation, PA dilation, stenting of PA stenoses (in patients with TOF or truncus arteriosus/PA conduit) to reduce RV loading, decrease RA pressure/tricuspid regurgitation (TR), improve systemic cardiac output; 2) Coil embolization of venous or arterial collaterals (Glenn, Fontan, TOF/PA) to reduce cyanosis or systemic ventricular volume load; 3) Fenestration closure (Fontan) to decrease risk to paradoxical embolus (spinal fusion); 4) Radiofrequency catheter ablations; 5) Temporary pacemaker insertion for sinus bradycardia (Fontan); 6) Lung scans to assess distribution of pulmonary blood flow (TOF, TOF/PA) before thoracotomy and potential one-lung ventilation; 7) Dobutamine "tune-up" for dilated cardiomyopathy; 8) Assessment of pulmonary hypertension and response to inhaled nitric oxide, nifedipine, or prostacyclin; 9) DNAase, nebulized antibiotics, nutrition (Kartagener's); and 10) cardiac magnetic resonance imaging (MRI) to assess RV function and quantify amount of PR (TOF; recommended for PV replacement before extensive elective surgery).

Outcomes of Common CHD Lesions and Repairs

As noted above, important residua and sequelae should be expected with most types of CHD and CHD repairs. Overall, surgical corrections can be classified as "physiologic," where the circulation is in series, cyanosis is corrected, but the result is a single ventricle repair (e.g., Fontan for tricuspid atresia or hypoplastic left heart syndrome [HLHS]) or the RV is functioning as the single ventricle (e.g., Mustard or Senning procedure for transposition). Virtually all physiologic repairs have significant long-term complications. In contrast, an anatomic repair not only corrects any cyanosis and renders the circulation in series but also has the RV and LV as pulmonary and systemic ventricle, respectively. Long-term complications are less likely after anatomic repair if the heart is structurally normal and the procedure was undertaken in a timely fashion with a structurally and functionally successful outcome (e.g., PDA, ASD, VSD). On the other hand, late and progressive sequelae are likely if a complex procedure (e.g., baffle, conduit, outflow tract reconstruction, arterial or AV valve repair) was required.

Aortic Coarctation

Problems in these patients include residual or recurrent stenosis at the coarct site, which can frequently be addressed by balloon dilation in the catheterization

laboratory. Persistent systemic hypertension (independent of any residual obstruction and at times difficult to manage) and LV hypertrophy can be found in approximately 25%–33% of repaired coarct patients. The incidence is higher in patients repaired later in childhood. An increased risk of sudden death is also present in this subset of coarct patients.

Atrial Septal Defect

As noted previously, most successful ASD repairs carry little risk of late complications. However, there is a modest but increasing risk of the development of pulmonary hypertension, beginning around the second or third decade of life, if the defect is closed late. Persistent atrial arrhythmias (flutter and fibrillation) are also more likely if closure is delayed until after 10–12 yr of age.

Ventricular Septal Defect

If the VSD is repaired early (first few months to the first year or two of life), myocardial and pulmonary function are likely to be normal subsequently, assuming that there are no residual VSDs of significance ($Qp/Qs < 1.5$), and no outflow obstruction, subaortic membrane, or heart block. However, in a small number of patients, LV dysfunction and/or pulmonary hypertension may be late problems. These are more likely after repair of a large defect, especially if closure was delayed to later in childhood.

Atrioventricular Canal Defects

These patients have large left-to-right shunts, excessive pulmonary blood flow, and hence CHF and are at risk for pulmonary hypertension. The repair of the most frequent types involves dividing the common AV valve and closing the ASD and VSD with either one or two patches. Frequently, the mitral valve (and sometimes the tricuspid valve) requires additional approximation and suspension of the divided valve apparatus. The most common problems after AV canal repair include valvular regurgitation (especially mitral), residual VSD, and occasionally pulmonary hypertension; the last is more likely Down syndrome patients.

Tetralogy of Fallot

TOF is perhaps the classic example of a lesion that is "fixed" but not cured. The majority of problems relate to abnormal RV loading (both pressure and volume) and to problems associated with RVOT reconstruction. Many of these issues are shared by other lesions that require RVOT reconstruction or the placement of an RV-PA conduit (that can subsequently develop obstruction), such as truncus arteriosus, pulmonary atresia, and the Rastelli procedure for transposition of the great arteries with pulmonary stenosis. Progressive RV dysfunction, along with the development of ventricular arrhythmias and increased risk of sudden death, are the major problems after TOF repair. Factors that have been associated with these problems and reduced long-term survival include older age (>4 yr) at repair, initial palliative shunting procedures (especially central shunts), and significant residual RV hypertension (RV:LV pressure ratio >0.5–0.75; often as the result of residual RVOT obstruction or distal PA stenoses) and/or volume loading of the RV (e.g., PR after attempted relief of RVOT obstruction with a transannular patch). Overall, recent long-term outcome data indicate that both pressure and volume loading of the RV are poorly tolerated over time.

Progressive systolic RV dysfunction can occur as a maladaptation to chronic pressure overload alone, although it is probably more likely when combined with volume load because of pulmonary regurgitation. It is a predictor of late morbidity and mortality. It may be manifest as clinically decreased exercise tolerance and signs of right-sided congestion, or may only be detected by exercise testing (decreased maximal aerobic capacity and endurance), or stress echocardiographic or radionuclide techniques (revealing decreased RV ejection fraction). Cardiac MRI has become particularly useful to quantify RV systolic function, RV volume, the degree of pulmonary regurgitation (PR) (not readily quantified by any other technique), and image potential sites of RVOT obstruction. We have also found the presence of significant TR to be a likely surrogate for the presence of substantial RV dysfunction.

The incidence of ventricular arrhythmias on ambulatory Holter or exercise testing is significant and increases with age; however, the exact prognostic significance is not known (these findings have not predicted sudden death in most series, although they are almost certainly linked to RV dysfunction). The ability to induce VT during programmed electrophysiologic stimulation, particularly in a symptomatic (palpitations, syncope) patient, is believed at the present time to be significant and an indication for ablation, antiarrhythmic agents, or an implantable cardioversion-defibrillator (ICD). Finally, a subset of TOF patients has what has been termed "restrictive" RV physiology on the basis of diastolic noncompliance. These patients are less likely to develop RV dilation and cardiomegaly, have less pulmonary regurgitation, function at a *higher* RV end-diastolic pressure, and are more likely to maintain exercise capacity and manifest a lower risk for ventricular arrhythmias.

As with all of these situations, it is difficult to accurately predict patients who will respond poorly to anesthesia and surgery. There is very little objective

information or study in these patients. It is therefore impossible to prescribe an algorithm or recipe for every patient. However, some concepts and approaches would seem to be valid in these patients. First, defining their risk factors for and their degree of RV dysfunction, as outlined above, is essential. Consideration should be given to interventional catheterization for significant lesions that appear to be amenable to improvement (e.g., RVOT or PA obstruction, residual VSD, collaterals causing LV volume loading, ventricular arrhythmias). The potential for positive pressure ventilation (which mechanically increases RV afterload and decreases RV filling by increasing intrathoracic pressure) to decrease cardiac output in patients with significant PR and RV dysfunction should be recognized, as should the ability of the acutely dysfunctional RV to compromise systemic cardiac output (via decreased RV output, as well as ventricular interdependence causing decreased LV filling and function). Thus, initial considerations are directed at optimizing and maintaining RV function. Factors that increase pulmonary vascular resistance should be avoided, especially in the setting of free PR and RV dilation. RV filling should be maintained, with the understanding that excessive volume loading may also be poorly tolerated. Drugs that significantly diminish RV contractility should be avoided if contractile dysfunction is a prominent feature. Factors that are detrimental to the RV myocardial supply-demand relationship need to be optimized; this is true of all types of patients but may need to be specifically pointed out in the patients with restrictive physiology, as their stiff, noncompliant RVs may be particularly susceptible to reductions in subendocardial oxygen delivery. From a practical standpoint, this means maintaining contractility and filling volumes (while attempting to avoid overdistention), keeping heart rate approximately normal, and maintaining RV oxygen delivery by maintaining blood pressure and oxygen carrying capacity (e.g., the combination of tachycardia, hypotension, acidosis, and anemia is particularly detrimental). Based upon the severity of preexisting dysfunction and the magnitude of the planned procedure, one should have a low threshold for invasive monitoring and postoperative care in an ICU setting. "Prophylactic" administration of inotropes to improve RV contractile performance should be considered.

Transposition of the Great Arteries

There are two distinct methods to correct transposition of the great arteries (TOGA): an atrial switch procedure (Mustard or Senning operation) or an arterial switch. In the former, a fairly complex atrial level baffle redirects pulmonary venous return across the atrium to the tricuspid valve (and hence to the RV and the aorta); another aspect of the atrial baffle carries systemic venous return across to the mitral valve (and hence to the LV and out the PA). Again, this is a prime example of a physiologic repair, where the circulation is established in series and cyanosis is removed; however, the RV and tricuspid valve are left in series with the aorta for life, and thus must work at systemic pressure and against systemic levels of afterload. The arterial level switch operation was pursued and perfected in large part a result of the long-term and late complications of atrial baffle procedures. Usually performed in the neonatal period, the great arteries are divided distal to their respective valves and reattached to the opposite, anatomically correct ventricle; in addition, the coronaries must be excised and reimplanted into the proximal neoaorta (formerly the pulmonary root).

Functional Outcome of the Atrial Switch

These patients have their RV as the systemic ventricle in a 2-ventricle physiologic repair. Many of these patients will self-report a reasonable functional status and are able to lead fairly normal lives into their third and fourth decade; the 15- to 20-yr survival may approach 80%–85%. However, the long-term prognosis for cardiac function is not good, and progressive deterioration of RV function, development of TR (the systemic AV valve), and signs and symptoms of right heart failure, arrhythmias, and sudden death are likely. In patients both with and without significant functional complaints, exercise testing demonstrates moderate to severe limitations in RV function and exercise response and hence overall exercise and aerobic capacity and peak heart rate and blood pressure responses in 50%–75% of adult patients. In addition to ventricular arrhythmias, these patients are likely (50%–60% non-sinus rhythm by 10–20 yr after the repair) to develop significant atrial tachyarrhythmias as well as sick sinus syndrome as they age; these may be preceded by the development of RV dysfunction or initially occur as an independent finding (presumably attributable to the extensive atrial suture lines and atrial distention). Pacemaker insertion may be indicated for sick sinus syndrome and as an adjunct to aggressive antiarrhythmic drug therapy. Radiofrequency ablation may be useful, although the success rate is less than with many other lesions (e.g., Wolff-Parkinson-White [WPW] syndrome), as the anatomy and mechanisms of these arrhythmias are often complex and multiple. Problems with the atrial baffle can be present or develop. Baffle leaks can result in intraatrial shunting and hypoxemia. Baffle obstruction of the systemic venous return can cause superior vena cava syndrome, hepatic congestion, ascites, and peripheral edema. Protein-losing enteropathy (PLE) can

occur occasionally in these patients. PLE is defined as an albumin level <3 mg/dL in the absence of liver or renal disease; other features include ascites, peripheral edema, abdominal pain, diarrhea, and lymphopenia. Hemodynamic features in many PLE patients include decreased RV function, decreased cardiac index, and increased systemic venous pressure (although intuitively attractive, it is not clearly always related to the degree of systemic venous hypertension). Obstruction of the pulmonary venous side of the baffle can result in pulmonary edema (if severe) or the development of pulmonary hypertension. End-organ dysfunction appears to be more likely the result of chronic low output and the other issues.

Functional Outcome of the Arterial Switch Procedure

In centers with extensive experience, the early hospital mortality is <3% (and <1%–2% in many), and actuarial analyses indicate 5–10 survivals >97%–98%. It is these results and the numerous problems and poor long-term outcome with the atrial switch operations described previously that have made the arterial switch operation the preferred procedure in most centers. Longer-term follow-up is just beginning to accrue, but intermediate results (teenagers) suggest that the risk for complications after the arterial switch is fairly small. Various echocardiographic, catheterization, and exercise data indicate clinical, hemodynamic, and functional performance indistinguishable from age-matched controls. Arrhythmias appear to be uncommon. There is a small incidence of supravalvular pulmonary arterial and aortic stenoses at the anastomotic sites; these are less common in the current era because of improved surgical techniques and can frequently be addressed by balloon dilation. However, there is the suggestion of at least two longer-term complications that will bear watching as this group of patients ages: 1) neoaortic valve (the anatomic pulmonary valve) regurgitation, and 2) coronary ostial lesions. At present, a 25%–30% incidence of neoaortic valve regurgitation, usually trivial to mild, has been reported; the development so far of severe regurgitation has been rare. On the other hand, experience with the Ross procedure (which autografts the pulmonary root into the aortic position) might suggest that progressive incompetency of the pulmonary valve in the aortic position could be a problem in the long term. The significance of the coronary ostial lesions, occurring in 3%–5% of patients based on coronary angiographic findings, is also unclear at present. There has been no evidence of infarction, and Holter, exercise testing, and echocardiography rarely show evidence of myocardial ischemia. However, it is possible that the coronary reimplantation establishes abnormal regional flow patterns or other responses that promote the development of ostial stenosis; for now, I believe that one should have an increased index of suspicion for the possibility of coronary stenosis, especially as these patients age.

Care of the Patient with Fontan Physiology

The Fontan operation (in all of its iterations) passively routes systemic venous return to the pulmonary arteries. Pulmonary blood flow and cardiac output are the result of the pressure differential between systemic venous return and the pulmonary artery (the "upstream" driving pressure) and the "downstream" pulmonary venous atrium/systemic ventricle. In the "ideal" Fontan circulation, the systemic venous or baffle pressure is approximately 10–15 mm Hg, and the pulmonary venous atrial (the functional left atrium) pressure is approximately 5–10 mm Hg; this leads to a transpulmonary gradient, or TPG, driving pressure of 5–8 mm Hg. At the most upstream point, optimal Fontan physiology depends on unobstructed venous return, adequate preload, patent anastomotic connections, and low intrathoracic pressure. At the level of the lungs and pulmonary circulation, it requires low mean pulmonary artery pressure (<15–20 mm Hg), low PVR (ideally <2 Wood units), unobstructed pulmonary arteries, normal lung parenchyma and alveolar ventilation, and minimal or no pulmonary vascular disease or pulmonary venous obstruction. At the level of the systemic ventricle, good Fontan function depends on sinus rhythm (primarily to maintain ventricular function and cardiac output), a competent AV valve, normal systolic and diastolic function, and no outflow obstruction. Any significant departure from these requirements can result in severe compromise. For example, ventricular dysfunction leading to an EDP of 15 mm Hg mandates a venous pressure of 20–25 mm Hg to achieve comparable pulmonary blood flow, ventricular filling, and cardiac output. Alternatively, an infectious pneumonitis that increases PVR will acutely decrease cardiac output (as the result of decreased filling of the ventricle) and subsequently require a significantly increase in the TPG to restore ventricular filling and cardiac output toward normal.

The use of mechanical ventilation in Fontan patients derives from similar considerations. There is little doubt that the majority of pulmonary blood flow in spontaneously ventilating Fontan patients occurs during inspiration and that it (and cardiac output) can be further augmented during negative pressure (iron lung) ventilation; in contrast, one can demonstrate either no flow or even a reversal of flow in the pulmonary arteries of Fontan patients during the administration of a positive pressure breath. Positive pressure ventilation at relatively high lung volumes can

directly increase PA pressures via transmission of increased intrathoracic pressure, and also by airway and alveolar distention (which compress adjacent blood vessels); excessive lung stretch may also induce a myogenic contractile response in pulmonary arterioles. On the other hand, hypoventilation (as might be expected to occur during anesthesia and also due to procedures associated with abdominal or thoracic compression) increases PVR via alveolar hypoxia, hypercarbia, and atelectasis; surgical stress responses may also increase PVR. The effects of positive pressure ventilation to decrease afterload on the systemic ventricle (via the Law of LaPlace) and thereby improve cardiac output, particularly of the dysfunctional ventricle, also need to be considered. Therefore, for most surgical situations, I believe that the sum of effects favors judicious use of positive pressure ventilation to provide an effective anesthetic while maintaining lung volumes and normal gas exchange. We would typically start at a somewhat greater than normal tidal volume and a reduced respiratory rate. Similar considerations apply to the use of positive end-expiratory pressure (PEEP)—overdistention is probably deleterious, whereas maintaining lung volume, gas exchange, and FRC is almost certainly beneficial to this circulation.

Although most patients report normal functional status with moderate exercise tolerance, this is not borne out by more objective measures. Systemic ventricular dysfunction, perhaps more likely with a systemic *right* ventricle (e.g., HLHS), is not uncommon. With longer term follow-up, progressive decline in New York Heart Association class, as well as decreased exercise tolerance, endurance, maximal aerobic capacity, heart rate response, and delayed recovery from peak exercise have all been described; overall as a group, their exercise reserve capacity is 50% or less that of age-matched control subjects. This inability to significantly increase pulmonary blood flow and cardiac output in response to exercise stress is attributable to several factors, including limited ventricular function reserve, inability to increase heart rate (sinus node dysfunction and other conduction abnormalities), and the inability to achieve preload augmentation; the last of these would appear to be the result of Fontan patients having high resting venous (and arterial) tone. Fontan patients are thus very dependent on the status of the pulmonary vascular bed for ventricular filling; the limited contractile and preload reserves increases dependency on heart rate to increase cardiac output. The implications of these findings for anesthesia and surgery have not been formally evaluated but are probably significant in light of the potential for anesthesia and surgery to be associated with increased PVR (hypoxia, hypercarbia, acidosis, stress hormones), myocardial depression (e.g., inhaled anesthetics), vasodilation (causing a critical decrease in preload), and hypovolemia (blood loss). There are other important complications and limitations in these patients. PLE may occur in 3%–15% of Fontan patients. There is an increased incidence of thromboembolism in Fontan patients, with reports of up to 20%–30% having one or more such events during the course of their life and a stroke rate of 2%–3%. Numerous abnormalities of procoagulant and anticoagulant factors have been measured, including increased factor VIII and decreased protein C and S; however, these are often accompanied by reduced levels of procoagulant factors, and it is unclear whether the measured abnormalities have any causal relationship to the observed incidence of thromboembolism. Other risk factors in these patients include increased venous pressure and stasis of flow in the right atrium or through the right atrial baffle, atrial dysrhythmias, and perhaps increased resting venous tone. The routine use of long-term anticoagulation remains controversial. Perioperatively, bleeding may be more significant in Fontan patients because of the presence of collaterals and high venous pressures, in addition to any deficiencies in procoagulant factors.

Arrhythmias such as atrial flutter, sick sinus syndrome, sinus bradycardia, and heart block occur in more than 20% of Fontan patients by 10 yr after surgery. By 15–20 yr, especially with older versions of the operation (extensive atrial suture lines, damaged SA node artery, chronic atrial hypertension) and in patients who were older at the time of Fontan operation, the probability of being free from atrial arrhythmias is $<$40%–50%. Recent data on the lateral tunnel version (although really only approximately 10 yr out for most of the patients) of the operation suggest that the incidence of atrial flutter and other tachyarrhythmias, as well as severe ventricular dysfunction, is substantially lower; interestingly, the incidence of sinus bradycardia and sick sinus syndrome was still approximately 10%–15%; it of course remains to be seen whether the outcome will worsen with longer follow-up.

The preoperative assessment and planning for Fontan patients follows directly from the above considerations. In addition to standard noninvasive tests, echocardiography is useful to assess ventricular function and Fontan pathway patency; cardiac catheterization may be indicated in patients with poor or deterioration in function, and radiofrequency ablation should be considered before major elective surgery in appropriate patients. Invasive monitoring should be considered based on the individual patient and the planned procedure. Central venous cannulation facilitates monitoring of venous filling and pulmonary artery pressure, as well as mixed venous oxygen saturation. One must, however, be aware of the risk of thrombosis and serious impairment to venous return. In operations with the potential for major fluid shifts and/or cardiopulmonary embarrassment (e.g., spinal fusion), we have found it helpful to place a balloon-tipped

pulmonary artery catheter, particularly because it can be wedged and thus the transpulmonary gradient can be measured directly. Placement is best done in the catheterization laboratory under fluoroscopic guidance (and with the availability of special guide wires) because of the abnormal anatomy and absence of pulsatile waveforms.

Again, no specific anesthetic regimen can be recommended. General principles include maintenance of adequate preload, ensuring normal gas exchange while minimizing mechanical effects on PVR and pulmonary blood flow, limiting significant increases in the stress response, preserving sinus rhythm and ventricular filling and contractility, and avoiding large increases in afterload. As might be inferred, appropriate afterload reduction (as long as myocardial perfusion is maintained) is usually well tolerated and perhaps even beneficial to this circulation. Support of ventricular function, perhaps preferably with an inodilator such as milrinone, or otherwise with dopamine, should be considered in patients with evidence of ventricular dysfunction and/or in whom the surgery and its consequences are extensive. As mentioned previously, end-organ dysfunction can be a major postoperative complication in older Fontan patients. Contributing factors presumable include chronic low organ perfusion resulting from limited cardiac output and high venous pressures, with superimposed acute deterioration from, for instance, anesthetics or blood loss. The liver and kidneys seem particularly susceptible. This problem is a further indication to make all attempts to optimize Fontan pathway flow and myocardial performance; agents such as fenoldopam, which even at very low doses increase mesenteric and renal blood flow, are theoretically attractive. Because of the apparent increased risk of deep vein thrombosis or thrombus formation in the baffle or atrial appendage, consideration should be given after surgery to the use of subcutaneous heparin or low molecular weight heparin, along with adequate hydration and early mobilization. Facilities for pacing (external, esophageal, and overdrive for tachyarrhythmias), and external cardioversion should be immediately available in the operating room.

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